This book is provided in digital form with the permission of the rightsholder as part of a Google project to make the world's books discoverable online.

The rightsholder has graciously given you the freedom to download all pages of this book. No additional commercial or other uses have been granted.

Please note that all copyrights remain reserved.

**About Google Books**

Google's mission is to organize the world's information and to make it universally accessible and useful. Google Books helps readers discover the world's books while helping authors and publishers reach new audiences. You can search through the full text of this book on the web at [http://books.google.com/](http://books.google.com/)
Clinical Strabismus Management
Principles and Surgical Techniques
CLINICAL STRABISMUS MANAGEMENT
Principles and Surgical Techniques

Arthur L. Rosenbaum, MD
Professor of Ophthalmology
Chief, Division of Pediatric Ophthalmology
Vice-Chairman, Department of Ophthalmology
Jules Stein Eye Institute
University of California, Los Angeles School of Medicine
Los Angeles, California

Alvina Pauline Santiago, MD
Visiting Assistant Professor
Department of Ophthalmology, Division of Pediatric Ophthalmology
Jules Stein Eye Institute
University of California, Los Angeles School of Medicine
Los Angeles, California
Associated Eye Specialists
Pasig City, Metro-Manila
Philippines

W.B. Saunders Company
A Division of Harcourt Brace & Company
Philadelphia • London • Toronto • Montreal • Sydney • Tokyo
Figure on the cover adapted from graphics produced by Orbit® 1.8 Gaze Mechanics Simulation, Eidactics, San Francisco. Courtesy of Joseph Demer, M.D., Ph.D., and Joel Miller, Ph.D.
To my mentors, Arthur Jampolsky, M.D., and Alan Scott, M.D., for decades of intellectual stimulation and true friendship.

To Pauline Santiago, M.D., for her intense loyalty, dedication, and sense of excellence.

To my wife Sandra and my son Steve, for always being by my side with love, encouragement, and understanding.

To Jane, Hal, Emily, and Betsy, for their love and support.

To my father, Harry, for lovingly being the ultimate role model, and to my mother, Evelyn, for her deep sense of love and family.

Arthur L. Rosenbaum, M.D.

To my teacher and friend, Arthur L. Rosenbaum, M.D., for his knowledge, experience, guidance, and trust.

To my mentors, Anthony R. Caputo, M.D., and Sherwin J. Isenberg, M.D., for unselfishly sharing their knowledge.

To my father Mon and my mother Lea, for instilling in me the value of education and inspiring excellence.

To my extended family, Grandma Hok Im, Reynato Santiago, M.D., Lolita and Lina Santiago, Gervacio Lim, Jr., the Manalilis, the Puezeo, and the Mirandas, for their love and support.

To my friends, mentors, and colleagues from the Associated Eye Specialists, for waiting, encouraging, and believing.

And finally, to the Almighty, for making all things possible.

Alvina Pauline Santiago, M.D.
Leonard Apt, MD
Professor of Ophthalmology, Director Emeritus, Division of Pediatric Ophthalmology and Strabismus, Jules Stein Eye Institute, University of California, Los Angeles School of Medicine, Los Angeles, California. Special Consultant in Pediatric Ophthalmology for the Los Angeles City Health Department and the Bureau of Maternal and Child Health, Department of Public Health, State of California, Los Angeles

Inferior Oblique Weakening Procedures: Technique and Indications

Albert W. Biglan, MD
Adjunct Associate Professor of Ophthalmology, University of Pittsburgh School of Medicine. Director of Ophthalmology, Children’s Hospital, Pittsburgh, Pennsylvania

Pattern Strabismus

Edward G. Buckley, MD
Professor of Ophthalmology and Director of Pediatric Ophthalmology and Strabismus, Duke University School of Medicine, Durham, North Carolina

Fadenoperation (Posterior Fixation Suture)

Jean D. A. Carruthers, MD
Clinical Professor, Department of Ophthalmology, University of British Columbia, Vancouver. Active Staff, Vancouver Hospital and Health Sciences Center, and Mount St. Joseph Hospital, Vancouver, British Columbia, Canada

Strabismus in Craniosynostosis Syndromes

Joseph L. Demer, MD, PhD
Professor of Ophthalmology and Neurology, University of California Los Angeles School of Medicine. Chief, Comprehensive Ophthalmology Division; Director, Ocular Motility Clinical Laboratory; Chair, EyeSTAR Program, Jules Stein Eye Institute, University of California Los Angeles School of Medicine, Los Angeles, California

Orbital Imaging in Strabismus Surgery: Clinical Applications of Computer Models for Strabismus

David C. Dries, MD
Assistant Professor, Texas A and M University College of Medicine, Temple. Scott and White Clinic, Temple, Texas

Inferior Oblique Palsy: Diagnosis and Management

James E. Egbert, MD
Associate Professor, Department of Ophthalmology, University of Minnesota Medical School, Minneapolis, Minneapolis, Minnesota

Factors Influencing Measurement and Response to Strabismus Surgery

Forrest Daryel Ellis, MD
Professor Emeritus of Ophthalmology, Indiana University School of Medicine, Indianapolis. Midwest Eye Institute, Indianapolis, Indiana

Selected Surgical Complications

Aldo Fantin, MD
Fellow, Pediatric Ophthalmology, University of Minnesota Medical School, Minneapolis, Minneapolis, Minnesota

Factors Influencing Measurement and Response to Strabismus Surgery

Douglas R. Fredrick, MD
Assistant Clinical Professor of Ophthalmology, University of California, San Francisco School of Medicine. Director, Pediatric Ophthalmology, San Francisco General Hospital, San Francisco, California

Serious Neurologic Disease Presenting as Comitant Esotropia

Robert A. Goldberg, MD
Assistant Professor of Ophthalmology, Chief, Division of Orbital and Ophthalmic Plastic Surgery, Jules Stein Eye Institute, University of California, Los Angeles School of Medicine, Los Angeles, California

Strabismus After Orbital Fractures and Sinus Surgery

Lisabeth S. Hall, MD
Assistant Professor of Ophthalmology, New York Medical College, Valhalla. Assistant Director, Pediatric Ophthalmology and Ocular Motility, New York Eye and Ear Infirmary, New York, New York

Strabismus After Orbital Fractures and Sinus Surgery

Latif M. Hamed, MD
Former Chief of Pediatric Ophthalmology, University of Florida College of Medicine, Gainesville. Medical Director, Florida Eye Specialist Institute, Gainesville, Florida

Strabismus After Adult Cataract Surgery
Elias I. Traboulsi, MD  
Head, Department of Pediatric Ophthalmology, Director, The Center for Genetic Eye Disease, Cleveland Clinic Foundation Eye Institute, Cleveland, Ohio  
*Congenital Fibrosis of the Extraocular Muscles*

Lawrence Tychsen, MD  
Associate Professor, Departments of Ophthalmology and Visual Sciences, Pediatrics, Anatomy, and Neurobiology, St. Louis Children’s Hospital at Washington University School of Medicine, St. Louis, Missouri  
*Infantile Esotropia: Current Neurophysiologic Concepts*

Barry N. Wasserman, MD  
Attending Staff Surgeon, Wills Eye Hospital, Philadelphia, and Chester County Hospital, West Chester, Pennsylvania  
*Selected Surgical Complications*

David R. Weakley, Jr, MD  
Associate Professor of Ophthalmology and Director of Pediatric Ophthalmology, University of Texas Southwestern Medical School, Dallas. Director, Pediatric Ophthalmology Outpatient Services, Children’s Medical Center, Dallas, Texas  
*Brown Syndrome*

Anne Ziffer, MD  
TLC Northwest Eye, Seattle, Washington  
*Monocular Elevation Deficiency (Double Elevator Palsy) and Monocular Depressor Deficiency (Double Depressor Palsy)*
The editors are to be congratulated for attempting and accomplishing the almost impossible task of assembling in one place the distilled personal knowledge and management decision-making process of many experts for a wide variety of strabismus problems. The editors (who are also contributing authors) have chosen an outstanding array of strabismus experts, from a variety of schools of thought (sometimes called cults) and let the chips fall where they may.

The common thread was the editors’ charge to each contributing author to first provide the theoretical basis and principles before displaying their personal thought process and management decision-making alternatives. This carries the discussion to a new level. Every conceivable strabismus disorder is discussed with a level of detail and depth that will reveal the thinking process one uses to arrive at a clinical decision. The reader, whether student, general ophthalmologist, or specialist, will be better able to understand the rationale of the author’s management approach. It is naturally unrealistic to expect overwhelming agreement among the authors or among the readers. However, this is timely, since other ophthalmologic specialties are undergoing a bottoms-up review and re-evaluation of the theoretical basis and management rationale.

Clinical evaluation techniques are covered in an organized way, including a complete review of evaluation of strabismus mechanics and a thorough discussion of how to differentiate mechanical from innervational problems. The reader will find uniquely separate detailed discussions of evaluation and management of torsion problems, as well as the roles of computer modeling and orbital imaging in problem solving.

Between the covers of this superb new book one also finds an in-depth coverage of complex topics such as reoperations, strabismus after scleral buckling, Duane syndrome, strabismus associated with orbital trauma, and others, which are fully elaborated.

The editors succeed in their efforts because of their conceptual vision, backed by their respected positions in the strabismus community that elicited cooperation from so many differing experts. The editors’ excitement for their task is reflected throughout the book, and is contagious. We congratulate and thank them for this unique and outstandingly excellent contribution to strabismus management.

Arthur Jampolsky, M.D.
The clinical practice of strabismus is a complex challenge requiring the collection and synthesis of patient examination data, followed by the consideration of a myriad of therapeutic possibilities. More complicated cases require a reasoning and judgment process that is often difficult for the student to completely comprehend.

The primary goal of this book is to provide an in-depth and detailed description of the various strabismic entities, followed by a review of the treatment options, in such a way that the reader can grasp the author's reasoning process as various alternatives are considered.

The book begins with a review of clinical examination techniques and newer laboratory testing procedures, which aid in precisely clarifying diagnoses. Gathering accurate clinical data is often difficult, and the examiner needs every assistance possible to accomplish this task. Some clinical tests are traditionally regarded as being in the realm of the subspecialist. These have been simplified and illustrated so that the reader will feel competent and comfortable in their performance.

We have purposely included authors who represent different schools of thought. We hope that this diversity will be one of the strengths of the book. Obviously, each contributor will not agree with every point raised by other authors.

We have included a section on surgical management that details newer and less commonly performed procedures. Color digital images of these surgical techniques are used whenever possible to provide maximum clarity.

This project has truly been a labor of love. We hope that our personal excitement and enthusiasm for strabismus “rubs off” in its pages. We are profoundly appreciative to each of our contributing authors for their diligent commitment to this project. All have strived to communicate details of their knowledge and personal decision-making process for providing the best patient care.

We are also grateful to Bartly J. Mondino, M.D., and Bradley R. Straatsma, M.D., for providing assistance and encouragement during this book’s development as well as during our entire academic careers. In addition, we would like to thank the following persons and institutions for their support and technical expertise: the Jules Stein Eye Institute (JSEI); the Medical Photography Division of the JSEI, photographers Charlie Martin, Michael Heneghan, and Dennis Thayer; Grace Galvan-Silva and Stephanie Adams; the University of California in Los Angeles (UCLA) Office of Instructional Development, Reed Hutchinson, photoillustrator; Lynne Olson, artist; David Cramer, M.D.; and Richard Lampert and Shelley Hampton of the W.B. Saunders Company.

Arthur L. Rosenbaum
Alvina Pauline Santiago
We are grateful to Helen and Marty Kimmel for their friendship and generous support. Without their help, our dream of creating this book would not have been realized.
CONTENTS

section one

STRABISMUS EVALUATION AND PRINCIPLES OF MANAGEMENT ............. 1

chapter 1
CHIEF COMPLAINT, HISTORY, AND PHYSICAL EXAMINATION .................. 3
ASHISH MEHTA, MD

chapter 2
UNDERSTANDING SENSORY EVALUATION ...... 22
DAVID G. KIRSCHEN, OD, PhD

chapter 3
TESTS OF MUSCLE FUNCTION ............ 37
ALVINA PAULINE SANTIAGO, MD,
and ARTHUR L. ROSENBAUM, MD

chapter 4
EVALUATION OF OCULAR TORSION AND PRINCIPLES OF MANAGEMENT .......... 52
PAUL H. PHILLIPS, MD,
and DAVID G. HUNTER, MD, PhD

chapter 5
FACTORS INFLUENCING MEASUREMENT AND RESPONSE TO STRABISMUS SURGERY ........ 73
JAMES E. EGBERT, MD, and ALDO FANTIN, MD

chapter 6
ORBITAL IMAGING IN STRABISMUS SURGERY ........................................ 84
JOSEPH L. DEMER, MD, PhD,
and JOEL M. MILLER, PhD

chapter 7
CLINICAL APPLICATIONS OF COMPUTER MODELS FOR STRABISMUS ................. 99
JOEL M. MILLER, PhD,
and JOSEPH L. DEMER, MD, PhD

section two

HORIZONTAL DEVIATIONS ..................... 115

chapter 8
INFANTILE ESOTROPIA: CURRENT NEUROPHYSIOLOGIC CONCEPTS ................ 117
LAWRENCE TYCHSEN, MD

chapter 9
DIFFICULT ESOTROPIA ENTITIES: PRINCIPLES OF MANAGEMENT ............. 139
EDWARD L. RAAB, MD, JD

chapter 10
SERIOUS NEUROLOGIC DISEASE PRESENTING AS COMITANT ESOTROPIA ...... 152
CREIG S. HOYT, MD, and DOUGLAS R. FREDRICK, MD

chapter 11
DIVERGENCE PARALYSIS ...................... 159
LUCIOUS LIM, MD

chapter 12
INTERMITTENT EXOTROPIA .................. 163
ALVINA PAULINE SANTIAGO, MD,
MALCOLM R. ING, MD, BURTON J. KUSHNER, MD,
and ARTHUR L. ROSENBAUM, MD

chapter 13
SELECTED EXOTROPIA ENTITIES AND PRINCIPLES OF MANAGEMENT ........ 176
STEPHEN P. KRAFT, MD, FRCSC

chapter 14
PATTERN STRABISMUS ...................... 202
ALBERT W. BIGLAN, MD

section three

VERTICAL DEVIATIONS ...................... 217

chapter 15
SUPERIOR OBLIQUE PALSY AND SUPERIOR OBLIQUE MYOKYMIA ................. 219
DAVID A. PLAGER, MD
NOTICE

Medicine is an ever-changing field. Standard safety precautions must be followed, but as new research and clinical experience broaden our knowledge, changes in treatment and drug therapy become necessary or appropriate. Readers are advised to check the product information currently provided by the manufacturer of each drug to be administered to verify the recommended dose, the method and duration of administration, and contraindications. It is the responsibility of the treating physician, relying on experience and knowledge of the patient, to determine dosages and the best treatment for the patient. Neither the publisher nor the editor assumes any responsibility for any injury and/or damage to persons or property.

THE PUBLISHER
Figure 1–13. Asymmetry of the red reflex in the Bruckner test. A, In patients with refractive errors, the brighter crescents in the red reflex suggest the diagnosis. Normal patients (bottom) have equal red reflexes in both eyes. With a small hyperopia, small brighter crescents are seen superiorly (second from bottom). This crescent increases with the amount of hyperopia (top). Myopic patients will exhibit a brighter crescent inferiorly (second from top). B, In patients with strabismus, a brighter reflex is seen in the deviated eye. In this patient with accommodative esotropia and alternate fixation, the hyperopic crescent is shown best in the top photograph. The middle photograph shows the patient fixing with the right eye. The brighter reflex is seen in the deviated left eye. With the left eye fixing, this brighter reflex is now observed in the deviated right eye (bottom). This test may be used as a screening tool even in patients without an obvious abnormality. ( Courtesy of Gerhard Cibis, MD.)

Figure 4–10. Double Maddox rod test for measuring subjective torsion. Maddox rod cylinders have been accurately marked so that a line appears horizontal when the mark is at 90 degrees. A, Patient with trial frame in place. All room lights are turned off to fully dissociate the eyes. Examiner holding 6-PD prism base down in front of patient’s left eye to disrupt fusion. Both Maddox rod lenses are initially oriented 5 to 10 degrees away from vertical. Patient has already adjusted the left side and is now adjusting the right lens. B, Patient’s view. Right eye is excyclotorted (patient perceives torsion). Patient rotates right lens until lines are parallel. C, Patient’s view. Adjustment is complete. Right Maddox rod is extorted, confirming presence of subjective excyclotorsion. Amount of torsion can be read directly from the axis scale on the trial frame after the patient has made the proper adjustment.
Figure 5-6. The anterior extent of the medial rectus insertion retracts toward the corneoscleral limbus after disinsertion. This can result in more than a millimeter of anterior displacement of the insertion site. A, Before disinsertion, the distance between the insertion and the limbus is shown to be 5.5 mm with the use of a Scott curved ruler. B, After disinsertion, anterior displacement was evident, showing a distance between the insertion and the limbus of only 4.5 mm using the same Scott ruler.

Figure 7-1. Knapp's 1861 ball and string ophthalmotrope reflects only extraocular geometry, ignoring contractile and elastic tissue properties and coordination of innervations.
Figure 7–2. Some types of data expressible in a biomechanical model. Inward-pointing arrows indicate data that are incorporated into a model. Outward-pointing arrows indicate studies suggested by modeling. Cadaveric magnetic resonance imaging (MRI) can take advantage of long imaging times to give near-microscopic resolution of connective tissue planes. SOV, superior orbital vein; Globe, globe and optic nerve; Sling, posterior sling portion of lateral rectus pulley. Clinical Alignment data are compared with model predictions. Immunohistochemistry identifies specific tissue types. Thin slices are then digitized and reconstructed to restore three-dimensional tissue relationships. Region of medial rectus (MRI) pulley is shown, with a quadrant of collagen (Coll) (blue) removed to reveal smooth muscle (SM) (pink). Alert Patient MRI was initially used to establish normal functional anatomy\textsuperscript{23} and is now used to study anatomic and contractile abnormalities and effects of surgery.\textsuperscript{10–14} Preoperative (PreOp) and postoperative (PstOp) scans are shown for a case of lateral rectus (LR) palsy treated by vertical rectus transposition. The belly of the transposed superior rectus (SR) resists lateral pull of the transposed insertion.

Figure 7–3. Anterior medial rectus showing collagen, elastin, and smooth muscle. Computer reconstruction is shown from two posteroinferior perspectives (some readers will be able to fuse the two images as a crossed-disparity stereogram). The medial rectus (brick red) with cut posterior edge and a piece of the globe (green) are shown. So that other structures would be visible, we decimated the collagen (blue), rendered it translucent, and deleted its inferomedial quadrant. Thus, collagen is actually denser than shown. For clarity, only the highest densities of elastin (orange) are shown. Elastin encircles the medial rectus. A band of smooth muscle (pink) also encircles the muscle and is particularly dense between the muscle and the orbital wall, perhaps serving to control the coupling of the medial rectus to the orbital wall. (Quick Time\textsuperscript{®} VR movies of this and other reconstructions are available at Internet address www.ski.org/JMMiller_lab/OTA proj.)
Figure 7–4. Diagrammatic representation of structure of orbital connective tissues. IO, inferior oblique; IR, inferior rectus; LPS, levator palpebrae superioris; LR, lateral rectus; MR, medial rectus; SO, superior oblique; SR, superior rectus; tndn, tendon. Coronal views represented at levels indicated by arrows in horizontal section.

Figure 7–6. A Clemente Clark Electric Hess Screen, modified by replacing each red fixation LED with a vertical row of three LEDs, allowing measurement of ocular torsion. The subject sits with chin (and if necessary, forehead) stabilized against a support 50 cm from the screen. Eyes are dissociated with red and green filter goggles. The subject indicates localizations using a hand-held green streak projector.
Figure 7–12. Patient TS, three-dimensional MR reconstructions of extraocular muscle paths in primary gaze. A, Preoperative MR image shows left superior oblique and left lateral rectus atrophy typical of chronic denervation. Note, in inset, posterior view showing unusually thin lateral rectus. B, Postoperative MR image shows little lateral displacement of superior rectus and inferior rectus belly (not visible in this view), with marked path inflections (arrow to superior rectus) as muscles course through pulleys to their transposed insertions.

Figure 22–1B. Intraoperative findings reveal a large sponge beneath lateral rectus causing restriction. After sponge removal, motility and restriction improved. Note tightness of the muscle in top left photo with sponge (arrow) under the muscle and laxity of the muscle after sponge removal (lower left). The large arrow in the lower right photo indicates the cut edge of the sponge. Small arrow indicates adhesions between sclera and muscle.
Figure 22-9. A Baerveldt device implanted between two rectus muscles. (Courtesy of M. Roy Wilson, MD.)

Figure 29-5. Ragged red fibers in orbicularis oculi muscle of a patient with mitochondrial myopathy (progressive external ophthalmoplegia). (Masson’s trichrome stain at 300× magnification.) (Courtesy of Narsing Rao, MD, University of Southern California, Doheny Eye Institute.)

Figure 30-10. Lagophthalmos due to poor Bell’s phenomenon with complications showing conjunctival edema and threatening corneal integrity.

Figure 30-11. Intraoperative view of forehead advancement in a child with Apert syndrome. (Courtesy of Don Fitzpatrick, MD.)
STRABISMUS EVALUATION
AND PRINCIPLES
OF MANAGEMENT
Chief Complaint

The most important and revealing component of the medical history is the chief complaint. Concise and clear questioning is required. The specific ocular or visual disturbance that led the patient to seek ophthalmologic care provides the first insight into the patient’s problem. If answers are vague, questions should be rephrased to clarify the patient’s or parent’s concern. Certain chief complaints are often heard from patients presenting to a strabismus practice. A few of these will be discussed within the framework of obtaining a history.

DIPLOPIA AND VISUAL CONFUSION

Patients complaining of double vision may actually be describing blurred vision rather than diplopia. If true diplopia exists, ascertain whether it is monocular or binocular. If the double vision is monocular, ocular disease other than a motility disturbance should be considered, such as corneal abnormality, lenticular opacification, or retinal pathology.

Binocular diplopia strongly suggests a strabismic origin. Patients with an obvious cause such as paralytic or restrictive strabismus often describe the location of two disparate images in the horizontal and vertical planes but have difficulty appreciating torsional misalignment. The latter may be described as slanted, but this term is also used to describe a combination of vertical and horizontal misalignment. To confirm torsion, patients should look at a vertical line (e.g., the edge of a door) and describe tilting. Patients who appreciate tilting should be carefully evaluated for torsion (see Chapter 4). Precipitating factors such as trauma or a cerebrovascular accident should be noted, and one should verify if specific events such as reading, hot weather, or fatigue precipitate the onset of intermittent diplopia.

If the same object of interest is seen by the fovea of one eye and the peripheral retina in the deviated eye, diplopia results. Most patients with diplopia become aware of a second image. Diplopia may be relieved when the horizontal and/or vertical deviations are neutralized with prisms in free space or with the synoptophore. The latter can also neutralize a torsional component of strabismus. In strabismus that is acquired after visual maturity (usually after age 7), patients who do not describe diplopia may be ignoring a second image, especially when the deviation is large; or they may have poor visual acuity in one eye so that two distinct images are not seen. Patients younger than age 7 frequently learn to suppress a second image.

Visual confusion results when the fovea of each eye sees two different objects that are interpreted by the brain as being in the same visual direction. Visual confusion is an expected sensory response in patients having an ocular deviation with good foveal function in both eyes. Patients, however, may not be able to accurately describe this phenomenon, much less volunteer the information. The clinician may need to ask specific questions to identify patients with visual confusion.

ASTHENOPIA

Asthenopia refers to symptoms of ocular fatigue or tiredness. It is usually associated with sustained near work but may also occur with distance vision and may be a result of strabismus or refractive problems or a combination of both. Refractive asthenopia results from an improperly corrected refractive error. Relation to a particular activity such as reading, or to a certain time of day, should be sought. Is there associated diplopia or visual blurring? Are symptoms relieved when the initiating activity is discontinued or when the patient reads with one eye covered?

ABNORMAL EYE MOVEMENTS

Abnormal eye movements may be described as “wobbly eyes” or “eyes that do not move together.” This type of complaint can usually be clarified by asking the parent or patient to point to the abnormal eye(s). “Wobbly eyes” may mean that a patient has nystagmus. “Eyes that do not move
together” may reflect an incomitant strabismus such as Duane syndrome. A complaint of abnormal eye movement should be explored further by asking about the presence of a preferred gaze position or whether an anomalous head posture is adopted.

**History**

Generally, questions asked during history taking should be based on the chief complaint. Certain basic information is required from any patient with a strabismic disorder. This information is summarized in Table 1–1.

If patients cannot recall specific strabismus surgical procedures, they usually remember whether surgery was performed in one or both eyes. This can guide the clinician in seeking clues during the ocular examination (e.g., conjunctival scarring). The direction of the deviation is usually not forgotten, especially if preoperative photographs are available. Photographs are invaluable for assessing an anomalous head posture.

The presence of associated neurologic signs and symptoms may provide crucial diagnostic assistance; these include headache, seizures, ataxia, acquired nystagmus, muscle weakness, fatigue, ptosis, and bowel or bladder incontinence. Variability in clinical features or worsening with fatigue suggests myasthenia gravis. Characteristics systemic abnormalities may be noted in patients with Duane syndrome, Möbius syndrome, and craniofacial anomalies (see Chapters 24, 26, and 30 for a more detailed discussion).

**Physical Examination**

**ASSESSMENT OF VISION**

Visual acuity testing in ophthalmology is akin to obtaining vital signs in other fields of medicine. It provides crucial information regarding visual function and is the key measure for monitoring the effectiveness of treatment in amblyopia. It helps predict preoperative and postoperative monocular fixation preference, which may influence management.

Complete occlusion is mandatory for monocular assessment of visual acuity. An occlusive patch should be used in any child without nystagmus. Children reported to have normal acuity in both eyes may in reality be profoundly amblyopic in one. Visual acuity reports from another examiner may have been obtained binocularly or with inadequate occlusion that allowed peeking through the good eye.

**Visual Milestones in Infancy**

At birth, a blinking response to bright lights should be present. A vestibulo-ocular reflex is observed by 7 days in a full-term infant. It may be induced by gentle, rapid rotation of the child’s head to one side (doll’s head maneuver). The normal response is a nystagmus with both a slow and fast (saccade) phase, which should subside in approximately 3 seconds. A blind or abnormally sighted child cannot easily inhibit the induced vestibular nystagmus, which may take more than 15 to 30 seconds to resolve.

In neonates, the best fixation target is the human face—if not the examiner’s then certainly the mother’s. Visual milestones may be delayed in prematurity and neurologic disease. Cerebral palsy may be associated with abnormal or absent saccades (generated as the fast phase of nystagmus). The normal fast phase may not be evident. Following movements may not be due to smooth pursuit but rather may represent hypometric saccades. These infants may be misdiagnosed as being blind because of poor saccadic excursion.

**Fixation Patterns**

As early as 3 months of age, some infants may show transient fixation and following responses. This progressively improves, and by 6 months of age the infant should clearly fixate and follow an object. At this age and thereafter, the words “central, steady, and maintained (CSM)” are used to describe fixation ability. “Central” refers to a corneal light reflex from a fixation light falling in the center of the pupil. The reflex is also considered normal if it falls in the same location in both eyes under monocular conditions. Steadiness of fixation is assessed with a muscle light held in front of the child as it is moved slowly. An accommodative target such as a small, thumb-sized toy is best coupled with the light. Nystagmus or oscillations result in unsteady fixation. “Maintenance of fixation” refers to the ability to keep the eye fixed on a target when either eye is covered.

The CSM method cannot reliably detect amblyopia unless strabismus with a fixation preference is present. Fixation
preference testing reliably detects amblyopia only if the deviation exceeds 10 PD. Otherwise, other clinical methods of assessing vision are required.

The 10-PD test is specifically designed to assess fixation preference in preverbal children who are not strabismic or who have small deviations. It may be performed either in the base-down or base-up position. The prism is placed in front of one eye while an appropriate accommodative target is presented at near and/or distance. Spontaneous alternation of fixation between the two eyes should be observed. If one eye is clearly preferred, that eye is covered to allow the other eye to assume fixation and be observed for maintenance of fixation. If fixation is held for more than 5 seconds, through a blink, or with a smooth pursuit, approximately equal acuity is present. Smooth pursuit may be elicited at near by moving the fixation target and at distance by rotating the patient’s head or swiveling the examining chair. The same procedure is performed on the other eye.

Orthotropic patients without a fixation preference on the 10-PD test will demonstrate no difference in recognition acuity between the two eyes. Orthotropic patients who have a fixation preference, or exhibit less than 10 PD of deviation, cannot be reliably concluded to have amblyopia based on fixation analysis alone.

A false impression of equal vision in a monofixator with peripheral fusion may be gained from this test. Alternating fixation may be observed after removing the eye from its facultative scotoma during presentation of the vertical prisms. If the patient experiences vertical diplopia, fixation may be switched, giving a false impression of equal acuity. Maintenance of fixation should be observed throughout a smooth pursuit. A poorer-seeing eye will be unable to perform this task, revealing its true visual level.

Alternatively, 25-PD base-in prism can be used in preverbal children. The test need not be performed with occlusion of either eye, or through a smooth pursuit. Either of these maneuvers may result in loss of cooperation and an unreliable result. The test assumes that fixation will be preferred in the eye without prisms before it, because prisms induce an esotropic shift and a consequent decline in acuity. If acuity is the same in both eyes, alternating fixation should be noted when the prism is moved from one eye to the other. The test can reliably predict the absence of unequal vision in orthotropic patients with free alternation after induced esotropia. If a strong fixation preference is found, the test may not be a reliable indicator of unequal visual acuity; a fixation preference may persist in treated amblyopes despite the same recognition acuity in both eyes.

Patients with infantile esotropia who demonstrate cross fixation can be reliably stated to have equal acuity only if switching of fixation occurs past the midline in either eye. If fixation preference persists after moving a target past midline into abduction, unequal acuity exists. The eye that does not pick up fixation is amblyopic.

Alternate Methods of Assessing Vision in Preverbal Children

Other methods of visual acuity assessment in preverbal children include optokinetic nystagmus (OKN) reflex testing, forced-choice preferential looking (FPL) techniques (e.g., Teller acuity cards), and sweep visual-evoked potentials (VEP) (Fig. 1–1).

The appropriate binocular response to the OKN stimulus is consistent with visual acuity of approximately 20/400 in the newborn. As the visual system matures, visual acuity correlates with an appropriate increase in the OKN response until it reaches 20/30 by age 20 to 30 months. Unfortunately, OKN testing has several limitations. The target stimulus must be presented under rigidly standardized test conditions specifying lighting and the speed of target presentation to stimulate both central and peripheral vision. These requirements make it difficult to repeat the test with the OKN drum in the clinical setting. Moreover, the OKN response requires both normal (sensory) input and (motor) output. An infant with a normal sensory system will have a poor OKN response if a motor problem exists. The response has been observed in infants without a visual cortex, suggesting that an extrageniculate system exists.

FPL techniques rely on the principle that an infant prefers...
to look at a pattern stimulus rather than a homogeneous target (Fig. 1–2). Reliable testing requires a cooperative child as well as a trained observer. Teller acuity cards measure resolution acuity in cycles per degree. This may overestimate Snellen recognition acuity. Fixation preference is a more reliable indicator of amblyopia than Teller acuity cards in patients with strabismus. The benefit of FPL techniques lies in their ability to sequentially follow patients with suspected subnormal vision, ensure that vision is developing appropriately in both eyes, and monitor amblyopia therapy.

Sweep VEP testing has been refined since its original description. However, despite convincing evidence of its validity in assessing visual acuity, its usefulness is limited because it is a sophisticated test requiring expensive equipment and technical expertise.

Assessment of Vision in Verbal Children

As children become verbal, recognition acuity may be assessed. Allen pictures or matching games (e.g., Sheridan Gardner or HOTV) may be used as initial recognition acuity tests, but the gold standard is to use Snellen optotype letters. As the child matures, illiterate E testing becomes possible. Allen pictures overestimate acuity when compared with the illiterate E or Snellen optotype acuity. Parents may practice the E game at home with the child before testing. Also, Snellen optotypes (letters or numbers) can be introduced even in a young child to familiarize the patient with the test.

Older children are able to memorize standard eye charts in most offices. This can be circumvented by asking patients to read backward or begin in the middle before proceeding in either direction. The Mentor BVAT system allows the random generation of acuity symbols. This is especially advantageous for amblyopic patients receiving treatment who make frequent office visits. Isolated targets overestimate acuity in patients bothered by crowding when targets are presented in a line. If a patient cannot understand where to start, an assistant should point to the symbol of interest without obscuring the rest of the symbols in that line.

Assessment of Vision in Nystagmus

Before assessing monocular vision in nystagmus patients, one must ascertain the smallest distance and near acuity targets visible with binocular viewing. Attention should be paid to anomalous head postures. Distance and near acuity testing should be repeated in the forced primary position to check for deterioration.

When assessing monocular vision in a patient with nystagmus, an occluder placed in front of one eye may cause nystagmus to worsen, leading to a decline in recorded acuity. Other means of monocular occlusion should be used that provide some form of peripheral binocular cues to prevent worsening of nystagmus but permit monocular assessment of central vision. Four methods are commonly used: remote occlusion, high plus lenses for fogging, neutral density filter, or American Optical (AO) vectograph testing. The method used to measure acuity should be noted, and subsequent testing should use the same method so that meaningful comparisons of visual acuity can be obtained.

Remote occlusion is most reliable in cooperative patients. The occluder is placed a certain distance in front of one eye but close enough so that the eye cannot see the acuity symbols presented. Alternatively, a translucent semiopaque (Spielmann) occluder may be used (Fig. 1–3). High plus lens occlusion is a simple yet effective technique. A lens at least +4.00 D more than the known or anticipated refractive error for a given distance can be used. If neutral density filters are used, the highest filter that does not appreciably worsen the nystagmus should be chosen. AO vectograph
testing requires wearing polarized glasses. The vectograph projects letters corresponding to those in the polarized glasses. The right and left eyes view letters in the right and left polarized lenses, respectively.

**SENSORY TESTING**

Sensory testing is an integral part of strabismus evaluation. Because one may not know a patient's motor status on initial consultation, it is advisable to begin with sensory testing, specifically stereoacuity testing. Patients with poorly controlled intermittent deviations may break down into a frank tropia if binocularity is disrupted even momentarily by any form of monocular occlusion (e.g., during visual acuity and cover testing). All sensory testing is performed with the appropriate refractive correction.

**Near Stereoacuity**

Numerous stereoacuity tests at near have been described in the literature. The Titmus fly stereotest is most readily available and perhaps the most familiar but has limitations (Fig. 1–4). The Titmus stereotest has monocular cues in the fly, the three animals, and at least the first row of circles. The most obvious of these clues is lateral displacement of the targets. True stereopsis may be confirmed by rotating the test target 180 degrees. The target that was elevated should sink or regress below the plane of the book. One may also verify by asking if lateral displacement is the basis of the patient's response. Rapid alternation of viewing by the two eyes enables a highly observant individual to appreciate "stereopsis." The same patients can give appropriate responses to rotation, but none will achieve more than 140 seconds of arc stereoacuity, suggesting that true stereopsis begins at levels better than this.

The level at which the wings of the fly are grasped suggests the stereoacuity level. Patients with normal stereoacuity (40 seconds of arc) will grasp the wings several centimeters above the Titmus test plane, whereas monofixators with less than normal stereoacuity will grasp the wings closer to the test plane. Stereopability of 40 seconds of arc or better is considered indicative of bifoveal fixation. Stereacuity worse than 40 seconds of arc suggests peripheral fusion.

Random dot stereograms (e.g., the TNO and randot E) are alternative tests for near stereoacuity (Fig. 1–5). These stereograms lack monocular cues but are more difficult to understand and complete. Red/green or polarized glasses need to be worn, and they may impede cooperation by some children. The TNO test is based on the anaglyphic (red/green dissociation) method. Many believe the red/green glasses are more dissociating than the polarized glasses of the Titmus test, contributing to decreased or absent stereoacuity measurements.

A few patients who are stereoblind on random dot testing exhibit legitimate stereoacuity on the Titmus test that is not based on monocular cues or alternation. The inability to perform on random dot testing may be due to (1) crowding effects from closely placed random dots in the stereogram; (2) a lack of monocular cues, preventing the vergence movements necessary to appreciate disparity—the basis of stereopsis; and (3) an inability of monofixators to resolve the numerous fusional ambiguities in the test.

The Frisby and Lang stereotests do not require glasses (Fig. 1–6). The Frisby stereotest is cumbersome and difficult for young children to understand. By the time most children are able to cooperate with this test, they are able to perform any of the tests requiring glasses. In contrast, the Lang test is useful in children because it depicts recognizable objects (e.g., Lang I: a star, cat, and car) at various levels of disparity. Stereopsis can be demonstrated, but the precise level of stereoacuity cannot be quantified.

**Distance Stereoacuity**

Two types of stereotests are available for distance stereoacuity testing. The older of these is the AO vectograph stereotest, which is dissociating because of the need to wear polarized glasses. Moreover, lateral displacement of circular targets provides monocular cues. More recently, the Mentor BVAT system was developed to assess distance stereoacu-
The test is a computerized system in which liquid crystal binocular glasses are connected to a microprocessor (Fig. 1–7). The glasses contain a liquid crystal shutter aperture for each eye that selectively blocks out light transmission. Each eye is presented with disparate images alternating at 60 Hz. The rapid alternation of these images allows simultaneous perception, because the frequency is higher than the binocular lateral critical flicker fusion level of 30 Hz. Two distance stereoacuity tests are included in this system: the randot tumbling E (Fig. 1–8) and contour circles (Fig. 1–9). Normal patients exhibit lower levels of stereoacuity with the randot E test.

Distance stereoacuity testing differentiates patients with poor control from those having good control in intermittent exotropia (see Chapter 12). A dynamic stereoacuity test that measures motion in depth has been described. Preliminary results suggest that this is a more sensitive measure of control in intermittent deviations.

Worth Four-Dot Test

If stereoacuity can be demonstrated with the tests discussed previously, Worth four-dot (W4D) testing need not be done to assess binocular function. It may, however, be used to define the size and location of a suppression scotoma in many patients with strabismus with less than normal stereoacuity (see later discussion). In the absence of stereo ability, the test may provide useful information if fusion can be demonstrated.

Worth, in the early 1900s, described using red/green glasses and four dots—one white, one red, and two green—to measure binocularity. The test may be used to determine the presence of fusion, suppression, and diplopia (Fig. 1–10). If two red or three green lights are seen, suppression is present. If four lights are seen, fusion exists but must be verified by the presence of motor fusion on cover testing. A patient who sees five lights has diplopia but must be distinguished from one who alternates rapidly between two red and three green dots. All other responses are equivocal.

The usefulness and reliability of the W4D test has been criticized for several reasons: (1) Reversal of the red/green
glasses changes the response from fusion to suppression, and vice versa, in some patients. For example, the higher contrast of the green images compensates for reduced contrast sensitivity in the ambyopic eye, changing the response depending on whether the green glass is in front of the amblyopic or the normal eye. (2) The luminance under which testing is performed is difficult to standardize in the clinical setting. (3) The poor quality of many commercially available red/green glasses permits the appreciation of monocular cues. (4) The test is dissociating because of the anaglyphic (color) nature of the test glasses used. The test may not reflect the true binocular status under everyday visual conditions. The response may be checked by reversing the color in front of either eye. If different responses are given, fusion is tenuous, suppression is possible, or the results are invalid.

A newer polarized version decreases the dissociating nature of the anaglyphic method. Fusion can be detected better, but the test is more difficult for children to understand. Preverbal children may be asked to touch the dots at near, but their responses are unreliable and poorly predictive of true sensory status.15

Despite criticisms, the W4D test remains useful for assessing binocularity in a cooperative patient when the results are unequivocal. If fusion cannot be demonstrated, however, it cannot be concluded with certainty that it does not exist because of the limitations just discussed. Peripheral fusion may still be demonstrated despite the absence of stereoaucity and W4D fusion if less dissociating tests such as Bagolini striated glasses are used.

The size of a suppression scotoma can be quantified using the W4D flashlight.16 To perform this calculation, the target size (W4D flashlight) presented at distance and near is measured along with the distance it is held from the patient. The size of the scotoma is calculated using the tangent angle ($\theta$) of the ratio of the object size ($a$) to object distance ($b$): $\tan \theta = a/b$ (Table 1–2). The degree of fusion may also be defined based on the size of the scotoma: a 3.0- to 6.4-degree scotoma size means peripheral fusion; 1.14 to 3.0 degrees indicates macular fusion; and foveal fusion is indicated if the scotoma is less than 1.0 degree. For example, a patient who fuses at 13 inches but suppresses at 72 inches has a scotoma size of 2.29 degrees and has macular fusion.

**Retinal Correspondence**

Retinal correspondence refers to the ability of the sensory system to appreciate the perceived direction of the fovea and other retinal elements in each eye relative to the other. In orthotropic patients, the object of regard stimulates each fovea simultaneously and therefore is localized subjectively as being straight ahead. The two eyes have corresponding retinal elements that have a common visual direction. Corresponding elements localize an object at the same point in space. The two foveas represent the highest degree of correspondence.

Normal retinal correspondence occurs in straight eyes (no tropia) under binocular conditions or when the patient’s objective and subjective angles of strabismus are the same. The objective angle is measured by the alternate prism cover test. The subjective angle is determined by measuring the amount of neutralizing prisms required for superimposition or fusion. Prisms are placed over the nonfixing eye until diplopia resolves, the direction of disparity changes (crossed to uncrossed), or fusion occurs. A more elaborate method of determining retinal correspondence employs the synoptophore (see also Chapter 2).

Anomalous retinal correspondence (ARC) is present when the objective and subjective angles are not equal. It is a sensory adaptation of the immature visual system to an abnormal motor position of the eye. It allows the child some semblance of binocular vision in the presence of strabismus and prevents diplopia. Suppression is an accompaniment of

<table>
<thead>
<tr>
<th>Distance Flashlight is Held (ft)</th>
<th>Scotoma Size (degrees)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>6.4</td>
</tr>
<tr>
<td>3</td>
<td>2.3</td>
</tr>
<tr>
<td>6</td>
<td>1.1</td>
</tr>
<tr>
<td>8</td>
<td>0.9</td>
</tr>
<tr>
<td>10</td>
<td>0.7</td>
</tr>
<tr>
<td>15</td>
<td>0.5</td>
</tr>
<tr>
<td>20</td>
<td>0.3</td>
</tr>
</tbody>
</table>
ARC with a facultative scotoma in the nonfixing eye. Facul-
tative implies that the scotoma can change to the other eye if fixation is switched, preventing diplopia.

There are two types of ARC: harmonious and unharmoni-
ous. Harmonious ARC occurs when the subjective angle is
zero. For example, a patient who has 20 PD of esotropia on
alternate cover testing but who reports a subjective fusion
response with Bagolini lenses (without neutralizing prisms)
has harmonious ARC. Unharmonious ARC occurs when the
subjective angle is greater than zero but less than the objec-
tive angle. This results from incomplete sensory adaptation,
a test artifact, or a change in the original strabismus angle.

In ARC, two different but related phenomena occur: First,
the fovea in the deviating eye loses the same visual direction
as the fovea in the fixing eye. Some tests of ARC evaluate
the relationship of the foveas to one another (e.g., afterimage
test and the amblyoscope). Second, the fovea of the fixing
eye shares a common visual direction with a peripheral
retinal element in the deviating eye. Other tests of ARC
determine the relationship of the fovea of the fixing eye to
the retinal periphery in the nonfixing eye (e.g., Bagolini
striated glasses and the amblyoscope).

The afterimage test labels the fovea of each eye with a
linear afterimage. Each fovea is stimulated separately (mon-
ocularity). The patient views a linear light filament while the
central zone of the light is occluded to allow the fovea to
fixate but remain unlabeled. The vertical afterimage is pre-
sented to the deviating eye, because suppression scotomas
usually occur along the horizontal meridian. A horizontal
afterimage will be obscured if placed in front of a deviating
eye with suppression. The vertical afterimage is presented
first because it is easier to recall. After the presentation of
afterimages, the patient recalls and draws his or her percep-
tion. In normal retinal correspondence a cross with a central
gap is seen. If there is esotropia and ARC, the afterimages
will be crossed; in exotropia with ARC, the afterimages will
be uncrossed. Patients with eccentric fixation should have
the afterimages placed directly on the fovea. This may be
done ophthalmoscopically using a visuscope target.

Bagolini striated glasses do not have dioptic powers but
have narrow striations running in one principal meridian,
oriented at 45 and 135 degrees during testing. These lenses
allow evaluation of retinal correspondence at distance and
near with minimal dissociation and reflect the binocular
status under everyday visual conditions. The patient’s re-
sponse is checked while fixing a light at near. Immediately
after the response, cover testing is done over the relatively
clear Bagolini lenses to check for a deviation. If the response
is unclear, the patient is instructed to pay attention to the
presence of small gaps in the line, especially toward the
center and asked to draw what was perceived. The various
responses are summarized in Figure 1–11.

ARC is not an “all or nothing” phenomenon. It may be
present under daily visual circumstances or manifest only
when tests dissociate the patient from the normal visual
environment. ARC found on afterimage testing is deep
seated. ARC found using Bagolini striated glasses is proba-
bly present under everyday conditions. Suppression, ambly-
pia, and ARC occur in children who acquire strabismus
before reaching age 7. Twenty percent. It is rare for these abnormalities to
develop later; their presence suggests childhood strabismus.

Four-PD Test for Foveal Suppression

The 4-PD base-out test is used to detect small suppression
scotomas associated with monofixation syndrome or as a
motor test of bifoveal fixation. The 4-PD base-out prism is
placed over one eye as the patient fixates an accommodative
target at distance. The farther the target, the more central
the area being tested. Testing is repeated with the prism over
the opposite eye.

In a bifoveal fixator the normal response is a conjugate
saccadic version movement in both eyes, followed by a
slower vergence recovery movement in the eye without the
prism. A 4-PD base-out prism over the right eye will cause
temporal displacement of the image on the retina in this
eye. Nasally directed saccades occur in the right eye, and
temporally directed ones occur in the left eye (version move-
ment in the direction of the apex of the prisms). Next, a
slower vergence movement directed nasally in both eyes
(convergence) establishes bifoveal fixation. The movement
is best observed in the eye without the prism.

When a suppression scotoma is present, a 4-PD base-out
prism placed in front of this eye will not elicit movement.
If placed in front of the fixing eye, the initial conjugate
saccade toward the apex of the prism is noted. The slow
recovery movement in the eye without the prism will not be
seen, however, identifying it as the eye with foveal suppres-
sion (Fig. 1–12).

Four atypical responses in normal patients have been
reported with this test: (1) The eye without the prism
makes no movement whereas the eye with the prism makes
the expected nasally directed version movement. This occurs
when version and vergence movements are equal but oppo-
site in direction in the eye without the prism or when the
version movement in this eye is too small to be appreciated.
(2) Normal movement is not observed in either eye until a
few seconds later. This occurs with inattention to the fixation
target or defective fusion. (3) Oscillating movements in the
same direction are seen in both eyes, followed by slow
recovery of fusion by disconjugate eye movements. (4) De-
lay in vergence movement is seen following the initial move-
ment, most apparent in the eye without the prism.

The second atypical response is akin to the response to a
4-PD base-out prism placed in front of an eye with a sco-
toma, except that the normal response is seen after a longer
observation period. Atypical responses 2 and 4 may be
suspected if a patient appreciates diplopia. A patient with a
true scotoma suppresses without diplopia. The most signifi-
cant cause of an atypical response is a defective fusion
reflex, seen with poor fusional convergence amplitudes (mo-
tor) and less than normal stereoacuity (sensory).

Postoperative Diplopia

In some situations, predicting postoperative diplopia may
be crucial in deciding on a treatment strategy. In the clinic,
prisms that simulate overcorrection are held before the pa-
tient’s eyes at distance and near. If no diplopia is reported,
no further testing is required; diplopia is unlikely. If diplopia
is described, the patient is given sufficient time to resolve it.
Prisms are placed on Janelli clips or on a trial frame for 15
to 20 minutes. If apparent diplopia resolves, true diplopia
is unlikely. If the patient remains diplopic, Fresnel add-on
prisms are tried and the patient reevaluated in a week.
Diplopia that resolves is unlikely to represent persistent postoperative diplopia. Postoperative diplopia that persists may be bothersome and require additional intervention.

**Fusional Amplitudes**

Knowing the patient’s fusional amplitudes provides insight into his or her ability to control an intermittent deviation. Base-out prisms are used for convergence, base-in for divergence, base-up for infravergence, and base-down for supravergence. Amplitudes are measured using rotary prisms or prisms held in free space (prism bar or loose prisms) as the patient fixes on an accommodative target at distance and near. Details of determining convergence and divergence fusional amplitudes are discussed in Chapter 12 on intermittent exotropia, where the information has proved most useful in the clinical setting.

Slow vergence movements dissipate slowly. Determining fusional divergence immediately after measuring fusional convergence may result in artificially low fusional divergence amplitudes. Measuring vertical fusional amplitudes alternately with horizontal amplitudes or waiting a few minutes in between measurements will circumvent this problem.

Normal distance vergence movements are approximately 20 PD for convergence; 6 to 8 PD for divergence; and 3 to 4 PD for vertical vergence. These figures increase by 6 to 10 PD for convergence and divergence at near. There is no significant difference in vertical fusional amplitudes between distance and near.

**MOTOR TESTING**

Careful measurement of the angle of deviation in strabismus is crucial in planning surgery. Cover tests are preferred clinically, of which the alternate prism cover test (measuring the manifest and latent deviation) is most reliable. If these cannot be performed in uncooperative children or because of poor fixation, light reflex tests may be used. These are less accurate than the alternate prism cover test in patients with good fixation.

**Light Reflex Tests**

Light reflex tests should only be used to measure the angle of strabismus in patients unable to cooperate with cover testing.

**Brückner Test.** The Brückner test detects the presence of ocular abnormalities by observing an asymmetric red reflex through the pupil (Fig. 1–13). It can identify patients with
strabismus, ametropia, lenticular opacity, or retinal pathology. The deviated or ametropic eye demonstrates a brighter reflex than the fellow eye, whereas lenticular opacities or retinal pathology cause either a more dull or a brighter than normal reflex. The test should be performed in a dimly illuminated room with coaxial lighting. A direct ophthalmoscope with a halogen light source is preferred. The child sits on the parent’s lap while the coaxial light is shone on the pupils. The examiner sits at approximately arm’s length from the patient. Normally, both pupils constrict and light reflexes in both eyes become equally dull. The corneal light reflex (Hirschberg) can be observed (described later). Amblyopia may be detected by observing the monocular response in the presence of an asymmetric reflex.\(^\text{51}\) If immediate maintained fixation occurs with pupillary constriction when moving the light from the fixing eye to the abnormal eye, amblyopia is unlikely. The Brückner test is unreliable in patients younger than 8 months, because asymmetrical dimming of the red reflexes occurs in 28% of normal infants.\(^\text{3}\)

**Hirschberg Corneal Light Reflex.** The Hirschberg\(^\text{24}\) corneal light reflex test relies on observing reflected light on the cornea from a light source held 14 inches away. If the eyes are deviated, the light reflex will fall on different locations in the deviating and fixing eyes. The reflex is displaced nasally in exotropia and temporally in esotropia. Each millimeter of decentration has classically been taught to equal 7 degrees or 15 PD of misalignment. As a general rule, if the pupils are 3.5 mm in diameter, a reflex displaced to the pupillary margin is 15 degrees; to halfway between the pupil and limbus, 30 degrees; and to just outside the limbus, 45 degrees. Studies have shown that the conversion from degrees to prism diopters is not linear for all degrees of displacement but is estimated to be 21 PD/mm of decentration.\(^\text{1,4}\)

**Krimsky Test.** The Krimsky test is an extension of the Hirschberg corneal light reflex test. It repositions the displaced light reflex to the center of the deviating eye with prisms. Prisms held in front of the deviating eye are preferred in patients with incomitant and paralytic deviations (see Chapter 5). The test is often performed at near in the primary position but may be done for other positions as well if the patient is cooperative. In sensory deviations with poor acuity and fixation in the involved eye, an intermediate testing (conversation) distance is used to estimate the target.

---

**Figure 1-12.** Response to 4 prism diopters base-out test for foveal suppression. A. When prisms are placed over the left eye, dextroversion occurs during refixation of this eye, indicating absence of foveal suppression. If a suppression scotoma is present, no movement will be observed from either eye. B. A subsequent slow fusional adduction movement of the right eye indicates absence of foveal suppression in the right eye. C. If the right eye stays abducted, the absence of adduction movement indicates foveal suppression in the right eye or (D) weak fusion. In the latter case, patients experience diplopia until refixation occurs spontaneously. (From von Noorden GK: Binocular Vision and Ocular Motility: Theory and Management of Strabismus. St. Louis, Mosby-Year Book, 1996. Reprinted with permission.)
angle for surgery. The Krimsky method underestimates the angle of strabismus when compared with the alternate prism cover test in deviations larger than 15 PD.1

**Cover Tests**

The motility evaluation involves measuring the deviation in gazes other than primary position at distance and near. If a horizontal deviation is present, distance measurements in side gazes and upgaze and downgaze are made to uncover lateral incomitance or A- or V-pattern strabismus. If vertical deviations are seen (including dissociated vertical deviations), right and left head tilt measurements are obtained (Fig. 1–14). If oblique dysfunction is suspected or present, deviation should also be measured in the oblique fields. It may be difficult to obtain accurate quantitative measurements in the oblique fields; qualitative assessment will suffice.

Cover tests should be done by eliminating all accommodative influence. This is accomplished by using not only an accommodative target with sufficient discernible detail but also the full hyperopic correction and a 20-ft (6-m) working distance. This distance permits only one-eighth diopter of accommodation, which may be considered negligible for clinical purposes. In addition, some patients (such as those with intermittent exotropia and tenacious proximal fusion) may require prolonged occlusion to suspend all convergence influence. At near, primary and downgaze deviations are always checked. If bifocals are worn, the near deviation is measured through the reading segment.

**Cover/Uncover Test.** The cover/uncover test is the first of the cover tests to be performed. If one suspects a deviating eye, the opposite eye is covered while the patient fixes on an accommodative target. Movement in the uncovered eye is consistent with a tropia. If no movement is observed, the cover is removed to allow the patient to view the target.
binocularly. The cover then is moved to the other eye. If no movement occurs, there is no tropia. Any movement seen in the eye under the cover is consistent with a phoria.

The test may also be used to determine subtle fixation preferences, especially in patients with intermittent exotropia who have equal visual acuity. The patient is allowed to remain binocular until he or she is no longer tropic. One eye is covered (e.g., the right eye). Under the cover, this eye assumes an exotropic position. While maintaining fixation on an appropriate target, the cover is removed from the right eye. If this eye picks up fixation instantaneously, and the left eye becomes exotropic momentarily before binocular fusion and straight eyes are achieved, the patient has a right eye preference and left intermittent exotropia. If the right eye picks up fixation without movement of the left eye, this represents either true alternating exotropia or right intermittent exotropia. This can be verified by placing the cover over the left eye after binocularity is reestablished and proceeding with a similar evaluation.

Simultaneous Prism Cover Test (SPCT). The SPCT measures the angle under normal binocular conditions and is important in accommodative esotropia and monofixation syndrome (less than 10 PD). In these patients, a larger angle may be uncovered by the alternate prism cover test. The SPCT should be performed before the alternate cover prism test.

The tropic eye is identified on the basis of observations on the cover/uncover test. Prisms in the amount estimated to neutralize the manifest deviation are selected. As one covers the fixing eye with an occluder, the prisms are placed in front of the deviated eye at the same time. If no movement is observed, the prisms chosen correspond to the manifest tropia. Alternate prism cover testing can proceed to uncover the total deviation.

Prism Under Cover Test (PUCT). Neutralization of a dissociated deviation, often vertical, is performed using the PUCT (Fig. 1–15). The dissociated vertical deviation (DVD) is neutralized by placing base-down prisms over the nonfixing eye under the occluder. As the patient views an accommodative target, the occluder is transferred to the fixing eye. Absence of movement in the vertical direction defines the endpoint of neutralization. In some patients, this endpoint cannot be identified. Neutralization may be considered when overshoot and refixation movements are equal. The same procedure is used for measuring DVD in the fixing eye. Patients must be forced to fix with the nonpreferred eye after occlusion of the dominant eye.

Alternate Prism Cover Test (APCT). The APCT uncovers the total (manifest and latent) deviation. Patients are not allowed to establish binocularity during alternate covering. Results of this test determine the amount of surgery required for most patients with strabismus. Neutralization occurs just before the deviation reverses. When it is difficult to judge neutralization, as in nystagmus or with large refixation movements, prisms that elicit an equal amount of overshoot and redress are considered the endpoint.

Factors Affecting Motor Measurements

Extrafoveal Fixation

Extrafoveal fixation can affect the measured deviation in strabismus. It occurs when there is a foveal pathologic process. An apparent strabismus may be caused by extrafoveal fixation, but cover testing reveals an absent or different deviation. Fixation may be assessed by presenting a visuscopie target with a direct ophthalmoscope (see also Chapter 2).

Angle Kappa

Angle kappa is the angle formed by the patient’s visual and pupillary axes. The visual axis, or line of sight, corresponds to the line connecting the object of fixation to the fovea. The pupillary axis is formed by a line that passes perpendicular to the center of the cornea. Angle kappa is important clinically because it may give the appearance of strabismus. Mean positive angle kappa ranges from 1.4 to 2.8 degrees, with emmetropes and hyperopes tending to have slightly larger angles kappa than myopes. A positive angle kappa occurs when the corneal light reflex is displaced nasally as a patient fixes monocularly on a light at near, giving the appearance of exotropia. A negative angle kappa occurs when the light reflex is displaced temporally, giving an esotropic appearance; this is common in high myopia. A positive angle kappa is more common. Most angles kappa are physiologic, but some, especially the larger angles, may be caused by retinal traction, as occurs in retinopathy of prematurity with temporal dragging of the macula and in Toxocara scarring with retinal folds and macular heterotopia. The vertical equivalent of an angle kappa secondary to macular heterotopia is usually the result of scar tissue formation in the posterior pole.

Primary and Secondary Deviations

A secondary deviation, usually seen in paralytic and restrictive strabismus, occurs when a patient fixes with the paralytic or restricted eye. It is larger than the primary deviation because of the increased innervation needed for a paretic or restricted eye to fix on a target. By Hering’s law, more innervation goes to the contralateral yoke muscle,
resulting in a larger deviation. This is significant when the patient has the potential to switch fixation postoperatively to the involved eye, allowing the secondary deviation to become manifest and causing symptoms such as asthenopia and diplopia.

Primary deviations are obtained with prisms over the nonfixing eye, and secondary deviations are obtained with prisms over the fixing eye. In many situations the primary deviation is the target surgical angle. However, in patients fixing with the involved eye who will not switch fixation to the contralateral eye (usually because of poorer vision), the target angle for surgery should be the secondary deviation.

**Antepodean Strabismus**

Antepodean strabismus may lead the examiner to believe that a measurement error has occurred. It is present when the patient has esotropia while fixing with one eye and exotropia when fixing with the other eye. This occurs with anisometropia and an improperly corrected refractive error. Some cases are believed to be due to unequal accommodation.

**“Latent” Strabismus**

At times a clear history is present but the deviation cannot be confirmed on examination. Although uncommon, this may occur in children with esotropia. If cover testing after adequate cycloplegia uncovers an esotropia, it is likely that the patient will demonstrate esotropia in the future. The OKN drum can be used before pharmacologic cycloplegia to uncover an esotropic deviation (Kushner BJ, personal communication, 1997). The child fixes the vertically rotating stripes of the OKN drum. When adequate fixation is present (normal OKN response), the rotating drum is brought closer to the patient. An esotropia in either eye suggests a tendency toward esotropia, which may become manifest on subsequent examination.

**Other Tests**

**Red Glass Test.** The red glass test is a useful subjective test for diplopia and for identifying subtle changes in deviation. Patients suspected of having masked bilateral superior oblique palsy may exhibit reversal of hypertropia in the oblique fields of gaze when the deviation is small. The test also detects secondary deviation in patients fixing with the paretic or restricted eye. While looking at a fixation light, the patient is asked whether separation is greater with red lenses over one eye compared with the other. If a disparity exists, a secondary deviation is present. This can be confirmed by measuring the deviation using the alternate prism cover test.

**Maddox Rods.** The Maddox rod is a lens system consisting of a series of parallel cylinders. The lens converts a point source of light into a single streak oriented perpendicular (90 degrees) to the direction of the Maddox cylinders. A single Maddox rod, usually red, is used to detect horizontal or vertical deviations. The Maddox rod is dissociating and cannot distinguish between phorias and tropias.

To diagnose a horizontal deviation, by convention the red Maddox rod running horizontally is presented before the right eye. The patient fixes on a point source of light and sees a single vertical red light in the right eye and a white spot of light in the left eye. The red line bisects the white light in orthotropia. If the red line is to the right of the white light (uncrossed), an esodeviation is present; if it is to the left of the light (crossed), an exodeviation exists. A similar procedure is used for vertical deviations by orienting the Maddox rod cylinders vertically.

Double Maddox rods for testing torsion are discussed in Chapter 4.

**Three-Step Test.** The three-step test for diagnosing isolated cyclovertical palsy was outlined by Parks’ based on the Bielschowsky head tilt phenomenon. The head tilt test utilizes the utricular reflex to help isolate the palsied, cyclovertically acting muscle. Three basic questions require answers in performing this test:

1. **Which eye manifests the hyperdeviation?** A hyperdeviation caused by a paretic muscle occurs when one of the two muscles responsible for depression—the superior oblique (SO) or the inferior rectus (IR)—is weak. One should be wary of a patient who manifests a hypodeviation, because this may be secondary to a paretic cyclovertical muscle in the opposite eye if fixation with the paretic eye is preferred. Identifying the eye with the hyperdeviation narrows the number of possible weak cyclovertical muscles from eight to four. For example, in right hypertropia (RHT) (or left hypotropia), weakness of the depressors of the right eye—the right SO and right IR—or the elevators of the left eye—the left superior rectus (SR) and left inferior oblique (IO)—can explain the observed deviation. Unopposed innervation of the direct antagonist of the depressors of the right eye—the right SR and right IO—leads to right hyperdeviation, whereas unopposed action of the depressors of the left eye—the left SO and left IR—leads to left hypotropia.

2. **Does hyperdeviation increase on lateral gaze?** Assessing the amount of hyperdeviation in lateral gaze allows one to determine whether an oblique or vertical rectus muscle is paretic. Anatomically, the oblique muscles have their greatest vertical action in adduction, whereas the vertical rectus muscles have their greatest vertical action in abduction (see also section on ocular rotations). Therefore, in the example just presented, of the four possible muscles involved from step 1, RHT that increases on right gaze could be due to a weak right IR or left IO; RHT that increases on left gaze could be due to a weak right SO or right SR.

3. **Is the hyperdeviation larger on right or left head tilt?** Normally, head tilting stimulates intorsion of one eye and extorsion of the other eye, allowing binocularity to be maintained. Right head tilt stimulates intorsion of the right eye (SR and SO) and extorsion of the left eye (IR and IO). Left head tilt does the opposite. If RHT increases on right tilt, the weak muscles could be the right SR or SO or the left IO or IR. If RHT increases on left tilt, the weak muscles could be the right IR or IO or the left SO or SR. The cyclovertical muscle palsy is identified by the one muscle identified by all three maneuvers. Note that, by the time one observes the effect of head tilting on the size of the vertical deviation, the choice of which cyclovertically acting muscle is paretic will be narrowed to an oblique muscle in one eye and a vertical rectus muscle in the
other. For example, if RHT (four weak muscles are the right SO and IR and the left IO and SR) increases on left gaze (two weak muscles are right SO and left SR) and right head tilt, the isolated cyclovertical muscle that is palsied is the right SO (see also Chapters 15 and 16).

In some clinical situations, three-step testing may lead to misdiagnosis of the cause of a vertical deviation. These causes include contracture of a vertical rectus muscle (e.g., thyroid ophthalmopathy), paresis of more than one vertical rectus muscle, DVD, previous vertical muscle surgery, skew deviation, myasthenia gravis, and small nonparalytic vertical deviations associated with horizontal strabismus.

To verify that one is not dealing with isolated cyclovertical muscle palsy, the following should be done:

1. Check for secondary deviation if paralytic or restrictive strabismus is suggested by the history.
2. Pay attention to ductions and versions, especially in the oblique fields of gaze. If three-step testing suggests isolated cyclovertical muscle palsy but versions demonstrate no underaction of this muscle or overaction of its antagonist, one should suspect the accuracy of the original diagnosis.
3. Perform alternate prism cover testing in the oblique fields of gaze to detect the reversal of hyperdeviation seen in masked bilateral SO palsy.
4. Determine if the vertical deviation is dissociated.
5. Elicit a history of previous surgery on a vertically acting muscle, periorbital trauma (orbital fracture), or thyroid disease.
6. Maintain a high index of suspicion for a history or findings suggesting skew deviation or myasthenia gravis. Patients with diabetes mellitus or hypertension can suffer small, otherwise clinically silent strokes that may result in skew deviation. Intermittent symptoms that worsen on activity and fatigue suggest myasthenia gravis.

**OCULAR ROTATIONS**

Observing a patient’s ocular rotations is a crucial part of the strabismic evaluation. Careful assessment of ductions and versions helps detect the presence of dysfunction in the horizontal, vertical, or oblique fields of gaze. Ductions are defined by the observation of monocular eye movement with the opposite eye occluded. Versions are binocular eye movements. Normal versions preclude the need to check ductions (Fig. 1–16). However, if versions are abnormal, monocular duction testing should be performed, especially in the field of limited ocular rotations.

When observing versions and ductions, six cardinal fields of gaze are usually assessed. These are gaze right, right and up, right and down, gaze left, left and up, and left and down. Assessment of ocular rotations in these fields takes advantage of the anatomic site at which the cyclovertically acting muscles have their purest vertical action. The torsional functions of these muscles are not evaluated in this schema. The vertical rectus muscles have their greatest (pure) vertical action when the eye is abducted 23 degrees from the midline, whereas the oblique muscles have their greatest vertical action when the eye is adducted 51 degrees from the midline. Some strabismologists prefer evaluation in nine gaze fields, as shown in Figure 1–16.

Version testing also helps to confirm or alert one to the presence of incomitant strabismus. For example, if an A pattern is apparent on cover testing, versions may reveal bilateral SO overaction. An alternate diagnosis should be sought if cover testing suggests an isolated SO palsy but versions demonstrate no SO underaction and/or IO overaction.

Duction and version tests are governed by Hering’s and Sherrington’s laws. Hering’s law states that whenever an impulse for eye movement is initiated, the yoke muscle in the contralateral eye receives the same amount of innervation to contract. Sherrington’s law of reciprocal innervation postulates that whenever a muscle receives an impulse to contract, its antagonist will receive an equal amount of inhibitory input to relax.

Underaction of a muscle on version testing may be secondary to an underlying deviation, an “inhibitional palsy,” true muscle paresis, or mechanical restriction. For example, if there is a deviation such as a large left exotropia, an

---

**Figure 1–16. Normal version test.** Nine-picture composite of normal version testing. The extraocular muscles involved in each gaze are identified. Strabismologists who talk about six cardinal gaze positions refer to (1) right, (2) left, (3) right and up, (4) right and down, (5) left and up, and (6) left and down. If any limitation in ocular rotation is observed, monocular duction testing should be performed. The same muscles involved in version testing are used in performing ductions. Duction testing is performed by covering one eye and allowing the patient to fixate an object in the different gaze positions.
apparent underaction of the right medial rectus muscle may be observed on left gaze. If true underaction of the right medial rectus muscle is present, the underaction should also be observed on ductions (which would not be expected in this case).

An “inhibitional palsy” results when equal innervation is sent to an agonist to contract and to an antagonist to relax. With inhibitional palsy of the contralateral antagonist in a patient with right SO palsy, less innervation is required to the antagonist of the right SO—the right IO—when the eyes look to the left and up (Sherrington’s law). This decreased innervation of the right IO results in decreased innervation to its yoke muscle in the left eye—the left SR, resulting in apparent underaction of this muscle when looking to the left and up (Hering’s law). When the right eye is covered, ductions will reveal normal functioning of the left SR. In contrast, underaction noted on both version and ductions suggests a true muscle paresis or mechanical restriction. These two conditions can then be differentiated by forcedduction and force generation testing.

Figures 1–17 and 1–18 demonstrate a grading scheme for IO and SO overaction and underaction. In the presence of horizontal strabismus the appearance of oblique dysfunction may differ. In esotropia, the vertical action of the oblique muscles is more evident whereas the abducting component may become more apparent in exodeviation (Fig. 1–19). If the examiner is not aware of a prominent abducting component, an oblique muscle dysfunction may be missed (see also Chapter 14).

Other abnormal eye movements that may be confused with oblique muscle dysfunction include upshoots and downshoots associated with the Duane and Brown syndromes. In Duane syndrome these do not represent primary oblique dysfunction but rather reflect slippage of the globe under the horizontal rectus muscle, usually the lateral rectus. In Brown syndrome the typical downshoot is secondary to a restrictive phenomenon or leash effect. These abnormal movements should be distinguished from primary overaction of the oblique muscles.

Pseudo–IO overaction has been described in three clinical situations: (1) after SR recession for dissociated vertical deviation, (2) in some Y and V syndromes, and (3) after anterior transposition of the IO. Pseudo–IO overaction after SR recession for DVD is due to fixation duress of the contralateral IO. For example, in looking to the right and up, a weakened right SR requires more innervation to look in this gaze field. By Hering’s law, this increased innervation also goes to the left IO, giving the appearance of left IO overaction.

In some cases of Y and V patterns, co-contraction of the lateral rectus muscles may be responsible for pseudo-overaction of the IO. These patients differ from those with true IO overaction, because hypertropia on direct side gaze and objective fundus torsion are not observed. IO weakening procedures do not resolve the apparent IO overaction or the pattern strabismus, but recession and elevation of the lateral rectus muscles (for exotropia) are effective.

Finally, an abnormal motility pattern simulating IO overaction may be seen in patients who have had previous anteriorization of the IO muscle. Excessive anteriorization (more than 2 mm anterior to the IR insertion) creates an increased depressing vector in the operated eye. This results in increased innervation to elevate the eye in abduction. By Hering’s law, this innervation also goes to the yoke IO in the opposite eye, resulting in apparent overaction. The situation is remedied by “weakening” the anteriorization by moving it more posteriorly (behind the IR insertion), recessing the IO, or performing a myectomy procedure.
CYCLOPLEGIC REFRACTION

The cycloplegic refraction is a cornerstone of strabismus evaluation. It enables one to uncover the full amount of hyperopia in patients suspected of having an accommodative component to their strabismus. In myopes, it prevents over-correction, permitting clear vision without using inappropriately excessive accommodation. If the patient has an ocular deviation, maximizing vision by dispensing the proper cycloplegic refraction provides optimal control of the deviation. Prescribing the appropriate cycloplegic refraction when indicated supersedes the need to balance the prescription. An improper or inadequate cycloplegic refraction can lead to misdiagnosis and inappropriate treatment.

There remains an ongoing debate as to what constitutes an appropriate regimen of cycloplegic agents. General guidelines, however, do exist. Any infant younger than 1 year of age should not receive “full-strength” drops because of the likelihood of systemic effects. For infants, a commercially available combination of cyclopentolate 0.2% and phenylephrine 1.0% (Cyclomydril), given in two doses 5 minutes apart, is preferred. This is usually effective even in patients with dark irides if the agents are instilled properly and if at least 40 minutes pass before retinoscopy is performed. If this is inadequate, a single drop of cyclopentolate 1% is added. Atropine 1%, cyclopentolate 1% and 2%, and the mydriatic phenylephrine 2.5% all should be used with caution in infants.

In children older than 1 year, cyclopentolate 1% is instilled in two doses 5 minutes apart. Using a topical anesthetic before instilling the cycloplegic agent augments comfort and efficacy. In esotropic children with more than +2.00 D of hyperopia after cyclopentolate, atropine refraction may uncover more than +1.00 D of additional hyperopia. Atropine 1% is instilled twice a day for 2 days before refraction. Parents should be informed of possible side effects from cyclopentolate and atropine.

Deciding whether the full cycloplegic refraction should be dispensed may be difficult. Myopic or astigmatic patients are given the full refraction except when the ophthalmologist chooses to over-minus an intermittent exotrope. A dispensing algorithm for hyperopia is outlined in Table 1–3. This table is only a guideline; each patient should be treated individually.

Dispensing decisions are difficult when accommodative esotropia is present in an older child. The full hyperopic correction may not be accepted because of visual blurring. In general, children younger than age 4 years with esotropia will accept, and should be given, the full hyperopic correction. Children older than age 4 years with esotropia may have a trial of the full cycloplegic correction but should be warned of visual blurring and the possible need to reduce the prescription. It is useful to direct these children to an optical shop that will replace the lenses free of charge if the prescription changes within a certain period of time (usually 3 to 6 months). If a child does not wear his or her hyperopic spectacle correction, atropine or homatropine drops may serve to increase compliance. Within 2 to 3 weeks the cycloplegic agent is slowly weaned. If the full hyperopic refraction is not tolerated, it is reduced by 1.00 to 1.50 D.

An interesting case is that of the hyperopic intermittent exotrope. Typically, providing a hyperopic correction worsens an exodeviation. Some intermittent exotropes may gain better control if the hyperopic correction improves visual

Table 1–3. Guidelines for Dispensing Refraction in Hyperopic Children

<table>
<thead>
<tr>
<th>Age (years)</th>
<th>Deviation</th>
<th>Dispense</th>
</tr>
</thead>
<tbody>
<tr>
<td>Younger than 1</td>
<td>Esotropia</td>
<td>Full hyperopic correction if ≥1.50 D</td>
</tr>
<tr>
<td>1–5</td>
<td>Esotropia</td>
<td>Full hyperopic correction if ≥1.00 D*</td>
</tr>
<tr>
<td>Older than 5</td>
<td>Esotropia</td>
<td>Full hyperopic correction if ≥1.00 D*</td>
</tr>
<tr>
<td>Younger than 5</td>
<td>No deviation</td>
<td>Full hyperopic correction if ≥3.50 D</td>
</tr>
<tr>
<td>Older than 5</td>
<td>No deviation</td>
<td>If cycloplegic refraction ≥3.50 D, dispense hyperopic correction 1.00–1.50 D less than this.</td>
</tr>
</tbody>
</table>

*If hyperopia is less than 1.00 D but accommodative esotropia is suggested, an atropine refraction should be performed.
comfort and acuity.26 Such patients usually have more than 2.00 D of hyperopia (see also Chapters 5 and 12).

Residual Esotropia

A child may manifest residual esotropia while wearing hyperopic spectacles. The effect of additional plus in reducing the deviation and its effect on visual acuity should be checked in older children before repeating the cycloplegic refraction. It is not sensible to correct additional hyperopia that is found on cyclopentolate or atropine refraction if this blurs vision. The use of atropine to help in adjusting to glasses is most effective in young children who do not accept the initial spectacle prescription. It is less useful in older children in whom additional hyperopic correction and/or chronic cycloplegia cause visual blurring. In a similar manner, one can attempt to reduce the hyperopic correction in a well-controlled accommodative esotrope by placing concave (minus) lenses in Janelli clips. If control of esotropia is not compromised and fusion status is unaffected, the hyperopic correction can be reduced.

Bifocal Dispensing

Hyperopic, esotropic children are provided with the full hyperopic correction measured at the initial visit. If the patient has residual esotropia on follow-up, the refraction is rechecked using atropine. Bifocals are indicated if orthotropia or monofixational esotropia is present at distance. Conceptually, bifocals are indicated if fusion can be demonstrated at distance but residual larger-angle esotropia is present at near. The level of stereopsis or peripheral fusion should be documented through the bifocals, because later decisions to continue using them rest on their ability to maintain or improve binocularity.

When dispensing bifocals in children, the type and location of the bifocal must be stated clearly. A D-segment or executive type that extends the full length of the optic should be used. The height at which bifocals are set depends on the patient's age, but the ultimate goal is for the child to spontaneously use the bifocal for near. In general, bifocals bisect the pupil in younger children and may be set at the level of the lower limbus or lower lid in older (school-age) children. The slab-off technique of grinding bifocals may be used when there is an associated small vertical deviation (e.g., induced prisms in anisometropia).

Fundus Examination

A complete fundus examination, including assessment of the periphery, should be performed on the initial visit. The size and color of the optic nerve head should be noted in seeking possible organic causes of decreased acuity. If the patient has a history of strabismus surgery, the retinal areas underlying the previously operated muscles are examined for evidence of perforation. Macular heterotropia may arise from cicatricial retinopathy of prematurity or more subtle lesions such as epiretinal membranes. If subtle retinal pathology is suspected, fundus contact lens examination can be helpful in cooperative individuals. If motility examination suggests oblique muscle dysfunction, the fundus is assessed for the presence of torsion.

ANOMALOUS HEAD POSTURE

Anomalous head posture may not be immediately obvious to the examiner and requires a systematic evaluation. Old photographs demonstrating the abnormality should be reviewed. The visual conditions under which anomalous head posture occurs should be determined and re-created in the clinic. Anomalous head posture may occur only when performing a visual task. Patients are observed while binocularly reading the threshold Snellen optotype at distance and near. In young children, appropriate targets such as a toy, cartoon video, or picture may be used for fixation.

The characteristics of anomalous head posture are noted at distance and near: face turn, chin elevation or depression, head tilt, or a combination of these. In cooperative patients, a hand-held orthopedic goniometer may be used to measure the amount of anomalous head posture. To measure face turn, one arm of the goniometer is directed toward the fixation target while the other is aligned with the sagittal axis of the patient's head. For chin elevation and depression, the goniometer is positioned with one arm perpendicular to the fixation target and the other directed parallel to the anteroposterior axis of the face. For head tilts, one arm is directed perpendicular to the floor and the other is held parallel to the axis of the face (Fig. 1–20). The amount of anomalous head posture in degrees can be read directly from the goniometer. Goniometer measurements are adequate for clinical purposes. If quantitative measurements cannot be made, an estimate should be recorded.

Causes of anomalous head posture can be divided into two major categories: ocular and musculoskeletal or neurologic (Tables 1–4 and 1–5).29, 47 Anomalous head posture should not be attributed to a nonocular cause until a thorough search.
for an ocular cause is completed. Most patients develop an anomalous head posture to gain some level of single binocular vision or fusion. Thus, diplopia may be reported in primary position but not in the preferred head position. Alternate prism cover testing will reveal a manifest deviation in gaze positions with diplopia but no deviation or latent deviation with the anomalous head posture if underlying strabismus is the cause. Fusion may be demonstrated in the preferred head posture but not in other head positions. Neutralizing the deviation in primary position with prisms (prisms in a trial frame or Janelle clips, a prism bar, or Fresnel prisms) should improve the head posture unless there is residual torsion that could not be corrected. Patching one eye should improve anomalous head posture due to strabismus but not if it is musculoskeletal or neurologic in origin.

Some patients develop anomalous head posture to improve visual acuity; this is often seen in patients with nystagmus and individuals having astigmatic refractive errors. In patients with nystagmus, vision may be checked in the forced primary position and the preferred head posture to confirm the clinical impression. Alternatively, one may place a 20 PD prism with its apex oriented in the direction of the face turn or chin position. Patients gaining better vision will exhibit improvement or resolution of the anomalous head posture. Patients with astigmatism who use a head tilt may have an improved head posture once the appropriate spectacle correction is worn.

Paradoxically, some anomalous head postures result in a larger manifest deviation (e.g., acquired SO palsy). This permits greater separation between two images and makes it easier to ignore the second image.

### Table 1–4. Ocular Causes of Anomalous Head Postures

<table>
<thead>
<tr>
<th>Strabismus</th>
</tr>
</thead>
<tbody>
<tr>
<td>Muscle paresis</td>
</tr>
<tr>
<td>Superior oblique palsy</td>
</tr>
<tr>
<td>Sixth nerve palsy</td>
</tr>
<tr>
<td>Third nerve palsy</td>
</tr>
<tr>
<td>Double elevator palsy</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Restrictive strabismus</th>
</tr>
</thead>
<tbody>
<tr>
<td>Thyroid related ophthalmopathy</td>
</tr>
<tr>
<td>Brown syndrome</td>
</tr>
<tr>
<td>Orbital wall fractures</td>
</tr>
<tr>
<td>Congenital fibrosis</td>
</tr>
<tr>
<td>Incomitant strabismus</td>
</tr>
<tr>
<td>Duane syndrome</td>
</tr>
<tr>
<td>A or V pattern</td>
</tr>
<tr>
<td>Torsional incomitance</td>
</tr>
<tr>
<td>Dissociated vertical deviation</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Nystagmus</th>
</tr>
</thead>
<tbody>
<tr>
<td>Idiopathic congenital nystagmus (congenital motor nystagmus)</td>
</tr>
<tr>
<td>Congenital nystagmus with congenitally blind eye</td>
</tr>
<tr>
<td>Congenital nystagmus with retinopathy of prematurity</td>
</tr>
<tr>
<td>Nystagmus compensation syndrome</td>
</tr>
<tr>
<td>Acquired nystagmus</td>
</tr>
<tr>
<td>Spasmus nutans</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Miscellaneous</th>
</tr>
</thead>
<tbody>
<tr>
<td>Congenital esotropia with ocular posture of Lang</td>
</tr>
<tr>
<td>Foveal fusion</td>
</tr>
<tr>
<td>Cosmetic</td>
</tr>
<tr>
<td>Ocular motor apraxia</td>
</tr>
<tr>
<td>Oblique astigmatism</td>
</tr>
<tr>
<td>Parafoveal/extrafoveal fixation to avoid foveal lesions</td>
</tr>
<tr>
<td>Transient head posturing with initiation of amblyopia therapy</td>
</tr>
</tbody>
</table>

### Table 1–5. Nonocular Causes of Anomalous Head Posture

<table>
<thead>
<tr>
<th>Congenital Muscular and/or Skeletal Torticollis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Associated with sternocleidomastoid mass or pseudotumor</td>
</tr>
<tr>
<td>Associated with bony abnormality</td>
</tr>
<tr>
<td>Klippel-Feil syndrome</td>
</tr>
<tr>
<td>Springel deformity</td>
</tr>
<tr>
<td>Bony abnormality of the cervical spine</td>
</tr>
<tr>
<td>Occipitocervical synostosis</td>
</tr>
<tr>
<td>Basilar impression</td>
</tr>
</tbody>
</table>

| Congenital shortening of the sternocleidomastoid |

<table>
<thead>
<tr>
<th>Acquired Nontraumatic Torticollis Secondary to Inflammation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sinusitis</td>
</tr>
<tr>
<td>Mastoiditis</td>
</tr>
<tr>
<td>Cervical adenitis</td>
</tr>
<tr>
<td>Retropharyngeal abscess</td>
</tr>
<tr>
<td>Myositis</td>
</tr>
<tr>
<td>Subluxation of atlantoaxial joint (Grisel syndrome)</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Neurologic Torticollis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Arnold-Chiari malformation</td>
</tr>
<tr>
<td>Cerebral palsy</td>
</tr>
<tr>
<td>Meningomyelocele</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Neoplasms Involving Cervical Cord or Cervical Spinal Vertebrae</th>
</tr>
</thead>
</table>

<table>
<thead>
<tr>
<th>Traumatic Torticollis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sandifer syndrome</td>
</tr>
<tr>
<td>Benign paroxysmal torticollis of infancy</td>
</tr>
<tr>
<td>Idiopathic</td>
</tr>
</tbody>
</table>

### REFERENCES


Good motor alignment (straight eyes) and improvement in or attainment of normal binocular vision are the goals of strabismus surgery. These are the benchmarks against which the success or failure of a given procedure are measured. In general, it is possible to have acceptable motor alignment with no functional binocular vision, but it is highly unlikely that normal binocular vision can coexist with poor motor alignment. Therefore, every means must be pursued to secure a functional correction.

A patient’s sensory status is considered crucial to the long-term stability of a successful surgical outcome. A thorough sensory evaluation defines a patient’s sensory (fusion) capabilities and adds valuable information for use in making an informed surgical decision. Sensory fusion (at least peripheral fusion) serves as the “glue” to maintain alignment. A surgically aligned eye with no meaningful vision relies exclusively on the balanced mechanical forces of the eye muscles to maintain ocular alignment. The incidence of postsurgical drift is significantly higher after surgery on eyes with very poor vision than on eyes with good vision. On the other hand, good sensory fusion with excellent stereopsis, present preoperatively, drastically lowers the risk of postsurgical drift. A continuum exists between these two extreme situations.

**The Normal Sensory System**

Several components of the normal sensory system function together to produce clear, comfortable binocular vision.

**VISUAL ACUITY**

Normal visual acuity is generally considered to be 20/20 (6/6) or better in each eye. The binocular sensory system functions best if acuity is 20/20 or better in each eye, but it can still function even with lower acuity. Normal acuity also implies normal monocular fixation in each eye. Because vision rapidly declines with any eccentricity of fixation away from the fovea (Fig. 2–1), normal acuity requires foveal fixation under monocular conditions.

**NORMAL RETINAL CORRESPONDENCE**

The organization of visual space normally requires input from both eyes. Points on each retina combine to determine the binocular visual direction of our percepts. Normal retinal correspondence (NRC) is the normal state in which the visual direction of each fovea is the same. The term retinal correspondence is somewhat of a misnomer because the relationship between the visual directions associated with the two foveas is determined at the level of the cerebral cortex, not the retina.

**FUSION: CENTRAL**

Bifoveal fixation is fundamental to normal sensory fusion. Worth divided fusion into three broad categories using a

![Graph showing how vision rapidly decreases as fixation moves away from the fovea. (Modified from Weymouth F, Hines D, Lawrence J, et al: Visual acuity within the area centralis and its relation to eye movements and fixation. Am J Ophthalmol 1928;11:947.)](image)
hierarchy of fusional abilities. **First-degree fusion**, also called *simultaneous perception*, requires that dissimilar targets be presented to each eye and be perceived at the same time in the same visual direction (Fig. 2–2, top row). **Second-degree fusion**, also called *flat fusion*, requires similar targets with dissimilar components specific to each eye. These monocular suppression checks—the dissimilar components—are used to determine if each eye contributes to the binocular percept (see Fig. 2–2, middle row). **Third-degree fusion**, or *stereopsis*, uses the same targets as second-degree fusion except that disparity is introduced (Fig. 2–2, bottom row). Some portions of one eye’s target are laterally displaced with respect to a similar component in the other eye. This lateral displacement (disparity) is converted into depth (three-dimensional perception or stereopsis) by the brain. High-grade stereopsis is one of the highest functions and is part of the definition of normal binocular vision. For clinical purposes, stereopsis equal to or better than 40 seconds of

---

**Figure 2-2.** Degrees of fusion. Top row, First-degree fusion or simultaneous perception is tested by presenting dissimilar targets to each eye and should be perceived at the same time in the same visual direction. Middle row, Second-degree fusion or flat fusion is tested by presenting similar targets with dissimilar components (monocular suppression checks) specific to each eye. Bottom row, Third-degree fusion or stereopsis is tested using similar targets with suppression checks (e.g., flowers) but with lateral displacement creating horizontal retinal image disparity (swing).
arc is considered normal. Most other fusion tests evaluate peripheral fusion because of the size of the test target. To test for bifoveal fusion, small targets requiring high degrees of stereopsis must be used.

**FUSION: PERIPHERAL**

Corresponding points in the retinal periphery have a slightly different function than those in the central visual field. Because the retina is organized into large receptive fields, the "grain" of the retina is coarser in its periphery. Consequently, visual acuity and stereopsis are reduced as more peripheral areas of the retina are stimulated. Peripheral fusion, however, plays a very important role in maintaining alignment. There are several clinical situations in which alignment is maintained despite a loss of central vision (e.g., age-related macular degeneration and central suppression), just as strabismus can occur when there is a severe loss of peripheral vision but central vision is unaffected. The monofixation syndrome is an excellent example of the importance of peripheral fusion. Patients have a manifest strabismus of up to 8 to 10 PD with no double vision and low-grade stereopsis. Under binocular conditions there is a central (foveal) suppression scotoma. These patients are usually without symptoms, and peripheral fusion is sufficient to maintain postsurgical ocular alignment.

**Sensory Anomalies**

**SENSORY ANOMALIES OFTEN ASSOCIATED WITH THE IMMATURE VISUAL SYSTEM**

**Amblyopia and Eccentric Fixation**

When patients with normal visual acuity fixate monocularly on a small target of interest, the fovea (the retinal area of highest resolution) is aimed directly at the target. The eye, however, is not perfectly still. It has a 10–20 minutes of arc fixation tremor that constantly moves the image over a small angular region around the fovea. If the normal physiologic fixation tremor is monitored over time, the time-averaged position of the fovea is centered on the fixation target (Fig. 2–3).

One to 2 percent of the population have amblyopia, defined as unilateral loss of vision that is due neither to uncorrected refractive error nor to detectable ocular disease. This is usually accompanied by strabismus and/or anisometropia. Commonly associated with the amblyope's visual deficiency is an inability to maintain steady foveal fixation. The fixation tremor of the amblyopic eye is 2 to 10 times larger than the normal fixation tremor. When amblyopic subjects monocularly fixate a small target of interest, the fovea is not directed steadily at the target and does not move symmetrically around it. The time-averaged position of the fovea, as measured by the Maxwell spot or Haidinger brushes, misses the fixation target. The amount by which it misses the target (on average) is the magnitude of eccentric fixation (EF) (see Fig. 2–3, diamonds).

The most widely accepted theory of the genesis of functional amblyopia stems from the work of Claud Worth. From his examination of about 1000 cases, he developed a sensory theory of amblyopia. Despite a paucity of scientific evidence, amblyopia was believed to develop from a sequence of events that begins with an insult to binocular vision early in life. This usually takes the form of strabismus and/or anisometropia. In either instance, binocular fusion is disrupted and suppression ensues to eliminate the diplopia associated with strabismus, or to eliminate a blurred image associated with anisometropia. Facultative inhibition (Worth's term for suppression under binocular condition), if allowed to persist, eventually becomes obligatory (i.e., demonstrable even when the fixing eye is closed). This inhibition presumably results in reduced sensitivity of the foveal area for resolution under monocular conditions, leading to functional amblyopia.

The early diagnosis and treatment of amblyopia are essential to effective visual rehabilitation. Amblyopia is one of the earliest consequences of a binocular vision insult due to strabismus or anisometropia and should be detected when examining an infant or small child.

**Suppression**

When the visual input from the two eyes is significantly different in visual direction (strabismus) or focus (anisome-
tropia), the fusional mechanism will often have difficulty integrating the two images. To avoid diplopia, suppression can occur, perceptually blocking the input from one eye.

Suppression is a complex subject. It is an active phenomenon that may partly block or, rarely, eliminate the total input from one eye. More often it is regional, eliminating a foveal image (esotropia) or a hemiretina (exotropia). Most authorities believe that suppression is an adaptive mechanism for avoiding diplopia or visual confusion. Depth of suppression is a concept denoting the ease or difficulty of "breaking through" the suppression to force perception of a normally suppressed target. Blinking and/or changing the lighting conditions are examples of techniques used to break through suppression.

Suppression is detrimental to the functional correction of patients with minimal binocular function. It reduces fusional vergence ranges for both sliding and step vergence and also reduces stereo awareness. In selected patients, orthoptics may be useful for stimulating binocular vision presurgically, such as patients with acquired esotropia in whom prism adaptation is being used to fully manifest the total deviation. Antisuppression orthoptics may also be useful preoperatively in cases of exotropia. Patients learn to appreciate pathologic diplopia, making it easier for them to fuse after surgery.

Antisuppression therapy in older patients with infantile esotropia, however, is controversial because of the possibility of causing constant, intractable diplopia. Stereoenhancement techniques involve breaking down central suppression and correcting fixation disparity and may be effective in orthotropic patients with reduced stereopsis.

Suppression as a sensory anomaly of the immature visual system is quite common in infantile and acquired esotropia. It is very selective, and usually takes the form of a D-shaped scotoma (Fig. 2–4A). It most often extends from the optic disc across the nasal retina, stopping in a vertical line at the fovea of the deviating eye. This scotoma is sufficiently large to accept the image of the fixation target, thereby preventing diplopia or visual confusion. The nasal retina beyond the disc and the temporal retina are functionally normal in visual direction and sensitivity.

The suppression seen in exotropia usually encompasses the entire temporal retina of the deviating eye (hemiretinal suppression) (see Fig. 2–4B). Most exotropes do not have diplopia and possess very little control of their deviation. Intermittent exotropia is especially challenging to treat, because suppression limits the benefit from both surgical and nonsurgical forms of treatment. Suppression of the temporal hemiretina keeps a vergence error from being perceived. Antisuppression orthoptics may be effective in exodeviations if they can restore the feedback mechanism (diplopia) that allows the patient to respond to the vergence error and control the eye position.

Anomalous Retinal Correspondence

Anomalous retinal correspondence (ARC) is a binocular condition in which the visual direction associated with the two foveas differs. Classically, ARC has been thought of as an adaptive mechanism used to avoid diplopia after a binocular insult such as strabismus has occurred (sensory theory). Rather than suppressing the image of the normally deviating eye, the cortex appears to have the ability to shift the visual directions associated with each retinal point in the central visual field of the deviating eye. Thus, the visual direction associated with the fovea of the normally fixing eye is the same as the image point in the deviating eye. This type of correspondence is known as harmonious ARC. With it, the strabismus patient has no diplopia and usually manifests low-grade stereopsis.

There are three other types of anomalous correspondence (other than harmonious) that are dependent on the definition of "point X" (Fig. 2–5). Point X is the point in the deviating eye that gives rise to a percept in the same visual direction as the fovea of the fixing eye. If point X coincides with the image point, the correspondence is labeled harmonious. If it lies between the image point and the fovea of the deviating eye, the correspondence is unharmonious. If point X is in the same hemiretina as the image point but farther away from the fovea than the image point, the correspondence is called paradoxical type I. If it lies in the hemiretina opposite to the image point, the correspondence is called paradoxical type II. (A more in-depth discussion is beyond the scope of this chapter. The interested reader is referred to the appropriate reference.)

The motor theory proposes that ARC is not an adaptation to diplopia but rather is related to the type of innervational
Figure 2-5. Types of retinal correspondence based on the position of point X with respect to the fovea. A, Normal: Point X coincides with F. B, Harmonious anomalous retinal correspondence (HARC): Point X coincides with T'. C, Unharmonious ARC (unHARC): Point X lies between F_L and T_L. D, Paradoxical type I: X lies in the same hemiretina as T_L but is farther from the fovea than T_L. E, Paradoxical type II: X lies in the opposite hemiretina as T_L. X indicates the point in the deviated eye that gives rise to a percept in the same visual direction as the fovea of the normally fixing eye (corresponding points, i.e., X and F_L). F_R, fovea of the right eye; F_L, fovea of the left eye; T, target; T', image of the target on the left retina.
pattern associated with the strabismus. The central concept in the motor theory hinges on the existence of "registered" and "nonregistered" eye movements. Registered eye movements (like versions) are associated with changes in egocentric direction, whereas nonregistered eye movements (like vergence) do not result in such changes. Therefore, strabismus caused by eye movements that alter registered innervational patterns would result in anomalous correspondence. Strabismus caused by eye movements that alter nonregistered innervational patterns would result in normal correspondence. Whereas the sensory theory is most consistent with harmonious ARC, the motor theory is equally robust for all types of anomalous correspondence.

ARC is a sensory anomaly often associated with the immature visual system afflicted with strabismus. Approximately half of all esotropes have ARC. ARC should be tested whenever possible, because the information obtained contributes to the functional treatment of strabismus. Preoperative or postoperative treatment of ARC is controversial, but its diagnosis helps the strabismus surgeon to avoid or minimize surgical complications such as postoperative drift. Postsurgical drift in esotropia is usually in the esodirection, as if to reestablish the original angle of anomaly (i.e., so point X and the image point coincide).

Horror Fusion

Horror fusion is a rare clinical finding associated with congenital strabismus, commonly with harmonious ARC. This term literally means "fear of fusion." Patients are unable to simultaneously perceive images presented to the right and left eye in the same place in space.

Horror fusion is often confused with suppression or central fusion disruption syndrome. However, it is diagnosed by a very specific set of responses during a synoptophore evaluation (Fig. 2–6). Neither the right nor the left eye's images ever disappear. The image is swept across the retina of the nonfixing eye with constant velocity. Initially the target appears to move with constant velocity, then speed up, slow to a stop, and then "jump over" the fixation target in the fixing eye before moving away with constant velocity.

This phenomenon is more easily understood when considering the patient with esotropia and harmonious ARC. The horopter is the loci of all points in objective space whose images fall on corresponding retinal points for a fixed amount of convergence. Patients with esotropia and harmonious ARC who exhibit horror fusion have a notch in their horopter. This notch is an irregularity in the shape of the horopter in the central visual field in which it "dips down" between the visual axes (Fig. 2–7). This unusual shape may represent an attempt by the brain to maintain normal correspondence in the central visual field while allowing the periphery to be abnormal. The notch demonstrates a point-to-area correspondence between the two eyes and reliably predicts the responses as the image is swept across the retina of the deviating eye. Because the target never disappears, horror fusion is not should be confused with suppression. It is different from central fusion disruption in that there is no history of closed-head trauma, and none of the speed effects described earlier in this section during synoptophore examination have been reported. Finally, horror fusion is seen only in patients who have had strabismus since early childhood.

SENSORY ANOMALIES OFTEN ASSOCIATED WITH THE MATURE VISUAL SYSTEM

Diplopia and Visual Confusion

Strabismus, when acquired as an adult, is usually accompanied by diplopia and/or visual confusion. Diplopia is
seeing a single object simultaneously in two different visual directions so that a single object of regard is perceived as two (Fig. 2–8A). Visual confusion implies seeing two different objects in the same visual direction (see Fig. 2–8B). Depending on the magnitude of separation of the images, visual confusion may be more bothersome in daily life than diplopia.

Most patients can tell the difference between the “real” and “extra” image and learn to ignore the diplopic image because it usually is off to one side and does not interfere with the real target of interest. In many long-standing cases of strabismus, patients learn to suppress or ignore the diplopic image. With visual confusion, however, the extra image is superimposed on top of or right next to the target of interest, making it difficult to ignore.

**Central Fusion Disruption**

Strabismus that results from significant closed-head trauma may lead to constant diplopia that persists even if the images are aligned on the fovea of each eye, either optically or surgically. Pratt-Johnson and Pratt-Johnson and Tillson reviewed nine cases of acquired strabismus in which the patients had “central disruption of fusional ampli-
tude.” As London and Scott correctly pointed out, of the nine cases reported, there was only one in which the patient had a true loss of fusion; all other patients could fuse but had no fusional amplitudes. Three patients who lost fusion and did not recover it were also reported. The investigators suggested calling the entity “sensory fusion disruption syndrome.” If we consider both groups to be describing the same clinical condition (albeit with subtle differences), and agree that the site of the deficit is probably in the cortex, the word “central” should be incorporated: “central fusion disruption syndrome” (CFDS).

Testing for CFDS requires a haploscopic-type instrument such as a major amblyoscope or synoptophore (see Fig. 2–6). Such an instrument allows correction of the horizontal, vertical, and torsional components of the strabismus. Neutralizing the torsional deviation is essential because many patients with closed-head trauma have associated unilateral or bilateral fourth nerve paresis. (The superior oblique muscle supplied by the fourth cranial nerve is an intorter.) Lack of fusion involving uncorrected torsion may masquerade as CFDS.

In patients with CFDS, fusion of first- or second-degree targets will not occur even when all components of the strabismus deviation have been neutralized. Despite aligning the images on both foveas, the images will be perceived as very close to one another or even overlapping. Patients often report that one of the images is still while the other is in constant motion, “swimming” around the stationary target seen by the other eye (Fig. 2–9). At no time does the patient describe even fleeting fusion. If there is no torsional component to the strabismus, fusion may also be tested in free space by holding neutralizing prisms before the patient’s eyes. The same response of “no fusion” is obtained in CFDS.

If the patient demonstrates an ability to fuse (after neutral-
ization of all components of the strabismic deviation), this is a favorable sign for successful surgical treatment of strabismus. Otherwise, serious thought must be given to the possibility of bothersome postsurgical diplopia, which may be intractable in some cases. London and Scott state that,
Figure 2–9. In patients with central fusion disruption syndrome, images may approximate each other but one remains stationary (solid car), whereas the other images constantly move around the stationary target.

“Recognition of this entity may spare the clinician and patient a disappointing search for [resolution of diplopia].”

If patients with CFDS undergo strabismus surgery and achieve good motor alignment, the diplopic images will then be closer together and may be harder to ignore. This may be more bothersome than when the images were more widely separated. Presurgical counseling about the chance of altered image positions after surgery is critical.

Partial field occlusion may provide some relief from bothersome diplopia in patients with CFDS who are surgically aligned. Total occlusion with a black contact lens may be used, but many patients are bothered by the total loss of visual field. Partial field occlusion has the advantage of obliterating the central diplopic image while allowing the use of peripheral vision from the partially occluded eye (Fig. 2–10).7,28,36

Specific Tests to Identify Sensory Anomalies Often Associated with Strabismus

REFRACTIVE CORRECTION

Before performing a sensory examination it is necessary to be sure that the patient is wearing the most hyperopic (least myopic) prescription, as determined by manifest non-cycloplegic refraction.21 If the child’s age prevents manifest
CLINICAL STRABISMUS MANAGEMENT

noncycloplegic evaluation, cycloplegic retinoscopy should be used. The use of noncycloplegic refraction is important, because the sensory system is exquisitely sensitive to small, uncorrected refractive errors that may not be prescribed in the absence of binocular vision concerns (especially intermittent exotropia). In adults, this procedure tests the function of the sensory system under normal daily conditions. Patients do not usually view the world through fixed dilated pupils and weakened accommodative systems. If the patient is not wearing the appropriate refractive prescription (either contact lenses or glasses), this should be obtained and worn for several weeks before the sensory evaluation.

AMBLYOPIA AND ECCENTRIC FIXATION

After the best corrected visual acuity has been determined, the next step in a sensory evaluation is to detect amblyopia and eccentric fixation (EF) (Fig. 2–11). If acuity is normal in each eye, no further fixation analysis is required. EF is not found in nonamblyopic patients. If amblyopia is present, a fixation analysis is needed to determine the presence or absence of EF. The most effective and readily available instrument is a visuscope or calibrated ophthalmoscope (Fig. 2–12). The rings are usually calibrated in prism diopters (PD). The direction, magnitude, and degree of unsteadiness of fixation should be noted. It is also important to determine if the unsteadiness involves the fovea. For example, a common notation of EF reads: OS: 3 PD of nasal EF with unsteadiness of approximately ± 2 PD, fovea involved.

If the reduced acuity is associated with EF, this equation relates visual acuity to the amount of EF:

\[
\text{MAR} = \frac{EF^a + 1}{a}
\]

where MAR = minimum angle of resolution, the reciprocal of the Snellen fraction. For example, acuity of 20/40 (6/12), MAR = 40/20 or 2. EF indicates the magnitude of EF in prism diopters. The sign convention is plus (+) for nasal and minus (−) for temporal EF.

This formula predicts the visual acuity loss resulting from EF. For example, a patient with 2 PD of nasal EF will have an expected acuity of MAR = 2 + 1 or 3 (60/20), the reciprocal of which in terms of Snellen fraction is 20/60 (6/18). Visual acuity should be about 20/60. This relationship, of course, is not absolute but is simply a guideline. If acuity in the sample case just presented was 20/200 (6/60), one should seek a pathologic cause of reduced acuity in addition to EF. If acuity is better than 20/25 (6/9), an error may have occurred either in determining the acuity (e.g., child cheating during the acuity measurement) or in measuring EF.

EF is a potential contaminant when evaluating the angle of strabismus and in determining the state of retinal correspondence. Not all tests for the angle of deviation and the angle of anomaly are affected by EF, only those requiring monocular fixation during some aspect of testing. Specific examples will be given when each of these topics is discussed below.

STRABISMUS EVALUATION

The unilateral and alternate cover tests are the preferred methods to detect the presence and magnitude of strabismus (see also Chapter 1). The angle of strabismus needs to be determined to know the eye position before performing any sensory tests. This is especially true in any situation in which the angle of deviation changes over time or in conditions such as accommodative esotropia and intermittent exotropia.

The tests chosen to define the presence or absence of strabismus will partially depend on EF. If EF is not present, any of the standard tests for determining the angle of strabismus may be used. If EF is present, the measured angle of deviation must be corrected for EF and the surgical plan may be altered based on these effects. The magnitude of the correction is expressed by the following formula:

\[
H_1 = H_m + EF
\]

where \(H_1\) is the true angle of deviation (in PD), \(H_m\) is the measured angle of deviation (in PD), and EF is the magnitude of EF (in PD). The sign convention is plus (+) for esotropia and nasal EF and minus (−) for esotropia and temporal EF. For example, if a patient has a cover test measurement of 20 PD of esotropia and 5 PD of nasal EF, the true angle of esotropia is 25 PD. This correction should be considered whenever testing requires monocular fixation.
during evaluation, as in the cover test. Hirschberg or (modified) Krimsky light reflex estimates of the angle of strabismus need not be corrected for EF because monocular fixation is not required during testing.

It is important to perform a unilateral cover test (UCT) before the alternate prism cover test (APCT) on each patient. For those patients suspected of having the monofixation syndrome, the UCT should include simultaneous prism cover testing (SPCT). In performing the UCT, the magnitude of the deviation must be estimated by observation or measured by prism neutralization. Any significant difference between the UCT and the APCT is suggestive of ARC.\(^\text{18, 42-45}\) Parks,\(^\text{31, 52}\) in his report on monofixation syndrome, described a subset of patients with small-angle esotropia whose SPCT results were less than 8 PD, but APCT revealed prism neutralization with more than 15 PD base-out prisms. Whereas Parks proposed foveal suppression with peripheral (NRC) fusion, other investigators believed that the findings can be explained by the presence of ARC.\(^\text{18, 42-45}\)

**RETNAL CORRESPONDENCE**

Determining the state of retinal correspondence may provide valuable information for determining the likelihood of obtaining normal binocular fusion after strabismus surgery.\(^\text{19}\) Constant esotropes with ARC have a very poor chance of gaining functional correction. Using orthoptics to eliminate ARC and promote normal fusion may be nonproductive. Intermittent exotropes, on the other hand, have a better chance of securing a functional correction with the help of vergence training that promotes fusion and eliminates ARC.\(^\text{34}\)

Tests for correspondence fall in two categories: direct measures of the angle of anomaly and measurements by calculation.\(^\text{24}\) The Bielschowsky afterimage (AI) test directly measures the *angle of anomaly*, defined as the separation in perceived space between two foveally placed images. To perform the AI test a horizontal line afterimage (with a gap in the middle) is centered on the fovea of the fixing eye under monocular conditions. Shortly thereafter a vertical AI is centered on the fovea of the deviating eye under monocular viewing conditions. With both eyes open, the patient then fixes a target on a calibrated scale. The horizontal AI is centered around the fixation spot while the patient is asked to identify the location of the vertical image (Fig. 2–13). If it intersects the fixation spot to form a perfect cross, the patient has NRC. If it is off to the right or left of fixation, the patient has ARC. The degree of separation of the images is a direct measure of the angle of anomaly (A).

Notice that placing afterimages on the fovea of each eye requires monocular fixation. Thus, if EF is present, a correction factor must be introduced. The same formula applied to the angle of deviation also applies to the angle of anomaly:

\[
A_i = A_m + EF
\]

where \(A_i\) is the true angle of anomaly (in PD), \(A_m\) the
measured angle of anomaly (in PD), and EF the amount of EF (in PD).

To determine the value for the angle of anomaly by calculation, two other angles must be measured: the objective angle of deviation (H) and the subjective angle of directionization (S). Angle A may be calculated using the formula:

\[ H = S + A \]

To be valid, H and S must be measured under the same conditions and distance (Fig. 2–14). For example, if the major amblyoscope (or synoptophore) is used, both the angle of deviation and the subjective angle should be measured using the instrument. If the red lens test (measure of angle S) is used, a cover test (measure of angle H) should be done at the same distance and under the same lighting conditions. Correction for EF must be made when necessary (tests requiring monocular fixation) when measuring angle H. Tests for subjective angle (S) do not require such correction.

The Bagolini striated lens test is often used to distinguish NRC from ARC. Proponents of this test contend that its major advantage is that it is a “natural” test, done in free space and with no artificial stimulus such as an afterimage. However, the incidence of ARC is much higher than with any other test. To perform the test, Bagolini lenses are placed before the eyes with the lens axes oriented at 45 and 135 degrees, respectively. While the person fixates on a penlight (or a transilluminator), two white lines are seen (Fig. 2–15). The patient is simply asked to describe the location of the two white lines with respect to the fixation light. If the cover test reveals strabismus at the time of testing, a single light and two white lines passing through the light (making a perfect X) indicates harmonious ARC (see Fig. 1–11). If two fixation lights are seen and are separated by the angle of deviation, each line has one of the two white lines passing through it, indicating NRC. The Bagolini striated lens test is also used to diagnose suppression. Because the Bagolini test is a measure of the subjective angle of directionization (angle S test), there is no need to account for EF.

DIPLOPIA ANALYSIS

A diplopia analysis includes a group of tests used to determine if the direction and magnitude of diplopia correlate with the patient’s eye position and state of retinal correspondence. If the patient has strabismus with NRC (the two foveas, when stimulated, give rise to a perception in the same visual direction) and no suppression, double vision should be experienced. The magnitude and direction of diplopia should be evaluated to see if it correlates with what is predicted from the type of correspondence. Similarly, if the patient has ARC, the type of ARC (harmonious, unharmonious, paradoxical type I, paradoxical type II) should be identified.

SENSORY EVALUATION

Before performing a sensory evaluation when a manifest deviation (tropia) is present, the angle of deviation (or the subjective angle of directionization in the case of ARC) must be neutralized with prisms. Prisms optically place the image of the target on the two foveas or on corresponding retinal points. In this way, the patient is given the best chance to manifest binocular fusion ability.

Sensory evaluation of a strabismic patient may be performed using a haploscopic-type instrument (synoptophore or major amblyoscope) or in free space. The synoptophore is the preferred instrument because it permits neutralization of the horizontal, vertical, and torsional components of the strabismus before beginning sensory evaluation. The targets used allow testing of first-, second-, and third-degree fusion (see Fig. 2–2). The test also affords an opportunity to break through any suppression and provides the best chance of fusion. During the presentation of fusible targets, the normally fixing eye should occasionally be occluded to ensure that the patient is fusing normally and not anomalously (Douse target test). When the tubes of the instrument are set at the patient’s angle of deviation and the patient reports fusion of second-degree targets, the normally fixing eye should be momentarily occluded and any eye movements of the fellow eye noted. If the deviated eye does not move, normal fusion is occurring. If there is movement, anomalous fusion or EF is present. Whether the patient fuses in the instrument, a sensory evaluation is also conducted in free space without the constraints (proximal accommodation and proximal vergence) imposed by the tubes of the instrument.

Sensory fusion in free space may be tested in a variety of ways depending on the complexity of the case and the patient’s ability to fuse. Second-degree fusion is typically tested with a red lens and penlight or by the Worth dot test. Before starting, the angle of deviation should be neutralized with loose prisms (hand-held or in a trial frame, prism bars, or rigid Fresnel prisms). With the prisms in place a cover test is performed to verify that motor fusion has been achieved.

In the red lens test, a red-colored filter is placed over the right eye. The patient is asked to fixate a white penlight and asked to describe what is seen (Fig. 2–16). If the response is one light, the color of the light should be identified: red or white indicates suppression, and pink, fusion. Vague responses such as red surrounded by white, alternating red and white, or half red and half white should be verified by other tests. The key feature in the analysis is simultaneously seeing the red and white light with color mixture. If suppression is reported, changing the stimulus conditions (fixation distance or lighting) or having the patient blink should be tried to break through any suppression and maximize fusion potential.

The Worth dot test is performed similarly except that red and green filters are used over the eyes. Use of these filters has been criticized as being dissociative, mitigating against fusion. This result may, however, be viewed in a positive way: if fusion can be demonstrated under such adverse conditions, then the fusion response must be strong. Conversely, the lack of a fusion response does not mean that fusion cannot be demonstrated by a different test.

The red lens, Worth dot, and Bagolini striated lens tests all may be used to evaluate the presence of suppression in free space. The image seen by the suppressing eye will not be seen under binocular viewing conditions. In the red lens test, a patient with suppression reports that either the red or white light is missing. In the Worth dot test, either two red
or three green lights will be seen at one time. In the Bagolini striated lens test, only one fixation light, or one of the white lines, will be appreciated at any one time. If suppression occurs when testing in free space, techniques to break through suppression and promote second-degree fusion should also be used.

Third-degree fusion is a higher grade of fusion and is more difficult to demonstrate. Its occurrence in patients with strabismus when the deviation is neutralized with prisms is a positive indication that correcting the strabismus will improve sensory function and favors long-term stability of the outcome. Traditionally, third-degree fusion is tested at near with a book test using contour stereotest targets. Random dot targets may be used, but many of these tests evaluate gross stereoeability (e.g., Lang stereotest). Stereopsis may also be tested using third-degree targets in the major amblyoscope. This test has some advantage in that it is done at optical infinity (because of the +5.00-D lenses in the tubes.
Physical Space

Figure 2-15. Bagolini striated lens test. The striations of the Bagolini lenses are oriented 45 and 135 degrees in front of the patient's eyes, and a fixation light is held 40 cm in the midline in front of the patient. The patient is asked to describe the position of the lines.

Perceived Space

Figure 2-16. Red lens test. By convention, the red lens is usually placed in front of the right eye and a fixation light is held at 40 cm in front of the patient. The esotropic patient with normal retinal correspondence shown here will perceive 2 lights: one red (seen by right eye) and one white (seen by left eye).

Prism Adaptation Test

Jampolsky 11 first described the prism adaptation test (PAT) as developed by his orthoptist Woodward. PAT was used to preoperatively predict which patients will develop residual esotropia after surgery. Overcorrecting prisms (the alternate cover test demonstrates esotropia with the prisms in place) are given in the office and worn for approximately 1 hour (Fig. 2–17). Prism acceptance is defined by a stable exodeviation or a slight convergence movement to attain bifoveal fixation (no movement on the unilateral cover test through prisms). Patients with either of these favorable reactions were believed to have the best chance of gaining a stable surgical correction. If the patient converged to overcorrecting prisms, becoming esotropic on cover testing, the likelihood of postsurgical esotropia was greater.

The PAT was later modified by “chasing” the amount of overcorrecting prisms required. If the patient converged to

the initial overcorrecting prisms, base-out prisms are added until a small stable exodeviation can be maintained.66

Presurgical use of PAT resulted in 82% success 6 months after surgery, compared with only 51% in a non–prism-adapted group.67 In the PAT group, the larger prism-adapted angle was the target angle for surgery. For example, if a patient with 20-PD esotropia stabilized after PAT at 30-PD esotropia, the target angle is 30-PD esotropia. There were no overcorrections in the PAT responders despite the increased amount of surgery performed. Patients who prism adapted to greater than 60 PD or who did not obtain a stable angle with peripheral fusion were operated on for the original angle of deviation.

A 5-year randomized prospective multicenter trial using PAT for acquired esotropia (PAT study) found that wearing prisms that neutralized the angle of deviation and waiting a sufficient time for rudimentary sensory fusion to develop offered the patient the best opportunity of attaining a stable postoperative angle.68 Eighty-nine percent of patients who were prism adapted before surgery achieved a stable alignment at the 6-month postoperative examination, of whom 69% attained rudimentary sensory fusion. In contrast, only 72% success was achieved in the non–prism-adapted group. From these results it appears that prism neutralization alone
**Why Treat Adult Strabismus?**

Strabismus surgery in adults can result in much more than just improvement in motor alignment and appearance. It may improve fusion as well as increase a patient’s binocular field. Peripheral fusion and expansion of the visual field offer benefits to adult patients in daily activities. Walking in unfamiliar areas and driving are two activities in which peripheral fusion and an expanded visual field are particularly helpful.

In a study of 24 patients (12 of whom had congenital esotropia) with no evidence of binocularity presurgically (as measured by the red lens test, Worth dot, or stereopsis), all patients had motor alignment of less than 8 PD and exhibited a fusion response on the Worth dot test after surgery. Half the patients had between 100 and 200 seconds of arc stereacuity on the Titmus test. In another series, 86% of 359 adult patients with no evidence of binocular response before surgery demonstrated a postsurgical fusion response on the Bagolini striated lens test after being aligned to within 10 PD of orthotropia.

In neither study was an attempt made to distinguish between normal and anomalous fusion. Four lights on the Worth dot test and perceiving an X on the Bagolini test with residual esotropia of less than 10 PD imply that the fusion measured was anomalous. This is supported by the very low-grade stereopsis achieved in one of these studies.

Expansion of the binocular visual field has been described as a benefit from treating adult strabismus. With the use of the Goldmann perimeter to measure the binocular visual fields, the vast majority of patients experienced an expansion of their visual field postsurgically. In one series, all 35 patients had expanded visual fields proportional to, and predicted by, the amount of surgical correction and the residual deviation, independent of the patient’s best corrected acuity.

(Without orthoptics) enhances presurgical fusion and increases the chance of fusion being attained after surgery.

**Conclusions**

In many clinical situations, examination in the office with tests such as the red lens test, the Worth dot test, and distance and near stereopsis tests provides most of the data required to assess a patient’s sensory status. (Many of these tests are discussed in detail in Chapter 1.) In some situations a more thorough sensory evaluation is desirable so that the ophthalmologist can gain an adequate idea of the benefits or disadvantages of proceeding with either orthoptics or surgery. Examples of such situations include the following:

1. In a patient with a history of constant strabismus, will bifoveal fixation or peripheral fusion be achievable after motor alignment?
2. In an adult who develops diplopia after a long-standing childhood strabismus, can orthoptics or surgery restores a previous area of suppression to relieve diplopia or visual confusion?
3. What type of retinal correspondence is present in a visually mature individual with a history of long-standing strabismus? There are rare occasions when a patient with ARC but straight eyes experiences intractable diplopia. The surgeon needs to be informed of this possibility, so that patients may be counseled appropriately.
4. If a torsional deviation is a major component of strabismus, will neutralization of the torsion permit fusion?
5. In patients with longstanding unilateral cataracts and strabismus, has central fusion disruption occurred that will cause permanent inability to fuse?
6. With strabismus occurring after severe head trauma, will the patient be able to fuse or will central fusion disruption prevent sensory fusion despite motor alignment?

**REFERENCES**

47. Maxwell J: On the unequal sensibility of the foramen centrale to lights of different colors. Rep Br Assoc Advan Sci 1856;Part 2:12.
Tests of muscle function are used in selected cases of incomitant deviation primarily to evaluate limited ocular rotations. The tests allow for a more precise diagnosis and provide valuable information for deciding between treatment options. The spontaneous improvement or progression and effects of surgery may be assessed.

These tests are used to differentiate between muscle weakness and restrictive causes of limited ocular rotations, and to distinguish between extraocular muscle paresis and true palsy. Clinically useful measures include the forced duction test (or passive forced traction test), the force generation test, saccadic velocity analysis, and dynamic magnetic resonance imaging (MRI). The concurrent use of edrophonium (Tensilon) with saccadic velocity testing has broadened the clinical usefulness and diagnostic accuracy of this test. Orbital imaging is discussed in Chapter 6.

Previously, electromyography (EMG) was also used to evaluate muscle function. EMG activity, however, may exist in a muscle that is still essentially paretic before true muscle force returns. The response is also suppressed by general anesthesia, except perhaps with ketamine and low doses of nitrous oxide. It continues to be of limited use in subtle cases of Duane syndrome to document aberrant firing of extraocular muscles when globe retraction is clinically difficult to observe or when the diagnosis is questionable. Chemodenervation therapy uses the EMG to confirm accurate toxin administration.

### Passive Forced Duction Testing

The forced duction test was first described for evaluating Duane syndrome in the 1900s. The use of intraoperative forced duction was not emphasized until 1964. Today the forced duction test is critical for the diagnosis of muscle palsies or restrictions. The test is valuable in specific cases of strabismus (Table 3–1) but is not useful if ocular rotations are full. It may be performed when the patient is under general anesthesia or in the office utilizing topical anesthesia.

A grading scale (e.g., a three- or four-point scale) may be used to assign some arbitrary value to the forced duction test.

### OFFICE FORCED DUCTION TESTING

The best results of forced duction testing in the office are obtained in cooperative adults and children aged 7 years or older. In preparing the patient for the procedure the ophthalmologist should clearly communicate the goals and expectations of the test. When properly performed the test is short and relatively painless.

A topical anesthetic such as tetracaine is applied, either as eye drops or with a cotton-tipped applicator at the area where the conjunctiva will be grasped. If there are no medical contraindications, 1:1000 epinephrine solution may be instilled to enhance anesthetic absorption.

The patient is instructed to cover one eye and, with the

<table>
<thead>
<tr>
<th>Table 3–1. Clinical Indications for Forced Duction Testing</th>
</tr>
</thead>
<tbody>
<tr>
<td>Trauma (e.g., orbital floor fracture)</td>
</tr>
<tr>
<td>Primary: restrictions ± paresis</td>
</tr>
<tr>
<td>Acquired: secondary contracture of antagonist</td>
</tr>
<tr>
<td>Endocrine ophthalmopathy</td>
</tr>
<tr>
<td>Congenital restrictions</td>
</tr>
<tr>
<td>Brown syndrome</td>
</tr>
<tr>
<td>Congenital fibrosis of the extraocular muscles</td>
</tr>
<tr>
<td>Möbius syndrome</td>
</tr>
<tr>
<td>Strabismus fixus</td>
</tr>
<tr>
<td>Postoperative restriction of motility</td>
</tr>
<tr>
<td>Strabismus procedures</td>
</tr>
<tr>
<td>Retinal detachment repair</td>
</tr>
<tr>
<td>Glaucoma implant procedure</td>
</tr>
<tr>
<td>Orbital surgery</td>
</tr>
<tr>
<td>Long-standing deviations with secondary muscle contracture</td>
</tr>
<tr>
<td>Transposition procedures</td>
</tr>
<tr>
<td>Orbital diseases</td>
</tr>
<tr>
<td>Tumors</td>
</tr>
<tr>
<td>Inflammation</td>
</tr>
</tbody>
</table>
Figure 3-1. Forced duction testing. Patient with limited upgaze is instructed to look as far up and out before forceps are used to grasp the globe. The examiner then moves the globe farther in this field following the muscle’s natural arc of rotation. Arrow indicates direction of forceps.

other, look in the direction of the muscle suspected of having limited rotation. It may be necessary to occlude the patient’s other eye with an orthoptic patch or an occluder. This ensures fixation with the eye being tested. The crucial question to answer is, “Can the forceps rotate the globe further than the patient can using maximal innervation in that gaze field?”

The patient is asked to look as far as possible into the field of gaze being tested; a fixation target will help the patient maintain gaze in this position, which ensures that maximum innervation is recruited for evaluating the suspected muscle’s function. There is frequently a tendency for an eye with limited ocular rotation to “fall off” or migrate back toward the primary position. If this occurs, the results of the forced duction test may be misleading because the examiner is able to rotate the eye passively (Fig. 3-1). Holding the lids apart with one hand, the examiner uses forceps (either the toothed variety or Pierce forceps) for globe fixation. The examiner should grasp as close to the limbus as possible opposite the side of gaze limitation. This area corresponds to that where Tenon’s capsule and conjunctiva are fused in one layer. This limits stretching of the conjunctiva and gives the examiner a firmer grasp in rotating the globe.66

Prior studies have found that patients better tolerate cotton-tipped applicators for fixation. This technique, however, is less sensitive, and inadvertent retroplacement of the globe may occur, invalidating the results of the test.37, 121 In doing the forced duction (note that this is a monocular test), it is important to follow the natural arc of globe rotation.28 Knapp69 stressed that pushing the eye into the orbit would give a false-negative result because full rotation may be simulated by retroplacement of the globe. For vertical rectus muscles, Jampolsky64 believes that the test should be performed in 23 degrees of abduction, following the anatomy of the vertical rectus muscles, instead of straight up or straight down. If the globe cannot be passively rotated farther than the patient can, a restriction is present. If passive rotation is possible, some degree of muscle paresis is likely.

Retroplacement of the globe may distinguish between inferior and superior restrictions to upgaze limitation.115 In an upgaze deficiency such as Brown syndrome, retroplacement of the globe during upward rotation exaggerates a restriction caused by the superior oblique muscle. If the restriction is lessened with retroplacement, it probably is located inferiorly. A “knife’s edge” sensation may be present when performing the traction test in Brown syndrome.112

Forced duction testing in the office increases diagnostic accuracy and helps in choosing a surgical strategy (Table 3-2). However, there will be patients who cannot cooperate fully during this test despite the most ideal situation. Stimulating the vagal response may cause syncopal attacks.66 In these situations the test may be done in the operating room just before surgery.

**INTRAOPERATIVE FORCED DUCTION TESTING**

It is instructive to perform intraoperative forced duction testing on all patients undergoing surgery, even those who are expected to have negative results. This trains the surgeon in the “feel of normal” passive forces.68, 69 More importantly, patients with limited ocular rotations before surgery or those in whom surgery may cause limited extraocular muscle movement should have a forced duction test done intraoperatively. They include patients in whom the forced duction test was first done in the office. Testing before surgical intervention confirms the results of office testing, whereas testing during surgery evaluates the need to modify the surgical procedure. For example, excessive medial rectus resections may cause limited abduction and postoperative overcorrection in contralateral gaze despite good results in the primary position. The medial rectus is an “unforgiving” muscle. Careful forced duction tests during surgery before the muscle is permanently secured to the globe prevents this type of lateral incomitance.

In cases in which a resection or a transposition procedure was done, it may be necessary to use an intraoperative adjustable suture technique (e.g., slip knot) to detect an iatrogenically caused restriction. If surgery caused too great a restriction, the muscle position may still be modified by slightly recessing the muscle before the final knots are tied.58, 66

In performing intraoperative forced duction testing, the muscle is passively rotated along its natural arc of movement. With the patient supine, the globe is slightly proptosed before carrying out the movement. In some cases the conjunctiva contributes significantly to the mechanical restric-

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Forced Duction</th>
<th>Force Generation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mechanical restriction</td>
<td>Restricted</td>
<td>Normal</td>
</tr>
<tr>
<td>Muscle palsy</td>
<td>Free</td>
<td>Absent</td>
</tr>
<tr>
<td>Combined paresis and</td>
<td>Restricted</td>
<td>Weak to absent</td>
</tr>
<tr>
<td>restriction</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Table 3-2. Results of Forced Duction and Force Generation Testing and Possible Diagnosis
tion, producing a characteristic “string or indentation sign” (Fig. 3–2 and see also Fig. 38–2).66, 87a

The results of forced duction testing before any surgical intervention should be compared with those obtained in the office and with the clinically observed limitation of ocular rotations. After the cause of the restriction has been identified and fibrous tissue has been excised, the offending agent (e.g., a scleral buckle) removed, or a contracted muscle detached from its insertion, the forced duction test is repeated. This confirms the identified cause and the surgical efficacy of releasing the restriction.

**FORCED DUCTION TESTING OF OBLIQUE MUSCLES**

Guyton11 described an exaggerated forced traction test that helps evaluate oblique muscle function on the operating table. The test does not follow the principle of rotating the globe in the normal physiologic field of rotation, as is normally done in the forced duction test described earlier in this chapter. It represents more of a stress test for the oblique muscles and probably explains why a high percentage of mechanical tightness is obtained, even in normal oblique muscles. The Guyton technique uses retroplacement and torsional maneuvers of the globe and permits a graded evaluation of oblique muscle tightness. Details of performing this test are discussed in Chapter 4. Plager’s modification is presented in Chapter 15.

**INTERPRETATION OF RESULTS**

**Qualitative Assessment**

Three types of restriction are recognizable by both qualitative and quantitative forced duction tests.99

**Absolute Restriction.** The first type, an absolute restriction (e.g., Graves ophthalmopathy or Brown syndrome), is the most obvious type of limitation. An immediate increase in resistance is felt when forceps are applied with just a small amount of passive forced duction movement. The examiner cannot rotate the globe past a certain point despite exerting maximum force.

**Uniform Restriction.** In a uniform type of restriction the examiner feels a linearly progressive increase in resistance to passive traction across the arc of rotation. Examples of this type include restrictions secondary to scar tissue or muscle contracture.

**Leash Phenomenon.** In leash phenomenon there is a range of normal resistance followed by a sudden increase toward the end of the field of gaze as the end of the “leash” is reached. This has been documented in certain cases of moderate contracture and fibrosis. It also is noted in long-standing extraocular muscle palsy, indicating that agonist muscle weakness may result in secondary mechanical restriction of the antagonist.98, 106 Jampolsky,95 the first to describe the surgical leash phenomenon, differentiated between leashes and reverse leashes in strabismus. A direct leash is due to scar tissue or stiffness of the muscle opposite the gaze limitation. A reverse leash is due to scar tissue or muscle tightness on the same side as the gaze limitation.

**Quantitative Assessment**

Several attempts have been made to quantify the amount of passive force from the extraocular muscles.20, 70, 91, 128, 129 Most of the instruments used for this purpose are bulky, difficult to use, and require special training. With the hope of gaining wider acceptance, recent modifications have been made, including attempts at decreasing cost, improving the mobility of instruments, and enhancing their ease of use.82, 99 Passive length-tension curves constructed in several studies reveal an almost linear relationship, requiring 0.3 to 0.5 g of force to produce a degree of rotation in anesthetized patients.20, 47, 91, 99, 110 With the patient awake, 0.5 to 1.6 g of force is needed for each degree of rotation.110, 119 Comitant esotropia and exotropia did not yield statistically significant differences from normal values.25, 96, 99, 128 This contrasts with findings obtained in patients with incomitant strabismus (usually of the restrictive type) with limited rotations.47, 72 In patients with restrictions (e.g., those with Graves disease), steeper curves show that 0.7 to 2.0 g of force is required to produce 1 degree of movement.99 Furthermore, surgical recession of medial rectus muscles produces larger changes in passive tension than equal amounts of recession of the lateral rectus in esotropic and exotropic patients.25, 90

**Common Pitfalls in Diagnosis**

**Patient Apprehension.** Apprehensive patients may squeeze their eyes and guard muscle movement in anticipation of the forceps grasping the globe. This precludes accurate interpretation of the forced duction test. If the patient does not adequately look into the field being examined, the antagonist muscle remains innervated. A resistance will be felt and misinterpreted as a restriction. Patient anxiety also may lead to complications such as conjunctival tearing and hemorrhage.58, 66 This is one reason why the test needs to be repeated before surgery in the operating room under anesthesia.

**Pharmacologic Effects.** In patients under general anesthesia, short-acting depolarizing agents such as succinylcholine (Anectine) cause a sustained contraction of the extraocular muscles that persists for 15 to 20 minutes after administration.27, 122 When this agent is used, the examiner should wait

---

*Figure 3–2. “String or indentation sign” in conjunctiva of a patient after multiple reoperations. Conjunctiva is contributing to restricted ocular rotations. (Courtesy of Arthur Jampolsky, MD.)*
20 minutes before doing the forced duction test. Some ophthalmologists compare the results of the forced duction in the suspected field of limited rotation to that in the opposite direction (the field of the antagonist) to compensate for the effect of succinylcholine, but the results may be difficult to interpret. Passive length-tension curves convert to straight parallel lines after intravenous administration of this agent. Lack of awareness of the use of succinylcholine or similar agents will yield false-positive results. Alternatively, nondepolarizing agents such as pancuronium may be used without affecting forced duction testing results.

Errors in Technique. Failure to follow the natural arc of muscle movement or pushing the globe in its socket can yield false-negative results by shortening the arc of contact. Proposing the globe too much, on the other hand, may give a false-positive result. Overstretching of tissues during surgery may lend a false sense of having relieved a restriction. Occasionally, release of a restriction has been reported after the forced duction test. 

Posterior Restrictions. Restrictions may occur far more posteriorly than conventional meticulous dissection can relieve. More than 90% of respondents in one survey experienced this type of restriction. This finding has been documented by computed tomography (Fig. 3–3).

False-Negative Test. Failure to recognize the presence of a restriction when one exists constitutes a false-negative test. This commonly occurs in cases of mild restriction but may be avoided if the surgeon has become adept at identifying normal forced ductions.

Coexisting Paresis and Restriction. Kushner reported the association of rectus muscle paresis and restriction in orbital floor fractures. One may diagnose limited rotations due to a restriction and overlook an associated muscle palsy. In these cases, force generation testing and saccadic velocity analysis provide useful adjunctive information. This problem is also encountered in long-standing paresis with secondary contracture of the antagonist and in unusual cases of thyroid ophthalmopathy.

Co-contraction Syndromes and Aberrant Innervations. Cases of co-contraction syndrome and aberrant innervation may yield a positive test when no abnormality exists on testing in the office. With anesthesia, the test results may be more reliable. Tongue documented her experience with forced duction testing in patients with Duane syndrome. In an esotropic type with Duane syndrome in the left eye, for example, forced duction testing on the left lateral rectus muscle is free on attempted abduction and paradoxically positive in adduction. The resistance to rotation is relieved when the patient is asked to fixate in abduction (left gaze) while the globe is rotated into adduction. Restriction in adduction is due to aberrant firing of the lateral rectus muscle in right gaze, whereas relieving this restriction on abduction is due to the absence of lateral rectus firing in left gaze (Fig. 3–4).

Force Generation Testing

For decades neurologists and orthopedists have assessed muscle function by estimating muscle force. By applying a counteracting force or resistance to a series of isometrically contracting muscles, the strength of the muscle or muscle groups is estimated. Contraction of active extraocular muscles formerly were documented with an oculomyodynamometer, but it was Scott who first described the application of force generation testing in strabismus for patients with lateral rectus paralysis. His report marked the transition to active force generation tests when he showed that the technique is a better investigative tool than the EMG.

OFFICE FORCE GENERATION TESTING

As in office forced duction testing, the patient needs to be reassured and informed about the goals and expectations of the procedure. The active force generation test cannot be done with a patient under anesthesia or heavy sedation because active cooperation is required. Topical anesthetic is applied in the usual manner. The forceps are positioned slightly anterior to the rectus muscle in question.

Potential complications include conjunctival hemorrhage and corneal abrasion. Conjunctival hemorrhage may occur as the conjunctiva is pulled against several grams of force. Corneal abrasion happens when the examiner’s grasp of the forceps is overcome by the generated muscle force and the forceps slip. The position of the forceps should be changed to the limbus opposite the duction effort to reduce the risk of corneal abrasion.

Although both techniques of force generation testing described are measures of isometric muscle forces, Kushner described the following modification. The patient is instructed to voluntarily look maximally in the field of action of the suspected paretic muscle before the surgeon grasps the eye at the limbus opposite the muscle and pulls against it to evaluate the force or, more appropriately, the resistance encountered.

The currently preferred technique for force generation testing is as follows. In lateral rectus palsy, for example, the patient is instructed to abduct the eye as far as possible. The surgeon grasps the globe slightly posterior to the limbus near the medial rectus insertion. While the patient looks in the lateral rectus muscle’s field of gaze, the examiner applies traction in the opposite direction and evaluates the resistance. Minimizing the amount of time that forceps are applied to the globe limits the risk of complications. The technique may also be done right after forced duction testing using the same forceps site. The conjunctiva near the limbus then is grasped only once (Fig. 3–5).

Differential intraocular pressure (IOP) readings are an indirect means of estimating generated muscle forces when ocular movement is limited by muscle contracture, as in restrictive thyroid ophthalmopathy. If a healthy agonist muscle contracts against a tight antagonist, IOP readings increase compared with the relaxed state. This technique is useful in the office and for children who will be uncooperative with invasive techniques such as the use of forceps to achieve fixation. With use of a pneumotonometer to provide recorded tracings of IOP, the amount of generated muscle force increases on eccentric gaze in cases of restriction, suggesting good muscle function.

Another innovative but indirect technique uses a spring dynamometer to distinguish muscle paresis from a restriction and is performed by recording the movement of the nonfixing eye at distance. By Hering’s law, equal and simultaneous innervation is received by corresponding yoke muscles.
Figure 3–3. A, Patient with limited ocular rotations of right eye in all gazes. B, Computed tomography demonstrates dense fibrous tissue posterior to the equator within the muscle cone. C, Forced duction testing revealed restricted ocular rotations in all directions. Figure shows restricted upgaze (top) and downgaze (bottom). (B from Rosenbaum AL, Urrea PT. Investigation of limited ocular rotations: Current status. Am Orthopt J 1987;37:1. Used with permission of University of Wisconsin Press and John Baker, MD. Photographs courtesy of John Baker, MD.)
Normal nonfixing eye movements were established as 12 to 13 degrees when the fixing eye moved 15 degrees. Paretic muscles moved less than 11 degrees, whereas those with restriction did not have movements differing from normal controls.

**LABORATORY FORCE GENERATION TESTING**

Scott used a strain gauge voltage output device to measure the amount of force generated against the degree of ocular rotation. He compared his results with EMG studies and found his to be a better method of quantifying generated muscle forces. Later, forceps were devised to measure extraocular muscle strength. Using this technique in awake patients, 60 to 80 g of isometric force is produced by a normal rectus muscle in extreme lateral or vertical gaze. Paretic muscles generate less force, whereas muscles acting against a restriction generate supranormal values.

Several laboratory devices have been devised to measure both passive and active forces of the extraocular muscles, with the ultimate goal of providing an objective basis for calculating the amount of strabismus surgery. They have not been widely used because of limited cost and availability.

**INTERPRETATION OF RESULTS**

**Qualitative Assessment**

Although force generation is a qualitative evaluation, this test remains crucial for making management decisions in cases of paresis versus palsy or the combined occurrence of paresis and restriction. Paresis means a weakened muscle, but the paretic muscle still is able to generate some movement or contraction. Complete palsy, on the other hand, means absence of innervation and the inability to generate any significant force.

If a muscle contracts normally in its field of gaze, the observer is unable to rotate the globe in the opposite direction. If the muscle is paretic, the examiner can rotate the globe but notes resistance. In a completely palsied muscle, the observer is able to rotate the globe with no resistance felt in the opposite direction. The results can be classified as normal, weak, or absent force generation. A three- or four-point scale may be used.

**Quantitative Assessment**

The gold standard in force generation testing is to measure the amount of force generated by a contracting muscle. With the knowledge that a normal extraocular rectus muscle

**Figure 3-4.** Paradoxical results of forced duction testing of a patient with Duane syndrome of the left eye. Top left, Lid retraction on adduction of the left eye and mild adduction deficiency. Top middle, Small exotropia in primary gaze. Top right, Markedly limited abduction of the left eye. Middle, Forced duction testing performed in left lateral gaze. The examiner is able to rotate the globe more in abduction (left gaze) without encountering any resistance. Bottom left, Forced duction testing reveals a restriction on attempted adduction of the left eye. Bottom right, Restriction is relieved (negative results) when the patient attempts to abduct the left eye. This paradoxical result of forced duction testing in Duane syndrome is due to the absence of lateral rectus muscle firing in abduction and anomalous lateral rectus muscle recruitment in adduction. (Courtesy of Andrea Tongue, MD.)

**Figure 3-5.** Force generation testing. The surgeon does not need to change the position of the forceps. While the patient looks as far up and out as possible, the examiner applies countertraction in the opposite direction. Arrow indicates direction of countertraction applied by examiner.
generates 60 to 80 g of isometric force, muscles generating forces less than this are considered to be paretic or underacting. A completely palsied muscle will register less than 5 g of force with Scott’s modified forceps. Mild paresis will generate 30 to 50 g, and moderate paresis will produce only 10 to 30 g of force. In the absence of any generated force, muscle transposition surgery may be required.

Common Pitfalls in Diagnosis

The most common error is a false-negative result, denoting normality, in cases of mild paresis. If saccadic velocity analysis also is done, mild paresis may be better documented. Lack of standards contributes to the confusion and explains why some surgeons are reluctant to use the procedure. As in forced duction testing, patient cooperation and apprehension are important factors that may affect the results.

Saccadic Velocity Analysis

HISTORICAL PERSPECTIVE

The term saccadic movement was introduced to describe the rapid changes in eye position that occur during fixation pauses while reading. Saccades are rapid eye movements between two points in space and can be horizontal, vertical, or oblique. With EMG recording, saccades are recorded as an intense, almost total firing or recruitment of the agonist muscle accompanied by relaxation or electrical silence of the antagonist in an “all or nothing” manner. Movements during the fast phase of nystagmus have similar characteristics. Rectus muscles are believed to be primarily responsible for generating these eye movements. The degree of slowing of saccades may be proportional to the extent of muscle weakness.

François and Derouck were the first to study eye movement velocity for evaluating paralysis and the return of muscle function. More accurate measurements were not obtained until the 1960s. The first-generation tests used electro-oculography (EOG) techniques and were widely used for the study and diagnosis of eye movement disorders (Table 3–3). Problems with EOG technology in the study of vertical saccades later became apparent. Artifacts believed to come from eyelid excursion or changes in tissue resistance altered the EOG tracings and made their interpretation difficult. This led to the development and use of the infrared technique, later followed by the scleral search coil technique.

QUALITATIVE OFFICE SACCADIC VELOCITY TESTING

The patient is instructed to look at two separate objects or fixation targets held horizontally apart for studying horizontal saccades or vertically for vertical saccades (Fig. 3–6). Movements of at least 20 degrees are optimal for obtaining a clinically useful measurement of saccadic speed. It is important to bring the eye to the field where the muscle in question has maximal function. For the test to yield good clinical information, saccades should be measured only in gaze fields where the patient has a full unrestricted range of motion. If one measures into the field of paresis, movement artifacts (and tracing artifacts if done in the laboratory) can be misleading. In a lateral rectus muscle palsy, for example, the eye should be brought into adduction so that the patient can maximally demonstrate abducting power while gener-
ing lateral saccades. This is a better maneuver than using primary gaze as a starting point and instructing the patient to look laterally; residual muscle function may be missed if the test is conducted in this manner. The larger the movement that the patient can execute, the easier it is to observe saccades.

The briskness of the generated saccades should be noted and the agonist and antagonist velocities compared. Saccades generated from the fellow eye in the same direction also provide an inherent normal control on which to base judgments. "Floating" saccades refer to the type of saccades generated by a palsied muscle. When these are seen, laboratory confirmation of the diagnosis is probably unnecessary.

### INDICATIONS FOR QUANTITATIVE SACCADIC VELOCITY ANALYSIS

Quantitative analysis of saccadic velocity, though providing valuable information, may not always be necessary. A clinical saccadic velocity test done in the office, especially in conjunction with other tests of muscle function (forced duction and force generation testing), may yield sufficient information for an accurate clinical diagnosis.

Some indications for laboratory saccadic velocity testing are listed below:

1. Subtle differences between mild paresis and a normal muscle may be difficult to discern clinically, even by the astute observer. Force generation tests may be equivocal, and saccades may appear normal clinically. In these cases the laboratory determination of saccadic velocity is more sensitive and serves as an invaluable adjunct for reaching a precise diagnosis.

2. In cases of paresis versus restriction, or when both occur, the test effectively identifies the paretic component. This usually occurs in association with either an orbital blow-out fracture with coexisting entrapment and rectus muscle injury or a paresis with secondary contracture.

3. In children or adults who otherwise will not cooperate with more invasive tests for muscle function involving forceps, saccadic velocity analysis may still be done successfully. The test is particularly important in cases of lateral rectus palsy, double elevator palsy, and lost or slipped muscles in children.

4. Subtle cases of Duane syndrome may exhibit mild slowing of adducting saccades without clinically apparent globe retraction or enophthalmos because of failure of relaxation of the lateral rectus muscle on adduction.

5. Lost or slipped muscles may be diagnosed by comparing saccadic differences between the agonist and antagonist muscles.

6. In the study of neurologic diseases that affect extraocular muscles such as myasthenia gravis, saccadic velocity analysis used in conjunction with edrophonium testing help confirm the diagnosis.

7. If the patient generates less than 20 degrees but more than 5 degrees of saccadic excursion, clinical observation alone may be insufficient to differentiate total from partial palsy and may require a laboratory evaluation (see also section on clinical applications).

### TECHNIQUES OF QUANTITATIVE SACCADIC VELOCITY TESTING

#### Electro-oculography

Electro-oculography (EOG) was first described for use with saccadic velocity analysis as early as 1955. Since then, many studies using EOG have followed. EOG is still a practical method despite its limitations (i.e., artifacts making interpretation difficult). In many institutions, EOG remains the most readily available of the three types of laboratory test for saccadic velocity analysis. The recordings are valid without head restraint, and the equipment is fairly inexpensive and does not require sophisticated technician training. The procedure is relatively free of discomfort and may easily be used in infants and children.

In recording horizontal saccades, skin electrodes are attached nasal to the medial canthus and temporal to the lateral canthus (Fig. 3–7). The indifferent electrode is placed on the forehead. For vertical saccades the electrodes are attached centrally on the lower lid and above the brow; the neutral electrode is placed on the skin lateral to the eye. Calibrated eye movements are obtained by having the patient perform voluntary saccades at a known distance.

#### Infrared Scleral Reflection Technique

The infrared scleral reflection technique records eye movements by monitoring the position of the corneal limbus. The technique more accurately reproduces horizontal saccades of 20 and 30 degrees than does EOG but continues to produce recording artifacts with vertical saccadic recordings. The detection of infrared light from the surface of the eye remains obscured by the lid margins both superiorly and inferiorty. In most subjects, the infrared method cannot detect eye movements greater than 10 degrees in upgaze or 20 degrees in downgaze. Artifacts from displacement of infrared receptors or improper positioning of the sensors become more apparent with increasing eccentric gazer.

#### Scleral Search Coil Technique

Because of the problems encountered with EOG and infrared techniques, the magnetic scleral search coil method gained prominence. It is regarded as the most sensitive, accurate, and "noise-free" method of measuring eye movements. A wire is embedded in a large-diameter soft contact lens annulus that fits flush to the surface of the globe outside the corneal diameter. The patient is centered in a magnetic field generated by large search coils. The technology, however, is expensive, and the test causes minor eye irritation and discomfort, requiring more cooperation than the previous two methods. These disadvantages limit its use with infants and children.

### INTERPRETATION OF RESULTS

#### Qualitative Assessment

Abnormal saccadic movements may be observed qualitatively. "Floating" saccades of the eyes into a muscle’s field of action suggest muscle palsy, whereas slowing of the entire
Figure 3-7. Electro-oculographic technique of saccadic velocity testing. A. Note placement of electrodes for recording of horizontal saccades. B. A similar procedure may be performed in children without the need for anesthesia. A pacifier or bottle usually is sufficient to gain patient cooperation. C. Electrode positions showing technique of recording horizontal saccades in both eyes and vertical saccades in left eye.

In infants and children who are too young to produce voluntary eye movements, saccades may be obtained by spinning the child in the arms of an adult to elicit vestibular nystagmus or by presenting an optokinetic tape or drum to produce optokinetic nystagmus (OKN). The rapid recovery phase of nystagmus is a saccade; it can be evaluated from the eye movement recording and compared with the fast phase of OKN generated in the opposite direction. In most clinical situations, saccades of 20 degrees provide more than adequate data for clinical use. A reduction in saccadic velocity suggests weakness of a muscle generating the saccades. Normal voluntary saccades are generally regarded as having the same agonist and antagonist velocities. In a comparison of the velocities from the agonist and the antagonist muscles, a difference of at least 20% suggests paresis, whereas a difference greater than this suggests a slipped muscle. A similar scheme is used for evaluating vertically acting muscles.

Quantitative Assessment of Saccades

Normal values need to be established by each laboratory. Depending on the literature cited, with EOG techniques, velocities of normal saccades of 20 degrees or more can range from 200 to 350 degrees/sec to as high as 370 to 550 degrees/sec. Ten-degree saccades were reported to be 235 to 315 degrees/sec. In most clinical situations, saccades of 20 degrees provide more than adequate data for clinical use. A reduction in saccadic velocity suggests weakness of a muscle generating the saccades. Normal voluntary saccades are generally regarded as having the same agonist and antagonist velocities. In a comparison of the velocities from the agonist and the antagonist muscles, a difference of at least 20% suggests paresis, whereas a difference greater than this suggests a slipped muscle. A similar scheme is used for evaluating vertically acting muscles.

Most studies that establish normal saccadic values are based on values in young adults. Horizontal saccade parameters correlated negatively with increasing age for larger saccades in the range of 35 to 40 degrees. The increase in latency of saccade values is greatest for larger saccades, becoming progressively less with smaller ones. The reduction in velocity is not seen for saccades of less than 20 degrees.
Vertical saccades are more difficult to study with both EOG and infrared techniques. Nonetheless, slowed and delayed vertical excursions have been shown using a 10-degree target. With scleral search coil techniques, decreased range and accuracy of vertical saccades and increased latency with senescence were documented even for saccades greater than 30 degrees. Because of the wide difference in normal velocities with different methods of recording, it is better to compare vertical saccades with saccades generated in the same direction by the contralateral eye.

Common Pitfalls in Diagnosis

**Errors in Technique.** Failure to perform saccadic velocity tests in the field where the extraocular muscles have some remaining function will yield recording and movement artifacts. These artifacts will make interpretation difficult if not erroneous.

**Pharmacologic Effects.** Generated saccades may be affected by various drugs including scopolamine, barbiturates, alcohol, methadone, benzodiazepines (e.g., midazolam, diazepam), alpha-adrenergic agents such as clonidine, and the antagonist idazoxan.

**Fatigue.** Normal patients may grow tired and lose concentration during the procedure. It is imperative that the examiner pay attention to the patient’s state of alertness while recording saccades. If fatigue is the cause of changes recorded in saccades and the patient is alerted, the changes should disappear.

**Time of Day.** The time of day has been shown to slightly affect saccadic velocities.

CLINICAL APPLICATIONS

### Paralytic Strabismus

In lateral rectus palsy, abducting saccades are slowed compared with adducting saccades (Fig. 3–8). In oculomotor palsy, slowed saccades have been documented in adduction, elevation, and depression, compared with normal saccades generated by the lateral rectus muscle. Recovery of the nerve or the muscle may be documented using serial analysis, as was shown in studies of lateral rectus palsy (see also Chapters 18 and 19 for discussions of third and sixth nerve palsy).

Downward saccadic velocities are mildly slowed in patients with superior oblique palsy when recorded in the adducted position. The difference between upward and downward saccades of more than 50% was strongly suggestive of superior oblique palsy. Other reports, however, failed to confirm these results. It is believed that oblique muscles do not play a major role in generating vertical saccadic velocity (see also Chapter 15).

Serial analyses will demonstrate a progressive reduction in the agonist-antagonist difference while muscle function improves. In these cases, surgery may be delayed until velocities have stabilized. In cases of contracture of the antagonist, simple version testing may reveal a decrease in rotations. If saccadic velocities were obtained, any progressive return of muscle function can still be analyzed. This is important in determining the timing of intervention. Significant return of muscle function may influence the selection of muscle resection as opposed to transposition surgery.

**Other Neurologic Disorders**

Myasthenic patients have been documented to have abnormally slowed average saccadic velocities of large-amplitude saccades with increased duration. Although not a universal finding in all patients with myasthenia gravis, reversal of saccadic fatigue by edrophonium appears to be a useful diagnostic feature (Fig. 3–9). This is consistent with transient reversal of the myasthenic muscle defect by edrophonium. Hypermetric saccades and increased saccadic gain have also been described after edrophonium administration and can probably be explained by an adaptive increase in innervation. False-positive results have been reported in healthy subjects exhibiting hypermetric responses after administration of edrophonium. Pure ocular myasthenics may also show a negative response to edrophonium. Tearing has artificially reduced the effect of edrophonium on saccades. The fatigue effect may be accentuated by inducing an OKN that represents repetitive movements. The nystagmus shows a small rolling pendular movement rather than a slow following pattern with rapid recovery saccades (Fig. 3–10).

Maximum saccadic velocity generated during the initial phase of a saccade is frequently normal to supranormal in myasthenic patients but this velocity cannot be

![Figure 3–8. Saccadic velocity tracings of lateral rectus palsy. Upward direction is adduction; downward direction is abduction. A, left, Peak velocity channels show reduced peak velocity in abducting saccades. A, right, Normal peak velocity channels are shown for comparison. Note equal peaks attained by adduction (medial rectus [MR] and abduction (lateral rectus [LR]). B, left, Position tracing shows slowed abducting saccades. B, right, Normal position tracing shown for comparison. (Courtesy of Ocular Motility Laboratory, Jules Stein Eye Institute, University of California, Los Angeles.](https://example.com/figure38.png)
maintained with large saccades. Intrasaccadic fatigue causes a sudden fall in saccadic velocity. The presence of a normal or supranormal maximum velocity at the start of a saccade means that a muscle functions during the initial phases and is paretic only later in the movement. In a related study, investigators have observed a quiver movement on attempted saccades even in the presence of severe ophthalmoplegia. The finding, although rare, was believed to be pathognomonic for myasthenia.

The constellation of the following findings suggests the diagnosis of myasthenia gravis: normal to supranormal peak saccadic velocities in the initial phases of saccade generation; quiver eye movements; fatigue effects or decrescendo intrasaccadic disorders; and the effect of edrophonium. These findings distinguish myasthenia gravis from progressive external ophthalmoplegia, internuclear ophthalmoplegia, and cranial nerve palsies (Table 3–4).

Patients with progressive external ophthalmoplegia

Figure 3-10. Optokinetic nystagmus (OKN) induced saccades in a patient with myasthenia gravis. L, left gaze; R, right gaze. A, Before edrophonium (Tensilon), a poor OKN response is shown with flattening of the tracing as fatigue sets in. B, After Tensilon administration, there is increased amplitude of OKN and recovery of left saccades. (Courtesy of Ocular Motility Laboratory, Jules Stein Eye Institute, University of California, Los Angeles.)
Table 3–4. Saccadic Velocity Analysis in Neurologic Disorders

<table>
<thead>
<tr>
<th>Saccadic Features</th>
<th>Myasthenia Gravis</th>
<th>Progressive External Ophthalmoplegia</th>
<th>Internuclear Ophthalmoplegia</th>
<th>Cranial Nerve Palsies</th>
</tr>
</thead>
<tbody>
<tr>
<td>Peak velocity</td>
<td>Normal to supranormal</td>
<td>Slowed</td>
<td>Slowed</td>
<td>Slowed</td>
</tr>
<tr>
<td>Average velocity</td>
<td>Decreased</td>
<td>Slowed in all directions at any phase</td>
<td></td>
<td>Slowed</td>
</tr>
<tr>
<td>Amplitude</td>
<td>Decreased</td>
<td>Decreased</td>
<td>Decreased</td>
<td>Decreased</td>
</tr>
<tr>
<td>Optokinetic nystagmus</td>
<td>Increased</td>
<td>No effect</td>
<td>No effect</td>
<td>No effect</td>
</tr>
<tr>
<td>Latency</td>
<td>±</td>
<td>Increased</td>
<td>±</td>
<td>Increased</td>
</tr>
<tr>
<td>Fatigue effects</td>
<td>+</td>
<td>±</td>
<td>±</td>
<td>±</td>
</tr>
<tr>
<td>Reversal with edrophonium</td>
<td>±</td>
<td>Increased</td>
<td>±</td>
<td>±</td>
</tr>
<tr>
<td>Other findings</td>
<td>“Quiver” saccades; intersaccadic variation</td>
<td>Abducting nystagmus</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

+, finding observed; ±, finding may or may not be observed; –, no effect or finding not observed.

138, 139 have slowed saccadic velocities in all directions at any phase of saccade generation. This pattern is bilateral and symmetric and is unaffected by edrophonium.60, 70, 75 In progressive external ophthalmoplegia, muscle fibers are already paretic at the beginning of a saccade so that normal saccadic velocities cannot be generated.

The classic constellation of findings in internuclear ophthalmoplegia includes slowing of adduction saccades with normal abduction velocity and abducting nystagmus (Fig. 3–11).24, 48, 60, 85 In internuclear ophthalmoplegia, the generated saccades are limited by a decreased amplitude of pulse innervation reaching the extraocular muscles.138 Möbius syndrome is characterized by sixth and seventh nerve palsy with associated horizontal gaze palsy. Saccadic velocity analysis shows normal vertical saccades, whereas horizontal movements are extremely limited with marked slowing of both adducting and abducting saccades.132

Restrictive Causes

Patients with restrictive forms of strabismus generally maintain normal saccadic velocities.66, 70 This has been demonstrated in thyroid ophthalmopathy,66, 61 fractures of the orbital walls without associated paresis,66, 81 and Brown syndrome63 (see also Chapters 21, 23, and 25).

If an orbital floor fracture is associated with inferior rectus paresis, a slow downward saccade will be obtained.61 Other causes of restriction that do not affect saccadic velocity include rectus muscle contracture, scarring due to multiple eye surgeries such as strabismus procedures, and retinal detachment surgery.60

Duane Syndrome

Abduction saccades may show findings similar to sixth nerve palsy owing to lack of recruitment of the lateral rectus muscle on abduction; however, adduction saccades also show mild to moderate slowing.80, 132 This is a key distinguishing feature between Duane syndrome and sixth nerve palsy, or even medial wall entrapment of the medial rectus, as a cause for abduction deficits.58, 80 The information is helpful in planning strabismus surgery in these patients, especially if surgery on the lateral rectus muscle is contemplated11, 58, 87 (see also Chapter 24).

Figure 3–11. Patient with internuclear ophthalmoplegia due to multiple sclerosis. Notice bilateral deficiency in adduction in this patient. The classic constellation of findings in internuclear ophthalmoplegia also includes slowing of adduction saccades with normal abduction velocity and abducting nystagmus (not shown).
Patients with synergistic divergence show saccadic pattern
trends similar to those seen in Duane syndrome. This
syndrome consists of monocular adduction palsy with a large
extropia. On attempted contralateral gaze, the eye rotate
into simultaneous abduction (divergence). Ocular torticollis,
with nystagmus occurring spontaneously and with optokinetic
and vestibular responses, has also been observed.

**Lost or Slipped Muscle**

When a muscle becomes lost or disinserted due to either
trauma or surgery, saccadic velocity analysis yields valuable
information. If the muscle has slipped, saccadic velocities are
reduced 20% to 50% or more from normal. Movement
of the globe does not stop at midline. Rotations may
be reduced but are not absent. The reduced saccade is usually
undetected clinically if saccades are greater than 100
degrees. “Floating” saccades are not seen. When the muscle
is located and reattached, saccadic velocities return to normal.
If the muscle has been lost, saccadic velocities may be
profundely reduced (see also Chapter 40).

**Evaluation of Surgery**

After muscle transposition surgery for palsied extraocular
muscles, increased saccadic velocities are recorded in the
field of action of the paralytic muscle, a finding not noted
after large resections alone. A posterior fixation procedure or the fadenoperation theo-
retically decreases the torque exerted by the muscle as the
eye is moved into its field of action. Following this para-
digm, Kushner postulated that the procedure should re-
sult in successive reductions in saccadic velocities as the
operated eye moves into its field of action. No theories
were proposed, but his study failed to disclose the expected
response after surgery on the medial rectus.

**Conclusions**

Tests of muscle function have proven valuable for as-
sessing the strabismic patient. Although laboratory tests of
muscle function permit a quantitative evaluation of strabis-
us, essentially the same information may be obtained by
clinically similar tests done in the office or the operating
room, without the need for complex laboratory instrumentation.
Because this book is designed for clinical use, the
details of conducting tests in the laboratory are not fully
discussed. The intent is to alert the clinician to simple ways of
evaluating muscle function to arrive at a more conclusive
diagnosis and choose the most appropriate surgical strategy.

**REFERENCES**

1. Abbott RL, Metz HS, Weber AA: Saccadic velocity studies in Möbius
2. Abel LA, Troost BT, Dell’Ossio LF: The effects of age on normal
saccadic eye movements in children of primary school age. Doc
1979;29:1150.
movements in man: I. Effects of the benzodiazipine receptor ligands
for the diagnosis of myasthenia gravis. Ann NY Acad Sci
velocity in normal subjects and myasthenic and nonmyasthenic
11. Bodi F, van Allen MW, Yarbrough JC: Duane syndrome: An electro-
12. Boghen D, Troost BT, Daroff RB, et al: Velocity characteristics of
13. Chioran GM, Yee RD: Analysis of electro-oculographic artifact during
vertical saccadic eye movements. Graefes Arch Clin Exp Ophthal
14. Ciancia AO, Garcia HA, Lavin R: Treatment of the lateral rectus
15. Cogan DG, Yee RD, Gittinger J: Rapid eye movements in myasthenia
16. Collewijn H, Erkelens CJ, Steinman RM: Binocular co-ordination of
17. Collewijn H, Erkelens CJ, Steinman RM: Binocular co-ordination of
18. Collewijn H, van der Mark F, Jansen TC: Precise recording of human
Clinical Implications, p 145. Elmsford, NY, Pergamon Press,
1975.
20. Collins CC, Scott AB, O’Meara DM: Elements of the peripheral
oculomotor apparatus. Am J Optom Arch Am Optom
21. Cook G, Stark L: The human eye-movement mechanism: Experiments,
adduction palsy and synergistic divergence: A clinical and electro-
23. Dodge R: Five types of eye movements in the horizontal meridian
oplegia: Recovery and plasticity. Invest Ophthal Vis Sci
intraoperative length-tension curves of human extraocular muscles.
In Campos EC (ed): Strabismus and Ocular Motility Disorders. Pro-
cedings of the 6th International Strabismological Association, Sur-
1990.
tent conduction block in myasthenia gravis and Guillain-Barré syn-
drome: An oculographic study with computer simulations. Arch Neu-
27. France NK, France TD, Woodburn JD, et al: Succinylcholine alteration
28. Francois J, Derouck A: Etude electrooculographique des paralysies
29. Fricker SJ: Use of velocity and acceleration measurements in the
evaluation of the strabismus patients. In Moore S, Mein J, Stockbridge
International Orthoptic Congress, 1975, p 113. New York, Stratton
Intercontinental Medical Books Corp, 1976.
30. Gilmour JY, Meyer JS: The neurologic examination and functional
movements in man: II. Effects of the alpha,-adrenergic receptor ligands
32. Goldstein JH: The intraoperative forced duction tests. Arch Ophthal-
mol 1964:72:5.
33. Guyton DL: Exaggerated traction test for the oblique muscles. Ophthal-


Rosenebaum VL: The clinical application of saccadic velocity testing. In Souza-Diaz C (ed): Smith-Kettlewell Symposium on Basic Sciences in Strabismus. "Mechanical and Tonic Factors on Strabismus Diagnosis and Surgery." Annex to the 5th Congress of the Conselho Latino...


Torsional strabismus occurs when the eye is abnormally rotated about the visual axis. Although evaluation of ocular torsion is clearly indicated when a patient complains of torsional diplopia, this assessment is often neglected in patients lacking this complaint. The vertical rectus and oblique muscles ("cyclovertical" muscles) are responsible for the torsional position of the eye. Malfunction of these muscles causes characteristic alterations in the torsional position of the eye. Therefore, an assessment of torsion adds another dimension to the diagnosis and management of all patients with vertical strabismus, even those who do not complain of torsional diplopia.

DEFINITIONS

Torsion refers to rotation of the eye about its visual axis. Intorsion occurs when the 12 o'clock meridian is rotated nasally, and extorsion occurs when the 12 o'clock meridian is rotated temporally. Objective (anatomic) torsion refers to anatomic rotation of the eye about the visual axis. Subjective torsion refers to the patient's perception of rotation that may result from anatomic torsion.

The torsional position of the eye should be distinguished from torsional movements. Torsional position applies to the eye when it is at rest (in any gaze position), whereas torsional movements apply to the eye in motion. For example, a patient with a superior oblique (SO) palsy has an abnormal torsional position (extorsion) of the involved eye both in primary position and in abduction. In contrast, a patient with dissociated vertical deviation (DVD) has normal torsional movements (extorsion) as the eye elevates. The rotary motion observed in torsional nystagmus is another example of abnormal torsional movement.

This chapter discusses the diagnostic and therapeutic value of assessing the torsional position of the eye. All further references pertain to the torsional position, not torsional movements, of the eye.

ANATOMIC TORSION

Anatomic torsion is the actual torsional position of the eye. It is not dependent on a subjective patient response. It is difficult to accurately determine the anatomic torsional position of the eye on external examination because there is no consistent landmark denoting the 12 o'clock meridian. It is helpful to compare the relative positions of two fundus landmarks, the fovea and the optic nerve. The actual axis of rotation of the eye is close to the visual axis emanating from the fovea. However, it is easier for most examiners to visualize the fovea moving relative to the optic nerve.

Bixenman and von Noorden analyzed fundus photographs from 50 normal subjects and found that a horizontal line through the fovea intersected the inferior half of the optic nerve in approximately 95% (Fig. 4–1). The eye was considered abnormally extorted if a horizontal line through the fovea passed below the lower margin of the optic nerve and abnormally intorted if this line was above the center of the disc (Fig. 4–2). If one uses this scale, normal torsional position could be anywhere within a range of 12.5 degrees of rotation. Relatively little variation was found in the torsional position of the two eyes in any given individual. An interocular difference greater than or equal to ¼ disc diameter in the foveal position relative to the optic nerve was considered abnormal.

ETIOLOGY

Clinically significant torsion may be associated with idio-pathic strabismus or can be caused by extraocular muscle, neurologic, or orbital disorders. Paralysis or overaction of any of the four cyclovertical
muscles in either eye will cause strabismus with a torsional component. Primary oblique muscle overaction associated with loss of fusion is probably the most common cause of anatomic torsion. A restrictive process involving cyclovertical muscles may also cause strabismus with a significant torsional component. Examples include thyroid-related orbitopathy, Brown syndrome, and orbital blow-out fractures. Local myotoxicity from retrobulbar or peribulbar anesthesia may induce scarring of the cyclovertical muscles. This has been implicated as a frequent cause of strabismus after cataract and retinal detachment surgery.4, 17, 22

Neurologic disorders are a frequent cause of torsion; fourth cranial nerve palsy is the most common. Brain stem disease may cause skew deviations, which are sometimes associated with abnormal torsion.2, 3, 7, 10, 20, 33 Patients with skew deviation generally have other neurologic signs in addition to ocular misalignment.

Ocular torsion may be secondary to anatomic orbital abnormalities seen in craniofacial malformations such as plagiocephaly (Fig. 4–3).5, 6, 19, 30 The induced torsion may be secondary to abnormal anatomic insertions of the extraocular muscles or abnormal torsion of the actual orbit. Some cases of congenital SO “palsy” may actually be caused by trochlear displacement secondary to orbital malformation.30

Figure 4–1. Average foveal position (dotted line) and range of normal (solid lines); noninverted (direct) view of the left eye. (Redrawn from Bixenman WW, von Noorden GK: Apparent foveal displacement in normal subjects and incyclotropia. Ophthalmology 1982;89:58.)

Figure 4–2. Noninverted (direct) view of the left eye. A, Incyclotorsion is present when the fovea is above the normal range in the direct view. B, Excyclotorsion is present when the fovea is anywhere below the normal range. (Redrawn from Bixenman WW, von Noorden GK: Apparent foveal displacement in normal subjects and incyclotropia. Ophthalmology 1982;89:58.)

Figure 4–3. Extorsion of the right orbit in a patient with right-sided plagiocephaly. Arrows indicate change in the path of the right eye compared with the normal path of the left eye in upgaze and downgaze. (From Diamond GR, Katowitz JA, Whitaker LA, et al: Ocular and adnexal complications of unilateral orbital advancement for plagiocephaly. Arch Ophthalmol 1987;105:381. Copyright 1987, American Medical Association.)
Theoretical Origin of Primary Oblique Muscle Overaction

Primary oblique muscle overaction is often associated with other types of strabismus in patients with poor fusion. Guyton and Weingarten propose that absent or poor fusion predisposes to abnormal torsion, which in turn causes primary oblique muscle dysfunction and the associated A and V patterns. The key to understanding this theory is the concept of muscle length adaptation.

There are two distinct mechanisms for maintaining normal ocular alignment. The better understood short-term alignment is maintained by rapid vergence movements, which are regulated by the nervous system in response to retinal disparity. Long-term alignment is controlled by muscle length adaptation in response to tonic nerve stimulation.

Muscle length adaptation is a remodeling process in which extraocular muscles that are stretched will lengthen physically by adding sarcomeres. Muscles that receive reduced tonic nerve stimulation also lengthen over time. In contrast, muscles that are slack (or that receive excessive tonic nerve stimulation) gradually shorten through the removal of sarcomeres. In normal subjects, muscle length adaptation is regulated by long-term tonic nerve stimulation.

When a patient suppresses one eye, there is no feedback loop to ensure that the eyes remain aligned. Thus, in some patients the suppressed eye will drift outward—a sensory exotropia. Initially this is caused by a failure of short-term vergence control of ocular alignment. Eventually, failure of long-term alignment control causes the lateral rectus (LR) muscle to shorten (by losing sarcomeres) and the medial rectus (MR) muscle to lengthen (by adding sarcomeres). Similarly, patients with loss of fusion have no feedback loop to maintain torsional alignment. The oblique muscles therefore may shorten or lengthen just as the horizontal rectus muscles remodel in a sensory exotropia. It is not known why some patients develop shortening of the inferior oblique (IO) muscle whereas others develop shortening of the SO muscle. It also is not clear why younger patients tend to develop sensory esotropias or why older patients have sensory exotropias.

If the IO muscles shorten, the patient develops extorsion. This, in turn, extorts the horizontal and vertical paths of each eye, causing hypertropia with adduction and a V pattern—findings typical of IO muscle overaction (Fig. 4–4). Alternatively, if the SO muscles shorten the patient develops intorsion, which intorts the horizontal and vertical paths of each eye. This causes hypotropia with adduction and an A pattern, findings typical of primary SO muscle overaction.

THE "EXAGGERATED TRACTION TEST": FORCED DUCTIONS FOR THE OBLIQUE MUSCLES

The exaggerated traction test detects tightness of the SO and IO muscles. When done properly, this test places the oblique muscles on maximum stretch by simultaneously retroplacing, torting, and rotating the globe. Note that forced ductions of the rectus muscles are best performed by pulling the eye forward to put these muscles on maximum stretch. In contrast, forced ductions of the oblique muscles are best done by retroplacing the eye. For example, to place the SO muscle on maximal stretch, the globe should be retroplaced and rotated superonasally.

Guyton initially described this test as a “two-handed” test. He subsequently modified the test to be performed with one hand—our preferred method. To perform the test, it is essential to securely grasp episclera and sclera nasally with 0.5-mm toothed forceps to feel the muscle tightness and prevent a conjunctival tear (Fig. 4–5). The eye is then fully retroplaced and rotated nasally. To test the SO muscle the globe is rotated superonasally. A “bump” is felt as the globe is rotated superotemporally over the taut SO tendon. The tendon in effect is “plucked” as if it were a guitar string when the retroplaced globe is rotated from the superonasal to superotemporal position. To test the IO muscle the globe is placed inferonasally, then rotated inferotemporally over the taut IO tendon.

The “bump” or tightness of the IO muscle is less prominent than that of the SO muscle, probably because there is more tissue between the IO muscle and the globe. All normal oblique muscles can be felt if the test is performed properly. Muscle tightness may be subjectively graded on a scale from 0 to 4+, with average SO muscle tightness graded as 1.5+...
and average IO muscle tightness as 1.0+. Good interexaminer correlation has been shown in grading oblique muscle tightness. Grading made with the exaggerated traction test correlate well with clinical overaction (but not underaction) of the oblique muscles. Patients with pure DVD do not have tight oblique muscles on exaggerated traction testing.

The exaggerated traction test should be performed during surgery before and after the oblique muscle is disinserted or tenotomized. Increased oblique muscle tightness can confirm the diagnosis of oblique muscle overaction. The test helps differentiate between hyperdeviations caused by IO overaction (IOOA), DVD, or rectus muscle contracture. In patients with combined IOOA and DVD, the amount of oblique muscle tightness should reflect the contribution of the IOOA to the deviation.

Intraoperative assessment of the completeness of an oblique muscle weakening procedure is probably the most useful application of the exaggerated traction test. After an SO tenotomy, no “bump” is felt as the globe is rotated from the superonasal to the superotemporal quadrant. Often after an apparently “complete” SO tenotomy the test will reveal residual tightness, indicating that some SO fibers were missed. Similarly, residual IO muscle tightness reflects residual fibers that were not disinserted.

Guyton performed the exaggerated traction test on patients who required repeat surgery for horizontal deviation several months after an SO or IO muscle weakening procedure. All of these patients had complete oblique muscle disinsertion confirmed by the lack of a “bump” on testing during initial surgery, as well as postoperative resolution of the oblique overaction. Surprisingly, these patients had normal oblique muscle “tightness” at the time of their second surgery. This was attributed to reattachment of the oblique muscle stump to intermuscular septum, Tenon’s capsule, or sclera, which occurs even after a complete oblique muscle disinsertion.

### Measuring Torsion

#### METHODS OF MEASURING ANATOMIC

**OBJECTIVE** TORSION

**Indirect Ophthalmoscopy**

Indirect ophthalmoscopy provides a simultaneous view of the optic nerve and fovea and is the most convenient way for the examiner to objectively assess torsion. The inverted view, however, can be a source of confusion. The examiner must become fluent in assessing torsion directly from the inverted view provided by the indirect ophthalmoscope, thereby eliminating the extra mental step needed to invert the image. All fundus images in the remainder of this chapter will be inverted to simulate the actual view through the condensing lens of the indirect ophthalmoscope.

In the “indirect ophthalmoscopy view” the horizontal line through the fovea will normally pass through the superior half of the disc. The line will appear to be above the disc when the eye is extorted and below the superior half of the disc when the eye is intorted.

Guyton proposed a grading system to quantify the amount of anatomic (objective) torsion observed during indirect ophthalmoscopy (Fig. 4–6). Based on a recalculation of the data obtained by Bixenman and von Noorden, the anatomic torsional position of an eye was considered normal when a horizontal line through the fovea passes within the inferior one third of the disc. In the indirect ophthalmoscopic view, a horizontal line through the fovea therefore will normally pass through the superior third of the disc. This “normal range of torsion” represents a rotational variability of 9 degrees (Fig. 4–7), which is slightly less than the 12.5-degree normal range defined by Bixenman and von Noorden. If the horizontal line is displaced ½ disc diameter above the superior disc border in the indirect ophthalmoscopic view, the eye is considered to have 4+ extorsion. Similarly, if the horizontal foveal line is displaced ½ disc diameter below the junction of the top and middle third of the disc, the eye is considered to have 4+ intorsion. Intermediate amounts of torsion are defined in Figure 4–6. Examiners not comfortable with the Guyton grading system can use a protractor attached to an indirect ophthalmoscope lens to quantify the ocular torsional position in degrees, although we have not found this step necessary in our practice.

To assess torsion objectively with the indirect ophthalmoscope, the patient is instructed to fixate on the headlight with the eye under study. The light intensity is set as low as possible to allow the patient to comfortably fixate on the light without squeezing the lids. To measure torsion in primary position, the head should be held straight without any turn or tilt. A horizontal line passing through the fovea is imagined by the examiner and anatomic fundus torsion graded as described in Figure 4–6. To assess torsion in other gaze positions, the head may be turned appropriately. For example, a measurement in right gaze can be obtained by turning the patient’s head to the left while maintaining fixation on the headlight. The head should never be tilted, because this will artifically alter the torsional position of the eye.

Owing to the 9-degree normal range of anatomic torsion, a patient may be considered “normal” despite a significant change in ocular torsion. For example, a patient with the horizontal foveal line at the superior disc border (indirect ophthalmoscopic view) can subsequently develop 9 degrees of intorsion in each eye and still be graded as “normal” (see Fig. 4–7). Comparing both eyes of a patient may provide a clue to abnormal torsion in these cases, because the torsional position of the eyes is normally symmetric. An interocular difference of ¼ disc diameter in the foveal position relative to the optic nerve implies abnormal torsion in one or both eyes, even if both eyes appear to be within the normal range. Information from the rest of the ocular motility examination should help identify the abnormal eye.

Reasonably good agreement was found among examiners in grading anatomic torsion by indirect ophthalmoscopy; estimates were within one unit 80% of the time. In addition, there is excellent agreement when indirect ophthalmoscopic grading is compared with fundus photograph grading, as described in the next section. Indirect ophthalmoscopy thus provides a rapid, convenient assessment of anatomic torsion which is accurate and reproducible enough for most clinical situations. The main pitfalls of this technique include sensitivity to improper head positioning and the inadvertent use of a bright headlight.

**Fundus Photography**

Torsion may be determined from fundus photographs that simultaneously include the fovea and optic nerve. To obtain...
Figure 4–5. One-handed exaggerated traction test for the superior oblique muscle, right eye. Upper diagram shows surgeon’s view; lower diagram shows orbital cross section, viewed from above. A, Nasal sclera securely grasped with 0.5-mm toothed forceps. Orbital cross section demonstrates normal laxity of superior oblique muscle and tendon. B, Globe is fully retropulsed. Note redundant conjunctival tissue in surgeon’s view, indicating significant retropulsion. Superior oblique muscle is now on stretch. C, Globe is fully adducted while still retropulsed. Superior oblique muscle remains stretched. D, Globe is rotated superonasally. Resistance to superonasal rotation is felt as superior oblique muscle is now maximally stretched.
Figure 4–5 Continued E, Globe begins to roll over or "pluck" stretched superior oblique tendon. There is simultaneously a slight downward and rotational movement of the globe as it rolls over the tendon. F, Final position. "Bump" was felt as superior oblique tendon slid into new position stretching across opposite side of equator. Globe rolls and moves slightly upward as maneuver is completed.

Figure 4–6. Grading system for estimating abnormal torsion by indirect ophthalmoscopy. Note that this is the view of the right fundus, as seen by the examiner during indirect ophthalmoscopy. Surrounding circle represents edge of indirect ophthalmoscope lens. The normal range for the fovea is between the dotted lines. The upper line is level with the top edge of the disc, whereas the lower line is level with the junction between the top and middle thirds of the disc. If the fovea is located at either end of normal range, "trace" torsion is present. Each additional eighth of a disc diameter is graded as 1+, 2+, and 3+ torsion. If the fovea is displaced one-half disc diameter or more from the normal range, 4+ abnormal torsion is present.
reproducible photographs it is essential that the photographer not allow the patient to adopt a head tilt. The patient should fixate with the eye being photographed. To measure torsion in degrees, the slide film is removed from the slide holder or negatives are studied directly. A horizontal reference is obtained by aligning the edge of the film with a straight edge. The angle between this horizontal reference and a line connecting the fovea with the optic nerve center can then be measured with a protractor (the optic nerve head-foveal angle).

Magnification factors introduced by the photographic technique and the patient's refractive error may affect some measurements of the optic nerve head (ONH)-foveal angle. One photographic method reduces the magnification effect. In this method, the horizontal dimensions of the ONH are measured in each eye. A vertical line through the fovea (distance A) is extended to its point of intersection with a horizontal line extending from the geometric center of the ONH (distance B) (Fig. 4–8). The lengths of these lines are measured with calipers and expressed in relative horizontal disc diameters for each individual eye. The tangent of the ONH-foveal angle is obtained by dividing distance A by distance B, and the actual angular measurement is calculated. Using this technique, Bixenman and von Noorden studied 50 nonstrabismic subjects and found that the fovea is located at a mean angle of 7.25 degrees below the horizontal meridian with a 95% confidence limit of approximately 0 to 12 degrees. This does not mean that the normal eye is physically extorted 7.25 degrees; rather, it reflects the normal anatomic position of the fovea below a horizontal line through the center of the optic nerve.

The main advantage of using fundus photography to grade torsion is that a ruler or protractor may be placed over a still photograph to accurately determine the amount of torsion. This is the most accurate method of measuring anatomic torsion. The main disadvantage of fundus photography is its sensitivity to improper head position. Other drawbacks are the time delay, increased cost, and inconvenience involved.
compared with indirect ophthalmoscopy. In addition, accurate fundus photographs are difficult to obtain in uncooperative children.

Patients with abnormal torsion may still be graded as “normal,” even when photography is used, owing to the large normal range. Despite the large interpatient variability, torsion photography has been shown to be a remarkably sensitive and specific means of diagnosing oblique muscle dysfunction.\(^1\)

**Blind Spot Mapping**

Locke\(^2\) described blind spot mapping as a method of determining anatomic torsion. This was one of the first methods for demonstrating abnormal ocular torsion in patients with cyclovertical muscle dysfunction. To perform the test as originally described, the patient is positioned 1 meter away from a tangent screen. Care is taken to avoid any abnormal head posture, and the blind spot is mapped with a 5-mm white test object. The foveal center is represented by the fixation target. Any abnormal torsional position of the eye will be indicated by heterotopia of the blind spot. A Goldmann visual field test may also be used for blind spot mapping using a 14° target.

The normal position of the disc on blind spot mapping is slightly lower than the point of fixation. Because torsional rotations are centered on the fovea, the disc is lower than the fovea when the eye is intorted and higher when it is extorted. Because the visual field is inverted, the blind spot will be above the normal position if the eye is intorted and below the normal position if the eye is extorted.

Blind spot mapping correlates well with fundus photography. The disadvantage of this technique is the time and cooperation needed to obtain an accurate map of the blind spot.

**Search Coils**

Search coils are a highly accurate method of measuring eye movements. In this technique, scleral contact lens electrodes are placed on the eyes and changes in eye position are detected by measuring alterations in the surrounding magnetic field. The search coil can measure torsional movements but cannot measure absolute torsional position. This technique is a valuable research tool, but it is too time consuming, expensive, and inconvenient for routine clinical use.

**Summary**

Objective (anatomic) torsional position can be measured by indirect ophthalmoscopy, fundus photography, and blind spot mapping (Fig. 4-9). Indirect ophthalmoscopy is the least quantitative but most easily applied method. The findings correlate well with those of fundus photography, and indirect ophthalmoscopy is our preferred method of as-

---

**Figure 4-9.** Three measures of abnormal extorsion of left eye. A, Indirect ophthalmoscopic view (as seen by examiner). B, Fundus photograph. C, Blind spot map. (Modified from Guyton DL: Clinical assessment of ocular torsion. Am Orthopt J 1983;33:7. Reprinted with permission of University of Wisconsin Press.)
METHODS OF MEASURING SUBJECTIVE TORSION

By definition, all methods of assessing subjective torsion require the patient to assess the torsional orientation of a target. The two methods most frequently used are the double Maddox rod and Lancaster red-green tests. Both are dissociative tests; that is, they disrupt fusion when performed properly. The less commonly used Bagolini striated glasses do not disrupt fusion.

Double Maddox Rod Test

In the classic double Maddox rod test the patient fixates on a point source of light while Maddox rod lenses are placed before each eye to produce two horizontal streaks. Each Maddox rod lens has a calibration mark initially placed at 90 degrees in a trial frame. By convention, a red Maddox rod is placed over the right eye to produce a red streak and a white Maddox rod is placed over the left eye to produce a white streak. If necessary, a vertical prism is placed in front of one eye to disrupt fusion. A patient with normal subjective torsion will perceive two parallel horizontal lines. If an eye has abnormal torsion, the lines will not appear parallel. The patient is instructed to rotate the appropriate Maddox rod lenses until the lines become horizontal and parallel. The degree of subjective torsion matches the final position of the Maddox rods. For example, if the patient rotates the right lens from 90 to 97 degrees, the right eye has 7 degrees of subjective extorsion.

The advantages of the double Maddox rod test are that it is easy to perform, it is quickly completed, and it provides a quantitative measurement of subjective torsion. However, there are several disadvantages: torsion can be measured only in primary position; torsion may falsely localize to the wrong eye; and the Maddox rod axis must be accurately marked (and the lenses properly placed in the trial frame) to avoid errors. As with all tests of torsion, proper head position is crucial.

In a study of patients with SO paresis, 83% localized subjective extorsion to the eye viewing through the red Maddox rod lens, irrespective of the side of paresis or fixation preference. This artifact is explained by increased light transmission and a better ability to perceive environmental cues through the white Maddox rod lens. The patient perceives the image viewed through the white Maddox rod lens as horizontal and localizes torsion to the dissociated image viewed through the red Maddox rod lens. These effects may be reduced by placing red Maddox rod lenses in front of both eyes and performing the test in a completely dark room to minimize environmental cues. With these modifications, 94% of patients correctly localized subjective extorsion to the paretic eye.

We recommend the following modified protocol for the double Maddox rod test (Fig. 4–10). Red Maddox rod lenses are placed over each eye in a trial frame. The lenses are oriented obliquely (calibration mark 5–10 degrees away from the vertical) to avoid falsely localizing torsion to one particular eye. If necessary, a 6-PD prism is placed base down over one eye to vertically separate the lines and prevent cyclofusion. The room is completely darkened, and the patient then adjusts the trial frame lenses to make each line horizontal (and both lines parallel).

Lancaster Red-Green Test

The Lancaster red-green test provides a diagrammatic representation of horizontal, vertical, and torsional deviations in the nine diagnostic positions of gaze. The following equipment is required: red-green goggles (large enough to cover the eyes in all positions of gaze); flashlights projecting a red and a green streak (Clement Clarke, Harlow, Essex, UK, and Columbus, OH, USA); and a screen (preferably calibrated) indicating the nine gaze positions.

The patient is seated 1 meter from the screen and asked to hold the head straight throughout the procedure (Fig. 4–11). The goggles traditionally are placed on the patient with the red lens over the right eye and the green lens over the left eye. The examiner projects the red streak on the wall in primary position (right eye “fixing”). The red streak is positioned slightly obliquely, and the examiner rotates it as instructed by the patient until it appears to be vertical. The patient then holds the green flashlight and is asked to position the green streak “in the same place as the red streak.”
The actual locations of the light bars are manually recorded, typically using red ink for the right eye and green or blue ink for the left eye. This procedure is repeated in nine positions of gaze. In each position the patient is first asked if the red streak appears to be vertical, and the green streak is not superimposed until the red streak is satisfactorily adjusted. The examiner and patient then exchange lights, and the procedure is repeated with the green light now representing the “fixing” left eye in each gaze position. The fixing eye is indicated on the diagram.

The Lancaster red-green test is interpreted as if the two streaks are direct projections from the foveas. Understanding this concept simplifies interpretation of the results. The left side of the plot indicates left gaze, and the right, right gaze. If the red streak is higher than the green, the right eye is higher (right hypertropia). If the red streak is rotated clockwise, the right eye is extorted, and if the green streak is rotated clockwise, the left eye is intorted. If the patient has esotropia, the visual axes cross and the patient will place the green streak (left eye) to the right of the red streak (right eye).

This direct correlation with eye position may confuse a novice, because the map is opposite to what a patient would report seeing with the more familiar red glass test. Another source of confusion is that, in esotropia, a V pattern will map out a letter A shape on the Lancaster red-green plot whereas an A pattern will map out a letter V shape. (In contrast, if a patient has exotropia, a V pattern will map out a letter V shape and an A pattern will map out a letter A shape.) With minimal experience in its use, however, the simple and direct correlation with eye position is a great asset of this test. A Lancaster red-green plot of a patient with a bilateral fourth cranial nerve palsy is shown in Figure 4-12.

The advantages of the Lancaster red-green test include the ability to assess torsion in all positions. The pictorial overview of motility is superior to that provided by prism-and-cover testing, which does not provide information on torsion. The test takes about the same amount of time to perform as prism-and-cover testing in nine positions. The test can be quantitative if done using a calibrated screen.

A disadvantage of the test is the potential for error in transcribing the streak position to paper. (However, Gonzales has developed a computerized version of the Lancaster red-green test that automatically records the results.) The Lancaster red-green test does not facilitate control of accommodation, which may cause variable horizontal deviations. Patients with poor vision in one eye may have trouble seeing
the corresponding streak. A patient with poor vision in the left eye, for example, may not appreciate the green streak. In some cases, placing the red lens over the eye with poor vision may help. The Lancaster red-green test is not useful when deep suppression and anomalous retinal correspondence are present. As with other tests of torsion, head position is crucial. To avoid artifacts the patient should be regularly reminded to move only the eyes when fixing in different gaze positions and to maintain a straight, fixed head position. Testing should be carried out in a dark room to fully dissociate the eyes and avoid environmental cues to fusion.

Another potential problem with Lancaster red-green testing is that a patient may localize more of the torsion to the left eye when the examiner is holding the red light (right eye fixing). The patient may then localize more torsion to the right eye when the examiner holds the green light (left eye fixing). Thus, it is important to record Lancaster plots with each eye fixing in the nine diagnostic gaze positions. Other patients consistently localize torsion to the nondominant eye. Clinical correlation with anatomic torsion is essential if there is any question about which eye is involved.

### Bagolini Striated Lenses Test for Cyclofusion

Bagolini striated lenses measure subjective torsion without disrupting fusion. These are transparent, plano lenses with microscopic etchings that produce a streak of light perpendicular to the axis of striations when viewing a point source of light. (The streak effect of the microscopic etchings may also be produced by carefully wiping a thumb print across a clean plano lens.) The test is done in a lighted room. In contrast to the double Maddox rod test, the surrounding environment is easily viewed through the lenses, allowing subjective torsion to be measured under nearly normal binocular conditions. To evaluate anomalous retinal correspondence, the etch marks are oriented at 45 and 135 degrees. However, to evaluate torsion, the etch marks should be oriented at 90 degrees to produce streaks at 180 degrees.32,35 If there is horizontal or vertical strabismus, a prism should be placed over one lens so that the streaks seen by the patient overlap. If only one horizontal line is perceived, the patient is able to fuse the streaks. If two lines are perceived, the patient is instructed to rotate one lens until the streaks are superimposed and horizontal. Note that in this test the prism is used to facilitate cyclofusion. This contrasts to the double Maddox rod test, in which the prism is used to disrupt cyclofusion.

Ruttim32 and von Noorden36 classify patients having subjective torsion on double Maddox rod testing into two groups. One group has no subjective torsion on Bagolini striated lenses testing. They suggest that these patients successfully use adaptive mechanisms (described in the next section) to compensate for torsional strabismus and that it therefore is not necessary to specifically address torsion when planning surgery. The other group does have subjective torsion on Bagolini striated lenses testing. These patients are unable to use adaptive mechanisms to compensate for torsional strabismus, and therefore torsion should be addressed when planning surgery.

The advantage (and disadvantage) of the Bagolini striated lenses test for cyclotropia is that the value obtained for subjective torsion reflects the combined factors of anatomic torsion and several different adaptive mechanisms. Measurements are affected by numerous factors and may be difficult to interpret. Reliable measurements are obtained only in primary position, and it is essential that the frames fit properly and that the lens “axis” orientation is at 90 degrees initially.

### Summary

Double Maddox rod testing and Lancaster red-green testing both measure subjective torsion. Double Maddox rod testing may measure torsion more accurately in primary position, but the Lancaster red-green test provides a qualitative measurement of horizontal, vertical, and torsional misalignment of the eyes in the nine diagnostic gaze positions. For this reason we generally find it more useful than the double Maddox rod test. With both tests, care must be taken to remove environmental cues, or torsion may falsely localize to the nonparetic eye. Despite this precaution, patients may still localize abnormal torsion to the nonparetic eye regardless of which eye is paretic. Other patients localize torsion to different eyes on different examinations, even though the anatomic torsion is in the same eye on each examination. In addition, patients with long-standing torsion may have developed one or more adaptations that reduce or eliminate subjective torsion despite the presence of significant anatomic torsion (discussed in the next section). Therefore, subjective torsional assessment alone cannot definitively localize abnormal torsion. The results of the double Maddox rod test and the Lancaster red-green test should be combined with anatomic torsional assessment and the findings of the ocular motility examination for proper interpretation.

Bagolini striated lenses testing measures subjective torsion without eliminating numerous adaptive mechanisms and therefore provides information regarding a patient’s ability to adapt to torsional strabismus. Many patients with subjective torsion on double Maddox rod (and Lancaster red-green) testing have no subjective torsion on Bagolini striated lenses testing. Thus, it is more difficult to localize the eye with abnormal torsion using Bagolini lenses. This test therefore is diagnostically inferior to the double Maddox rod test and the Lancaster red-green test. The Bagolini test identifies a subset of patients who lack the ability to use adaptive mechanisms to compensate for torsion, and it may help in planning surgery for these patients.32,35 However, we believe that it is important to consider torsion when planning surgical correction for all patients with vertical strabismus, and we rarely use the Bagolini striated lenses test to measure subjective torsion.

### Sensory and Motor Adaptations to Torsion

Abnormal anatomic torsion is common in patients with strabismus. Overaction or paresis of any of the cyclovertical muscles will cause this disorder. However, patients rarely complain of torsional diplopia unless they have an acute cyclovertical muscle palsy of recent onset. Subjective torsion thus occurs much less frequently than anatomic torsion.
MECHANISMS OF SENSORY AND MOTOR ADAPTATION

Several mechanisms of sensory and motor adaptation to torsion combine to make torsional diplopia rare. They include torsional anomalous retinal correspondence (ARC), receptive field characteristics, torsional motor fusion, unique psychological adaptations, and suppression.

Anomalous Retinal Correspondence

Children with anatomic torsion may develop an ARC type of reordering of the retinal meridians at the occipital cortex. Anatomic torsion may be obvious in these patients; yet subjective torsion may not be demonstrable. Children with an onset of anatomic cyclodeviation before age 6 years rarely exhibit subjective torsion on double Maddox rod testing. In contrast, most patients with an onset of cyclodeviation after age 7 will have subjective torsion. It is possible that patients with adult-onset cyclodeviations can acquire this ARC type of sensory adaptation over many years.

Receptive Field Characteristics

Normal adults can fuse 8 degrees of cyclodisparity using sensory fusion. Adults with anatomic torsional strabismus can use this ability to fuse the tortured images without experiencing torsional diplopia. This ability to fuse torsionally disparate images derives from the receptive fields in the peripheral retina being large compared with those in the central retina. When an eye is tortured, the peripheral retina is displaced more than the central retina (Fig. 4–13). Therefore, patients with significant torsional strabismus may still have substantial overlap of corresponding receptive fields, enabling them to fuse. In contrast, horizontal or vertical misalignments displace the entire retina by the same amount. Therefore, a small amount of horizontal or vertical disparity between two eyes (representing only a few minutes of arc) will not allow overlap of corresponding retinal fields, disrupting fusion.

The double Maddox rod and Lancaster red-green tests are dissociative measures that may detect subjective torsion in patients who, under normal conditions, fuse torsionally disparate images. Patients who are using torsional sensory fusion to adapt to a new-onset torsional strabismus may initially complain of spatial distortion similar to that induced by corrective spectacles with an oblique axis of astigmatism. Most patients adapt to this distortion after days to weeks.

Motor Fusion

In the past, torsional motor fusion was thought to be negligible. However, when measured with large-field stimuli, 8 degrees of cyclovergence have been demonstrated in normal subjects. This amount of motor cyclovergence, combined with the 8 degrees of sensory cyclofusion, allows normal subjects to fuse up to 16 degrees of cyclodisparity. This considerable fusional range is another reason for the rarity of torsional diplopia in patients with cyclodeviations.

Psychological Adaptation and Suppression

Psychological adaptations to environmental cues may also reduce subjective torsion. Horizontal and vertical cues from familiar objects in the environment may lessen the subjective torsion experienced by a patient. Thus, a patient with an acute-onset SO paresis may not notice torsion when viewing monocularly with either eye. If that patient is unable to fuse the images when viewing binocularly, however, he or she will usually experience torsional diplopia. If the patient is able to fuse, these psychological adaptations will enable him or her to perceive the environment as "upright." It is not known whether vestibular or proprioceptive information may contribute to psychological adaptations.

Many patients with early-onset cyclodeviations suppress one eye and therefore do not have subjective torsion. These patients may not appreciate the streak corresponding to the suppressed eye on double Maddox rod or Lancaster red-green testing.

Clinical Applications

CLINICAL APPLICATIONS OF SENSORY ADAPTATIONS

Performing the double Maddox rod test and the Lancaster red-green test in a dark room reduces environmental cues, thereby eliminating psychological adaptations, sensory fusion, and motor fusion. Many patients who do not complain of torsional diplopia may therefore demonstrate subjective torsion when these tests are used. However, patients with long-standing torsional strabismus often have ARC or other poorly understood adaptations that reduce the amount of subjective torsion measured by these tests, even when they are performed properly.

Patients with childhood-onset cyclodeviation and the ARC type of adaptation may experience a “paradoxical” type of
torsional diplopia when the cyclodeviation is corrected.\textsuperscript{18} For example, a patient with anatomic extorsion from congenital SO palsy may have subjective intorsion if the torsional position of the eye is surgically corrected to the anatomically normal range. If the patient is able to fuse the images, he or she may complain of spatial distortions. The monocular and binocular adaptations described previously will generally alleviate this "paradoxical" type of torsional diplopia in a matter of days to weeks.

When anatomic torsion is present, subjective torsion may provide important clues to the time of onset of a cyclodeviation.\textsuperscript{18} The absence of subjective torsion is most consistent with a childhood onset. If subjective torsion is present but less than expected, a long-standing cyclovertical deviation is likely. Similar amounts of subjective and anatomic torsion are consistent with a recent onset.

The Lancaster red-green test may simultaneously demonstrate anatomic and subjective torsion. When an eye is intorted, the rectus muscles are displaced and the horizontal and vertical paths of the eye will be similarly intorted. For example, if an eye is extorted, the MR muscle is displaced upward, the LR downward, the superior rectus (SR) temporally, and the inferior rectus (IR) nasally. The horizontal and vertical paths of that eye will characteristically be altered as the eyes traverse the nine positions of gaze (see Fig. 4–4). This alteration is readily apparent on the Lancaster red-green plot and reflects the amount of anatomic torsion. In contrast, the amount of torsion indicated by each light streak reflects the amount of subjective torsion and is therefore affected by some sensory torsional adaptations. Figure 4–14 shows a Lancaster red-green plot that demonstrates approximately equal amounts of subjective and anatomic torsion consistent with recent-onset torsional strabismus. Figure 4–15 shows a Lancaster red-green plot in which the amount of anatomic torsion exceeds the amount of subjective torsion, suggesting a more remote onset of torsional strabismus.

**Figure 4–14.** Lancaster red-green plot of an acute onset right superior oblique palsy. The horizontal and vertical paths of the right eye (black dotted lines) are extorted, indicating anatomic extorsion of the right eye. In each gaze position, the extorsion of the streak representing the right eye is similar to the amount of anatomic extorsion, indicating correspondence of subjective with anatomic extorsion. (Solid "red" line = right eye; dashed "green" line = left eye. Small black dots are separated by 15 PD. Large black dots indicate fixation target for nine diagnostic gaze positions [left eye fixing].)

**Figure 4–15.** Lancaster red-green plot of a congenital right superior oblique palsy. The horizontal and vertical paths of the right eye (black dotted lines) are extorted, indicating anatomic extorsion of the right eye. The streaks representing the right and left eyes are upright in each gaze position, indicating that sensory adaptations have eliminated all subjective torsion. (Solid "red" line = right eye; dashed "green" line = left eye. Small black dots are separated by 15 PD. Large black dots indicate fixation target for nine diagnostic gaze positions [left eye fixing].)

### CLINICAL APPLICATIONS OF TORSION ANALYSIS

#### Superior Oblique Muscle Palsy

The SO muscle is an intorter whose torsional field of action is greatest in abduction. SO muscle paresis causes anatomic extorsion, which is most marked with abduction of the affected eye. An acute SO muscle paresis may cause 2 to 7 degrees of anatomic extorsion, corresponding to a trace to 1+ extorsion by the Guyton grading system (Fig. 4–16).\textsuperscript{13} Extorsion may increase over time due to secondary IO muscle overaction. Anatomic extorsion will occur in both eyes of patients with bilateral SO paresis. The total amount of cyclodisparity will often exceed 10 degrees, with 1+ to 2+ anatomic extorsion in each eye. As discussed previously, a corresponding amount of subjective torsion will be measured in acute cases, with less predictable amounts in chronic cases due to sensory adaptations.

Assessment of torsion is helpful in the diagnosis of unilateral SO palsies, acute or chronic bilateral SO palsies, and asymmetric bilateral SO disease. It can also provide clues to the duration of illness and help guide the surgical approach.

A typical acute unilateral SO palsy is easily diagnosed by the characteristic incomitant vertical strabismus. However, chronic deviations may become comitant (the so-called spread of comitance). Some of these patients may prefer to fixate with the paretic eye and may confuse the examiner when they present with a comitant hypodeviation of the unaffacted eye. The presence of anatomic extorsion of the fixing eye is a valuable clue to the correct diagnosis.

Patients with acute bilateral SO palsies typically have trace to 1+ anatomic extorsion in both eyes and often more than 10 degrees of excyclotorsion on double Maddox rod testing. The Lancaster red-green test may demonstrate many of the features of bilateral SO palsy, including subjective extorsion of both eyes that is worse in downgaze, extorsion...
of the muscle paths of both eyes (see Fig. 4–12), hypertropia of each eye in adduction, and V-pattern esotropia. In chronic cases, subjective torsion will be less than anatomic torsion.\(^\text{16}\)

If bilateral SO palsies are asymmetric, patients may not present with typical signs.\(^\text{24}\) Hypertropia of the less involved eye may be “masked” by the larger hypertropia in the other eye. These patients may appear to have a unilateral SO palsy in the more severely involved eye. The presence of bilateral extorsion may be a valuable clue to the true diagnosis.

Torsion considerations may influence the surgical treatment of SO palsies. Consider the surgical approach to the following cases:

1. **An adult patient presents with vertical and torsional diplopia secondary to a right SO palsy.** Physical findings include a right hypertropia and 5 degrees of right fundus extorsion. A right IO muscle recession, a left IR muscle recession, or a right SO tuck will induce intorsion, reducing torsional disparity between the eyes.\(^\text{16}\) These surgical options simultaneously treat the vertical and torsional strabismus. In contrast, a right SR muscle recession or right IR muscle resection will induce further extorsion. These procedures treat the vertical deviation but will adversely affect the torsional deviation and therefore should be avoided.

2. **An adult presents with chronic right SO palsy with spread of comitance, right hypertropia, 3+ anatomic extorsion of the right eye, and 3 degrees of subjective right extorsion.** A right SR muscle recession may be performed to treat the vertical deviation because the chronic SO palsy has produced secondary SR muscle contracture. In this patient, the SR muscle should be displaced temporally to increase its intorting effect, counteracting some of the extorsion caused by the SR recession.

3. **An adult patient with bilateral SO palsy describes torsional diplopia secondary to a V pattern and extorsion that is greatest in downgaze.** Cover testing shows esotropia with a 10-PD V pattern. Bilateral SO tucks, or an MR recession combined with Harada-Ito\(^\text{21}\) procedures, will treat the esotropia and the extorsion. However, bilateral MR muscle resections with downward transposition would exacerbate the extorsion—poor surgical choice when extorsion is a significant factor.

A Harada-Ito procedure specifically corrects extorsion by isolating the anterior (torsional) fibers of the SO tendon and tightening the fibers by reinserting them 6 to 8 mm posterior to the LR muscle insertion.\(^\text{21,29}\) Anatomic torsion assessment by fundus examination is a way of quantifying this procedure intraoperatively. The optimal surgical goal is slight overcorrection (approximately 1+ intorsion), achieved by placing

---

**Figure 4–16.** A, Indirect ophthalmoscopic view of the left fundus of a patient with left superior oblique paresis showing 1+ extorsion. B, After left inferior oblique myectomy and left superior oblique tuck of 15 mm, the left fundus was grossly intorted, accompanying a large postoperative occurrence of Brown syndrome. C, After a left superior oblique tenotomy, the fundus was back to a normal torsional position, and the patient’s symptoms had resolved. (From Guyton DL: Strabismus surgery decisions based on torsion findings. In: Anterior Segment and Strabismus Surgery, New York: Kugler, 1997. Reprinted with permission.)
the horizontal foveal line at a level that bisects the disc at its center. Although general anesthesia induces approximately 2 degrees of extorsion in patients with SO paresis, this relatively small artifact does not affect the endpoint. The Harada-Ito procedure can be more precisely quantified if it is done using adjustable sutures. Postoperative Lancaster red-green testing is used to guide the suture adjustment. The desired endpoint is 3 to 5 degrees of intorsion in primary position, with more intorsion in upgaze and less in downgaze. This overcorrection is essential because the effectiveness of the Harada-Ito procedure diminishes in the weeks following surgery.

**Vertical Rectus Muscle Palsy**

Patients with vertical rectus muscle paresis may have trace to 2+ anatomic torsion. The amount of subjective torsion is variable, reflecting the duration of the muscle paresis. The relatively small torsional action of the vertical rectus muscles contributes to some of the abnormal anatomic torsion, but the compensatory increase in innervation of the ipsilateral oblique muscle is more important. For example, the IR muscle normally is responsible for depression and a small amount of extorsion in primary position. Weakness of the IR muscle causes hyperdeviation and a small amount of intorsion of the paretic eye. The ipsilateral SO muscle receives increased compensatory innervation to depress the paretic eye and reduce the vertical deviation. This exacerbates anatomic intorsion of the paretic eye in primary position.

Patients with chronic cyclovertical muscle paresis may develop a “spread of comitance.” Anatomic torsion assessment is valuable for identifying the paretic muscle. Consider the following case:

_A 68-year-old retiree, complaining for years of vertical diplopia, has a comitant right hypertropia. The right eye has no torsion, whereas the left eye has anatomic and subjective extorsion. Forced ductions are normal._

Considering the vertical deviation, the patient may have weakness of one of the following four muscles: right IR, right SO, left SR, or left IO. Anatomic intorsion on the right would support a diagnosis of right IR muscle paresis, whereas anatomic extorsion on the right would support right SO muscle weakness. Anatomic intorsion on the left would support an impression of left IO muscle weakness. In this case, anatomic extorsion on the left is present, making chronic left SR muscle paresis the likely diagnosis. Another cause of left hypotropia with extorsion is left IR muscle contracture, which can be distinguished from left SR muscle palsy by forced duction testing.

**Primary Oblique Muscle Overaction**

Primary oblique muscle overaction is characterized by a vertical deviation that is greatest when the involved eye adducts (Fig. 4–17A, B). By definition, there is no evidence of a previous oblique muscle palsy. Primary IO muscle overaction is associated with V-pattern horizontal deviations and anatomic extorsion. Primary SO muscle overaction correlates with A-pattern horizontal deviations and anatomic intorsion. Patients generally have significant anatomic torsion, often 3+ to 4+ by the Guyton grading system, corresponding with as much as 20 degrees of torsion (Fig. 4–17C, D). Patients who appear to have less anatomic torsion than 20 degrees before developing the disorder. For example, if a patient with horizontal foveal lines that intersect the superior disc borders on indirect ophthalmoscopy subsequently develops SO overaction, as much as 18 degrees of intorsion (9 degrees in each eye [see Fig. 4–7]) may occur before abnormal anatomic intorsion is detected.

Anatomic torsion is more useful than subjective torsion when diagnosing and managing primary oblique muscle overaction. Primary oblique muscle overaction generally occurs gradually in patients with poor fusion. Consequently, there is minimal if any subjective torsion, and patients rarely complain of torsional diplopia. However, patients with A-pattern strabismus and high-grade stereopsis may develop disabling torsional diplopia after an SO muscle weakening procedure.

Assessing anatomic torsion can help distinguish primary oblique muscle overaction from DVD and from pseudo-overaction of the obliques seen in patients with Duane syndrome, the tight LR syndrome, and large-angle horizontal strabismus. In patients with primary oblique muscle overaction, the amount of anatomic torsion in primary position often correlates with the amount of oblique muscle overaction. Patients with DVD and with pseudo-overaction of the obliques typically have normal torsional eye positions in primary position (Fig. 4–18). A patient may have a combination of primary oblique muscle overaction, DVD, and/or pseudo-overaction of the obliques. The amount of anatomic torsion in primary position often reflects the relative contribution of true oblique muscle overaction to the deviation.

An assessment of anatomic torsion contributes to the surgical management of A- or V-pattern horizontal deviations. If a patient with an A- or V-pattern horizontal deviation has no abnormal torsion or oblique muscle overaction, horizontal muscle surgery combined with the appropriate vertical displacement of these horizontal muscles is the procedure of choice. Vertical displacement of the horizontal rectus muscles to collapse an A or V pattern increases the torsional deviation. For example, in a patient with V-pattern esotropia, displacing the MR muscle downward will increase the amount of extorsion. Therefore, this procedure is only recommended in patients without abnormal anatomic torsion or significant oblique overaction (except perhaps when fusion is not a consideration). If a patient with an A- or V-pattern horizontal deviation has significant torsion, horizontal muscle surgery is combined with an oblique weakening procedure to decrease the abnormal torsion (see Fig. 4–17).

In some patients with oblique overaction, versions may not correlate with anatomic torsion. In our experience, anatomic torsion is a more reliable indicator of oblique muscle overaction than is version testing, so the decision to perform oblique muscle surgery with horizontal surgery is weighted toward the results of objective torsion evaluation.

**Dissociated Vertical Deviation**

Dissociated vertical deviation is characterized by torsional movements of the deviating eye. However, the torsional position of the eye in primary position is normal. DVD may simulate IO muscle overaction because it often occurs
with ocular adduction when the nose occludes the eye. Torsional assessment helps distinguish DVD from primary oblique muscle overaction. Some patients have DVD combined with primary IOOA. The amount of anatomic torsion in primary position reflects the relative contribution of IOOA in patients who have both DVD and IOOA.

Assessment of anatomic torsion contributes to the surgical management of DVD. A patient who has DVD and significant IOOA (as indicated by more than 1+ extorsion) should be treated by a recession/anteriorization of the IO muscles to correct both IOOA and DVD. In contrast, patients with significant DVD and minimal IOOA (as indicated by minimal extorsion in primary position) are treated by SR muscle recessions.

A different scenario is enacted in the following case: a 7-year-old girl has 20 PD of esotropia, with a 15-PD A pattern, 2+ anatomic intorsion, and bilaterally symmetric DVD. Patients with DVD combined with SO overaction generally have anatomic intorsion. In this patient, the SO overaction is reducing the amount of manifest DVD. Therefore, an SO weakening procedure should be avoided because it may exacerbate the DVD. Upward displacement of the MR muscles should be avoided because this will worsen the intorsion. If both the horizontal deviation and DVD must be addressed, patients should have horizontal rectus muscle surgery and SR muscle recessions. The SR muscle recessions will improve the A pattern as well as the DVD. McCall and Rosenbaum have suggested SO weakening procedures combined with SR muscle recessions in some patients with both DVD and SO muscle overaction. In this case, the A pattern is small enough that SO weakening is probably not necessary.

**Postoperative Residual Hyperdeviations**

Postoperative assessment of anatomic torsion helps evaluate the response to surgery. This can be especially helpful in cases with residual hyperdeviations after seemingly appropriate surgical correction of IOOA or DVD. Consider the following case:

An 8-year-old boy with a history of 30 PD of esotropia and presumed 2+ IO muscle overaction of both eyes had bilateral MR and IO muscle recessions. Postoperative horizontal alignment was satisfactory. However, he was referred for evaluation of residual hypertropia in adduction of each eye. If postoperative anatomic extorsion is present in primary
position, the patient has residual IOOA, probably secondary to incomplete disinsertion of the IO muscle fibers during the recessions. If surgical correction is indicated, disinserting and recessing the remaining attached IO muscle fibers would be the procedure of choice. However, if postoperative anatomic intorsion is present, the patient may have DVD that was incorrectly diagnosed as IOOA and treated by IO muscle recessions. If DVD is confirmed and surgical correction is indicated, the DVD should be treated by judicious SR muscle recessions.

Brown Syndrome

Brown syndrome is characterized by limited elevation of the affected eye in adduction secondary to restricted passage of the SO tendon through the trochlea. The affected eye typically has anatomic intorsion in primary position and attempted upgaze (Fig. 4–19). In some cases, the affected eye has normal torsion in primary position, but most of these eyes exhibit anatomic intorsion on attempted elevation.

The anatomic torsional position of the eye in primary position can guide the surgical management of patients with Brown syndrome. Patients with anatomic intorsion in primary position (more than 1+ intorsion by the Guyton grading system) probably have a tight SO tendon and may be managed by SO tenotomy, although we now prefer an SO tendon spacer or a “chicken suture” in these cases.

Brown syndrome patients with normal torsion in primary position may have an inelastic SO tendon that limits passive upgaze. The SO tendon may not be tight when the eye is in primary position. In these cases, an SO tenotomy carries a significant risk of creating SO palsy. A short spacer or “chicken suture” is strongly preferred in these cases. An SO tenotomy combined with an IO weakening procedure is also acceptable, but it is more difficult to grade this surgery and much more difficult to reverse the procedure if necessary.

Thyroid Eye Disease

Thyroid eye disease with tight IR muscles may cause 1+ to 2+ anatomic extorsion (Fig. 4–20). As the induced extorsion develops gradually, these patients rarely complain of torsional diplopia.

It is important to consider the torsional consequences of surgical intervention in patients with thyroid eye disease. Large bilateral recessions of the IR muscles may suddenly induce significant intorsion, causing postoperative torsional...
diplopia. After IR muscle recessions, unopposed tight SR muscles and increased use of the SO muscles as depressors exacerbate the intorsion, especially in downgaze. Temporal displacement of the IR muscles at the time a large recession is performed will reduce postoperative intorsion. However, this may aggravate an A-pattern exotropia in downgaze that is caused by unopposed SO function. For this reason, some surgeons suggest nasal displacement of a large IR recession, despite the torsional consequences, to prevent the A pattern.

Strabismus After Injection of Local Anesthetic Agents for Eye Surgery

Intramuscular injection of local anesthetics is an increasingly recognized cause of strabismus after eye surgery, including cataract extraction and scleral buckling. There are three clinical patterns that may result from this type of injury, depending on the extent of damage and the cellular response:

1. If fibrosis involves a large portion of the muscle, a restrictive pattern occurs, with the strabismic deviation greatest opposite the field of action of the involved muscle. Forced ductions will be positive in these cases.
2. If the fibrosis involves a small segment of the muscle, an overaction pattern occurs, with a strabismic deviation greatest in the field of action of the involved muscle. Forced ductions will be positive only if the deviation is large, usually more than 30 PD.
3. If muscle degeneration is extensive and fibrosis minimal, or if there is damage to the nerve innervating the
muscle, a paretic pattern occurs. Forced ductions may be negative unless the antagonist develops a secondary contracture.

Torsion is an invaluable aid in identifying the involved muscle in patients with postsurgical strabismus. Because surgical recession of the involved muscle will frequently cure the strabismus, it is essential to accurately identify this muscle (see also Chapter 28). Consider the following cases:

A 76-year-old patient underwent cataract extraction with intraocular lens implantation in the right eye. Local anesthesia consisted of a single retrobulbar injection of lidocaine and bupivacaine. She noted double vision as soon as the patch was removed. The diplopa subsequently worsened, with significant oblique and torsional components. The Lancaster red-green plot is shown in Figure 4–21. There is a right hypertropia greatest in downgaze, consistent with overaction of the right IR muscle. There is also subjective extorsion of the right eye, which is greatest in adduction, the torsional field of action of the IR muscle. Fundus examination confirmed + anatomic extorsion in primary position. The Lancaster red-green plot also shows a V-pattern esotropia, consistent with IR overaction (adduction is a tertiary action of the IR muscle). Thus, the horizontal, vertical, and torsional deviations all implicated the right IR muscle. Forced duction testing at the time of surgery confirmed a tight right IR muscle, although this is not always noted when an overactive rather than a restrictive pattern is present. The right IR muscle was recessed 15 mm. Figure 4–21B shows the postoperative Lancaster red-green plot. The hypertropia, esotropia, and extorsion were all corrected by recessing only the involved muscle.

A 75-year-old patient underwent cataract extraction with intraocular lens implantation in the left eye. Local anesthesia consisted of a retrobulbar injection of lidocaine and

**Figure 4-21.** Lancaster red-green plot of a 76-year-old patient, 7 months after cataract extraction in the right eye. (Solid “red” line = right eye; dashed “green” line = left eye. Small black dots are separated by 15 PD. Large black dots indicate fixation target for nine diagnostic gaze positions.) A, Before strabismus surgery, left eye fixing. Right hypertropia with V-pattern and right extorsion. [Dotted lines connect plots of left and right eyes in each gaze position.] B, Postoperative appearance, following 15-mm right inferior rectus muscle recession. Left eye fixing. Hypotropia, V pattern, and extorsion are corrected. (From Guyton DL: Strabismus surgery decisions based on torsion findings. In: Anterior Segment and Strabismus Surgery, New York, Kugler, 1997. Modified with permission.)

**Figure 4-22.** Lancaster red-green plot of a 75-year-old patient who developed a left hypertropia after cataract extraction in the left eye. (Solid “red” line = right eye; dashed “green” line = left eye. Small black dots are separated by 15 PD. Large black dots indicate fixation target for nine diagnostic gaze positions.) A, Appearance before strabismus surgery, with right eye fixing. A left hypertropia is present in all gaze directions, with extorsion most notable on left gaze. B, Appearance 1 day postoperatively, after left inferior oblique muscle recession, with right eye fixing. Significant improvement is seen in alignment, with resolution of diplopia. (Modified from Hunter DG, Lam GC, Guyton DL: Inferior oblique muscle injury from local anesthesia for cataract surgery. Ophthalmology 1995;102:501.)
bupivacaine. The patient noted diplopia 6 weeks after cataract surgery. The Lancaster red-green plot, obtained 18 months after surgery, is shown in Figure 4–22. There is a left hypertropia in all gaze directions that could be explained by overaction of either the left SR muscle or the left IO muscle. Note that there is extorsion of the left eye greatest in left gaze—the torsional field of action of the left IO muscle. Fundus examination confirmed 1+ anatomic extorsion of the left eye in primary position. The pattern was most consistent with overaction of the left IO muscle secondary to intramuscular anesthetic injection. In this case, the patient developed a comitant vertical strabismus with no history suggesting a preexisting SO muscle paresis. Although the vertical deviation alone did not allow identification of the affected muscle, torsion findings provided valuable additional information. Recession of the left IO muscle relieved the diplopia. Figure 4–22B shows the postoperative Lancaster red-green plot, which demonstrates resolution of the strabismus.

A 52-year-old woman underwent cataract extraction and intraocular lens implantation in the right eye. Local anesthesia consisted of peribulbar injections of lidocaine and bupivacaine superonasally and inferotemporally. The patient noted diplopia 6 weeks after surgery. The Lancaster red-green plot is shown in Figure 4–23. There is a right hypertropia greatest in downgaze. The most common cause of this vertical pattern is overaction of the right IR muscle. However, note that there is right intorsion greatest in downgaze. The torsional deviation is inconsistent with IR muscle overaction, which causes extorsion. Fundus examination confirmed 2+ intorsion of the right eye that increased in abduction—the torsional field of the SO muscle (Fig. 4–24). These findings are most consistent with overaction of the right SO muscle secondary to intramuscular injection during the superonasal peribulbar block. In this case, failure to assess fundus torsion might have led to inappropriate surgery on the IR muscle. Exaggerated forced ductions, performed at surgery, confirmed tightness of the right SO muscle. A right SO tenotomy (with a “chicken suture”) relieved the vertical and torsional strabismus (see Figs. 4–23B and 4–24).
Conclusion

In patients with strabismus, torsional diplopia is rare (due to multiple levels of sensory adaptation), but anatomic torsion is common and may provide valuable clues to the proper diagnosis. Subjective torsion is best measured with the double Maddox rod test (in primary position) or the Lancaster red-green test (in all nine gaze positions), whereas anatomic (objective) torsion is best appreciated using indirect ophthalmoscopy. Comparison of subjective and anatomic torsion can help determine the time of onset of cyclovertical strabismus. The exaggerated traction test may provide additional information as to the cause of torsional and cyclovertical deviations. In many patients with otherwise confusing or indeterminate vertical deviations, fundus torsion assessment provides the missing piece of the puzzle before surgery, helping to differentiate the many disorders that can cause torsional misalignment. In everyday clinical practice, accurate characterization of fundus torsion is an invaluable tool for the evaluation and management of all cases of vertical strabismus.

REFERENCES

Surgeons are trained to use their skills in the most effective manner while exposing their patients to the least amount of risk. Strabismus surgeons achieve this by employing a precise surgical plan individualized to the amount and type of ocular misalignment and by using the fewest possible surgical interventions. In treating patients with a comitant deviation, most strabismus surgeons use empirical surgical formulas and tables based on the amount of ocular misalignment to determine how much surgery to perform. Accurate determination of the angle of deviation is essential. In this chapter we discuss the many factors that can result in erroneous measurements of ocular deviation and present ways of avoiding or minimizing them. Other factors that influence the response to surgery, including timing, surgical technique, ocular and orbital biomechanics, and individual patient characteristics, are also discussed.

Testing Factors

The basic strabismus deviation is measured while fixation is maintained by the dominant eye, accommodation controlled, and all fusion-vergence eliminated under normal visual environmental conditions. To achieve this, the examiner must control fixation and accommodation and completely suspend fusion. To obtain accurate fixation, the patient must be using the fovea for fixation. Refractive errors should be corrected to maximize visual acuity in each eye.

The underlying principle behind the use of a 20-ft testing distance (estimate used for infinity), wearing of the full hyperopic correction, and providing an accommodative target is to eliminate all influence of accommodation on strabismus measurements. Although this ideal situation cannot be entirely realized, it should be the goal when objectively measuring the strabismic deviation. Unless all accommodation is controlled, variable measurements will be obtained because of varying accommodative efforts required using different distances and targets. In performing the alternate cover test, it is imperative that all fusional vergence mechanisms—divergence and convergence—be suspended while the effects of accommodation are controlled.

TYPE OF TARGET FOR FIXATION

To ensure accurate fixation and eliminate accommodative effort, a target of regard with recognizable and resolvable contours should be presented. The target should have sufficient detail, sustain the patient's interest, and be larger than the patient's threshold acuity. For example, a patient with 20/50 Snellen acuity should be presented with a 20/70 letter as a fixation target. A muscle light does not provide clear contours and is not a suitable fixation target. Deviations can vary by more than 3 PD in as many as 35% of esodeviations and 25% of exodeviations when measurements using accommodative and nonaccommodative targets are contrasted. The patient should wear the maximum hyperopic or least myopic spectacle correction so that no accommodative effort is required at the 20-ft testing distance.

DISTANCE BETWEEN PATIENT AND TARGET

The standard 20-ft test distance is designed to eliminate any meaningful accommodative effort, because only 0.13 D of accommodation is required at this distance in the emme-
tropic patient. The distance at which the fixation target is placed contributes to the amount of deviation measured. Exodeviation was increased by 2 to 40 PD in 34 of 105 patients when the fixation target was moved from 100 to 20 ft.7 The type of exodeviation did not influence the frequency of increased deviation at the increased distance. Kushner23 confirmed the frequent increase in the size of the exotropic deviation when fixing on an accommodative target at distances greater than 20 ft. This was proposed as a frequent cause of undercorrection after surgery for intermittent exotropia, because the target angle for surgery may be underestimated. Shortened examination lanes (less than 20 ft, and 20-ft mirrored systems) have been used by many ophthalmologists. Patients with exodeviations measured significantly less in 10-ft lanes and 20-ft mirrored systems than when measurements were made with the standard 20-ft testing distance. Esotropic patients with a high accommodative convergence/accommodation (AC/A) ratio often exhibited higher distance deviation in 10-ft lanes than in 20-ft lanes. The difference was often greater than 5 PD, which could affect the amount of surgery performed.27

METHOD OF TESTING USED TO OBTAIN MEASUREMENTS

Alternate Cover Tests and Monocular Occlusion

The most effective ways to suspend fusion are the alternate cover test and prolonged monocular occlusion.34, 54 The alternate prism cover test suspends fusional convergence and divergence, permitting measurement of the misalignment to within 1 PD.18 In performing the alternate cover test, the patient is never allowed to regain fusion while the cover is transferred from one eye to another.54

Prolonged monocular occlusion was developed by Marlow24 to uncover the full amount of heterophoria. This is needed to completely eliminate tenacious fusion in patients who have developed strong fusional vergence. Owing to the long duration of occlusion (3 to 7 days), the test is not practical. However, shorter periods of monocular occlusion such as 45 minutes have been effective, especially in patients with exotropia.10, 23, 28 The basic angle of deviation can increase by more than 5 PD after prolonged occlusion in patients with exotropia.23, 28 Many patients with distance-near disparity will have equalization of the distance and near deviations after occlusion.23, 28

Light Reflex Tests

Some patients are too immature to cooperate with alternate cover testing. The deviation then may be estimated using corneal reflex tests such as the Hirschberg55 and Krimsky22 methods. Hirschberg found that each millimeter of decentration of the corneal reflex corresponds to 7 degrees of deviation of the visual axis. Whereas the true relationship between degrees and prism diopters is trigonometric, for angles less than 100 PD, every 2 PD is approximately equal to 1 degree. Later studies using photographic calibration established a conversion factor of 21 PD/mm of displacement.5, 8

Krimsky22 modified the Hirschberg method by placing a prism over the fixing eye so that the light reflex in the deviating eye was centered. When performing the test, the examiner must be in front of the deviated eye to avoid parallax errors in observation.54 Although this improves the precision of corneal light reflex estimation, it prevents the fixing eye from assuming the primary position. The misalignment is measured when the fixing eye is in a secondary position of gaze. For comitant strabismus this will not influence the amount of deviation. For incomitant strabismus, however, dissimilar measurements of the magnitude of deviation are obtained with the fixing eye in the primary and secondary positions. To allow measurement with the fixing eye in primary gaze, the prisms can be placed over the deviating eye (modified Krimsky method). With both techniques it is difficult to observe the corneal light reflection through prisms.

We recommend measuring angle kappa first when using corneal reflection tests in immature patients. An increased prevalence of positive angle kappa is found in esotropia, and an increased prevalence of negative angle kappa is found in exotropia.25 We also suggest coupling an accommodative target to the fixation light.12 For Hirschberg measurements, we correlate each millimeter of decentration with 21 PD of misalignment. When using the Krimsky method, we prefer to place the prisms over the deviated eye.

PRISM PLACEMENT

Errors in quantitative measurements of strabismus can also be induced by improperly placing prisms while measuring the deviation.51 Plastic prisms should be held in the frontal plane rather than the Prentice position (Fig. 5–1). The measurement error introduced by using the Prentice position increases as prisms greater than 20 PD are used. When deviations exceed the amount of the largest available prisms, the examiner should not stack prisms. Light rays cross the interface between the two prisms at a much greater angle than the calibrated angle of incidence, producing a deviation larger than the sum of the labels of the stacked prisms. Unfortunately, it is inaccurate to split the angle of misalignment between the two eyes by holding prisms up in front of each eye. Table 5–1 gives the combined effect of several possible pairs of prisms when held in front of each eye in their calibrated positions. It is obvious that substantial measurement errors occur when a more than 20-PD prism is added in front of each eye.51 Prisms may be stacked, however, if they are used to simultaneously measure vertical and horizontal misalignment, which are independent of each other.21 The prisms’ power is also influenced by the distance prisms are placed in front of the eye. The prisms’ power needed to neutralize a deviation with near fixation will increase as the prisms are held farther from the cornea.52

TORSIONAL DEVIATION

The axes of Fick remind us that the eyes rotate in the vertical, horizontal, and torsional planes. Prisms are unable to correct misalignments around the torsional y-axis. Torsional alignment can be assessed using both subjective and objective techniques. Commonly used subjective measurement methods include the Maddox rod and Bagolini striated lenses.54 Objective measurements can be obtained by indirect
FACTORS INFLUENCING MEASUREMENT AND RESPONSE TO STRABISMSURGERY • 75

MINIMUM DEVIATION POSITION

FRONTAL PLANE POSITION

Figure 5–1. Three common positions for ophthalmic prisms. Left, Prentice position; center, minimum deviation position; right, frontal plane position. (From Thompson JT, Guyton DL: Ophthalmic prisms: Measurement errors and how to minimize them. Ophthalmology 1983;90:204. Reprinted with permission.)

ophthalmoscopy and fundus photography. The synoptophore can neutralize the deviation in all three dimensions. The usefulness of this instrument has been stressed in evaluating patients who are unable to fuse in free space with prisms. Patients who demonstrate preoperative fusion after neutralization of torsion will benefit from surgical correction of the torsional imbalance (see also Chapters 2 and 4).

Patient Factors

ACCOMMODATION

Since the introduction of the concept of accommodative esotropia by Donders, retinoscopy using maximal pharmacologic cycloplegia has been an essential part of modern strabismus management. This sentiment was echoed by Jumpolsky, who reiterated that accommodation must be controlled by uncovering and correcting full hyperopic refractive errors in patients with strabismus. Full cycloplegia with 1% atropine can uncover more than +1.00 D of hyperopia than when using cyclopentolate (see also Chapter 1).

Miotics, in particular echothiophate iodide (Phospholine iodide), have been used to distinguish children whose strabismus can be corrected by spectacles from those who will require surgery despite correction of a hyperopic refractive error. This method has proved ineffective. Glasses reduce the deviation more than echothiophate iodide. We do not recommend using miotics as a diagnostic tool.

The full correction of hypermetropia can have varying results in exodeviations. Classically, correcting hyperopia will worsen an exodeviation. If there is significant blurring of retinal images, however, correcting hypermetropia will improve fusional convergence and lessen the amount of exodeviation. Some patients with exotropia and uncorrected hypermetropia may appear to be esotropic until the refractive error is corrected.

AC/A RATIO

The AC/A ratio refers to the amount of accommodative convergence exerted per unit of accommodation. It may be measured clinically with the gradient method or the

Table 5–1. Actual Deviation in Prism Diopters When One Horizontal Prism is Placed in Front of Each Eye

<table>
<thead>
<tr>
<th>Left Eye Prism (Labeled Value)</th>
<th>10</th>
<th>12</th>
<th>14</th>
<th>16</th>
<th>18</th>
<th>20</th>
<th>25</th>
<th>30</th>
<th>35</th>
<th>40</th>
<th>45</th>
<th>50</th>
</tr>
</thead>
<tbody>
<tr>
<td>10</td>
<td>20</td>
<td>22</td>
<td>24</td>
<td>26</td>
<td>29</td>
<td>31</td>
<td>36</td>
<td>41</td>
<td>47</td>
<td>52</td>
<td>58</td>
<td>63</td>
</tr>
<tr>
<td>12</td>
<td>22</td>
<td>24</td>
<td>26</td>
<td>29</td>
<td>31</td>
<td>33</td>
<td>38</td>
<td>44</td>
<td>49</td>
<td>55</td>
<td>60</td>
<td>66</td>
</tr>
<tr>
<td>14</td>
<td>24</td>
<td>26</td>
<td>29</td>
<td>31</td>
<td>33</td>
<td>35</td>
<td>40</td>
<td>46</td>
<td>52</td>
<td>57</td>
<td>63</td>
<td>69</td>
</tr>
<tr>
<td>16</td>
<td>26</td>
<td>29</td>
<td>31</td>
<td>33</td>
<td>35</td>
<td>37</td>
<td>43</td>
<td>48</td>
<td>54</td>
<td>60</td>
<td>66</td>
<td>72</td>
</tr>
<tr>
<td>18</td>
<td>28</td>
<td>31</td>
<td>33</td>
<td>35</td>
<td>37</td>
<td>39</td>
<td>45</td>
<td>51</td>
<td>57</td>
<td>63</td>
<td>69</td>
<td>75</td>
</tr>
<tr>
<td>20</td>
<td>30</td>
<td>33</td>
<td>35</td>
<td>37</td>
<td>39</td>
<td>42</td>
<td>47</td>
<td>53</td>
<td>59</td>
<td>65</td>
<td>71</td>
<td>77</td>
</tr>
<tr>
<td>25</td>
<td>36</td>
<td>38</td>
<td>40</td>
<td>43</td>
<td>45</td>
<td>47</td>
<td>53</td>
<td>59</td>
<td>66</td>
<td>72</td>
<td>79</td>
<td>85</td>
</tr>
<tr>
<td>30</td>
<td>41</td>
<td>44</td>
<td>46</td>
<td>48</td>
<td>51</td>
<td>53</td>
<td>59</td>
<td>66</td>
<td>73</td>
<td>80</td>
<td>87</td>
<td>94</td>
</tr>
<tr>
<td>35</td>
<td>47</td>
<td>49</td>
<td>52</td>
<td>54</td>
<td>57</td>
<td>59</td>
<td>66</td>
<td>73</td>
<td>80</td>
<td>87</td>
<td>95</td>
<td>103</td>
</tr>
<tr>
<td>40</td>
<td>52</td>
<td>55</td>
<td>57</td>
<td>60</td>
<td>63</td>
<td>65</td>
<td>72</td>
<td>80</td>
<td>87</td>
<td>95</td>
<td>104</td>
<td>113</td>
</tr>
<tr>
<td>45</td>
<td>58</td>
<td>60</td>
<td>63</td>
<td>66</td>
<td>69</td>
<td>71</td>
<td>79</td>
<td>87</td>
<td>95</td>
<td>104</td>
<td>113</td>
<td>123</td>
</tr>
<tr>
<td>50</td>
<td>63</td>
<td>66</td>
<td>69</td>
<td>72</td>
<td>75</td>
<td>78</td>
<td>86</td>
<td>94</td>
<td>103</td>
<td>113</td>
<td>123</td>
<td>133</td>
</tr>
</tbody>
</table>

heterophoria method. The heterophoria method is defined by the equation:

$$\frac{AC}{A} = IPD + \frac{\Delta N - \Delta D}{D}$$

where IPD is the interpupillary distance, $\Delta N$ the alternate prism cover test measurement at near (33 cm), $\Delta D$ the alternate prism cover test measurement at 6 m (20 ft), and $D$ the fixation distance (33 cm) in diopters of accommodation. The gradient method uses concave lenses at 6 m (20 ft) or convex lenses at 33 cm. Lenses are inserted in trial frames in strengths up to 3 D. As soon as the test target is seen, the alternate prism cover test is repeated. The ratio is calculated using the formula:

$$\frac{AC}{A} = \frac{(\Delta + L) - (\Delta - L)}{D}$$

where $\Delta + L$ is the alternate prism cover test measurement with concave (plus) lenses, $\Delta - L$ the alternate prism cover test without lenses, and $D$ the dioptric power of the lenses used. Other methods such as the graphic technique and fixation disparity method may be used but are not discussed in this chapter.\(^5\)

The heterophoria method can result in a falsely elevated AC/A ratio owing to convergence at near and is dependent on the IPD. A study of 39 patients with accommodative esotropia who were followed from childhood to adulthood showed lessening esotropia because of an increased IPD as the face matured.”\(^6\)

The term  **clinically high AC/A ratio** describes patients who show a greater amount of esodeviation at near than at distance.\(^6\) Several unrelated types of strabismus can give the appearance of a “clinically high AC/A ratio.” The patient with V-pattern esotropia should have the near deviation measured in the straight-ahead and not the downgaze position to be sure that the V pattern and accommodation factors are separated. Scott and associates\(^1\) have identified partially accommodative esotropia with greater near than distance deviation who prism-adapt to the amount of esodeviation at near. It is illogical to attribute their distance-near disparity to high AC/A ratio because the deviation can be ameliorated with prisms.

Patients with intermittent esotropia at distance may be orthotropic at near. This lack of exodeviation at near satisfies the criteria for a “clinically high AC/A ratio” and can be misleading.\(^6\) Whereas some patients may have a true high AC/A ratio, Duane\(^6\) found that monocular occlusion resulted in unmasking of exotropia at near and classified this as simulated divergence excess exotropia. Kushner\(^2\) found that these patients may exhibit an increase in the amount of exodeviation at near with +3.00-D lenses and also after monocular occlusion. Kushner does not believe that these patients have a true high AC/A ratio. Rather, the marked increase in esodeviation at near is due to the inability of the convergence fusion mechanism to control the preexisting masked exophoria at near as well as the added amount of exophoria induced by the normal AC/A ratio when viewing through +3.00-D lenses. This extra amount of exophoria prevents the patient from controlling the previously masked exophoria at near. The term  **tenacious proximal fusion** was suggested for patients who have intermittent exotropia at distance and no misalignment at near on the alternate cover test but who exhibit an increased angle of exotropia at near with monocular occlusion.

**HIGH REFRACTIVE ERRORS**

The peripheral prismatic effects of corrective spectacles introduce an artifact when measuring strabismus.\(^4\) Plus lenses decrease, whereas minus lenses increase, the measured deviation (Fig. 5–2). This effect is clinically significant with corrective lenses of more than 5.00 D. Tables 5–2 and 5–3 provide the actual deviation when the measured deviation and power of the spectacle lenses are known.\(^12\) These tables are placed in our examination rooms to facilitate their use. This artifact from the peripheral lens of spectacles may also be reduced by using lenses in trial frames and moving them until they are centered in front of the visual axis of the deviating eye.\(^1\)

**NEUROLOGIC OR ORBITAL DISEASE**

Neurologic or orbital disorders may result in variable strabismus deviations. They include cerebral palsy, myasthenia gravis, and thyroid orbitopathy.\(^3\)

There is a higher than normal frequency of strabismus in patients with cerebral palsy, ranging from 15% to 62%.\(^14\) Developmentally delayed children have an increased effect from the same amount of surgery than do normal children.\(^3\) This happens despite the common use of Krimsky measurements that usually underestimates the deviation compared with prism and cover testing. Bilateral medial rectus recession for esotropia resulted in satisfactory alignment in only 39% of developmentally delayed children, compared with 73% of normal children after a mean follow-up of 24 months.\(^39\) Parents should be informed of the potentially poorer outcome, and perhaps postpone surgery.\(^39\) Dysesthetic or variable strabismus is seen in 30% of strabismic patients with cerebral palsy.\(^6\) While fixing on an accommodative target, the child may exhibit esotropia, followed by orthotropia, and then exotropia. These variable angles are accompanied by slow tonic vergence movements. This type of strabismus does not usually evolve to a more constant devia-
FACTORS INFLUENCING MEASUREMENT AND RESPONSE TO STRABISMUS SURGERY

PLUS LENSES

measured deviation is less than the true deviation

MINUS LENSES

measured deviation is greater than the true deviation

Figure 5–2. The effect of spectacle lenses on the measured deviation ($\Delta_m$) with respect to the true deviation ($\Delta_t$) in horizontal strabismus. Note that plus lenses always decrease the measured deviation and minus lenses always increase the measured deviation. (From Scattergood KD, Brown MH, Guyton DL: Artifacts introduced by spectacle lenses in the measurement of strabismic deviations. Am J Ophthalmol 1983;96:439. Reprinted with permission.)

Although this observation warrants further study, we continue to recommend strabismus surgery only after amblyopia has been fully treated.

ANATOMIC FACTORS

Axial Length and Globe Size

Many authors have sought geometric ocular factors that could alter the response to surgery. The smaller size of neonatal eyes has been invoked as an explanation for poor predictability of surgical results in these cases. Most ocular growth occurs in the postequatorial sclera, half of it during the first 6 months of life. Although the distance from the insertions of the extraocular muscles to the limbus is 80% of that in adults, the insertions are closer to the equator (Fig. 5–3). Three- to 4-mm recession of the horizontal rectus muscles may place the muscles posterior to the equator in a patient younger than 6 months. Axial length measurements correlate closely with the surgical dose-response relation. Larger eyes have a lesser response to the same amount of surgery than do smaller eyes. This correlation, however, was regarded as not clinically important because of the much stronger influence of preoperative deviation. The clinical responses of esotropic patients who had recession of both medial rectus muscles posterior to the equator were analyzed. Three of 28 patients developed consecutive exotropia and underaction of the medial rectus muscles. In all 3 the medial rectus muscles were recessed more than 1.5 mm posterior to the equator. Table 5–4 may be used to estimate the location of the equator based on

CHILDHOOD STRABISMUS

Restoring normality in a child with strabismus means attaining orthotropia, asymptomatic heterophoria in all positions of gaze, bifoveal fixation, normal stereacuity, and normal motor fusion. Unfortunately, there are some types of strabismus that preclude the development of bifoveal fixation after strabismus surgery. Except for anecdotal case reports, the goal of restoring bifoveal fixation in children with infantile esotropia has proved elusive. A subnormal degree of binocularity can nonetheless be obtained in the majority of patients with infantile esotropia. Fortunately, children with acquired esotropia or intermittent exotropia have the potential to completely regain bifoveal fixation.

AMBYLOPIA

Patients with amblyopia have a greater chance of postoperative overcorrection and undercorrection after strabismus surgery. In one study no difference was found in surgical outcome between patients having surgery after amblyopia therapy was completed and others operated on during amblyopia therapy (as long as treatment continued after surgery).
<table>
<thead>
<tr>
<th>Measured Tropia (PD)</th>
<th>-1</th>
<th>-2</th>
<th>-3</th>
<th>-4</th>
<th>-5</th>
<th>-6</th>
<th>-7</th>
<th>-8</th>
<th>-9</th>
<th>-10</th>
<th>-12</th>
<th>-15</th>
<th>-20</th>
<th>-30</th>
</tr>
</thead>
<tbody>
<tr>
<td>5</td>
<td>5</td>
<td>5</td>
<td>5</td>
<td>4</td>
<td>4</td>
<td>4</td>
<td>4</td>
<td>4</td>
<td>4</td>
<td>4</td>
<td>4</td>
<td>4</td>
<td>3</td>
<td>3</td>
</tr>
<tr>
<td>10</td>
<td>10</td>
<td>10</td>
<td>9</td>
<td>9</td>
<td>9</td>
<td>9</td>
<td>8</td>
<td>8</td>
<td>8</td>
<td>8</td>
<td>7</td>
<td>7</td>
<td>6</td>
<td></td>
</tr>
<tr>
<td>15</td>
<td>15</td>
<td>14</td>
<td>14</td>
<td>13</td>
<td>13</td>
<td>13</td>
<td>12</td>
<td>12</td>
<td>12</td>
<td>12</td>
<td>11</td>
<td>10</td>
<td>9</td>
<td></td>
</tr>
<tr>
<td>20</td>
<td>20</td>
<td>19</td>
<td>19</td>
<td>18</td>
<td>18</td>
<td>17</td>
<td>17</td>
<td>16</td>
<td>16</td>
<td>15</td>
<td>15</td>
<td>13</td>
<td>11</td>
<td></td>
</tr>
<tr>
<td>25</td>
<td>24</td>
<td>24</td>
<td>23</td>
<td>22</td>
<td>22</td>
<td>21</td>
<td>21</td>
<td>20</td>
<td>20</td>
<td>19</td>
<td>18</td>
<td>17</td>
<td>14</td>
<td></td>
</tr>
<tr>
<td>30</td>
<td>29</td>
<td>29</td>
<td>28</td>
<td>27</td>
<td>27</td>
<td>26</td>
<td>26</td>
<td>25</td>
<td>24</td>
<td>23</td>
<td>22</td>
<td>20</td>
<td>17</td>
<td></td>
</tr>
<tr>
<td>35</td>
<td>34</td>
<td>33</td>
<td>33</td>
<td>32</td>
<td>31</td>
<td>30</td>
<td>30</td>
<td>29</td>
<td>28</td>
<td>27</td>
<td>25</td>
<td>23</td>
<td>20</td>
<td></td>
</tr>
<tr>
<td>40</td>
<td>39</td>
<td>38</td>
<td>37</td>
<td>36</td>
<td>36</td>
<td>35</td>
<td>34</td>
<td>33</td>
<td>33</td>
<td>32</td>
<td>31</td>
<td>29</td>
<td>26</td>
<td>23</td>
</tr>
<tr>
<td>45</td>
<td>44</td>
<td>43</td>
<td>42</td>
<td>41</td>
<td>40</td>
<td>39</td>
<td>38</td>
<td>37</td>
<td>37</td>
<td>36</td>
<td>35</td>
<td>33</td>
<td>30</td>
<td>26</td>
</tr>
<tr>
<td>50</td>
<td>49</td>
<td>48</td>
<td>47</td>
<td>45</td>
<td>44</td>
<td>43</td>
<td>43</td>
<td>42</td>
<td>41</td>
<td>40</td>
<td>38</td>
<td>36</td>
<td>33</td>
<td>29</td>
</tr>
<tr>
<td>60</td>
<td>59</td>
<td>57</td>
<td>56</td>
<td>55</td>
<td>53</td>
<td>52</td>
<td>51</td>
<td>50</td>
<td>49</td>
<td>48</td>
<td>46</td>
<td>44</td>
<td>40</td>
<td>34</td>
</tr>
<tr>
<td>70</td>
<td>68</td>
<td>67</td>
<td>65</td>
<td>64</td>
<td>62</td>
<td>61</td>
<td>60</td>
<td>58</td>
<td>57</td>
<td>56</td>
<td>54</td>
<td>51</td>
<td>46</td>
<td>40</td>
</tr>
</tbody>
</table>

*To account for the prismatic effects of myopic spectacles, use the measured tropia value (y-axis), and the myopic spectacle power (x-axis), to determine the actual deviation.
Table 5-3. Artifact From Peripheral Prismatic Effects: Hyperopic Spectacle Power*

<table>
<thead>
<tr>
<th>Measured Tropia (PD)</th>
<th>+1</th>
<th>+2</th>
<th>+3</th>
<th>+4</th>
<th>+5</th>
<th>+6</th>
<th>+7</th>
<th>+8</th>
<th>+9</th>
<th>+10</th>
<th>+12</th>
<th>+15</th>
<th>+20</th>
<th>+30</th>
</tr>
</thead>
<tbody>
<tr>
<td>5</td>
<td>5</td>
<td>5</td>
<td>5</td>
<td>6</td>
<td>6</td>
<td>6</td>
<td>6</td>
<td>6</td>
<td>6</td>
<td>7</td>
<td>7</td>
<td>8</td>
<td>10</td>
<td>20</td>
</tr>
<tr>
<td>10</td>
<td>10</td>
<td>11</td>
<td>11</td>
<td>11</td>
<td>12</td>
<td>12</td>
<td>13</td>
<td>13</td>
<td>13</td>
<td>14</td>
<td>16</td>
<td>20</td>
<td>40</td>
<td></td>
</tr>
<tr>
<td>15</td>
<td>15</td>
<td>16</td>
<td>16</td>
<td>17</td>
<td>18</td>
<td>18</td>
<td>19</td>
<td>19</td>
<td>20</td>
<td>21</td>
<td>24</td>
<td>30</td>
<td>60</td>
<td></td>
</tr>
<tr>
<td>20</td>
<td>21</td>
<td>21</td>
<td>22</td>
<td>22</td>
<td>23</td>
<td>24</td>
<td>24</td>
<td>25</td>
<td>26</td>
<td>27</td>
<td>29</td>
<td>32</td>
<td>40</td>
<td>80</td>
</tr>
<tr>
<td>25</td>
<td>26</td>
<td>26</td>
<td>27</td>
<td>28</td>
<td>29</td>
<td>29</td>
<td>30</td>
<td>31</td>
<td>32</td>
<td>33</td>
<td>36</td>
<td>40</td>
<td>50</td>
<td>100</td>
</tr>
<tr>
<td>30</td>
<td>31</td>
<td>32</td>
<td>32</td>
<td>33</td>
<td>34</td>
<td>35</td>
<td>36</td>
<td>38</td>
<td>39</td>
<td>40</td>
<td>43</td>
<td>48</td>
<td>60</td>
<td>120</td>
</tr>
<tr>
<td>35</td>
<td>36</td>
<td>37</td>
<td>38</td>
<td>39</td>
<td>40</td>
<td>41</td>
<td>42</td>
<td>44</td>
<td>45</td>
<td>47</td>
<td>50</td>
<td>56</td>
<td>70</td>
<td>140</td>
</tr>
<tr>
<td>40</td>
<td>41</td>
<td>42</td>
<td>43</td>
<td>44</td>
<td>46</td>
<td>47</td>
<td>48</td>
<td>50</td>
<td>52</td>
<td>53</td>
<td>57</td>
<td>64</td>
<td>80</td>
<td>160</td>
</tr>
<tr>
<td>45</td>
<td>46</td>
<td>47</td>
<td>49</td>
<td>50</td>
<td>51</td>
<td>53</td>
<td>55</td>
<td>56</td>
<td>58</td>
<td>60</td>
<td>64</td>
<td>72</td>
<td>90</td>
<td>180</td>
</tr>
<tr>
<td>50</td>
<td>51</td>
<td>53</td>
<td>54</td>
<td>56</td>
<td>57</td>
<td>59</td>
<td>61</td>
<td>63</td>
<td>65</td>
<td>67</td>
<td>71</td>
<td>80</td>
<td>100</td>
<td>200</td>
</tr>
<tr>
<td>60</td>
<td>62</td>
<td>63</td>
<td>65</td>
<td>67</td>
<td>69</td>
<td>71</td>
<td>73</td>
<td>75</td>
<td>77</td>
<td>80</td>
<td>87</td>
<td>96</td>
<td>120</td>
<td>240</td>
</tr>
<tr>
<td>70</td>
<td>72</td>
<td>74</td>
<td>76</td>
<td>78</td>
<td>80</td>
<td>82</td>
<td>85</td>
<td>88</td>
<td>90</td>
<td>93</td>
<td>100</td>
<td>112</td>
<td>140</td>
<td>280</td>
</tr>
</tbody>
</table>

*To account for the prismatic effects of hyperopic spectacles, use the measured tropia value (y-axis), and the hyperopic spectacle power (x-axis), to determine the actual deviation. From Hansen VC. Common pitfalls in measuring strabismic patients. Am Orthopt J 1989;39:3. Reprinted with permission.
Muscle Length-Tension Properties

Beisner theorized that the effect of rectus muscle resections is caused by alteration of the length-tension curve. Passive length-tension studies of the horizontal rectus muscles found no relationship between preoperative deviation and the response to surgery in patients with exotropia and infantile esotropia. A relationship was, however, found between length-tension properties, the preoperative deviation, and the response to surgery in acquired esotropia. Future understanding of length-tension relationships in the awake, alert patient should lead to more accurate surgery.

Surgical Factors

Timing. Patients who achieve satisfactory alignment before age 2 years have a better binocular outcome than those whose eyes are aligned after this time. It should be noted, however, that even adults with infantile esotropia who have never undergone previous surgical correction can develop subnormal binocularity after surgery. In patients with accommodative esotropia, a positive correlation was found between the development of normal binocularity and prompt correction of strabismus soon after its onset. Surgery should not be delayed in a child or adult who is medically stable and has a reproducible angle of deviation.
Table 5-4. Estimated Location of the Equator Underneath the Medial Rectus Muscle Based on Axial Length and Corneal Diameter

<table>
<thead>
<tr>
<th>Axial Length (mm)*</th>
<th>Corneal Diameter (mm)*</th>
</tr>
</thead>
<tbody>
<tr>
<td>10.00</td>
<td>10.50</td>
</tr>
<tr>
<td>17.00</td>
<td>8.60</td>
</tr>
<tr>
<td>17.50</td>
<td>8.98</td>
</tr>
<tr>
<td>18.00</td>
<td>9.35</td>
</tr>
<tr>
<td>18.50</td>
<td>9.73</td>
</tr>
<tr>
<td>19.00</td>
<td>10.11</td>
</tr>
<tr>
<td>19.50</td>
<td>10.48</td>
</tr>
<tr>
<td>20.00</td>
<td>10.86</td>
</tr>
<tr>
<td>20.50</td>
<td>11.24</td>
</tr>
<tr>
<td>21.00</td>
<td>11.62</td>
</tr>
<tr>
<td>21.50</td>
<td>11.99</td>
</tr>
<tr>
<td>22.00</td>
<td>12.37</td>
</tr>
<tr>
<td>22.50</td>
<td>12.75</td>
</tr>
<tr>
<td>23.00</td>
<td>13.13</td>
</tr>
<tr>
<td>23.50</td>
<td>13.51</td>
</tr>
<tr>
<td>24.00</td>
<td>13.89</td>
</tr>
<tr>
<td>24.50</td>
<td>14.28</td>
</tr>
<tr>
<td>25.00</td>
<td>14.66</td>
</tr>
<tr>
<td>25.50</td>
<td>15.04</td>
</tr>
<tr>
<td>26.00</td>
<td>15.42</td>
</tr>
</tbody>
</table>

Based on formula:

\[
\text{Limbus to equator (in mm)} = 2 \frac{\pi}{360} r \left( \frac{\beta}{2} \right)
\]

where \( r \) = radius of globe and

\[
\beta = \text{angle subtended by limbus—equator arc}
\]

and \( r = \frac{c^2 + (A + t - a, \alpha - c)^2}{2(A + t - a, \alpha - c)} \)

*Assumes \( K = 43.50 \); scleral thickness = 1 mm.


Figure 5-4. The difference between cord (b) and arc (a) measurements from the limbus. (From Scott WE, Martin-Casal A, Braverman DE: Curved ruler for measurement along the surface of the globe. Arch Ophthalmol 1978;96:1084. American Medical Association. Reprinted with permission.)

Figure 5-5. A, Comparison of the different measuring instruments: (1) calipers, (2) Scott curved ruler, and (3) prototype small curved ruler. The inset illustrates the different radius of curvature for the standard Scott curved ruler (11.0 mm) versus the prototype small curved ruler (9.5 mm). The white arrow indicates the point on the end of the prototype small curved ruler used to mark the sclera. B, Intraoperative measurements to recess the left medial rectus muscle 5.0 mm in a 6-month-old boy with esotropia. Prior to disinsertion, the distance from the corneoscleral limbus to a muscle hook pulled taut behind the medial rectus insertion was measured as 5.5 mm. After disinsertion, the asterisk shows the mark 5.5 mm posterior to the corneoscleral limbus, almost 2 mm behind the residual stump of the postinsertional stump. The desired 5.0 mm recession was added to the measured distance between the medial rectus insertion and the corneoscleral limbus prior to disinsertion, 5.5 mm, for a total measurement of 10.5 mm posterior to the corneoscleral limbus. Points 1, 2, and 3 represent the marks made by calipers, the Scott curved ruler, and the prototype small curved ruler, respectively, measuring 10.5 mm from the corneoscleral limbus.
Artifacts Introduced in Surgery. The strabismus surgeon may introduce artifacts during surgery. The anterior extent of the medial rectus insertion retracts toward the corneoscleral limbus after disinsertion (Fig. 5–6). This can result in more than a millimeter of anterior displacement of the insertion site. Many surgeons measure the distance of the anterior aspect of the insertion before disinserting the muscle from the globe. The muscle can then be recessed a predetermined amount posterior to the original site of the insertion before anterior displacement (by measuring recession from the limbus). Surgeons may also inadvertently distort the insertion site by applying traction on the globe during suture placement. This produces a V-shaped deformity, which results in anterior displacement of the insertion. If the amount of recession is measured from the anteriorly displaced V deformity, less recession will be performed than was planned (Fig. 5–7).

Sutures should be placed no more than a millimeter from the end of the insertion. Placing the sutures too far posterior will impose a resection effect on a recessed muscle.

Conclusions

For patients with comitant strabismus, empirical formulas and tables are used by a majority of strabismus surgeons. The surgeon can most accurately apply these guidelines by obtaining an accurate assessment of the patient’s preoperative misalignment. This is best performed using the appropriate testing conditions and measurement techniques to uncover the basic deviation.

The strongest determinants of the response to surgery are the type of strabismus and the preoperative deviation. The interval between the onset of strabismus and initiation of treatment may also influence the outcome. The surgeon can minimize the variability of the response to surgery by standardizing the surgical technique and through awareness of artifacts that may be introduced during the operative procedure.

REFERENCES

FACTORS INFLUENCING MEASUREMENT AND RESPONSE TO STRABISMUS SURGERY

In complex cases of strabismus, even a thorough clinical examination may not lead to a correct diagnosis and treatment plan. Orbital imaging has particular value for clarifying congenital anatomic anomalies, traumatic injuries, complications of endoscopic surgery, and orbital masses.

What kind of information can orbital imaging provide? Modern high-resolution imaging can demonstrate the path of a lost or detached extraocular muscle (EOM), an extirpated or avulsed segment of EOM, entrapment of an EOM or connective tissue in an orbital fracture, tumorous swelling or infiltration of EOMs and other orbital tissues, and supernumerary or absent EOMs. We have been particularly interested in imaging EOM cross sections as an index of elasticity and to appreciate changes in cross section with gaze as an index of contractility, as well as abnormal EOM paths.

History of Orbital Imaging

Orbital imaging was first used in strabismus to determine EOM paths, which were needed to mathematically model the mechanics of binocular alignment. Imaging protocols for orbital tissues were developed initially for plain film radiography in monkeys and radiographic computed tomography (CT) in humans. These studies yielded the surprising finding that the paths of the rectus EOM bellies remain stable relative to the orbit despite shifts in gaze. Magnetic resonance imaging (MRI) with three-dimensional reconstruction also showed the paths of normal rectus EOMs to be stable throughout the oculomotor range and that, during contraction, EOM cross-sectional area increases and the point of the maximum cross section shifts toward the origin of the EOM. Thus, noninvasive MRI can provide an index of functional EOM contractility. If tissue composition can be assumed to be normal, cross sections in the primary position are an index of EOM elasticity.

Qualitative MRI evidence of superior oblique (SO) atrophy was initially reported in trochlear palsy. High-resolution MRI then exploited improved orbital imaging protocols to demonstrate normal changes in SO cross section with gaze and the atrophy and loss of gaze-related contractility occurring in SO palsy. Interestingly, many cases of “SO palsy,” diagnosed by the familiar “three-step test,” do not exhibit SO abnormalities on MRI. Furthermore, this finding has been replicated by several investigators. Correlations of MRI with surgical exploration of the reflected SO tendon show that laxity of the tendon, occasionally observed during surgery for SO palsy, is secondary to atrophy of the muscle belly even in congenital cases. Lateral rectus atrophy has been demonstrated in abducens palsy. Reduction in the volume of EOMs innervated by a paretic oculomotor nerve may be demonstrated by MRI. “Dynamic MRI,” with imaging repeated during fixation on targets in multiple directions of gaze, has been used to determine the contractility of normal and injured EOMs in traumatic strabismus.

Principles and Techniques

GENERAL PRINCIPLES OF ORBITAL IMAGING

High-quality orbital images may be obtained with most modern scanners. Although interest may focus on one orbit, it may be best to image both for comparison. To judge the orientation of orbital structures relative to the head, one must include in the image a head landmark such as the interhemispheric fissure. An MRI scan typically takes several minutes to acquire, during which time patient motion will degrade the images. Image space is fixed relative to the scanner, so that patient movement results in “partial volume averaging” and has the effect of smearing moving tissues such as the globe and EOMs across the image. Any substantial body movement, even of the arms or legs, can disturb the magnetic field and degrade the image of immobile tissues. Fixation targets must be provided and the patient instructed to fix them accurately during the study. Pulse sequences should be selected so as to minimize scanning time and the...
motion artifacts caused by blinks and fixation instability. Image planes that best demonstrate the orbital structures of interest should be chosen; in most cases, this means that closely spaced coronal image planes are acquired using a surface coil.

An MRI “scan” yields a set of rectangular images, each representing a parallel “slab” of tissue that is summed along the thickness of the slab. The term dynamic MRI has been used to describe several such sets of static images, each acquired with the eyes in a different gaze position and assembled into a movie (“cine”) to highlight gaze-related changes in orbital structures. Scanning does not differ from ordinary MRI, and the term dynamic is a misnomer. The phrase “fast MRI” has been used to refer to a distinctive MRI technique in which the positions of the moving eyes are coordinated with image acquisition at rates up to 5 images/sec to yield “stop-action” pictures. Thus, fast MRI samples extraocular dynamics. The technique is challenging, and image resolution is modest. Truly unique information could be derived from fast MRI if it were feasible with fast saccadic eye movements, during which one would expect to see large changes in EOM paths and cross sections.

Orbital imaging, like other modern tomographic methods, builds images from signals emitted by voxels, which are small volume elements of tissue. The intersection of a voxel with the image or slice plane is a pixel, or picture element. Pixels are usually small, yielding high resolution in the plane of the slice. The third voxel dimension is the plane thickness, which is usually much larger, yielding low resolution orthogonal to the slice plane. Each voxel is assigned an intensity value derived from acquired x-ray (CT) or radio wave (MRI) signals. A modern orbital image slice is commonly 3 mm thick with a 10-cm field of view (FOV) and is divided into 256 × 256 pixels, giving an elementary voxel of 390 × 390 × 3000 μm. Better resolution (i.e., smaller voxels) is obtainable by reducing the FOV (e.g., to 4 cm on the GE Horizon scanner), increasing matrix density (e.g., to 512 × 512), and reducing slice thickness (e.g., to 2 mm on the GE Horizon Scanner). The 3-T research MRI scanner (available at the University of California at Los Angeles) is expected to achieve slice-plane resolution of better than 78 μm and a slice thickness less than 1 mm, providing near-microscopic resolution. Modern CT scanners employ even smaller voxels. However, because the signal-to-noise ratio (SNR) varies linearly with voxel volume, small voxels yield weak signals. Images with a low SNR appear grainy. Repeated MRI excitations and signal averaging improve the SNR, ideally by the square root of the number of repetitions (excitations), but at the cost of increased scan time. Unfortunately, long scan times increase the likelihood of motion artifacts, particularly in orbital imaging. Increasing the radiation intensity or exposure time may improve CT resolution but, in turn, raises a safety issue.

Attention to a few technical aspects can dramatically improve the quality of orbital imaging using the equipment found in nearly every clinical center. The first such issue is head positioning: the patient’s head must not move during imaging, even if images are acquired in multiple directions of gaze. It is usually sufficient to stabilize the head of a cooperative patient with pads and tape. In special situations a dental fixation device might be considered. As explained earlier, the best resolution is obtained with features of interest in the scan plane. If we are interested in EOM cross sections, for instance, their long axes should be at least roughly perpendicular to the scan plane. To image rectus EOMs and the SO muscle together, a good compromise is the quasi-coronal plane perpendicular to the axis of the orbit, as illustrated in Figure 6–1. Of course, this requires scanning each orbit separately. To scan both orbits together (e.g., to determine their relative orientations), the head must be oriented vertically. This places both the SO and medial rectus (MR) cross sections optimally in the scan plane but the lateral rectus (LR) cross section in an oblique plane. Spiral CT scanning in a true coronal plane requires careful attention to patient positioning to image the orbits.

The second major technical issue in orbital imaging is gaze control. To define EOM contractility over a range of gaze angles, each must be defined and maintained with fixation targets (see Fig. 6–1). In any case, control of fixation is necessary to minimize movement artifacts. It is necessary to remind the patient to maintain the gaze stable and the body immobile during imaging and to make clear which fixation target is to be used at each phase of imaging. Eye closure does not eliminate eye movement. MRI technicians must communicate with the patient over the intercom be-
between image acquisitions to ensure stable head position and proper fixation. Accurate fixation cannot be maintained without fixation fixation, which must be placed within the scanner. The nonfixing eye should be occluded, since the proximity of fixation targets might otherwise result in diplopia and alternating fixation. Each patient’s range of comfortable, stable fixation must be individually recognized, recalling that maintenance of stable gaze for a prolonged period can be fatiguing for all and is particularly difficult in the presence of certain disorders. Straight-ahead fixation can seldom be maintained for longer than 4 to 5 minutes, which is a limiting time for a single MRI scan. To achieve stable fixation, it is necessary to minimize factors that stimulate blinking; contact lenses must be removed and air exchange in the scanner minimized to reduce corneal drying. Eccentric targets usually cannot be placed farther than 20 to 25 degrees from primary position. Even normal young individuals develop intolerable gaze nystagmus and oscillopsia during prolonged fixation at larger eccentricities. Elevation is the most difficult eccentric position for most patients. Images should be viewed as they are acquired so that acquisitions can be repeated if necessary. CT scanning requires less time than MRI; coronal imaging of the whole orbit in 1-mm image planes is possible within 12 to 30 seconds. In children or other subjects who cannot maintain stable gaze because of sedation or lack of comprehension, the deep orbital paths of the EOMs, which are not significantly influenced by gaze direction and which have excellent intrinsic contrast against the orbital fat, can probably be best imaged clinically using high-resolution CT.

A third technical issue in orbital imaging is optimizing resolution. It is important to use as large a pixel matrix as possible. To maximize useful resolution the FOV of the CT or MRI scanner should be reduced to a size just large enough to contain the orbit or orbits of interest. High-quality orbital CT scans or MRI scans can be set to an FOV of 10 cm or less. The 20- to 25-cm FOV typically used for brain imaging is certainly suboptimal for EOM imaging. Image slice thickness should be minimized; high-quality clinical CT images are currently 1 mm thick, and high-quality MRI scans are 2 to 3 mm thick. Because between-slice resolution is poor to begin with, it is best to have no gap between MRI “slabs,” but a gap of 1 to 2 mm is acceptable for CT. MRI is greatly enhanced by surface coils, which are placed on the face to maximize reception of the radio signals emitted by orbital tissues. By improving the SNR, surface coils dramatically increase resolution by reducing the size of voxels that can be employed in orbital imaging without excessive image graininess. Surface coils also restrict the depth of tissue imaging to a distance approximately equal to the diameter of the coil, minimizing image artifacts arising from more distant tissues. A 3-inch-diameter surface coil is optimal for orbital imaging, because it has ideal depth of penetration and a nearly optimal SNR. One or two coils may be placed over the orbits so that the patient can view through the center of each coil. Modern MRI scanners (e.g., recent revisions of the General Electric 1.5-T Signa) allow two 3-inch coils to be used as a phased array, receiving signals from alternate tissue excitations and imaging both orbits simultaneously with an even better SNR. The general-purpose surface coils supplied with most scanners may be used for orbital imaging. Special-purpose surface coil arrays are under development for orbital imaging, promising further improvements in SNR and greater clinical convenience.

**TECHNIQUE OF ORBITAL MAGNETIC RESONANCE IMAGING**

Consider the following example of an MRI procedure designed to obtain quasi-cross-sectional images of the rectus EOMs in one eye. First, the range of comfortable fixation for the scanned eye is determined and perhaps five targets are affixed, centered inside the scanner at the top, bottom, left, right, and center of that range. Because the bore of the scanner is narrow, precise target location will depend both on the position of the scanner gantry and the sagittal angle of the patient’s head. The effect of the scanner table position can be controlled by consistently setting the table at landmark position with the projected crossed reference light beams at the midcorneal position of the scanned eye. It is useful if the technician or physician first lies supine in the gantry position to be assumed by the patient during scanning, and affixes targets centered on straight-ahead gaze. Bright-colored adhesive disks containing a central black cross work well. The nonfixing eye is patched. The patient then is positioned supine on the scanner table. The patient is instructed to close the scanned eye and turn the head away from the scanned eye by about 23 degrees, so that the center of the closed eye is directly beneath the crossed reference light beams. Noise-attenuating earphones are placed in the patient’s ears, and the patient’s head is stabilized with wedge-shaped foam pads and adhesive tape. A surface coil is placed around the scanned eye, without blocking any of the targets, and held in place with adhesive tape. Finally, the scanner table is moved to its operating position and the patient is asked to verify the centration and visibility of the target array. This is a crucial step, because the short viewing distance to the targets on the scanner bore converts small linear positioning errors into large gaze-angle errors and because it is very difficult to maintain uniform sagittal head positioning among subjects. The position of the adhesive targets may be adjusted if necessary before scanning. The ventilator fan in the scanner is adjusted to the lowest air flow setting to minimize blinking.

Typical scanner settings for the preferred T1-weighted imaging sequence are summarized in Table 6–1. The T1 pulse sequence displays the orbital fat as bright and the globe, connective tissues, and muscles as dark. To optimally place the high-resolution image planes relative to the orbits, a brief (40–90 seconds) axial localizer scan is done at low resolution and a large enough FOV (typically 12 × 12 cm) so that it can be centered on the orbit of interest. Because even the localizer scan can provide useful anatomic information if eye movement is avoided, the axial localizer is usually performed with fixation in primary gaze. An axial image through the plane of the horizontal rectus EOMs is acquired as a guide to placing a set of contiguous 3-mm-thick, medium-resolution coronal image planes that span the anteroposterior extent of the orbit from just anterior to the equator to the annulus of Zinn. This requires 12–13 image planes of 3-mm thickness in most cases. Finally, the desired quasi-coronal images are acquired at high resolution with a smaller FOV (typically 8 × 8 cm), taking additional time for 1.5 to 2 excitations to improve the SNR. A T1-weighted pulse
sequence requiring 3.5 minutes provides a set of 13 contiguous, 3-mm-thick coronal images in two excitations with an FOV of 8 cm and a matrix of 256 × 256 pixels. This provides resolution of 312 μm in scan planes and an image that is generally free of noticeable noise. To reduce acquisition time, the direct coronal scan plane is used instead of the arbitrary oblique plane setting. The patient is asked to rest with the eyes closed, and scanning is repeated at another fixation. Imaging the inferior oblique (IO) muscle, if indicated, is best achieved by separate image acquisitions in the sagittal plane using otherwise identical scanner settings. Sagittal image planes are also located using a coronal localizer scan.

Qualitative and quantitative scan interpretation is best done by a strabismologist familiar with the patient. If the images are printed on film, the magnification and centration should be consistent across gaze positions to accurately compare the locations and sizes of EOMs and other orbital structures. Films may be superimposed against a lighted viewer to highlight gaze-related changes in EOM position and cross-sectional area. The scanner’s analysis console may be used to measure locations and cross-sectional areas with the image still in digital format. Image analysis software is available to automatically determine the cross-sectional areas and area centroid coordinates of EOMs after these have been manually outlined using a computer cursor. Quantification may not be necessary for clinical purposes.

### TECHNIQUE OF ORBITAL COMPUTED TOMOGRAPHY

Computed x-ray tomography has benefited from recent improvements in both quality and utility for studying the EOMs. Conventional CT scanners rotate the x-ray source and one or more detectors around the patient in one plane at a time. Spiral CT scanners rotate the x-ray source and numerous detectors as the scanner moves the patient, producing a spiral path around the long axis of the patient. CT can achieve excellent resolution in arbitrarily chosen planes. The General Electric CT/i spiral scanner can produce contiguous 512 × 512 coronal orbital images with a slice thickness of 1.0 mm, spanning the orbit in 12 to 15 seconds. This substantially exceeds the speed and resolution of MRI. Drawbacks of x-ray methods include low contrast of EOMs against the sclera and orbital tissues and x-ray exposure of the lens and lacrimal gland. Spiral CT can estimate the contractile and trophic states of extraocular muscles and may be preferred by patients who are claustrophobic or cannot tolerate a prolonged study. Bony relationships are probably better demonstrated by CT than by MRI.

### Functional Anatomy of Normal Extraocular Muscles

The paths of the rectus EOM bellies are remarkably stable in the orbit over the full range of gaze. The MR images in Figure 6–2A show a normal orbit in the five cardinal positions of gaze. Sagittal images in primary gaze, upgaze, and downgaze demonstrate characteristic bowing of the relaxed rectus EOMs toward the orbital wall, likely due to the traction of pulley tissues and the pressure of compartmentalized orbital fat (see Fig. 6–2B). Rectus EOMs pass through pulleys, structures composed of connective tissue and smooth muscle that are coupled to the orbital wall and located approximately in a coronal plane just behind the equator of the globe. With respect to determining pulling directions the pulleys act as rectus muscle origins, and only the anterior muscle segments between the pulleys and insertions move with the eye. Pulley locations may be estimated from the area centroids of EOM cross sections in a plane about 3 mm anterior to the junction of the globe and optic nerve. It is difficult to discriminate anterior tendinous EOMs from the adjacent underlying sclera, and posterior slices give poor estimates of pulley position because of the conical shape of the orbit. An area centroid is the "center of mass" of a two-dimensional cross section, reasonable here as its effective location.

In normal subjects, this analysis yields the remarkable finding that pulley coordinates (Fig. 6–3, Table 6–2) are highly uniform among subjects. The 95% confidence intervals for rectus muscle pulley locations in cross-sectional planes are less than ± 1.9 mm. Many strabismus patients have normal pulleys but others do not; and, in some of the latter, pulley abnormalities account for the strabismus. Precise pulley locations are crucial for correct binocular fixation in three-dimensional space and mislocalization of the pulleys is a cause of incomitant strabismus. Three-dimensional reconstructions of high-resolution MRI data vividly show the stability of rectus EOM paths relative to the orbit over the largest sustainable gaze shifts (Fig. 6–4). The only structure seen to move is the junction of the globe with the optic nerve. (Of course the tendinous muscle insertions move with the globe but they are usually not discriminable as we have explained.)

The stability of posterior EOM paths in the orbit is a consequence of pulley stiffness. Nevertheless, unlike the cartilaginous trochlea, rectus pulleys are soft tissue structures and move slightly with changing gaze. Average pulley shifts with gaze are plotted in Figure 6–3. The inferior rectus (IR) pulley remains relatively immobile with horizontal gaze.
Figure 6–2. MRI scan of a normal orbit. A, Images of the eye in straight ahead gaze and in approximately 20 degrees of cardinal eccentric gaze. Imaging planes are perpendicular to the orbital axis. Paths of the rectus extraocular muscle bellies are stable. The dotted line shows the plane of sagittal imaging for B. B, Sagittal images in primary gaze, upgaze, and downgaze. Note the curved or bowed paths of the vertical rectus extraocular muscles, particularly where the muscle is relaxed. The dotted line indicates the anterior border of the inferior oblique (IO) in upgaze. Note the posterior shift of the IO in downgaze. SO, superior oblique; SR, superior rectus; IR, inferior rectus; ON, optic nerve; UPAB, up abduction; UPAD, up adduction.
Figure 6–3. Average positions of pulleys of rectus extraocular muscles (relative to right orbital center and viewed as if facing the subject) for normal subjects in a plane 3 mm anterior to the globe/optic nerve junction in upgaze, downgaze, abduction, adduction, and primary position. Note the nearly complete stability of the medial rectus (MR) pulley. The inferior rectus (IR) pulley is seen to shift medially in upgaze and laterally in downgaze, probably reflecting coupling of the IR to the inferior oblique in Lockwood ligament. The lateral rectus (LR) shifts up in downgaze and down in upgaze, probably reflecting the mutual coupling of the superior rectus (SR) and LR in the lateral levator aponeurosis. Error bands represent ± 2 SD (95% confidence intervals). (Redrawn from Clark RA, Miller JM, Demer JL: Location and stability of rectus muscle pulleys inferred from muscle paths. Invest Ophthalmol Vis Sci 1997;38:227.)

Figure 6–4. Three-dimensional reconstruction of the globe (GL), optic nerve (ON), and rectus extraocular muscles (SR, IR, LR, MR) in mid orbit in the nine cardinal positions of gaze. Images were obtained by averaging coronal magnetic resonance images of four normal subjects. Note that the mid-orbital paths of the rectus extraocular muscles do not shift noticeably relative to the orbit with these large gaze shifts; only the junction of the optic nerve with the globe appears to move. The unseen tendinous insertions of the rectus muscles, anterior to the pulleys, move with the globe during gaze shifts. Abbreviations as in Figure 6–1. (From Miller JM: Functional anatomy of normal human rectus muscles. Vision Res 1989;29:223. Reprinted with permission from Elsevier Science.)
shifts but moves superiorly as it straightens in downgaze and bows inferiorly in elevation, as expected. We did not expect to find horizontal displacement, but the IR pulley shifts medially 1.1 mm in upgaze and laterally 1.3 mm in downgaze. Perhaps the coupling between the IR and IO, called Lockwood ligament, provides an explanation: contracting in upgaze, the IO draws its belly medially and pulls the IR pulley in the same direction, whereas in downgaze, relaxation of the IO allows the IR to relax laterally.

The IO pulley shifts in the orbit with vertical gaze, moving anteriorly in upgaze and posteriorly in downgaze. It moves about 3 mm for 40-degree gaze shifts. This is about one third the movement of the IR insertion, suggesting that the IO path stabilization is different from that of the rectus muscles, perhaps through a balance of couplings to the orbit and the IR. Possibly the neurofibrovascular bundle performs this function. In any case, the IO path is unique in that it shifts to approximate the shortest “great circle” path over the globe. This shift would occur in the absence of any pulley-like coupling.

Trophic and contractile states of EOMs may be assessed by MRI measurements of cross sections. Relative trophic states can perhaps be compared most easily by contrasting maximum cross-sectional areas for a given eye position. Contraction is associated with an increased cross-sectional area and a posterior shift in the point of maximum cross section. Visual inspection of EOM cross sections may be sufficient for clinically assessing atrophy or loss of contractility. Figure 6–5A shows that, in primary gaze, the normal SO has its maximum cross-sectional area in mid orbit. In depression, the maximum cross section increases and its plane moves posteriorly in the orbit. In elevation, the maximum cross section decreases and moves anteriorly. Contractile change in the SO is plotted in Figure 6–5B, which shows that SO contractility is greatest in the posterior orbit.

### Some Clinical Applications

#### EXTRAOCULAR MUSCLE PARALYSIS

There are two typical MRI findings in chronic EOM paralysis: a reduced muscle cross section and subnormal change in muscle cross section with gaze shifts. These effects are best evaluated in the plane of maximum muscle cross section, which moves toward the origin as the EOM contracts. We take a reduction in maximum cross section to be an index of EOM atrophy and a reduced gaze-related change in maximum cross section as an index of contractile impairment. For many clinical purposes these indices are sufficient, although for biomechanical modeling we would want to know the relationships between the indices and EOM forces (see Chapter 3). Figure 6–6 illustrates both of these effects for a paralyzed left LR muscle. Similar results have been obtained for EOMs denervated by oculomotor nerve damage.

Contractility is of course lost immediately on EOM denervation. The time course of atrophy is not known.

Figure 6–7 shows these effects in SO palsy: the affected muscle has a reduced cross section and shows no contractile thickening in downgaze. In congenital SO palsy, no muscle may be visible on MRI. Objective confirmation of SO palsy is particularly useful because the actions of the SO and its cyclovertical fellows are complex. We have found that clinical diagnosis of SO palsy by the usual “three-step test” is unreliable, compared with MRI evidence of loss of contractility. Figure 6–8 plots maximum SO cross section against contractile change in cross section (with gaze shift from 22 degrees of elevation to 22 degrees of depression) in 16 normal orbits of 12 volunteers and 34 orbits of 17 patients with hypertropia in whom SO palsy (2 bilateral, 15 unilateral) had been diagnosed by a pediatric ophthalmologist. Conventional diagnostic findings included hypertropia that increased in contralateral gaze and with ipsilateral head tilt, ipsilateral SO underaction on version testing, and excyclo-tropia. Statistical cluster analysis showed that, in clinically diagnosed cases of SO palsy, the SO muscles fell into two nonoverlapping clusters: 9 had subnormal cross-sectional area and contractility (“true SO palsy”), and 10 had cross-sectional areas and contractilities in the normal range (“masquerading SO palsy”). All clinically normal fellow muscles in cases of unilateral SO palsy also exhibited normal size and contractility (see Fig. 6–8). Thus, about half of the eyes diagnosed by expert strabismologists as SO palsy did not contain any muscles with abnormalities apparent on MRI. Computer simulation shows that abnormal pulley positions alone can produce the clinical findings of SO palsy.

It has been claimed that bilateral asymmetrical SO palsy may be “masked,” in that it is difficult to discover the
Figure 6-5. Cross-sectional area data on the normal superior oblique (SO) muscle obtained from coronal MRI scan images of 16 orbits of 11 subjects, plotted against anteroposterior position in the orbit. Data were corrected geometrically to a plane perpendicular to the longitudinal axis of the muscle. Error bands represent 95% confidence intervals. A, Mean cross-sectional area of the SO, as obtained from primary gaze, 23 degree elevation, and 23 degree depression. Note that in depression the plane of maximum cross-sectional area moves posteriorly in the orbit; in elevation, the maximum cross-sectional area decreases and moves anteriorly in the orbit. B, Change in cross-sectional area of the SO from 23 degrees upgaze to 23 degrees downgaze. Note that the greatest contractile change in SO cross section is posterior within the orbit.
Figure 6-6. Coronal MRI scan of the mid orbit of a patient with left lateral rectus (LR) paralysis due to ischemic injury to the intrafascicular portion of the abducens nerve. In the normal right orbit, the lateral rectus has normal extent. The cross-sectional area increases in abduction and decreases in adduction. In the abnormal left orbit, the LR has a smaller cross section in primary gaze that does not increase in abduction. MR, medial rectus.

Figure 6-7. Coronal MRI scan of the mid orbit of a patient with unilateral traumatic superior oblique (SO) palsy. Note that the cross section of the affected SO is smaller than its normal fellow. Whereas the normal muscle exhibits contractile thickening with gaze shift from upgaze to downgaze, the paralyzed muscle does not.

Figure 6-8. Maximum superior oblique (SO) cross-sectional area plotted against contractility in 16 normal control muscles; 7 extraocular muscles with confirmed SO palsy; in 12 muscles with masquerading SO palsy; and in 10 clinically normal SO muscles whose fellows had SO palsy. The zone of palsy is marked with dotted lines.
Figure 6-9. Coronal MRI scan of both orbits of a young man with a chordoma enveloping the left abducens nerve, producing left lateral rectus (LR) paralysis manifested by large-angle esotropia. Imaging was repeated in primary, left, and right gaze, demonstrating normal LR contractile thickening on the right and absence of it on the paralyzed left. An unexpected finding was atrophy and loss of contractility of the left superior oblique (SO).

Paresis on the less affected side until after surgical correction of the more affected eye. It has been further claimed that some masked SO palsies cannot be detected before surgical correction of the initially hypertropic eye. These claims may have arisen from erroneous diagnoses of SO palsy. The concept of masked bilateral SO palsy is questionable, because simple surgical overcorrection of a hypertropia can readily be misinterpreted as a contralateral SO palsy.

Combined neurogenic pareses may be demonstrated by MRI in complex cases such as the patient illustrated in Figure 6–9. This young man rapidly developed an abducens paralysis with large-angle esotropia due to a chordoma of the skull base. LR paralysis was obvious clinically from gaze limitation and slowing of abducting saccades and was confirmed, along with LR atrophy, by MRI. Not clinically suspected, however, was atrophy and loss of contractility of the ipsilateral SO muscle. Knowledge of the SO palsy was important in achieving an excellent surgical result by augmented vertical rectus muscle transposition.

EXTRAOCULAR MUSCLE HETEROTOPY

Abnormal rectus EOM paths resulting from heterotopic pulleys can cause A- and V-pattern strabismus. Superior displacement of the LR relative to the medial rectus can readily be understood as a cause of an A pattern (Fig. 6–10). Similarly, V patterns may be caused by inferior displacement of the LR relative to the medial rectus or nasal displacement of the IR relative to the superior rectus (Fig. 6–11). Figure 6–12 shows a coronal spiral CT scan of both orbits in a 6-year-old girl who had marked A-pattern esotropia associated with medial displacement of the superior rectus muscles and superior displacement of the LR muscles bilaterally. Muscle heterotopies need not exhibit mirror symmetry in the two orbits.

Myopic strabismus fixus is a severe form of esotropia characterized by restricted abduction and progressive axial myopia. It may be unilateral or bilateral. Krizok and colleagues have given convincing evidence that myopic strabismus fixus is due to a large inferior displacement of the LR muscle, depriving the eye of normal abducting action and converting the LR to a depressor. Such a case is seen in Figure 6–13, illustrating coronal MRI of the right orbit in a patient with –30 D of axial myopia and a right eye fixed in approximately 100 PD (45 degrees) of esotropia and 30 PD (17 degrees) of hypotropia. Figure 6–13 shows that the LR has a path near the IR and that the vertical rectus EOMs are also located nasal to the center of the globe. The actual directions of pull of the rectus EOMs relative to the globe are shown in white arrows in Figure 6–13. Conventional strabismus surgery, involving resection of the LR muscle, only increased the patient’s hypotropia without correcting the esotropia, as might have been anticipated from the MRI scan.

SEVERED AND EXTRIPATED MUSCLES

Extraocular muscles may be injured by transection, partial avulsion, or denervation. Appropriate surgical planning requires determination of the contractile potential of the EOM. Obviously, a normally contractile EOM that has been partially or completely disinserted should be repaired by reinsertion. A transected EOM retaining contractile function should be reanastomosed. On the other hand, if the neuromuscular junction has been avulsed, leading to loss of contractility, then muscle function is permanently lost and other EOMs should be transposed. MRI can provide the information needed to plan management.

An example of a traumatized EOM that retained contractility is given in Figure 6–14. In this case, although the left MR had been transected as a complication of endoscopic ethmoidectomy, the posterior muscle belly continued to exhibit contractile thickening on attempted adduction. It thus seemed sensible to treat the large-angle esotropia by reanastomosing the severed muscle using a bridge of temporalis fascia. Unfortunately for another patient (Fig. 6–15), EOM damage during endoscopic sinus surgery was so extensive that the residual EOM was no longer contractile. In this case, large portions of both the right MR and right SO had been extirpated during the sinus surgery.

The patient shown in Figure 6–16 exhibited a large hypertropia after direct trauma to the orbit. Sagittal reconstruction with MRI showed avulsion of a large segment of the ipsilat-
Figure 6-10. A-pattern exotropia due to superior displacement of the lateral rectus muscles. A, Clinical photographs of patient showing A pattern and bilateral overdepression in adduction. B, Coronal MRI scan of the mid orbits showing superior displacement of the lateral rectus relative to the MR muscles, the cause of the A pattern.

Figure 6-11. Coronal MRI scan demonstrating nasal displacement of the inferior rectus muscle (arrow) associated with V-pattern esotropia in a patient.
**Figure 6–12.** Coronal spiral CT scan of a 6-year-old girl who was orthotropic in downgaze but had 60 prism diopters of esotropia in upgaze. The superior rectus (SR) muscles are displaced nasally, and the lateral rectus (LR) muscles are displaced superiorly in each orbit. This pulley heterotopy accounts for the A-pattern esotropia. MR, medial rectus; IR, inferior rectus.

**Figure 6–13.** Coronal MRI scan demonstrating inferior displacement of the lateral rectus (LR) and relative nasal displacement of the vertical rectus muscles associated with myopic strabismus fixus. The actual pulling directions of the rectus muscles, relative to the globe, are shown by white arrows. Note that the inferior path of the LR converts it to a depressor and deprives the eye of abducting force. SR, superior rectus; MR, medial rectus; IR, inferior rectus.

**Figure 6–14.** Axial MRI scan demonstrating preserved contractility with horizontal gaze shifts in a patient whose left medial rectus (MR) muscle had been inadvertently transected as a complication of endoscopic sinus surgery. Note the contractile thickening of the MR in adduction. LR, lateral rectus.
Figure 6–15. Coronal MRI scan demonstrating avulsion of extensive portions of both the right medial rectus (MR) and superior oblique (SO) muscles as a complication of endoscopic ethmoidectomy. Residual portions of the extraocular muscles were noncontractile with horizontal gaze shifts.

eral IR. This was judged irreparable by direct reanastomosis and instead was managed by inferior transposition of the horizontal rectus EOMs.

ENTRAPMENT

A blow-out fracture of the orbit may result in entrapment of EOMs and orbital connective tissues within the fracture or within the maxillary or ethmoidal sinuses. Although MRI does not show the bone fractures themselves, it can vividly demonstrate abnormal EOM paths. However, the mere presence of an orbital wall fracture does not necessarily explain restricted ocular motility. An example of prominent restriction is seen in Figure 6–17 in which the IR muscle is folded back on itself in an inferior blow-out fracture. Note that fragments of the fracture would be better demonstrated using CT than MRI, because bone is transparent to the latter imaging technique.

MASS EFFECT

Diagnosing a large tumor requires no special imaging techniques. Smaller masses that can impair ocular motility may escape detection unless high-resolution imaging methods are used. For example, a small cyst in the posterior part of the SO tendon may limit elevation in adduction, producing the misalignment pattern associated with Brown syndrome (Fig. 6–18). This patient had a palpable click, followed by release of restriction to elevation in adduction.

The patient in Figure 6–19 had restricted elevation of the right eye after several strabismus surgeries. High-resolution orbital MRI unexpectedly disclosed a cyst (probably an epithelial inclusion cyst) near the IO muscle, which was not apparent clinically. Its surgical excision relieved the restriction to elevation.

Figure 6–16. Sagittal reconstruction of coronal MRI scan demonstrating avulsion of a large segment of the inferior rectus (IR) muscle. Missing segment is denoted by double-headed arrow. ON, optic nerve; SR, superior rectus; LPS, lateral palpebrae superioris.
Figure 6–17. Coronal MRI scan with sagittal reconstruction demonstrating the right inferior rectus (IR) muscle folded back on itself through an inferior wall blow-out fracture into the maxillary sinus. SR, superior rectus; LR, lateral rectus; MR, medial rectus; ON, optic nerve; LPS, lateral palpebrae superioris.

Figure 6–18. Sagittal MRI scan of the right orbit parallel to the superior oblique (SO) muscle belly illustrating a cyst in the posterior part of the SO tendon in a patient with right Brown syndrome. ON, optic nerve; MR, medial rectus; SR, superior rectus; IR, inferior rectus.

Figure 6–19. Coronal MRI scan of the right orbit of a patient who had restricted elevation of the right eye after multiple strabismus surgeries. Note the prominent inclusion cyst near the right inferior oblique muscle. LR, lateral rectus; SR, superior rectus; MR, medial rectus; IR, inferior rectus.
Conclusions

High-resolution orbital imaging can be a useful diagnostic tool in cases of complex strabismus, revealing unexpected pathophysiologic mechanisms and aiding rational surgical planning. Imaging should be considered in complex cases as an adjunct to the clinical examination and other ancillary tests.

REFERENCES

The diagnosis and treatment of strabismus are based mostly on simple heuristics, shared experience, and intuition. Consequently, unusual and complex cases of strabismus may be difficult to manage. A description of the mechanisms of binocular coordination—a model—attacks these problems in a new way: experience, intuition, and experimental findings go into building and testing the model; and then the model, less impaired by unfamiliarity and complexity, is used in case management.

A model-driven approach, able to coordinate broad ranges of laboratory research and clinical experience, can accelerate progress in the diagnosis and treatment of strabismus. Without analytical methods it is difficult to develop useful new procedures, particularly for complex, cyclovertical disorders. Indeed, such developments have been infrequent. As prosthetic extraocular muscles (EOMs) become practical, their use will require judgments for which computational analysis provides the most appropriate guidance.

Actions of the oblique EOMs are complex, and dysfunction is often not obvious from inspecting ocular ductions and versions. It is possible that most strabismus is complex, involving abnormal contractile and elastic forces and path changes in several muscles, their innervations, and their connective tissue suspensions. In many cases of congenital and traumatic strabismus, the primary lesions are obviously multiple and complex. But even in isolated palsy there will generally be secondary changes in other muscles; it is well known that muscles adapt to changes in their working length. Innervational changes due to plasticity of neural control centers are less well understood but are probably important in many disorders. It may be that strabismus diagnoses are typically simple (and so, it would follow, incomplete) because they are based on limited, poorly controlled measurements of alignment. Such measurements may be the norm because it is impossible to assimilate more adequate data without computational help.

**Orientation to Strabismus Models**

What types of analytical models are available to help with strabismus diagnosis and treatment?

**EMPIRICAL GENERALIZATIONS**

A type of model familiar to strabismus surgeons is the empirical generalization, examples of which are the familiar tabular surgical "dose-response" relationships, and the computer programs of Russman, Konen and Russman, and Simonsz. Whether developed informally or with computerized databases and statistical techniques, empirical generalizations summarize experience; they are models of observations. Empirical generalizations are probably the basis of professional competence in most fields. However, because they are so closely related to experience, empirical generalizations may seem correct even if flawed. Thus, they may impede fundamental progress by lending a sense of understanding without explaining the causes of observed patterns. Nevertheless, empirical generalizations have been, and will continue to be, of great value in strabismus management.

**EXPERT SYSTEMS**

An expert system is a different kind of model, not of the topics of interest themselves (e.g., gaze angles, muscles) but of the inferences and judgments of human experts. Expert systems are psychological models. Their strength is derived from their extreme domain specificity, which at the same time limits their generality. These systems are perhaps best thought of as ways to distribute extant professional expertise rather than as means to fundamentally advance a field. Expert systems may also find roles in strabismus education and practice.
HOMEOMORPHIC MODELS

Neither empirical generalizations nor expert systems can put strabismology on a scientific basis, because neither treats the subject matter in terms of mechanisms, that is, lawful relationships among subprocesses or parts of the oculomotor system. Nonmechanistic models have difficulty dealing with new situations, because there may be no way to know if their similarity to known situations is merely superficial, and they tend not to have implications outside the problem area for which they were developed.

A homeomorphic model is a mechanistic model that has the “same form” as the system modeled; that is, the model has parts that correspond to physiologic structures, and the interactions of model parts reflect physiologic processes. A homeomorphic model can deal with arbitrary new situations, so long as they can be expressed using the model's terms. Successful predictions validate the model. Unsuccessful predictions tend to specify the physiologic research needed to improve the model.

BIOMECHANICAL MODELS

Biomechanical models of strabismus are homeomorphic models that focus on the globe, connective tissues, extraocular muscles, and innervations. The first biomechanical models were the familiar ophthalmotropes of Ruete, Wundt, and others (Fig. 7–1). Computer models have a critical advantage over the old ophthalmotropes in that their behavior is constrained only by our understanding of extraocular biomechanics, not by the materials and mechanisms feasible in a table-top physical model.

Biomechanical models assume that the complex behavior of muscles can be understood in terms of simple arrangements of elastic and force-generating elements, that complex orbital connective tissues may be represented by simpler elasticities, and that the orbital contents can be analyzed in isolation from the rest of the organism. Little attention is paid to nonmechanical factors, not because they are unimportant but because they are largely separable from the mechanics. In any case, nonmechanical factors must act through the eyes' mechanics. Thus, understanding extraocular mechanics facilitates, by “subtraction,” the study of nonmechanical factors.

Orbital geometry, muscle and connective tissue mechanics, and innervations are intertwined in ways that generally cannot be understood without a model of orbital biomechanics. Robinson laid a foundation for biomechanical models of ocular alignment with a system of equations and computational procedures based on balancing the static forces of extraocular muscles and orbital tissues and oriented to simulate strabismic disorders and their surgical correction. Variation in tension across the width of muscles and Lockwood's ligament, an elastic connection between the inferior rectus (IR) and inferior oblique (IO), have been modeled. The Robinson model has been tested against clinical data. Miller and Robinson's model provided muscles with innervation-length-tension relationships having “slack” and “leash” regions, made muscle sideslip a function of tension, allowed globe translation, and provided full binocularity. This computational model was called SQUINT.

There are two modern models derived from this line of work. Simonsz reviewed much of the existing data on orbital geometry and tissue stiffness and conducted several new studies to develop a model that runs on a palmtop computer. It achieves simplicity of use, rapidity of calculation, and clinical utility by limiting abnormalities and manipulations to eye size, muscle origins, insertions, length-tension relationships, and innervations. The second model is a Macintosh application called Orbit, which provides a user-friendly interface to SQUINT, hiding much of SQUINT's complexity rather than restricting its generality. The National Institutes of Health/National Eye Institute (NIH/NEI) has continued to support the development of SQUINT and the physiologic research on which it is based.

Physiologic Basis

A biomechanical model in isolation is only an academic exercise. With strong links to physiologic research, however,
Clinical applications of computer models for strabismus • 101

A biomechanical strabismus model can have both clinical utility and scientific validity. As a clinical tool it can help assimilate patient data, aid diagnosis, and clarify treatment options. As a scientific theory or hypothesis, such models propose explanations of normal and abnormal binocular alignment. Both roles cast the model as a construction or abstraction based on various kinds of data (inward pointing arrows of Fig. 7–2). Equally important is a model's role in influencing where investigators look for relevant data and what tools they use. Biomechanical strabismus models have been particularly fruitful in this regard (outward pointing arrows of Fig. 7–2). In particular, the failure of early strabismus models to predict the outcomes of transposition surgery led to the concept of extraocular muscle pulleys and testing of that concept by magnetic resonance imaging (MRI) before and after transposition surgery (see Fig. 7–2, lower right panel).

**Extracocular Imaging**

Muscle forces and axes of rotation are central variables in biomechanical strabismus models. A muscle's axis of rotation gives the direction in which it tends to spin the globe, and the muscle force gives the magnitude of that tendency. Modern extracocular imaging was motivated by modelers' needs for accurate descriptions of muscle paths, the determinants of those paths, and muscle cross sections and contractilities. We used MRI to demonstrate that the paths of rectus muscle bellies are stable in the orbit throughout the oculomotor range and showed that MRI can estimate EOM contractility and atrophy. Below, we analyze a case that had been diagnosed as oblique muscle dysfunction, in which MRI demonstrated normal oblique muscle contractilities and cross sections. Extraocular imaging is the focus of Chapter 6.

**Immunohistochemistry and Electron Microscopy**

In vitro study of extracocular tissues has provided histologic and structural details to supplement in vivo imaging. Human cadaveric orbits were decalcified, imbedded in paraffin, and cut in 10-μm sections. Three interleaved sets of sections were treated, respectively, with Masson's trichrome stain, which visualizes muscle and collagen; an immunohistochemical stain for smooth muscle α-actin; and van...
Gieson’s elastin stain. The slices then were mounted and digitally photographed for computer reconstruction (Fig. 7–3).

Perhaps the most interesting result of these immunohistochemical studies is the prevalence of smooth muscle cells in the connective tissue around the rectus EOMs in the vicinity of the globe equator (constituting what an anonymous NEI grant reviewer called “innervated connective tissue”). This smooth muscle helps reconcile the existence of mechanically significant connective tissue constraints on muscle path with the informal intraoperative observation that EOM bellies do not seem to be firmly fixed to the orbit. We have since traced a sympathetic projection to mid-orbital smooth muscle from the ipsilateral superior cervical ganglion and have found evidence of parasympathetic innervation.

Further evidence that these tissues function to control muscle paths comes from electron microscopy, which shows pulley collagen to be extremely dense and organized in an unusual, crisscrossed configuration suited to high internal rigidity. On the basis of these studies, we have created an overall scheme of human extraocular connective tissue (Fig. 7–4).

**RECTUS MUSCLE PULLEYS**

From the complex arrangement of tissues diagrammed in Figure 7–4 we developed the concept of muscle pulleys. A muscle pulley consists of a ring or sleeve of collagen, elastin, and smooth muscle that encircles an EOM and is coupled to the orbital wall and other connective tissue structures by similar tissues. Tendons and muscles travel through pulleys by sliding inside thin collagenous sheaths that telescope within the pulley sleeves. The pulleys are coupled to the orbit and in straight-ahead gaze are located near the globe equator in Tenon’s fascia.

Pulleyshave important implications for EOM function. Whereas the direction of pull (or axis of rotation) of an EOM was once thought to be determined by its anatomic origin in the annulus of Zinn (along with its point of tangency with the globe and the globe center), according to the pulley model the functional origin of an EOM is its pulley (Fig. 7–5). The path length of an EOM, which determines its stretch and therefore its tension, is increased by deflection through the pulley. Despite being stiffened by elastin, pulleys are compliant and potentially subject to actions of their suspensory smooth muscles, as well as changes in EOM tension. Although no functional role of pulley smooth muscle is proved, several roles are plausible. The smooth muscles might simply increase the stiffness of Tenon’s fascia and the pulley suspensions to provide a constant load and geometry for the ocular motor system. Simulation shows that binocular alignment is highly sensitive to pulley position. Thus, it is possible to imagine that smooth muscle tension is modulated to refine binocular alignment or assist in slow eye movements such as convergence.

**BINOCULAR ALIGNMENT MEASUREMENTS**

Alignment data routinely collected in the clinic are often not useful for modeling. The following are guidelines for collecting quantitatively useful data:

1. The eyes must be completely dissociated, with fixing and following eyes specified.
2. Fixing eye gaze angles must be specified. Thus, target positions must be defined, and the patient’s head position controlled.
3. A sufficient number of gaze angles over a large enough field must be included to reasonably constrain the simulation. Clinical data are modeled by adjusting model parameters to fit. If there are insufficient data, many different sets of model parameters (corresponding to different diagnoses) may fit the data, so that modeling is indeterminate. If the range of fixation is too small, distinguishing features of the pattern of misalignment (deviation) will not appear in the data. We routinely collect data for 21 fixations with each eye over a ±30° field.
degrees $\times \pm 30$ degrees field. Nine fixation points might be considered a minimum. (The Clement Clark Electric Hess Screen [Fig. 7–6; Harlow, England] is convenient to use and adequate in the preceding respects.)

4. Measurement of objective torsion should be attempted. Torsion is simply the third coordinate (with horizontal and vertical angles) needed to specify eye rotation, and measuring it provides more data to constrain modeling. Torsional measurements may be of particular use in discriminating oblique muscle disorders. Unfortunately, subjective torsion measurements may be unstable, and it may not be clear which is the tortured eye. Objective measures (e.g., ophthalmoscopy or fundus photography) in nonprimary positions may be inconvenient to acquire in the clinic.

Clinical Applications

WHEN IS A BIOMECHANICAL MODEL USEFUL IN TREATMENT PLANNING?

We compared predictions of the Orbit model with published surgical dose recommendations for comitant esotropia and exotropia and found that model predictions agreed with published recommendations. Interestingly, modeling showed that these simple types of horizontal strabismus respond to surgery independently of their mechanical, innervational, or combined etiology—surely part of the reason for the broad success of the recommendations. This illustrates how modeling may be used to clarify traditional methods, a laudable scientific goal, but also shows that there is little reason to use a biomechanical model with simple, familiar types of strabismus when traditional tables supplemented by clinical judgment will suffice.

HOW TO USE A BIOMECHANICAL MODEL IN DIAGNOSIS

Modeling has three steps:
1. Simulate the preoperative disorder.
2. Apply surgical manipulations to the preoperative simulation.
3. Compare simulated and actual surgical outcomes.

One may ask many questions of a strabismus model—what are the muscle forces? innervations? muscle paths?—and compare these predictions to measured or desired values. In strabismus management, binocular alignment is usually a central concern, and so we usually compare predictions and measurements of alignment. Accordingly, the Orbit model is designed to predict results of a dissociated eye alignment test such as the Hess, Lancaster, or prism-cover test. We use a format similar to that of the Hess and Lancaster tests, which shows intended gaze (the gaze angles of the fixing eye, reflected across the midline), positions,
Normal Eye

Conventional Model

Pulley Model

Figure 7–5. Muscle actions differ in conventional and pulley models. A muscle’s axis of rotation (indicated with “spin” arrows) is the axis the eye tends to rotate about when the muscle contracts. A and B, In a conventional model, the muscle acts as though it were inserted at its point of tangency with the globe. In elevation, it is as though there were a hinge at this point, and the axis of rotation remains roughly fixed with respect to the orbit. C and D, In a pulley model, the muscle passes through a connective tissue pulley that is stabilized with respect to the orbital wall and determines the effective muscle origin. In elevation, the axis of rotation tilts with the eye. Note that muscle paths can be the same in the two models, although mechanical actions are different.

of the following eye (the eye moving passively under the innervations determined by the fixing eye), and the overall pattern of misalignment (deviation) for 21 positions over a ± 30 degrees × ± 30 degrees field (see Fig. 7–7).

Hundreds of parameters are needed to describe the anatomy and physiology of the eyes, but it is impractical to measure more than a few of them in a given patient. Such situations, familiar in the life sciences, require a normative assumption: a patient is assumed to be normal except when there is evidence to the contrary. Thus, we begin a simulation with a description of normal eyes based on physiologic studies, such as those described earlier. An alignment chart for normal eyes is shown in Figure 7–7.

To demonstrate modeling procedures, consider patient AT, a 19-year-old white man who suffered whiplash, wrist, and chest injuries in an automobile collision but no direct head injury or loss of consciousness. During recovery he developed incomitant left hypertropia of 3 degrees (6 PD) in left gaze, increasing to 17 degrees (30 PD) in right downgaze, and a right head tilt of 7 to 10 degrees. Excyclotorsion of 6 degrees was measured by double Maddox rods. A forced head tilt test to the left was strongly positive. Left superior oblique (SO) palsy was diagnosed. Figure 7–8A shows AT’s preoperative alignment data, slightly smoothed to help focus on general modeling principles rather than case idiosyncrasies. Actual cases of strabismus often involve secondary muscular and innervational changes. Subjective measurements of torsion are shown, but, because of their unreliability, we only consider them informally.

We begin analysis with simulated normal eyes (Fig. 7–7). The aim of preoperative simulation is to bring simulated eye alignment into congruence with the clinical data (Fig. 7–8A). What is the biomechanical mechanism of the “SO palsy” of Figure 7–8A; is it simply that the left SO fails to contract, or is there more? Contractile muscle force (developed force) is produced under control of innervation. Interpreting the diagnosis literally, we suppose that left SO contractile force is zero (we enter “0” in the appropriate place in the model). The model predicts the preoperative alignment shown in Figure 7–8B. Although this preliminary simulation captures
Figure 7-6. (See Color Plate 4.) A Clemente Clark Electric Hess Screen, modified by replacing each red fixation LED with a vertical row of three LEDs, allowing measurement of ocular torsion. The subject sits with chin (and if necessary, forehead) stabilized against a support 50 cm from the screen. Eyes are dissociated with red and green filter goggles. The subject indicates localizations using a hand-held green streak projector.

some features of the clinical data (see Fig. 7–8A) including limitation of downgaze, particularly in adduction, and ex-cyclorotation, particularly in downgaze, magnitudes are systematically different from the data of Figure 7–8A. Thus, we do not yet have an adequate simulation of the patient’s preoperative disorder.

What else do we know or suspect about AT’s disorder? Here, modeling is similar to traditional diagnosis: if we have test results, we use them; otherwise we proceed on general experience or hypothesis. Here we suspect that the denervated left SO has become atrophic, that is, its elastic force is subnormal as well as its contractile force. If appropriate orbital images were available, we could estimate the reduction in elastic force by comparing the cross-sectional area of the paretic left SO with that of the normal right SO.11 We could also verify our contractile force assumption, as described in Chapter 6. However, orbital images were not available in this case, so, on the basis of experience with similar cases, we supposed that left SO elastic strength was 50% of normal. Running the modified simulation gives a good match to the clinical data except for small horizontal and vertical offsets of the entire alignment pattern. We assume that these are due to small, almost negligible abnormalities in rectus muscle lengths, either idiosyncratic or secondary to the palsy. Thus, we make some adjustments (~1 mm) in lateral rectus (LR), medial rectus (MR), and superior rectus (SR) muscle lengths. Figure 7–8C shows the resulting simulation, which is a good match to AT’s clinical alignment pattern.

We now have a reasonable simulation of patient AT’s preoperative strabismus, which is also a biomechanical diagnosis: the left SO is denervated and has atrophied—it develops no contractile force and only half of normal elastic force. Everything else, including AT’s right eye, is normal or close to normal. Modeling shows the implications of our hypotheses: that, in this case, SO palsy with atrophy and slightly abnormal rectus muscle lengths can account for the patient’s binocular misalignment. The preoperative simulation also serves as the starting point for treatment analysis.

TREATMENT SIMULATION

A year and a half after patient AT’s accident, he underwent strabismus surgery. Measured and simulated postoperative alignment are shown in Figure 7–9. In both the patient and model, the left IO was moved 5 mm posterior to the temporal insertion of the IR muscle and 1 mm temporal to it, and the right IR was recessed 4.5 mm. It can be seen that the simulation was a good predictor of surgical outcome. One month postoperatively, AT no longer reported diplopia and alignment was orthotropic except for a small, asymptomatic right hypertropia in extreme right upgaze.

ROLE IN TEACHING AND CHANGING PATTERNS OF CLINICAL PRACTICE

Schools of Thought

The philosopher of science Thomas Kuhn9 described a prescientific stage in which, lacking a unifying scheme of thought, or paradigm, practitioners in a field form intellec-
A Unifying Framework for Theory and Practice in Strabismus

We know of only one approach that might emerge as a paradigm for strabismus. Although most would probably agree that biomechanics may ultimately be the correct way to analyze binocular alignment, many argue that complexities of extraocular architecture, vagaries of posttraumatic and postsurgical healing, plasticity of central innervation, and individual differences would make such analyses impractical. We have already touched on some ways in which modeling has driven studies that have clarified extraocular architecture (see Fig. 7–3). Biomechanical effects of healing can be similarly studied. Central plasticity may be evaluated with the help of an extraocular model to infer changes in innervation that would account for observed changes in alignment. Individual differences present problems that will need to be addressed on several fronts. First, tomographic imaging can provide indices of EOM elasticity and contractility, and also of orbit and globe anomalies that affect extraocular geometry (see Chapter 6). Some features of extraocular connective tissue can be inferred from images of EOM paths as a function of gaze. Second, characterization of extraocular parameters by gender, race, and age would account for some of the variation between patients.

Patterns of specialization and referral, evolved in an era in which human health was considered priceless, will surely change. It will become more difficult to justify the cost of sending patients to distant experts, and so local specialists—ideally in consultation with experts—will become responsible for more sophisticated diagnoses and treatments. Instead of moving patients to experts, expertise will be brought to local specialists. Without accurate communication, however, it will be difficult for remote experts to...
Figure 7–9. Patient AT, postoperative alignment of left eye (right eye fixing). A, Postoperative clinical data. B, Simulated postoperative alignment. The fit is good, showing that the model accurately predicts surgical outcome and, indirectly, that our preoperative simulation (see Fig. 7–8C) was reasonable.

provide more than hints. Accurate communication and precise definition of terms are also necessary for multicenter clinical trials of new approaches in strabismus.

If patient data are to be shared, quantification must be well defined. Do all clinicians mean the same thing by “minus 3 gaze limitation?” Even with alignment measurements, quantification may be nominal. For instance, deviations by prism and cover testing do not typically specify the intended gaze angle at which nonprimary deviations were measured, or even which eye looked through the prisms. Well-defined quantification is essential for both biomechanical modeling and reliable communication.

Strabismus nomenclature is an ill-defined mixture of terms such as hypertropia and V pattern, which describe symptoms (observations), and terms such as underaction, overaction, and contracture, which sound etiologic, as if they point to mechanisms underlying observations but usually are just alternative descriptions of symptoms. Etiologic terms may seem convenient descriptions of complex observations; for instance, “SO palsy” is used to mean “limited depression and excyclorotation, particularly in adduction.” But these terms become barriers to clear thinking in cases, for example, when the SO muscle is perfectly normal. Biomechanical modeling can help by defining precise etiologic terms, distinguishing for example among contractile muscle strength, elastic muscle strength, and relaxed muscle length, which have differing effects on motility. These are not distinguished by referring to “weak” and “strong” muscles.

C A S E S T U D I E S

Heterotopic Pulleys and Pattern Strabismus

A-pattern strabismus is horizontal strabismus in which the eyes are relatively divergent in downgaze (or relatively convergent in upgaze). A patterns are usually attributed to oblique muscle dysfunction. However, simulation shows that significant displacement of rectus muscle pulleys, that is, heterotopic pulleys, can also produce these patterns.

Patient TL has A-pattern strabismus, which might be attributed to “overacting SOs” (Fig. 7–10A). However, coronal MRI clearly shows pulley heterotopy (Fig. 7–11), and simulation shows that this pulley disorder can account for the pattern of misalignment (see Fig. 7–10B).

We have identified significant pulley heterotopy in 11 of 12 patients studied so far with A- or V-pattern strabismus. Simulations are consistent with the idea that abnormal directions of rectus pull due to pulley displacements may cause or contribute to misalignments typically diagnosed as oblique muscle overactions or underactions. This insight has been useful in avoiding futile surgery on oblique muscles.

It has been suggested that abnormal rectus pulley positions might be a result of torsional malposition of the eyes. Although we cannot rule this out in all cases, evaluation of pulley positions in 14 orbits of seven exocylotropics subjects with SO palsy confirmed by MRI showed that the only significant pulley abnormality was MR pulley elevation, which averaged 1.1 mm. An isolated pulley displacement could not be the result of ocular torsion, particularly because the MR pulley has the firmest coupling to the orbital wall. Perhaps the upward displacement of the MR is due to movement into the space created by loss or atrophy of the adjacent SO.

Vertical Rectus Transposition With Posterior Augmentation Sutures

Since at least the 1970s, conventional vertical rectus transposition for LR rectus palsy has been augmented with posterior sutures, pulling the lateral borders of the transposed SR and IR muscles to the borders of the paralyzed LR. Only recently, however, has this procedure been analyzed and published. OrbitTM simulations provided an analysis of the procedure’s success in increasing horizontal range of gaze with little loss of vertical range. Postoperatively, MRI clearly shows inflections in the paths of transposed EOMs at locations that approximate pulley sites that are demonstrated grossly and histologically (Fig. 7–12B). Patient TS, a 33-year-old white man, experienced horizontal diplopia before surgical resection of a chordoma and both horizontal and vertical diplopia afterward. The tumor enveloped the abducens nerve, which was almost certainly lost to the

提供更多的-than hints。准确的沟通和精确的术语定义也是多中心临床试验新方法在斜视中的必要条件。

如果患者数据要共享，量化必须是明确定义的。是否所有临床医生对“-3度偏斜”有同样的理解？甚至与对准测量不同，量化可能只是名义上的。例如，棱镜和遮盖试验通常不具体指出非主要偏斜是在非主要偏斜时哪只眼睛透过棱镜。明确的量化对于生物力学建模和可靠通信是必不可少的。

斜视术语是一个定义不明确的混合物，如垂直眼位偏斜和V模式，描述症状（观察），而术语如薄弱，过强，和收缩，听起来像是病因的，好像它们指向观察机制。病因术语似乎方便描述复杂的观察；例如，“SO麻痹”用来指“限制性内转和外斜旋转，特别是在内转时”。但这些术语成为对思维的障碍，例如，当SO肌肉是完全正常的。生物力学建模有助于定义精确的病因术语，区分例如收缩肌肉力量，弹性肌肉力量，和松弛肌肉长度，这些对移动性有不同的影响。这些不是通过指称“弱”和“强”肌肉来区别的。

案例研究

异位滑车和模式斜视

A-型斜视是水平斜视中，眼睛在下视时是相对外转（或上视时是相对内转）。A型图案通常归因于斜肌功能障碍。但是，模拟显示，重要的肌肉滑车的移位，即异位滑车，也可以产生这些模式。

患者TL有A-型斜视，可能归因于“过动SOs”（图7–10A）。然而，冠状MRI清楚显示滑车位移（图7–11），并且模拟显示，这个滑车障碍可以解释这个模式的失对准（见图7–10B）。

我们已识别出11名12名已研究的A-或V型斜视患者具有明显的滑车移位。模拟与想法一致，即异常的肌肉的拉力方向可能由于滑车移位导致斜视，通常被诊断为斜肌过强或过弱。这个见解在避免对斜肌进行无用的手术上是有用的。

有人提出，异常的肌肉滑车位置可能是由眼球的旋转偏差引起的。虽然在所有情况下，我们不能排除这一点。在14只具有SO麻痹的临床中的7个眼睛的脉轮位置的评估中，MRI表明，只有显著的滑车异常是MR滑车升高，平均1.1毫米。单一滑车的移位可能不是由于眼球的旋转的，特别是因为MR滑车对眼球的壁有最牢固的耦合。也许MR滑车的上的位移是由于它对空间的移动导致的，这个空间是由相邻的SO和眼周组织的丢失或萎缩造成的。

垂直肌转位与后部增益缝合

自20世纪70年代以来，传统垂直肌转位治疗LR麻痹已经使用了后部缝合，将转位的SR和IR肌的外侧边缘拉到麻痹LR的边缘。只有最近，这个程序才被分析和发表。OrbitTM模拟提供对程序成功的分析，增加了水平眼位范围，且能保持很少的垂直眼位范围的丢失。术后，MRI清楚显示转位EOM的路径的弯曲，在位置靠近转位的滑车，这些位置被显微镜和组织学上证明（图7–12B）。患者TS，一个33岁的白人男性，在一畸瘤术后经历了水平复视，并且手术后还出现了水平和垂直复视。肿瘤包围了眼展神经，这在手术前几乎可以确信丢失了。
Figure 7-10. Patient TL, binocular alignment before strabismus surgery. A, A-pattern clinical binocular alignment measurements. B, Simulated misalignment due to the patient's heterotopic pulleys, eyes otherwise normal. Left panels show positions of the left eye with right eye fixing, and right panels show positions of the right eye with left eye fixing.
Eight weeks after chordoma surgery, strabismus surgery was performed, consisting of left SR transposition to the superior end of the left LR insertion, with an augmentation suture on its superior edge 8 mm posterior to the new insertion, and left IR transposition to the inferior end of the left LR insertion, with an augmentation suture on its inferior edge 9 mm posterior. Postoperative alignment 6 weeks later shows excellent improvement in horizontal range with little sacrifice of vertical range (Fig. 7–15A). Images obtained from MRI performed the same day are shown in Figures 7–12B (three-dimensional reconstruction) and 7–14B.

To simulate the patient’s strabismus surgery begins with the preoperative simulation of Figure 7–13B. We then simulate the left SR and left IR surgery done on patient TS (transposition is simulated by moving model insertions; an augmentation suture is estimated as a recess-resect, half the distance of the suture from the insertion). We suppose that the transposed muscles each stretch 5 mm and that their pulleys stretch 7 mm medially secondary to transposition. MRI shows some recovery of left MR and left SO contractility (see Fig. 7–14), on the basis of which we restore full left MR contractility and set left SO contractility to 50% of normal. Thus, we hypothesize that, whereas innervation to the left LR was permanently lost, innervation to the left MR was only temporarily impaired and left SO innervation was only partly impaired by surgical trauma. The resulting simulation gives a reasonable match to postoperative alignment measurements except in extreme left gaze (see Fig. 7–15B).

We cannot be sure why the simulation fails to match the clinical data in left gaze: there could be unsimulated
Figure 7-13. Patient TS, binocular alignment after chordoma resection but before strabismus surgery. A, Clinical measurements. Left eye shows severely limited abduction characteristic of left lateral rectus palsy, and V pattern characteristic of left superior oblique palsy. Right eye shows a large secondary deviation. Only a few right eye positions could be measured because of the limited left eye fixation range. B, Simulation (see text) is a good representation of measurements.
orbital idiosyncrasies, errors in the clinical alignment data, or limitations in the biomechanical model. Perhaps the patient’s preexisting horizontal diplopia is a clue, but we do not have enough information about it to form a hypothesis. Modeling can reduce, but not entirely eliminate, clinical uncertainties.

**Conclusions**

Predictions from computer models, confirmed by MRI in strabismus surgery patients, have shown the importance of the complex arrangements of connective tissues that support the extraocular muscles and determine their directions of pull. Immunohistochemical studies have shown that the mid-orbital connective tissues surrounding EOMs and coupling them to the orbital walls contain smooth muscle cells having sympathetic, parasympathetic, and nitric oxide mediated projections. Tracer and stimulation studies showed that the superior cervical ganglion is the source of the sympathetic projection. Thus, there is a substrate to support excitatory and inhibitory modulation of EOM pulling directions.

Already these findings have changed our understanding of extraocular muscle coordination, providing, for instance, an orbital substrate for Listing’s law. Future research will determine if this system acts to refine eye alignment, aid vergence, or simply optimize the stiffness of the pulley-like connective tissue structures. As the clinical importance of extraocular connective tissues is clarified, it will be natural to study systemic connective tissue disorders, first as tests of emerging models and then as cases in which new concepts can provide clinical guidance. Conditions such as the Marfan and Williams syndromes, well characterized at a molecular level, are strongly associated with strabismus and would seem to be a natural starting point for studying the relationship of abnormal connective tissue to strabismus. More general biomechanical models must be developed to reflect mechanically important structures such as the neurofibrovascular bundle and the lateral levator aponeurosis and such strabismus surgeries as muscle-splitting procedures. Based on Robinson’s work, the current Orbit model uses simplified “string” muscles and tendons. Intrinsically two-dimensional effects (e.g., the nonuniform distribution of force across the tendon’s width due to tendon bending) are treated as special cases. Representations of muscle and tendon surfaces are only for appearance (e.g., muscles can intersect). In a better model, tissue surfaces would be part of the biomechanical analysis, so that EOMs would deflect accurately when crossing (as when the SO emerges from the...
CLINICAL STRABISMUS MANAGEMENT

LEFT EYE (right eye fixing) UP RIGHT EYE (left eye fixing)

40 - - - - - - - - 40
3D - - - - - - - - 3D
20 - - - - - - - - 20
O • • • • • • • • •
F.
O • • • • • • • • •
40 t -40 t -40 t -40 t
L.

Horizontal Gaze, deg longitude

AB 40 30 20 10 0 -10 -20 -30 -40 AD AD-40 -30 -20 -10 0 10 20 30 40

Figure 7-15. Patient TS, binocular alignment after strabismus surgery. A, Clinical measurements. Left eye shows much improved horizontal range with little sacrifice of vertical range. Right eye shows secondary deviations, much decreased compared with Figure 7-13A. B, Simulation provides a good match to measurements, except in left gaze (see text).

REFERENCES


anterioredge of the SR). This would result in more accurate simulations (particularly of some surgeries) and more realistic graphic representations.

The scientific value of SQUINT and its successors lies in their ability to test—and embody in a useful summary form—physiologic findings, clinical data, and theories of extraocular function. As imaging data are more widely used to characterize contractile and trophic states of EOMs, and to describe structural abnormalities of orbits, it becomes more important to integrate these data with biomechanical modeling results. This will make possible better case simulations and better tests of the models.
SECTION 2

HORIZONTAL DEVIATIONS
Strabismus is one of the most prevalent health problems among children in the Western hemisphere, affecting 5 in every 100 U.S. citizens, or some 12 million persons in a population of 245 million. Infantile strabismus (i.e., strabismus starting in the first year of life) will affect about 1% of full-term, healthy newborns and a much higher percentage of newborns who suffer perinatal difficulties due to prematurity or hypoxic/ischemic encephalopathy. Taking into account this small but high-risk population, the overall prevalence of infantile strabismus is closer to 2%.

Of all subtypes of human strabismus, infantile strabismus may be the most important but least understood. Early-onset strabismus, first and foremost, stigmatizes children into their adult years by depriving them of the many benefits bestowed by normal binocular vision. It is important to clinicians because it is a leading cause of visual loss due to amblyopia and often requires multiple surgical procedures to restore proper eye alignment. Yet, despite the restoration of good to excellent ocular alignment, bifoveal fusion is seldom acquired. It is important to vision scientists because, in addition to eye misalignment, it is accompanied by improper development of stereopsis, motion processing, ocular fixation, and eye tracking, defects not found commonly in children whose strabismus begins after infancy. This constellation of sensory and motor deficits constitutes an infantile strabismus syndrome or complex.

More than 90% of infants who become strabismic develop an esotropic (convergent) misalignment of the visual axes, as opposed to exotropic misalignment. In the older literature the term congenital esotropia was used and, more recently, essential infantile esotropia has emerged. The terms are interchangeable, but infantile esotropia is used here for simplicity. Our focus in this chapter is the esotropia complex, although much of what is discussed on the following pages applies equally to infantile exotropes.

First addressed is causation, both historical notions and current neurophysiologic concepts. This is followed by a brief description of classic infantile esotropia and then variants on this classic presentation. A detailed discussion is included of diagnostic motor and sensory signs, which is followed by practical recommendations for medical and surgical treatment.

Causation

HISTORICAL PERSPECTIVE

Historical hypotheses regarding the mechanism of infantile esotropia suffer from two general weaknesses. The first is a lack of anatomic or physiologic specificity. To say that esotropia is caused by an excess of tonic convergence is a tautology unless we can explain the origin of this excess. Second, there is a lack of comprehensiveness: a failure to account for the timing, the constellation of ocular motor signs, the perceptual deficits, and the predicted response to treatment. Although it might be reasonable to postulate that the esotropia itself reflects a primary ocular myopathy, this cannot account for its high prevalence in infants who suffer delayed development of the cerebral cortex, for the sudden emergence of esotropia at the onset of cortical binocularity in normal infants, for nystagmus and conjugate asymmetries of pursuit, for deficits in motion processing, or for the stubborn persistence of microstrabismus despite early and precise eye muscle surgery.

Nature Versus Nurture

Historical viewpoints on the cause of infantile esotropia may be divided along a nature-nurture axis (Fig. 8–1). Worth (1903) was a strong “nature” proponent, postulating an inborn and irreversible defect of fusion. Although the
central nervous system location was not specified by Worth, it is reasonable to translate this as a congenital defect of disparity-sensitive, binocular neurons in the striate cortex. Crone (1973) postulated a similar primary dysfunction in the development of binocular sensitivity.

The leading proponent of the "nurture" hypothesis was Chavasse (1939). He contended that the neural components necessary for normal binocular fusion are present in strabismic individuals at birth but that the development of fusion postnatally is impeded by "obstacles" (abnormalities) of optical input (e.g., monocular cataract, hypermetropia) or muscular output (e.g., cranial nerve palsy). Chavasse couched his argument in pavlovian terminology, binocular vision being viewed as a reflex conditioned in the early postnatal period. A key point in his argument was that the strabismic patient could develop normal binocularity if the input or output impediments were removed by early therapy. Costenbader (1961) and Parks (1975), as consistent advocates of early surgery to restore eye alignment, also subscribe to the principles of the nurture hypothesis.

**Visual Pathways Versus Motor Pathways**

Opposing viewpoints on the origins of infantile esotropia are also arrayed along a visual-motor axis. Worth (1931) and Crone, who postulated a congenital deficit in binocularity, may be placed at the visual cortex end of this axis. A majority of other hypotheses fall into a vague middle ground between the visual cortex and extraocular muscles. Snellen, Scobee, Mindel, and Porter define the muscle end of this axis.

**Visual Cortex**

Keiner postulated a defect of cortical binocularity compounded by direct subcortical "light tonus" inputs. At birth, illumination of the temporal retina presumably drove the eyes nasally. Keiner's writing repeatedly touched on the topics of unstable infant vergence, a vague nasally directed bias, and what may be interpreted as a defect of disparity sensitivity. Keiner firmly believed that esotropia develops postnatally: "All children are born with a potential to squint and an almost total dissociation of the two eyes. Congenital squint does not exist; strabismus cannot occur until the light stimulus is able—in connection with the stage of development of the reflex paths—to produce a motor effect."

Flynn proposed a "neurodevelopmental synaptogenesis" model of infantile strabismus. The anatomic site is the cortical subplate of the occipital lobes: "Strabismus is, in its clinical phenomenology, the expression of the abnormal connectivity existing in the central nervous system. Primitive synaptic connections, probably programmed during the wait in the subplate layer of the cortex and executed then or after the neurons arrive at their destination in the cortical layers, do not disappear and, as a result, abnormal connections persist within the cortex, a true dystrophy of connections."

Flynn goes on to hypothesize a coexisting abnormality of the splenium of the corpus callosum, causing "abnormal connections between the maculae and the peripheral visual fields." The combination of an abnormal subplate and callosal connections is believed to bring about esotropia: "this abnormal array of binocular connections between the cortical representation of the macula area of the fixating eye and the extensive area of the peripheral, nonmacular retina of the nonfixating eye constitutes the abnormal disparity signal to the vergence motor system . . . the vergence control system receives a veritable torrent of binocular signal between very disparate areas of the visual fields, resulting in the abnormal ocular posture we observe."

Helveston has offered a "sensori-motor arch" model in...
which the defect causing esotropia "... seems to occur at the cortical center for motor fusion (keystone of the arch). When visual input 'meets' this imperfection at the age of maturation for ocular motility at 2 to 4 months, the 'down stream' signals emanating from the imperfect (absent) motor fusion center in the cortex are flawed. This flawed message is analyzed by the brain stem. This produces incompetent neural signals, which are then sent from the brain stem to the eye muscles via the cranial nerves. This results in misalignment (almost always esotropia) secondary to the unmodified or unchecked expression of the most exuberant innate ocular motor response—convergence!"

Subcortical Visual Pathway
Jampolsky’s 51, 52 (1978) "bilateral monocular esotropia" hypothesis is thematically related to the thinking of Keiner. "I offer the hypothesis that very early neonatal visual influences may be responsible for motor misalignment and anomalous motor development. Light stimulus in the premature insufficiently developed eyes (with yet incompletely resolved media diffusers in the vitreous and lens) fulfills the essential overall diffusion stimulus criterion—the chain of exaggerated monocular and binocular dominances with altered muscle tonus." According to this model, diffuse light excites a primitive brain stem motor reflex, driven by the nasal hemiretinas, which evokes bilateral, monocular adduction movements. Lang’s 64, 65 (1984) hypothesis similarly cites subcortical light inputs as the mechanism for infantile esotropia. The subcortical inputs cause “fixation on the nasal side of both foveas, and the eyes assume a convergent position. Only with development does fixation return to the fovea, but it has a tendency to slide over to the nasal retinal half.” The delay in myelination of the optic nerves may be a factor, as well as “difficulty in coordination between ocular and vestibular influences.”

Brain Stem Vergence Motoneurons

The notion of a primary defect of vergence motoneurons—"excessive tonic convergence"—has attracted the largest number of proponents. Duane 44 (1897) ascribed the esotropia to an excess of subcortical "convergence tonus" unopposed by cortical influences. Parinaud 66 (1899) and Adler 1 (1945) pointed to a primary anomaly of convergence innervation that produces, over time, secondary changes in the medial rectus muscle attachments. Parks 63, 64 (1975) noted that the mechanism for excessive convergence is unknown. Von Noorden 79 (1988) has proposed that "a delay in the development or a permanent defect of motor... vergences in a sensorially normal infant causes esotropia during the vulnerable first three months of visual immaturity under the influence of factors that destabilize the oculomotor equilibrium." These factors are "uncorrected hypermetropia and anisometropia already mentioned by Worth, excessive tonic convergence, an abnormally high AC/A ratio; or anomalies of the neural integrators for vergence movements. ..."

Brain Stem Vestibulo-ocular Pathway
Doden’s 33 (1958) implicated an unspecified lower brain stem vestibulo-ocular mechanism for esotropia, based on the observation that strabismus is highly associated with fixation nystagmus. Ciancia’s 94 hypothesis resembles Doden’s, in that children who suffer acquired pontocerebellar lesions are thought to often exhibit a combination of nystagmus and divergence paralysis that may mimic the signs of infantile strabismus.

Cranial Nerves
Snellen 66 (1913) believed the primary defect to be partial paralysis of the nerves to the extraocular muscles. The nerve palsy was presumed to be subtle enough that it does not cause a clinically paralytic incomitant esotropia. Similarly, Wright 114 proposed that the cause of infantile esotropia is a "... maturational delay of lateral rectus muscle function relative to medial rectus function," based on hypothetical delayed myelination along the lengthier path of the abducens nerve from the brain stem to the orbit. "If the motor imbalances persist for several weeks, binocularity is disrupted and abnormal sensory motor adaptations develop."

Extraocular Muscles
Scobee 88 (1948) argued that infantile esotropia is caused by inborn defects of the extraocular muscles and their tendinous attachments. Mindel 74, 75 (1980) appears to partly share the views of both Snellen and Scobee. Mindel postulates that the cause of esotropia is a relative excess of en grappe acetylcholine receptors on the medial rectus muscles. A secondary increase in such receptors would be expected after chronic excitation of the muscles from any cause. Mindel implies a primary excess. Rather than an excess of muscle receptors, Porter 88, 90 postulated that convergent strabismus may result from a relative excess of tonic fibers within immature medial rectus muscles.

CURRENT NOTIONS OF VISUAL CEREBRAL MECHANISMS
The afferent visual system operates in the first months of life not as a binocular system but as two parallel, overlapping, monocular visual channels. 12, 39, 98 At birth each channel displays, at the level of the primary visual cortex, a directional bias favoring nasally directed motion. 27, 29 The available electrophysiologic evidence suggests that each eye actually drives visual cortical neurons that will respond to either nasally directed or temporally directed target motion but that in the first months of life only the nasally directed pole responds robustly and connects to eye tracking (pursuit and optokinetic) motoneurons. 100, 101 Stated another way, the development of temporally directed pursuit tracking depends on the development of binocular connections in the visual cortex. 59, 120 If binocularity fails to develop, pursuit remains permanently biased for nasally directed motion (Fig. 8–2). Similar mechanisms could be operating to bias vergence motoneurons in a naso direction. For example, in normal adult humans and monkeys, when the visual motion of a target differs in magnitude or direction in the two eyes, the motor responses of the two eyes also differ, so that the movement of each eye matches the visual motion seen by that eye. 57, 68 The disconjugate motor response is proof that the brain is capable of keeping monocular information sege-
Figure 8-2. Neural network diagrams that summarize findings in strabismic monkeys and provide an anatomic and physiologic basis for asymmetric pursuit, latent nystagmus, and motion visual-evoked potential (VEP) asymmetries in infantile strabismus. Paucity of binocular connections between ocular dominance columns in cortex is the only abnormality required to produce the behavioral asymmetries. Signal flow is initiated by a moving stimulus in the monocular visual field, which evokes a response in visual area neurons. In both the normal and strabismic brain, visual area neurons drive pursuit area neurons in each cerebral hemisphere that encode only conjugate, ipsilaterally directed pursuit. In infantile strabismus, binocular connections between visual area neurons are deficient, so that only ipsilateral (nasally directed) visual area neurons are connected to the pursuit area, evoking strong nasalward pursuit. In examples shown, viewing is through the right eye. Bold line indicates active neurons and neuronal projections. Left diagram, from top to bottom. When viewing monocularly, normal monkeys display symmetric responses to nasally directed versus temporally directed target motion in the temporal or nasal visual hemifields. Retinal ganglion cell fibers from the nasal and temporal hemiretinas (eye) decussate at the optic chiasm (chi), synapse at the lateral geniculate nucleus (LGN), and project into alternating rows of ocular dominance columns in striate cortex (rectangles). In each cerebral hemisphere, columns representing the nasal hemiretinas (temporal visual hemifield) occupy slightly more cortical territory than those representing the temporal hemiretinas (nasal hemifield), but each column contains neurons sensitive to nasally directed versus temporally directed motion (half circles shaped like the matching hemifield; arrows indicate directional preference). Binocular connections between neighboring columns of opposite ocularity are numerous, linking neurons with similar orientation/directional preferences (diagonal lines between columns). Nasally or temporally directed motion viewed by the right eye excites visual area neurons that prefer either direction of motion. Visual neurons preferring nasally directed motion project to the left hemisphere pursuit area; visual neurons preferring temporally directed motion project to the right hemisphere pursuit area. Note that for temporally directed visual area neurons to gain access to the right pursuit area, they must connect via a binocular connection to a visual area neuron in the other eye's (left eye's) column that in turn makes a direct connection to the right pursuit area. Right diagram, in infantile strabismus, a paucity of connections between visual area columns produces a strong asymmetry. When viewing through the right eye, the only visual neurons that have direct access to a pursuit area are those that encode nasally directed (leftward) pursuit. Latent nystagmus is the result of an asymmetry of visual input to pursuit areas; when viewing with the right eye, the strabismic patient is preferentially activating the left pursuit area and deactivating the right pursuit area. Motion VEP asymmetry in strabismus may be due to feed forward/feedback circuits between pursuit area and visual area; nasally directed neurons in each hemisphere are linked by a common connection to the pursuit area, allowing coherent resonance to build a robust response. Temporally directed neurons are present and can be activated but they are isolated, and thus incoherent, call, corpus callosum, through which visual area neurons in each hemisphere project to opposite pursuit area; re, right eye; tf, temporal field; nr, nasal retina; le, left eye; nf, nasal field; tr, temporal retina.
Cortext convergences are increased. To the contralateral midbrain medial rectus motoneurons, extrastriate cortex. The pre-motoneurons could, in turn, project from nasal columns exceeding that from temporal columns, a phenomenon is more pronounced in primates who are binocularly defective, in which case a chronic bias favoring nasal vs. temporal oculardominance columns (ODC). Visual neurons from nasal hemiretinae of right eye decussates and synapses through lateral geniculate nucleus (LGN) to drive nasal ODC, and neuron from temporal hemiretinae of left eye synapses through LGN to drive temporal ODC. Visual connections to nasal ODCs (solid lines) are established earlier, outnumber, and are more robust than connections to temporal ODCs (dotted lines). Nasal ODCs provide drive to premotor neurons in cortex, which in turn drive motor neurons in contralateral midbrain (brain stem) medial rectus subnucleus. Temporal ODCs provide similar drive to pontine abducens nucleus. Activity in nasal ODCs exceeds that from temporal ODCs, producing greater drive to medial rectus (+++) as opposed to lateral rectus (+). Net effect is convergence bias. Any congenital, perinatal, or neonatal insult that slows cerebral development would unfairly punish inputs/outputs from less-robust temporal ODCs, accentuating the convergence bias.

Figure 8–3. Signal flow diagram showing how set point for tonic vergence could be influenced by inequalities in aggregate activity of nasal vs. temporal oculardominance columns (ODC). Visual neuron from nasal hemiretinae of right eye decussates and synapses through lateral geniculate nucleus (LGN) to drive nasal ODC, and neuron from temporal hemiretinae of left eye synapses through LGN to drive temporal ODC. Visual connections to nasal ODCs (solid lines) are established earlier, outnumber, and are more robust than connections to temporal ODCs (dotted lines). Nasal ODCs provide drive to premotor neurons in cortex, which in turn drive motor neurons in contralateral midbrain (brain stem) medial rectus subnucleus. Temporal ODCs provide similar drive to pontine abducens nucleus. Activity in nasal ODCs exceeds that from temporal ODCs, producing greater drive to medial rectus (+++) as opposed to lateral rectus (+). Net effect is convergence bias. Any congenital, perinatal, or neonatal insult that slows cerebral development would unfairly punish inputs/outputs from less-robust temporal ODCs, accentuating the convergence bias.

Cerebral mechanisms unrelated to visual motion or pursuit might also produce a nasal vergence bias. In each occipital lobe of monkeys with natural infantile esotropia, nasal retinal-driven (temporal hemifield) oculardominance columns occupy about 10% more territory than temporal retinal-driven (nasal hemifield) columns, and the metabolic activity of the nasal columns is approximately 20% higher than that of the temporal columns. The set point for tonic vergence could depend on something as simple as the aggregate activity of nasal versus temporal columns. In this scheme, a primary set of connections might project from nasal retinal columns in the striate cortex to vergence pre-motoneurons in the extrastriate cortex. The pre-motoneurons could, in turn, project to the contralateral midbrain medial rectus motoneurons (Fig. 8–3). The putative neural rule would be that, if activity from nasal columns exceeds that from temporal columns, a convergence signal is increased.

A discussion of cerebral mechanisms should also note the analogy between medial rectus hypertonicity in esotropia and skeletal muscle hypertonicity in cerebral palsy. In cerebral palsy, damage to the somatic motor regions of the cerebral hemispheres results in disinhibition of spinal motoneurons and flexor muscle contractures of the arms and legs. In infantile esotropia, damage to oculomotor regions of the cerebral hemispheres might cause disinhibition of brain stem vergence neurons and flexor (medial rectus) hypertonicity of the eyes. If binocular disparity signals were unavailable, the imbalance in innate flexor-extensor (medial rectus/lateral rectus) tone would need to be only a few percentage points to bias the system toward esotropia.

**NEUROANATOMIC FINDINGS IN INFANTILE ESOTROPIA**

What has clinical observation and basic science taught us about the visual cortex in infantile strabismus? Hubel and Wiesel, in 1977, reported a series of experiments in monkeys describing the functional architecture of the normal primary visual cortex (striate cortex or area V1). The primary cortex can be divided into layers containing neurons with different response properties, organized in columns such that alternating columns receive input from only the right or the left eye. When signals were recorded from these neurons, the majority responded to both eyes, implying the presence of binocular connections between oculardominance columns (ODCs), although the binocular connections themselves could not be visualized using available anatomic methods. Subsequently, Crawford and von Noorden produced artificial strabismus in infant monkeys and recorded an absence of binocular responsiveness when the animals were tested as adults. These important experiments proved that eye alignment in infancy is necessary for the development of binocular responsiveness. However, the experiments left several major questions unanswered: (1) Were these animals an appropriate behavioral model of human infantile esotropia? (2) What was happening structurally to neurons within ODCs that could account for the loss of binocular responses? (3) Was loss of binocular responsiveness accompanied by metabolic alterations in the ODCs?

To answer these questions, we studied macaque monkeys (originally described by Kiorpes and Boothe) that had an onset of natural alternating esotropia at age 4 to 6 weeks (the equivalent of 4 to 6 months in human development). The animals were determined to be a good behavioral model of human infantile esotropia (Table 8–1): the classic deficits in binocu-

<table>
<thead>
<tr>
<th>Table 8–1. Functional Deficits in Infantile Esotropia</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Fusion Deficits</strong></td>
</tr>
<tr>
<td>Absence of disparity vergence (motor fusion)</td>
</tr>
<tr>
<td>Lack of two-dimensional fusion and stereopsis</td>
</tr>
<tr>
<td>Subnormal binocular visual-evoked potential response</td>
</tr>
<tr>
<td><strong>Motion/Pursuit Deficits</strong></td>
</tr>
<tr>
<td>Asymmetric monocular tracking</td>
</tr>
<tr>
<td>Asymmetric monocular motion visual-evoked potential</td>
</tr>
<tr>
<td>Asymmetric motion perception</td>
</tr>
</tbody>
</table>
lar sensitivity (motor and sensory fusion) and a second set of more recently described but equally characteristic deficits in motion processing (as measured by motion visual-evoked potentials) as well as pursuit eye tracking. The motion/pursuit deficit is manifest as a strong bias favoring nasally directed target motion when viewing with either eye.

Layers 2 and 3 of the primary cortex in normal monkeys are known to contain neurons important for fine stereopsis, whereas layer 4b contains neurons most responsive to targets moving in specific directions at specific speeds. Therefore, the functional deficits in fusion versus motion processing in the strabismic animals implied structural deficits in the neural circuitry in different cortical layers. To examine cortical connections at different V1 layers, we performed the double-labeling experiments shown in Figure 8–4. A tracer (in this case a large sugar, biotinylated-dextran-amine [BDA]), was injected into ODCs. The tracer was taken up by individual neurons and actively transported to their axonal branches to reveal binocular connections on target neurons in other ODCs. A second set of labels (wheat germ agglutinin-horse-radish peroxidase and cytochrome oxidase) was used to reveal the ocularity of the neurons, that is, whether they resided in the right or left eye ODC.

Table 8–2. Functional-Structural Correlation in Infantile Esotropia

<table>
<thead>
<tr>
<th>Functional Deficit</th>
<th>Structural Deficit</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lack of fusion and binocular visual-evoked potential</td>
<td>Lack binocular connection layers 2 &amp; 3</td>
</tr>
<tr>
<td>Asymmetric pursuit and motion visual-evoked potential</td>
<td>Lack binocular connection layer 4b</td>
</tr>
<tr>
<td>Alternating suppression</td>
<td>Inhibition metabolic activity layer 4c</td>
</tr>
</tbody>
</table>

Quantitative analysis of the cortex in these animals revealed the findings summarized schematically in Figure 8–4 and in Table 8–2. Natural alternating infantile esotropia is associated with striking abnormalities of cortical wiring and abnormal suppression of ODC connections between neighboring ODCs. Monkeys with infantile esotropia had few

Figure 8–4. Neuroanatomic abnormalities found in area V1 of monkeys with natural infantile esotropia who alternated fixation and had normal visual acuity in both eyes: lack of binocular connections and nasotemporal inequalities of metabolic activity. A. Strabismic monkey has a paucity of horizontal connections for binocular vision; neurons are connected within individual ocular dominance columns (ODCs), but there are few connections to neighboring ODCs of the opposite ocularity. Connections to other ODCs of the same ocularity (e.g., right eye to right eye ODCs) are not shown for the sake of clarity. Layer 4c of right eye ODCs in the left V1 (i.e., contralateral eye or nasal retina ODCs) stains more intensely (dark shading) for cytochrome oxidase activity, indicating higher metabolic activity in "nasal" ODCs. B. In normal monkeys, neurons in layers 2 and 3 form part of the parvo-interblob pathway, which plays a major role in fine (random dot) stereopsis. Neurons in layer 4b form part of the magnocellular pathway, which plays a major role in motion perception and pursuit eye movement.
horizontal connections between neighboring ODCs. The lack of binocular horizontal connections was especially apparent in layers 2 and 3 and layer 4b. The cortex of the strabismic animals also showed inhibition of metabolic activity (as measured by cytochrome oxidase) in columns driven by the ipsilateral eye (i.e., temporal retina) in each cerebral hemisphere. The inhibition was most apparent at the input layer of the cortex—layer 4c. This implies that the nasal columns, which receive input from the contralateral eye, were able to suppress activity in the neighboring temporal columns that receive input from the ipsilateral eye. This nasotemporal inequality in metabolic activity may be due to heightened competition between ODCs caused by uncorrelated activity of each eye's input. 72–93

But why does suppression of temporal ODCs in both cerebral hemispheres occur? The reasons are not entirely clear but may be related to the fact that nasal inputs (i.e., inputs from the contralateral eye to each primary cortex), are established earlier in development and slightly outnumber (by a ratio of about 53:47 in monkeys) inputs from the ipsilateral eye. 115, 116 The heightened competition between ODCs caused by conflicting binocular images (and alternating suppression) may unfairly punish the less numerous ipsilateral inputs. Another way of describing the metabolic abnormality is to say that alternating strabismus is associated with partial shutdown of about 50% of the neurons in each area V1.

Are these cortical metabolic and wiring abnormalities primary or secondary? It is reasonable to consider the possibility that cortical abnormalities could be primary and cause the motor signs of infantile strabismus. First, the cortex is the initial locus in the central nervous system at which visual signals from the two eyes are combined 47 and a combination of visual signals is necessary to generate the vergence error commands that guide eye alignment. The second reason is the strong clinical linkage between perinatal insults to the immature cerebral cortex and subsequent development of the ocular motor signs of infantile strabismus. (A representative list of studies supporting this statement is shown in Table 8–3.) However, linkage does not prove causation, and it is entirely possible that the cortical abnormalities documented in the strabismic monkeys were purely secondary—the result of abnormal binocular experience.

**Clinical Characteristics**

**CLASSIC PRESENTATION**

The paradigmatic infant who develops strabismus begins to manifest a chronic esodeviation of the visual axes at 2 to 3 years of age. Infants with strabismus are prone to hypertensive injury to the dorsal motion-processing area of the extrastriate cortex, the watershed zone for all three major cerebral vessels. Maternal smoking as well as drug and alcohol abuse perturb cerebral development and are associated with increases in the risk of amblyopia or strabismus equivalent to those associated with prematurity or hypoxia. 40, 73, 90

**EPIDEMIOLOGY AND RISK FACTORS**

**CLASSIC PRESENTATION**

Both genetic and environmental factors appear to play a role in the causation of esotropia. As an example of genetic factors, in the study reported by Tychsen and Lisberger 120 in 1986, the strabismic subject who had the most severe pursuit/motion processing asymmetry had two siblings with infantile strabismus. Nonstrabismic kindreds in pedigrees of infantile strabismus have been found who manifest nasally directed biases of pursuit not present in the normal population. 106 Large-scale studies have documented that 20% to 30% of children born to a strabismic parent will themselves develop strabismus. 25, 40, 54, 70, 102 Maumenee and associates 70 analyzed the pedigrees of 173 families containing probands with an onset of infantile esotropia before age 6 months in the absence of major refractive error. 70 The results from this analysis suggested a mendelian codominant model (full expression of both alleles of a pair in a heterozygote) with an admixture of autosomal recessive cases.

As for environmental factors, the prevalence of strabismus and amblyopia is substantially higher in low-birth-weight, premature infants 26, 60, 61, 67, 123 or those who suffer perinatal hypoxia (see Table 8–3). 38, 60 Infants born weighing less than 1500 g have a prevalence of amblyopia and strabismus seven times that of normal-weight term infants. 60 Infants weighing less than 2500 g at birth have a prevalence of strabismus four times that of normal-weight infants. 60 The risk of strabismus increases roughly 4% for each 100-g decrease in birth weight under 5 pounds. 40

The increased risk of strabismus in these infants is probably due to the maldevelopment of binocular connections in the visual cortex and the downstream effects of this damage on cerebral ocular motor-related neurons. The occipital lobes in newborns are especially vulnerable to damage from hypoxia. 116, 123 The striate cortex is susceptible to hypoxic injury because it has the highest neuron-to-glia ratio in the entire cerebrum 49 and the highest regional cerebral glucose consumption. 66 Premature infants frequently suffer ischemic damage to the optic radiations (periventricular region) near the occipital trigone. 25, 24 Both premature and full-term infants are prone to hypertensive injury to the dorsal motion-processing area of the extrastriate cortex, the watershed zone for all three major cerebral vessels. Maternal smoking as well as drug and alcohol abuse perturb cerebral development and are associated with increases in the risk of amblyopia or strabismus equivalent to those associated with prematurity or hypoxia. 40, 73, 90

<p>| <strong>Table 8–3. Abnormalities of Cortical Development Predisposing to Infantile Strabismus</strong> |</p>
<table>
<thead>
<tr>
<th>Type</th>
<th>Prevalence Strabismus</th>
<th>Author(s)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Occipitoparietal hemorrhage and/or leukomalacia</td>
<td>57%</td>
<td>Pike et al., 99 1994</td>
</tr>
<tr>
<td>Intraventricular hemorrhage with hydrocephalus</td>
<td>100%</td>
<td>Tamura and Hoyt, 104 1987</td>
</tr>
<tr>
<td>Very low birth weight infant (&lt;1500 g)</td>
<td>33%*</td>
<td>van Hof-van Duin et al., 123 1989</td>
</tr>
<tr>
<td>Down syndrome</td>
<td>21%–42%</td>
<td>Hiles et al., 14 1974</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Shapiro and France, 97 1985</td>
</tr>
</tbody>
</table>

*Additional 17% of infants had persistent asymmetric OKN.
Figure 8–5. Serial photographs of the same infant taken at 1-month intervals spanning ages 1 to 8 months, showing absence of esotropia at age 1 to 2 months and onset of esotropia at age 3 to 4 months. Esotropia increased in magnitude after onset. Infant was referred by the pediatrician at age 8 months and operated on the same month.

4 months of age (Fig. 8–5).\textsuperscript{5, 6, 13, 100} Transient episodes of misalignment may precede this by several weeks and may account for the history often given by the parents that the eyes crossed “at birth.” Chronic esotropia in the neonatal period is rare.\textsuperscript{5, 6, 13, 100} If it is well documented by good serial photographs or ophthalmologic examination, the major concern is Duane syndrome or neonatal sixth nerve palsy, not classic infantile esotropia.\textsuperscript{108}

Classic infantile esotropia is constant and cosmetically conspicuous, typically exceeding 20 PD on corneal light reflex measurement.\textsuperscript{21, 26, 71, 129, 130} The infant cross-fixates (Fig. 8–6) so that the targets of interest presented to the left of the head elicit fixation with the right eye and targets presented to the right of the head elicit fixation with the left eye.\textsuperscript{32} The cross-fixation pattern may be so habitual as to obviate normal abduction of either eye under casual viewing conditions. However, if the examiner covers one eye, the infant cannot employ cross fixation and must abduct the viewing eye to track targets into the far temporal portion of the monocular field. The infant may be reluctant to abduct

Figure 8–6. Cross-fixation behavior in infantile esotropia. A, When viewing targets that are placed well to the right of the midline of the head, the infant prefers to fixate with the left eye. B, When viewing targets placed to the left side of the head, the infant prefers to fixate with the right eye.
the full excursion of the orbit but generally will abduct briskly to the midline or slightly beyond, verifying that lateral rectus/abducens nerve function is grossly intact. The esotropia is comitant (more or less the same magnitude) in right gaze, left gaze, up gaze, and down gaze (Fig. 8–7). Refraction using cycloplegia classically reveals a small amount (+2.00 D or less) of hypermetropia.

When one eye is covered and the infant is induced to attentively fix on a small accommodative target held at midline or in slight abduction (not a trivial task for the examiner), latent nystagmus may be observed. The nystagmus is seen as a repetitive series of abducting jerks, directed to the right when the infant views with the right eye and to the left when viewing with the left eye. The nystagmus is easy to miss if the infant is inattentive or keeps the eye in the nasal orbit (which dampens or eliminates the nystagmus). With the fixing eye in the nasal orbit, the infant will display an alternating head turn: the face is turned to the right when viewing with the adducted right eye, and to the left when viewing with the adducted left eye.

**SPECTRUM OF CLINICAL PRESENTATIONS**

A substantial proportion of infants who develop strabismus becomes esotropic beyond age 2 to 4 months. As many as 10% may not display a constant esotropia until as late as 9 to 12 months of age. These children usually do exhibit an intermittent esodeviation earlier in infancy.

The magnitude of the strabismus may increase in the first few weeks or months of observation, and the angle can vary depending on the level of attention. In general, the greater the degree of attentiveness, the larger the esotropia. The presence and severity of latent nystagmus varies. In about 20% of infants, latent nystagmus may be clinically undetectable. In a smaller number (approximately 5%), latent nystagmus is severe enough to result in variants of the "nystagmus blockage" phenomenon, in which the angle of esotropia may increase greatly when the infant attentively fixates. In these cases, the infant uses convergence to dampen nystagmus and improve acuity.

Incomitance may also be observed; the most common type is a V pattern, in which esotropia is greater in down gaze and less in up gaze. V-pattern infantile esotropia is commonly, but not invariably, associated with overaction of the inferior oblique muscles.

Other variants the clinician may encounter include either a combination of refractive (hypermetropic) and "baseline" infantile esotropia or high accommodative convergence/accommodation ("high AC/A") esotropia with infantile esotro-
The combined refractive-infantile esotropes show a substantial reduction in the total angle of strabismus when wearing hypermetropic spectacles (Fig. 8–8). Spectacles alone, however, seldom restore full orthotropia. The high AC/A-infantile esotropes will have 10 to 15 PD more esodeviation when fixating near, as opposed to distant, targets. Infantile microesotropes (infants who have small, cosmetically undetectable angles of esodeviation) rarely are diagnosed in infancy unless they display obvious latent nystagmus. More commonly, the diagnosis is made later in childhood when defective stereopsis and amblyopia are detected and cover testing for microtropia is easier to perform. The presence of latent nystagmus and dissociated vertical deviation (DVD) in these cases allows the examiner to retrospectively date the onset of strabismus to the first year of life.

**Diagnosis**

Infants who develop strabismus begin to exhibit a constellation of ocular motor signs: (1) esotropia, with or without strabismic amblyopia; (2) pursuit asymmetry; (3) latent fixation nystagmus; (4) motion visual-evoked potential (VEP) asymmetry and motion perception abnormalities; (5) a face turn and abduction deficit; and (6) vertical deviation. An understanding of the developmental mechanisms underlying these signs simplifies their detection and interpretation.

**ESOTROPIA**

Before age 3 to 5 months, many normal infants show unstable vergence movements of the eyes with some degree of esotropic or exotropic eye misalignment (Fig. 8–9). The magnitude and frequency of eye misalignment systematically diminish until a vergence “lock” appears to be established in normal infants at some time in the third to fifth postnatal month. It is likely that binocular connections in layers 2 to 4 of the visual cortex play a crucial role in establishing this lock.

Parental reports regarding the onset of esotropia are notoriously inaccurate. It is the exceptional parent of a child with infantile esotropia who does not insist that the eyes crossed at birth, despite firm clinical evidence to the contrary. As

---

*Figure 8–8. Nine-month-old infant who had infantile esotropia and superimposed refractive esotropia (refractive error = +4.50 D in both eyes). Spectacles for the full amount of the refractive error were prescribed, which reduced the total magnitude of the strabismus by about half—from 50 PD to 25 PD. Surgery was performed for the nonrefractive component of the esotropia (i.e., 25 PD).*

*Figure 8–9. Photographs of infant taken at age 1 week and age 6 months. A. An esodeviation of the eyes is obvious in the photograph taken at 1 week. The deviation was variable, and doll’s head (vestibulo-ocular reflex) maneuver elicited normal abduction of each eye. B, The transient (physiologic) esodeviation resolved. Follow-up at age 4 and 6 months revealed orthotropia and normal eye movements.*
stated earlier, constant esotropia in the neonatal period is more commonly due to sixth nerve palsy or Duane syndrome.  

The presence of strabismic ambylopia is assessed by using the “fix, follow, maintain” criteria. The simplest method is to judge the degree to which the infant objects to the covering of each eye in turn. Judgment of the relative acuity of each eye can also be made on the basis of how far the infant will follow a target under conditions of binocular viewing. The target is moved from a nasal to a temporal position with respect to the fixing eye. An esotropic infant with equal visual acuity in the two eyes (i.e., a “cross-fixator”) should follow the target until it reaches the midline, at which point fixation switches from one eye to the other. If there is significant asymmetry in vision, the infant will tend to follow with the better-seeing eye well into abduction before relinquishing fixation to the ambyopic eye.  

The induced vertical tropia test is a useful supplement. A 15-PD base-down prism is placed alternately before the eyes. The examiner should see the same behavior when the prism is suddenly introduced into the visual axis of each eye. The infant either makes a vertical saccadic eye movement to look at the displaced target through the prism or maintains fixation on the horizontal plane with the eye that is not viewing through the prism. The infant normally may employ either strategy. Amblyopia is implied when the infant consistently prefers to fix with one eye; thus, upward saccades are always generated and the eye remains upwardly deviated when the prism is introduced before the dominant eye and upward saccades are never generated when the prism is introduced before the ambyopic eye.  

In infants who have conspicuous latent nystagmus, a crude method of detecting ambylopia is to gauge the amplitude of the nystagmus jerks while the eyes are covered in turn. Generally, the eye with worse acuity will have larger-amplitude abducting jerks.

Pursuit asymmetry

Infants in whom normal binocularity fails to develop exhibit asymmetric horizontal pursuit. When one eye is occluded and a hand-held toy is moved from temporal to nasal before the fixing eye, pursuit is smooth (Fig. 8–10). Pursuit is absent or jerky (cogwheel or low gain) when the target moves nasal to temporal. The movements of the two eyes are conjugate (i.e., when fixing with the nonfixing eye, hence the term latent) (Fig. 8–12). The nystagmus will be missed if the examiner cannot view the “fix, follow, maintain” criteria. When attempting to fixate a small stationary target, the eyes drift nasally with respect to the fixing eye (the velocity of the slow drift and the number of corrective fast-phase jerks are accentuated by covering the nonfixing eye, hence the term latent) (Fig. 8–11). The nystagmus is detectable as a brief series of small jerks that are temporally directed with respect to the fixing eye. The nystagmus will be missed if the examiner cannot get the infant to fixate attentively.  

As is true with pursuit asymmetry, the direction of the nystagmus reverses instantaneously with a change in the fixing eye: the direction of the slow drift is always nasally directed with respect to the fixing eye, and the movements of the two eyes are conjugate (i.e., when fixing with the right eye, both eyes drift leftward and jerk rightward). The nystagmus persists into adulthood despite surgical correction of strabismus and thus serves (as does the pursuit asymmetry) as a permanent marker of abnormal binocular motion neuron development.  

Studies of motion perception and smooth pursuit, in children and adult patients who had infantile strabismus, suggest a visual mechanism for latent nystagmus. Individuals who have the nystagmus tend to perceive stationary targets as though they were moving nasally at low velocities. The defect of cortical motion circuitry causing the misperception also causes the pursuit and fixation motor systems to remain switched-on for nasal pursuit. The inappropriate pursuit takes the eyes off the target, visual acuity is degraded, a saccadic refixation occurs, and the cycle is repeated. Cognitive adaptation to the motion defect takes place very early in life, as indicated by a lack of awareness of motion misjudgments and the absence of oscillopsia. Figure 8–2 shows the circuitry defects and summarizes recent neuroanatomic and neurophysiologic findings from studies of strabismonic monkeys who have the nystagmus and nasally biased pursuit.  

Eye movement recording and waveform analysis are not necessary to diagnose the nystagmus clinically. Viewing the optic disc with a direct ophthalmoscope (while the patient fixes a small target with the nonoccluded eye) is a sensitive means of detecting the nystagmus jerks when they are not apparent on gross observation (Fig. 8–12). In total darkness, the velocity of nystagmus decreases to an average of 30% of that recorded when the eyes fixate in light. Persistence of nystagmus in total darkness does not indicate that it has a “motor” origin. Rather, it suggests that chronic visual drive can produce long-term adaptations in motoneurons that persist when the visual drive is temporarily removed.
The prevalence of latent nystagmus in classical infantile esotropia probably approaches 100%, but no large-scale studies using quantitative recordings have been done. Gross clinical observations indicate a prevalence exceeding 50%. Small-scale studies using eye movement recording indicate a prevalence exceeding 95%.

The velocity of latent nystagmus varies from one individual to another, as does the magnitude of infantile esotropia. Infants with high-velocity latent nystagmus (greater than 1.5 degree/sec) often have large, variable angles of esotropia. They may superimpose bouts of convergence upon a baseline of esotropia. The convergence dampens the nystagmus velocity and improves visual acuity, hence the terms nystagmus blockage syndrome and nystagmus compensation syndrome. Rather than representing a unique entity, nystagmus blockage syndrome appears to represent the extreme end of the spectrum of latent nystagmus and esotropia. Augmented amounts of strabismus surgery are needed to counteract the strong esodrive in this situation, which fosters recurrence of the esotropia.

Although latent nystagmus is usually associated with infantile esotropia, it may result from any visual lesion that interrupts binocular development in the first 6 months of life. Examples include monocular cataract, infantile glaucoma, corneal leukemia, marked anisometropia, and infantile constant exotropia or hypertropia. Latent nystagmus and the pursuit asymmetry occur infrequently in individuals who have no strabismus or amblyopia (primary maldevelopment of the visual motion pathway). The primary maldevelopment has a prevalence approaching 5% in children with Down syndrome and in first-degree nonstrabismic relatives in pedigrees in which multiple individuals have classic infantile esotropia. These observations suggest that genetic factors play an important role in development of the visual pursuit/motion pathway.

Latent fixation nystagmus is the only form of nystagmus...
that reverses direction instantaneously with a change of the fixing eye. For this reason, it is easily distinguished from all acquired forms of nystagmus. Latent fixation nystagmus is the most common pathologic nystagmus encountered by ophthalmologists (see Chapter 31).

**MOTION VISUAL-EVOKED POTENTIAL ASYMMETRY**

Motion VEPs provide additional evidence that the directional asymmetry of pursuit and latent nystagmus is due to cortical maldevelopment. Esotropic infants have asymmetries in their VEP response to horizontally oscillating stimuli (Fig. 8–13), responding robustly to only one direction of horizontal motion when viewing monocularly. The responses are directionally inverted by 180 degrees in the two eyes, analogous to the nasotemporal asymmetry of eye movements and motion perception. The motion VEP asymmetry tends to resolve in esotropic infants who have early surgical realignment of the eyes but persists in children and adults with uncorrected esotropia. The motion VEP asymmetry is not present in children who develop strabismus after infancy. It serves as an additional diagnostic marker for maldevelopment of binocular vision and an indicator documenting repair of the maldevelopment after early strabismus surgery (see later).

**FACE TURN AND ABDUCTION DEFICIT**

Infants with latent fixation nystagmus and the pursuit asymmetry prefer to view targets by placing the eye at a nasal position in the orbit. This is achieved by turning...
Funduscopy using direct ophthalmoscope is a sensitive method for detecting small-amplitude latent nystagmus not visible by gross observation. Left, Examiner's view of child's left fundus while child fixates distant target with the right eye. Right, Examiner's view of the child's right fundus while child fixates with the left eye. The light of the direct scope acts like an occluder over the nonfixing eye. The examiner focuses on a retinal vessel or the optic disc. Latent nystagmus is evident as rhythmic jerks of the fundus (arrows). The direction of the jerking in the fundus will appear opposite to that of the anterior segment of the eye.

The face toward the fixing eye (Fig. 8–14). Eye movement recordings indicate that, with the fixing eye in the nasal orbit, the velocity of nystagmus decreases an average of 25%. The reduced nystagmus velocity improves visual acuity. A consistent face turn in one direction often indicates amblyopia in the eye that is in a more temporal position in the orbit (the left eye in an infant with a right face turn).

Infants who have esotropia may appear as though they have limited abduction. The abduction deficit, if moderate, may be overcome by vigorously rotating the head (doll's head maneuver or vestibulo-ocular reflex). Frequently, marked degrees of abduction deficit are not fully overcome by rotation. The clinical notion that the abduction deficit is due to chronic medial rectus muscle contracture is reasonable, given the fact that alternate patching of the eyes over several days considerably improves abduction. A failure to improve abduction by patching indicates aplasia of the abducens nerve (Duane syndrome) or abducens palsy.

Figure 8-12. Motion visual evoked potential stimulus and response. The stimulus was a vertically oriented sinusoidal-contrast grating that oscillated left and right several times per second on a video screen. F1 (asymmetric) signals are evoked from cortical motion neurons in young normal infant human and monkey (shown) before development of binocular vision. A strong F1 response persists in uncorrected infantile strabismus. F2 (symmetric) signals represent the response of children and adults who have normal binocular vision. Time in graph on right progresses from top to bottom.
Figure 8-14. Infants who have latent nystagmus and the pursuit asymmetry prefer to view targets by placing the eye at a nasal position in the orbit. This is achieved by turning the face toward the fixing eye, which reduces nystagmus velocity. A, A 6-month-old infant with esotropia who turns the face to the right when viewing with the right eye and to left (B) when viewing with the left eye. A 7-month-old infant with esotropia viewing with (C) right eye and (D) left eye. Alternating face turn implies near equal acuity in the two eyes. A consistent face turn in one direction often indicates amblyopia in the eye that is opposite the direction of the face turn (the left eye in an infant with a right face turn).

DISSOCIATED VERTICAL OR HORIZONTAL DEVIATION

Dissociated vertical deviation (DVD) is characterized by an upward-directed slow movement of the nonfixing eye. The hallmark of the deviation is that it violates Hering’s law of equal innervation. The fixing eye does not move, or moves minimally, whereas the eye with the DVD is moving up under cover and down when uncovered as much as 10 degrees. DVD has been subdivided into several variants: dissociated hypertropia, dissociated hyperphoria, and dissociated horizontal (exo)deviation. The last term is used when the main vector of the movement is directed temporarily rather than upward. Wilson has termed this the dissociated strabismus complex.

Studies of pursuit and motion perception in individuals with DVD have revealed vertical asymmetries analogous to those that are horizontal. Patients who have DVD are more sensitive to upward-directed motion, measured as better pursuit of upward-directed moving targets and misperception of downward target velocities. The vertical deviation of the eyes in DVD changes markedly depending on whether the patient is erect, supine, or supine in a hyperextended (“head hanging”) neck position, indicating that static vestibular (otolithic) mechanisms may play a role. Taken together, these studies suggest a combined visual cortical and brain stem mechanism for DVD. Failure to develop normal binocularity leads to persistence of immature monocular circuits, one of which is visual and biased for upward motion and the other vestibular. In push-pull fashion these promote upward motion of one eye and relative downward motion of the other. When the weak binocular cortical connections of the strabismic patient are perturbed (e.g., by dissociating the eyes using a cover test or a neutral-density filter), the monocular visual/vestibular biases become manifest.

The reported prevalence of DVD in infantile esotropia ranges from 76% to 88%. It is nearly always
bilateral, but of differing magnitude in the two eyes. It is rarely detected in infants. Typically DVD appears in preschool-age and school-age children who have had horizontal muscle surgery to correct esotropia earlier in life. Head tilt is a frequent finding and is most often manifested as tilting of the head toward the shoulder that is contralateral to the eye exhibiting the greatest DVD amplitude when the head is upright. Pratt-Johnson postulated that the head tilt in DVD was an attempt to compensate for the torsional micro-nystagmus that frequently accompanies latent nystagmus. Precise eye movement recordings in adult patients who have DVD support this notion. The excyclo-elevation movement of the nonfixing eye in the DVD patients was highly similar to the cycloverision behavior of normal humans who were tested under conditions that evoked disparity-induced vertical vergence.

CONSTANT INFANTILE EXOTROPIA: A NEURO-OPHTHALMIC DISORDER

The ophthalmologist must be particularly diligent in ruling out neuro-ophthalmic abnormalities in any infant presenting with constant exotropia, as opposed to esotropia, in the first 12 months of life (Fig. 8–15). This dictum does not apply to infants who display early-onset intermittent exotropia, nor does it apply to normal infants younger than 3 to 5 months of age who display a transient physiologic exodeviation in early infancy.

The ratio of infantile esotropia to constant infantile exotropia at our institution is greater than 10:1. Unlike the majority of infants with esotropia, more than 90% of those with constant exotropia have significant eye or brain abnormalities such as optic nerve hypoplasia, morning glory anomaly of the optic disc, retinoblastoma, microcephaly, infantile spasm, encephalomalacia, or static encephalopathy. Similar findings were reported by Zak in infants with constant exotropia. The true ratio of infantile esotropia to exotropia in our demographic area most likely exceeds 10:1, because it is reasonable to assume that physicians would be less likely to refer classic infantile esotropes and more likely to refer the early-onset exotropes.

Thus, constant exotropia in infancy should be considered unusual enough to warrant careful neuro-ophthalmic examination for a relative afferent pupillary defect, a visual field defect (which may be tested using the evoked-saccade method), ptosis or other evidence of third nerve palsy, anomalous optic discs, nerve fiber layer loss, a history of seizures, or failure to thrive (see also Chapter 13).

Treatment

NONSURGICAL MANAGEMENT

Glasses

A good refraction with full cycloplegia (e.g., using 2.5% phenylephrine [Neo-Synephrine] and 1% cyclopentolate) should be performed on all esotropic infants. Covering one eye at a time during retinoscopy helps to ensure that the examiner remains accurately aligned with the visual axis. A skiascopy rack containing an array of lens powers, ranging from 0.5 to 15 D, speeds the refraction in fussy infants. The examiner may occasionally have to restrain an infant with the help of an assistant to ensure reliable results.

Spectacles are generally prescribed when the degree of hyperopia exceeds +2.50 D and/or when anisometropia exceeds +1.50 D. Any cylinder of +0.50 D or more should also be given. Spectacles should be prescribed for myopia exceeding −4.00 D. The rationale for correcting moderate to high hyperopia is to eliminate a significant refractive esotropia superimposed upon the “baseline” infantile exotropia. Correction of a high degree of myopia is logical in that (1) the clearer the images seen by the infant, the greater the likelihood of accurate fixation and, hence, obtaining accurate strabismus measurements; and (2) minus lenses may alter the accommodative demand and the infant’s strabismus angle, especially when fixing near targets.

Oclusion Therapy for Amblyopia

If amblyopia is detected, occlusion therapy is instituted after the first office visit (Fig. 8–16). If a strong fixation preference for one eye is detected, high-percentage occlusion (e.g., 90% of waking hours) is prescribed using opaque skin patches (Opticlude or Coverlet). The infant is reexamined in several weeks to gauge improvement and to rule out occlusion-induced amblyopia of the initially dominant eye. The general rule of thumb is 1 to 2 weeks of high-percentage occlusion per year of life before reassessment. If such frequent checks are impractical, lower-percentage occlusion may be used each day. Alternatively, some regimen of alternate occlusion may be employed; for example, the dominant eye can be patched 4 days and the amblyopic eye for 1 day in repeated cycles. If spectacles were prescribed (and the infant will keep them on), an opaque felt occluder with side shield (Patchworks) may be worn affixed to the glasses in lieu of a skin patch. The endpoint of patching is to produce a pattern of freely alternating equal vision. However, if this goal is not achieved in a timely manner (usually owing to
resistance on the part of the infant), strabismus surgery should not be deferred.  
When the examiner concludes that the infant has equal or nearly equal visual acuity in the two eyes, a strategy of alternate patching up to the date of surgery has some distinct advantages.  
First, if the examiner has missed a fixation preference during office examinations, alternate patching will help correct strabismic amblyopia by ensuring that, at least part of the time, the infant is forced to view exclusively with the weak eye. Second, alternate patching reduces the occurrence of strabismic suppression, because the infant does not view with abnormal binocular vision. Third, alternate patching may reduce the amount of chronic medial rectus contracture because the infant cannot cross-fixate and so is forced to abduct the viewing eye to track targets into the temporal visual hemifield. And, fourth, prolonged alternate occlusion (i.e., over a period of months) has been shown to reduce the nasotemporal motion processing deficits of infantile strabismus.  
A side benefit of prescribing alternate occlusion up to the day of strabismus surgery is that it keeps the child’s family actively involved in the treatment and eager to carry through with the surgical procedure so as to end the burden of patching.

SURGICAL MANAGEMENT
Rationale for Early Surgical Correction
The results of several clinical studies imply that defects of cortical binocular wiring in infantile esotropia can be repaired, in some infants to a remarkable degree. Careful psychophysical experiments found that a substantial proportion (41%) of infants whose eyes were aligned to within 8 PD in the first 16 months of life had the restoration of random-dot stereopsis on follow-up years later and that those whose eyes were aligned at 12 months who achieved stereopsis (49%) tended to achieve a finer grade.  
Similar and even more impressive results have been documented in a subset of infants operated on at age 4 months; 60% attained fine-grade random-dot stereopsis.

In addition to stereopsis, it appears that the defects in the motion pathway can also be repaired in a substantial number of strabismic infants. The bar graph of Figure 8–17 shows data from two groups of human infants who were operated on before age 18 months. Testing 3 to 6 months postoperatively showed that infants whose eyes were aligned within 10 PD of orthotropia tended to show a return of symmetric motion sensitivity, a finding not as apparent in infants whose eyes were poorly aligned.

Taken together, the stereopsis and motion VEP results show that early treatment can repair cortical binocular functions in a substantial number of strabismic infants. The mechanisms likely include both the restoration and strengthening of horizontal binocular connections between ODCs, brought about by eliminating conflicting activity in neighboring ODCs and by restoring a normal pattern of cooperative binocular activity. The flow diagram of Figure 8–18 is
okn, allowing more time for repair in the form of restoration of symmetric binocular connections.111

**Surgical Timing and Preoperative Measurements**

When the surgeon has documented that the infant has a constant esotropia exceeding 12 PD, surgical realignment should be carried out as soon as is practical for the surgeon and family (assuming there are no major cardiopulmonary problems that would pose a high risk for general anesthesia). Surgery should be deferred until the ophthalmologist is convinced that at least two sets of reasonably high-quality, reproducible measurements of esotropia have been obtained. “Reproducible” here means that the measurements agree to within a range of 5 to 10 PD. Ideally, the angle of the strabismus is measured using the alternate prism cover test to gauge the full magnitude of any combined esotropia and esophoria. Prism cover testing is done carefully for distance and near fixation in primary position wearing any prescribed spectacle correction. (Prism cover measurements for distance fixation may be difficult to obtain before age 9 to 12 months, and a Krimsky or Hirschberg corneal light reflex estimate may be substituted.) Assessment of the deviation is also performed in side and vertical gazes to rule out restriction, palsy, and significant oblique muscle overaction or underaction. With highly uncooperative infants the ophthalmologist may have to rely exclusively on measurements made using the Krimsky method, or, with a very large angle of esotropia, the Hirschberg reflex.

**Preoperative Counseling**

A neurophysiologic basis for correcting esotropia was addressed in the preceding discussion. The surgeon should also provide a physiologic rationale to the family: the opportunity for brain repair with some recovery of three-dimensional vision, motion vision, accurate eye tracking, reduction of nystagmus, and elimination of conflicting images that promote amblyopia. The explanation is reduced to simple terms, conveyed briefly but clearly by either the surgeon or his assistants, verbally or in a hand-out. For most parents the physiologic rationale will make more sense than the old notion that “the eye muscles are too strong (or too weak) and need adjusting.” The parents should be told up front of the possibility that reoperation may be needed in the months and years ahead. Infants with esotropia have required, on average, 1.9 to 2.6 operations to achieve stable alignment with some motor fusion.14, 21, 36, 50, 83, 129

Sample answers to typical questions posed by parents of a strabismic infant are listed below.

- Why do my child’s eyes cross? “The brain circuits for eye alignment are miscalibrated, and send the wrong commands to the eye muscles.”
- How does operating on the muscles fix the problem? “We can straighten the eyes by repositioning the muscles on the eyeballs. By doing this, we restore more normal activity to the brain circuits, and they can repair themselves to get the eyes to stay locked together.”
- Why is patching being used up to the day of surgery? “The connections that keep the eyes locked together form best when the signal from both eyes is equally
strong. We need to strengthen (by unilateral patching) the signal from the weaker eye.” Or, “We need to keep strong (by alternate patching) the signals from both eyes.”

- Can you fix it with one surgery? “Probably not. The average child requires at least 2 surgeries, spaced months to years apart. The brain circuit may initially respond well and keep the eyes tracking together. But often the strengthening is not enough to permanently lock the eyes together, so that it may be necessary to reposition other muscles to keep the eyes within the range that the circuit can handle. The microsurgery is anatomically very precise. The unknown factor is how much the brain circuits can recalibrate.”

- How soon should you operate? “As soon as is convenient for your family. The earlier in life we operate, the more plastic the brain connections, and the better the repair.”

- Is the surgery just cosmetic? “Definitely not. The major goal of this surgery is to restore what nature intended: both eyes locked on target so that your child develops good depth perception and proper eye tracking, and avoids double vision. When the eyes are misaligned, your child has a visual brain conflict and has to shut down half of the visual cells. The cosmetic improvement is important but is a side benefit.”

### Anesthetic and Operative Considerations

Infants who were markedly uncooperative during office examinations may benefit from a brief examination under anesthesia at the beginning of surgery. The cycloplegic refraction can be rechecked and a detailed examination of the peripheral fundus performed, if warranted. After antiseptic preparation and draping, forced duction testing is done to rule out restrictive myopathy. In infants with large-magnitude infantile esotropia, the surgeon may feel moderate resistance of the globe to abduction under anesthesia. This is attributed to mild medial rectus contracture.

The surgical strategy most frequently employed is recession of both medial rectus muscles. For esotropia greater than 60 to 70 PD, botulinum toxin may be injected under direct visualization into one of the maximally recessed muscles to augment the effect of the recession. Alternatively, the surgeon may perform “three muscle surgery”—resection of a lateral rectus muscle along with the bimedial recessions. If the infant displays an A or V pattern of 15 PD or more, the medial rectus muscle tendons can be displaced vertically relative to their normal insertions or, if the pattern is accompanied by substantial oblique muscle overaction, oblique muscle weakening is carried out in lieu of transposition (see Chapter 14). The optimal approach to the muscles to ensure immediate postoperative cosmesis and comfort is through the conjunctival fornix. Large muscle recession can be safely and simply performed in infants using a “hang-back” technique. Eye patches are not necessary at the conclusion of surgery.

Because children with infantile esotropia are likely to require more than one operation, the surgeon should exert considerable effort to make the first and any subsequent surgical experiences as pleasant for the family as possible. The perioperative event that is best remembered and most distressing to parents is vomiting. In children younger than age 2 years, the prevalence of postoperative nausea and vomiting is low—about 20% or less. Children at much higher risk of vomiting include those with a history of motion sickness (prevalence greater than 90%) and those older than 4 years (prevalence 80%). To significantly reduce nausea and vomiting, the anesthesiologist should avoid using narcotic agents. In addition, a nonsteroidal anti-inflammatory agent such as ketorolac may be given once the intravenous line has been inserted, along with one of the newer serotoninn-antagonist antiemetics (granisetron [Kytril] or ondansetron [Zofran]). In school-age children, using propofol after induction with nitrous oxide and halothane may further reduce the frequency of postoperative vomiting.

Immediately after recovering from the anesthetic the child is permitted to roam and play as desired. Parents can bathe and wash the child’s face without undue concern, especially if a fornix approach was used for the incision. Antibiotic-corticosteroid ointment (e.g., Cortisporin) may be applied at bedtime for the first week after surgery. If instilling the ointment proves difficult, the parent can apply it to the child’s lashes after the child is asleep. Occlusion therapy is discontinued for at least the first week after surgery. Spectacles, if prescribed, should be worn.

### Follow-Up Regimen

At the first postoperative visit, typically 3 to 10 days after the procedure, the surgeon checks the visual acuity, rules out an afferent pupillary defect, and ensures that a good red reflex is visible from both fundi. The conjunctival incisions are examined using a penlight (it is not necessary to pry open the eyelids). In infants, alignment is assessed using the Krimsky or Hirschberg method, and versions are examined to verify the absence of gross underaction and a “slipped muscle.” In older, cooperative children a prism cover test may be performed in primary position. Use of antibiotic-corticosteroid ointment is discontinued.

A second postoperative appointment is scheduled for 3 to 4 months hence. If amblyopia is present, occlusion therapy can be reinstituted. If marked overcorrection or undercorrection is noted, the child may be seen sooner, but reoperation seldom will be seriously considered until 3 to 4 months have elapsed. If alignment is optimal (within 8 PD of orthotropia) and acuity is equal in the two eyes, subsequent follow-up visits are spaced at about 6-month intervals up to age 6 years. Thereafter, the risk of strabismic amblyopia begins to diminish, and the stable patient may be seen once a year. After age 10, visits are on an as-needed basis.

Occlusion therapy and more frequent follow-up visits are reinstituted whenever the child exhibits recurrent amblyopia. Spectacles are prescribed for significant refractive error and/or refractive esotropia. Reoperation is performed when a constant or poorly controlled intermittent esotropia or exotropia exceeding 12 PD is detected. Reoperation also is indicated for conspicuous oblique muscle overaction, DVD, or dissociated horizontal deviation.

### ACKNOWLEDGMENTS

This work was supported in part by NIH grants EY02687 and EY10214-01A2 and an unrestricted grant to the Department of Ophthalmology and Visual Sciences from Research to Prevent Blindness, Inc.


77. National Society to Prevent Blindness: Vision Problems in the U.S.: Data Analysis, Definitions, Data Sources, Detailed Data Table, Analysis, Interpretation, 1980.


DIFFICULT ESOTROPIA
ENTITIES: PRINCIPLES OF MANAGEMENT

EDWARD L. RAAB, MD, JD

Several convergent ocular deviations are discussed in this chapter, along with the author’s specific management techniques. For these entities, differences between children and adults are not prominent but a natural separation exists to some extent. Accommodative esotropia and its decompensated form, as well as consecutive esotropia, occur mostly in children. The principles of treatment of the nonaccommodative form of intermittent esotropia, decompensated monofixational esotropia, and surgical overcorrections and undercorrections are applicable to patients of any age.

The cited references are a small but representative portion of the available literature on these subjects. Each has its own valuable bibliography, to which the interested reader is referred for further information.

INTERMITTENT ESOTROPIA

Intermittent esotropia presents in several general settings (Table 9–1). Common to all of them is that a manifest esodeviation is present at some times but not at others. The distinction among these varieties lies in the nature of the trigger for the esotropic interval.

Neurologically unstable patients demonstrate intermittency that essentially reflects the extreme variability of their strabismus (which less typically may be exotropia). The general view is that such patients cannot gain lasting improvement through surgery because of the difficulty of obtaining accurate measurements and because of a frequent overresponse to even conservative amounts of muscle alteration. At some later stage the esodeviation becomes less variable in size and more often manifest, allowing more accurate surgical planning. There is frequently a superimposed accommodative component and possibly amblyopia (anisometropic more so than strabismic), which require attention in the usual ways.

Intermittency as a feature of accommodative esotropia is dealt with in other parts of this chapter, along with the rare cyclic esotropia. The remainder of this section is concerned with intermittent esotropia as a separate, nonaccommodative entity (true intermittent esotropia).

Clinical Characteristics and Diagnosis

This form of intermittent esotropia presents as a recurring temporary decompensation of esophoria, which otherwise is controlled by fusional divergence. The greater the predominance of “tonic” or other nonaccommodative convergence impulses over those for divergence, the larger is the magnitude of the esophoria and the less likely that it will remain latent. The patient’s own attempts at control result in varying degrees of asthenopia, which ultimately cannot be sustained.
Discouraging accommodative effort does not alter the angle of esphoria. This is the major differentiating feature from the accommodative variety. The refractive error frequently is myopic and in hyperopic patients is most often no more than +2.50 D. Amblyopia is infrequent and mild.27

Fusional divergence may be subnormal to excellent, the latter attained through constant effort to compensate for the imbalance. The problem is that, for a particular patient, the available fusional divergence amplitude finally becomes inadequate for the amount of esophoria, particularly during periods of general stress or subnormal health.50

Because compensation may remain adequate for a long interval until the patient seeks attention, the onset of the underlying esphoria usually cannot be dated. The usual time for these patients to present for treatment is late in the first decade of life, in contrast to the typical onset of accommodative esodeviations before age 2 years.50 Diplopia and better grades of stereopsis are more commonly associated with an onset after age 8 years, implying a long past period of secure binocularity. Accompanying features such as inferior oblique overaction and dissociated vertical deviation are not prominent.27 There are no hereditary or gender-specific considerations.

Some observers have noted imperfect stereopsis and mild amblyopia despite the acknowledged fusion capacity of these patients.27 Even when they do not demonstrate the manifest small esoshift that identifies the most typical form of monofixation syndrome,26 nothing negates the possibility that these binocular sensory imperfections identify a primary monofixation syndrome independent of the motor anomaly, rather than an adaptive consequence.

A separate category consists of an acutely presenting esotropia that is constant from onset. Such patients raise a suspicion of intracranial neoplasm, mandating consideration of neurologic investigation and imaging studies. It is significant that such cases are known to be orthotropic, rather than esophoric, before the onset of the deviation50 (see Chapter 10).

## Treatment

Treatment is directed at eliminating symptoms or realigning the frequently crossed eyes. Especially in cases with larger angles (whether phoria or tropia), nonoperative treatment will most often prove to be ineffective.

Expanding fusional divergence may be possible through orthoptics, but only to a limited extent and with very rigorous efforts. Once the esotropia tendency overcomes previously successful attempts to maintain proper alignment, stimulation of divergence through less natural means is unlikely to be successful even in the relatively few patients who might persist in carrying out this therapy.

Wearing compensating prisms decreases the demand for fusional divergence. This may provide symptomatic benefit but would tend to promote or perpetuate the manifest esotropic state by eroding the patient’s own control mechanism.51 Reports that antiaccommodative measures are helpful in some patients with this type of intermittent esotropia suggest that the necessary diagnostic distinction has not been made.50

Surgical correction is effective. Recession of both medial rectus (MR) muscles has been used most often, but the surgeon who prefers unilateral recession/resection should feel free to elect this procedure. In those cases with an A or V pattern or oblique dysfunction, it seems logical to modify the procedure to include horizontal rectus reinsertion with offset or oblique muscle weakening, as the surgeon may consider appropriate. Authors on this subject emphasize that the full size of the esodeviation is the correct measure of the amount of surgery, regardless of whether the deviation is usually manifest or latent but symptomatic.27

## ACCOMMODATIVE ESOTROPIA

One type of accommodative esotropia results from an anomalous relationship between the central innervational controls of accommodation and convergence in the presence of only a modest need for accommodation (the so-called high accommodative convergence/accommodation, or AC/A, ratio).11,27 The second major category includes patients with normal linkage of these functions that is overstressed by an excessive demand, that is, substantially above-average hyperopia (refractive accommodative esotropia).40 These two forms occur with about equal frequency (Table 9–2).39,40 However, the separation between them is far from discrete, with much overlap in the associations of high and normal AC/A ratios and varying degrees of hyperopia.40,42

An occasional patient will show an identical clinical picture when the functional disturbance actually is subnormal accommodative ability, calling for additional innervational output for the same demand and producing obligatory over-convergence.8 This last variety simulates the high AC/A variety of accommodative esotropia and calls for the same treatment approach. The conscientious practitioner need not make this distinction to effectively treat such a patient. There is evidence that some highly hyperopic patients are protected from accommodative esotropia despite an appropriate unaided focusing effort due to a low AC/A.49

Strabismus, in general, is thought to be inherited in 30% to 70% of cases. Some studies indicate that the most likely mode of transmission is autosomal dominant with incomplete penetrance.12 However, in accommodative esotropia, it is evident that some highly hyperopic patients are protected from accommodative esotropia despite an appropriate unaided focusing effort due to a low AC/A.49

Some studies indicate that the most likely mode of transmission is autosomal dominant with incomplete penetrance.12

<table>
<thead>
<tr>
<th>AC/A</th>
<th>Raab50</th>
<th>Parks29</th>
</tr>
</thead>
<tbody>
<tr>
<td>Normal</td>
<td>137 (50%)</td>
<td>289 (43%)</td>
</tr>
<tr>
<td>High</td>
<td>137 (50%)</td>
<td>387 (57%)</td>
</tr>
<tr>
<td>Totals</td>
<td>274</td>
<td>667</td>
</tr>
</tbody>
</table>

*Based on comparison of distance and near alignment.
Clinical Characteristics and Diagnosis

The hallmark of accommodative esotropia is the onset of an intermittent esodeviation between ages 1 and 3 years in a neurologically normal child; it is elicited only when accommodative innervation is stimulated. This definition includes the possibility of accommodative esotropia occurring in aphakic children who attempt to deal with hyperopic blur despite the lack of any accommodative response. Such cases have been observed, particularly when only a distance correction is worn.

Particularly in the intermittent phase, amblyopia is not prominent unless the hyperopia is anisometropic. In the latter case, amblyopia results because the untreated patient supplies accommodation only to the extent that will clear the focus of the less hyperopic eye. Even when fusion capacity cannot be tested reliably, the existence of at least peripheral fusion should be presumed in most successfully treated cases.

Variations on the typical picture are a presentation before age 1 year, either alone or in rapid sequence with surgically corrected primary infantile esotropia (Fig. 9–1), and a masked accommodative esotropia noted as an additional anomaly in a patient with typical intermittent exodeviation. Also, the diagnosis is compatible with myopia; such individuals by definition would fall into the nonrefractive, high AC/A ratio part of the spectrum.

Controversy has arisen concerning the degree of precision required in measuring the AC/A ratio. No method of measurement quantifies the innervational output for these two functions. Whereas most observers favor the gradient method to precisely estimate the ratio, the distance-to-near alignment comparison, based on the change in accommodative demand, has remained useful for clinical purposes even without accounting for the interpupillary distance in the analysis. However, this may give misleading information. There are data indicating that some patients with accommodative esotropia whose near deviation exceeds that at distance actually have low AC/A ratios as demonstrated by the gradient method. This phenomenon has been termed “nonaccommodative convergence excess”.

Cycloplegic refraction and measurement of the strabismus angle at distance and near fixation are key parts of the examination. Even before knowledge of the refractive error, a useful and quick indicator of accommodative esotropia is a favorable response of the deviation at near fixation to empirically chosen +2.50 D or +3.00 D spherical lenses worn in a trial frame, provided that preliminary determination of visual acuity does not suggest myopia. Whatever the AC/A ratio and the magnitude of hyperopia, a reduction of the esodeviation as determined by prism and alternate cover testing, or even by gross observation, identifies the case as at least partially accommodative. The onset after age 6 months of an intermittent deviation in a neurologically normal child is presumptive evidence of accommodative esotropia and in combination with the “quick +3.00-D test” is conclusive.

The examiner must also be alert for a possible nonaccommodative component of the esotropia and for additional motility anomalies such as inferior oblique overaction and dissociated vertical deviation. Although the last two entities have been considered prominent in cases of infantile (congenital) esotropia, they also occur with important frequency in diverse forms of strabismus. In accommodative esotropia they occasionally require surgical correction despite adequate control of the horizontal deviation.

There are no major confounding entities in the differential diagnosis, because compensatory head positions, palpebral fissure variations and incomitancies are not present. The distinction from essential intermittent esotropia has been detailed in the previous section.

Figure 9–1. One-year-old infant with esotropia (left) that was eliminated by wearing the full cycloplegic correction for hyperopia (right). This deviation began within 2 months after 6.0-mm recessions of both medial rectus muscles for 65 PD of infantile esotropia at 6 months of age. Except for the earlier than usual onset, the features were entirely typical of accommodative esotropia.
In the early stage, before any deterioration of purely accommodative esotropia to a partially nonaccommodative or “mixed” state (discussed later), optical correction of substantially all of the patient’s hyperopia by spectacles or contact lenses is the most widely accepted initial treatment (see Fig. 9–1). The hyperopic correction that enables the patient to control any manifest esodeviation is the practitioner’s target. I have found it best to prescribe the complete cycloplegic correction (or within 0.50 D for errors above +5.00 D) initially, whether or not this will ultimately be needed, reasoning empirically that the earliest possible gain of control facilitates a subsequent reduction of correction and that this is preferable to possibly undercorrecting and having later to “catch up.” At the usual age at onset of 2 to 3 years, experience indicates that reduction of the correction is not feasible until much later.

Bifocal additions are the usual method of managing the uncompensated increment of accommodative overconvergence for near fixation that characterizes patients with a high AC/A ratio. When the near measurement in a newly presenting case is obviously at least 10 PD larger than at distance, initial prescription of a bifocal is better than waiting to learn whether correcting hyperopia is sufficient to compensate the esotropia at distance. This method is in keeping with the objective of providing immediate control. Younger patients typically require the full add of +2.50 or +3.00 D as the first choice, although it is often possible to progressively decrease this component sooner than the correction for hyperopia.

Subsequent arbitrary reductions in either the distance or near component of the prescription are to be discouraged; measurement of the esodeviation with the contemplated change of prescription before the patient’s eyes is the recommended method. It is not necessary to demonstrate that hyperopia under cycloplegia has correspondingly decreased. This will occur in many cases but, in a comparable number, hyperopia will be noted to be the same or even to have fractionally increased. This finding is not a reason to withhold an indicated reduction in plus lens power. It is merely an emphatic reminder that optimal treatment consists of the minimally indicated strength determined by actual trial.

It has been my practice to limit any single reduction to 0.75 or 1.00 D, even if a brief trial indicates that more reduction could be attempted. Separate determinations are made for the distance and near components in patients wearing bifocals. The least add that would be meaningfully effective is about +1.25 D. Once begun and judged successful, reduction is attempted again every 4 to 6 months; this is a rather cautious approach compared with the shorter intervals for spectacle power reduction or substitution of miotics in rapidly decreasing concentrations that have been advocated.

Although many parents report occasional, fleeting observations of loss of alignment control by their children after a prescribed reduction, this most often is just the initial step in the desired further expansion of fusional divergence that this treatment seeks. However, occasionally it is necessary to resume the prior prescription for another interval.

Anticholinesterase miotics are an alternative method of facilitating accommodative effort. Dosage schemes are available from many sources.64 The efficacy of these agents is variable, limiting their value for long-term management or for initial diagnostic trial. A further disadvantage applies to anisometropic hyperopia, because facilitation of accommodation will be directed to the needs of the less hyperopic eye, doing little to remove a source of amblyopia. Furthermore, miotics do not compensate for an astigmatic component of the refractive error.

One setting in which using miotics has proved useful is in the infant with minimal hyperopia and no crossing of the eyes except at near fixation who requires a bifocal for control but who will not tolerate this device. This appears to be logical, but encouraging increased effort to enforce bifocal wear is preferable. When compliance and lax parental enforcement of spectacle use lead the ophthalmologist to consider these agents, miotics usually have not proved adequate; their administration merely substitutes one parent–child conflict for another. The ophthalmologist should consider whether this is the real issue when children “would not wear glasses.”

The dangers of retinal detachment and lens opacification have been overstated for the age group in which these drugs have been used to treat strabismus. Rarely is it necessary to discontinue this treatment because of pupillary excrescences, the description of which seems less alarming when they are called “tags” rather than cysts. It is good practice to repeat the initial warning concerning interaction with succinylcholine—used as a muscle-relaxing adjunct to general anesthesia—at most, if not all, visits. Pediatric anesthesiology practitioners appear to be well aware of this point.

A vexing therapeutic question arises when a fully accommodative esotropia persists well beyond its expected time of spontaneous disappearance at age 8 to 10 years. This is seen in 30% or more of cases and does not appear related to the level of initial hyperopia, subsequent changes in hyperopia, a familial tendency to strabismus, or associated cyclovertical ocular muscle anomalies.69

When single-strength glasses are sufficient for control, contact lenses have been a satisfactory solution. For those patients heavily dependent on bifocals, the same option applies but usually yields less consistent results. Here, too, some degree of latitude is required at an acceptably small risk, on the basis of reports that bifocals offer no ultimate advantage even when the eyes cross at near fixation, so long as there is fusion at distance when the hyperopic correction is supplied.36

Miotics as a temporary supplement for those occasions when the patient would prefer not to use spectacles do not significantly jeopardize secure control. Daily administration is unnecessary when 0.125% echothiophate iodide (Phospholine Iodide) is prescribed. Four or five, or occasionally three, single doses per week provide the desired effect equally well. When periodic cycloplegic refraction is scheduled, it is not necessary to discontinue the medication beforehand.81

When optical or drug treatment can succeed, surgery to realign the eyes is highly controversial and has been advocated by only a small contingent.6 13 It has not been widely embraced by the community of strabismologists. The consecutive exotropia that usually results from such an approach may open the door to multiple operations with disappointing results. This is quite different from the need to surgically correct deterioration of a partial or complete nonaccommodative esotropia (discussed in the next section).

There are patients with a high AC/A ratio, or with nonac-
commodative convergence excess, whose eyes, while acceptably straight at distance, continue to overconverge at near. If the overconvergence does not produce symptoms or cause cosmetic distress, allowing it to persist is justifiable as long as distance alignment and fusion remain satisfactory. If the ophthalmologist contemplates surgery, recession of the MR muscles is the favored choice. Whether this results in esodeviation at distance remains conjectural.

Posterior fixation sutures on the MR muscles, alone or combined with recession, have been proposed for the patient with a high AC/A ratio and minimal esotropia at distance. Although this may be effective, it is best avoided because of the need for a rather extreme posterior placement of the second insertion, a surgical feat whose difficulty and potential dangers tend to be understated.

The sequelae of decompensation and persistence of accommodative esotropia have been considered complications. They are more correctly regarded as disappointing but expected features of this entity, notwithstanding excellent management. Amblyopia is usually not severe unless partly due to late or inadequately treated anisometropia. Patients with nonaccommodative convergence excess require close scrutiny, because their rate of decompensation is substantial.

DECOMPENSATED ACCOMMODATIVE ESOTROPIA

Decompensation of accommodative esotropia describes the evolution of a purely accommodative deviation into one that has both an accommodative component and a portion that is not corrected by relief of accommodative effort. This implies that something has occurred to change the original, purely accommodative condition. Another term for this occurrence is deterioration.

The usual but imprecise explanation for this phenomenon is that the MR muscles, through excessive innervation, are anatomically or physiologically changed. However, a case presenting initially with both accommodative and nonaccommodative features may actually have evolved in the reverse order, namely, accommodative esotropia superimposed on the infantile variety. The distinction is not particularly important for making management decisions but could limit expectations for the binocular sensory outcome.

Some authors have described decompensation as a complication peculiar to the high-AC/A subclass of accommodative esotropia. Other studies, however, have not confirmed this observation. Although neglected accommodative esotropia would carry a predisposition to decompensation under the etiologic theory discussed earlier, most strabismologists also have observed its development even with impeccable treatment and conscientious compliance, and despite an initially favorable response. An onset before age 2 years appears to be a prominent risk factor.

Clinical Characteristics and Diagnosis

The patient presents with a two-component esodeviation. Whether the AC/A ratio is normal or high, there is an esotropia at both distant and near fixation that even the full cycloplegic hyperopic correction, with or without a bifocal addition at near, does not reduce to 8 PD or less of manifest deviation. Eight prism diopters of esodeviation is considered the upper limit at which peripheral (monofixational) fusion is possible. If decompensation is total, differentiating the condition from acute-onset nonaccommodative esotropia is possible only if the original condition and its gradual evolution have been documented. Once the patient has decompensated while on maximal antiaccommodative treatment, subsequent reversal of the process cannot be expected.

Measuring the esotropia angle in the usual way will demonstrate the characteristics described earlier. A simple step in the examination may act as a check when decompensation appears to be present. With any AC/A ratio, a deviation at near that is reducible by fixating through additional +3.00 D to less than the distance correction with full cycloplegic correction indicates an additional uncompensated element of hyperopia. Clearly, no determination of decompensation should be made until this possibility is ruled out.

Treatment

Prisms in addition to hyperopic correction are a possible but unpopular nonsurgical measure. As with wearing prisms in any manifest strabismus, they do not restore alignment. In contrast to their use in paretic deviations or postoperative overcorrections, prisms are not a temporizing measure until the alignment hopefully corrects spontaneously.

Surgery is the preferred approach and is justified by the likelihood of not only restoring motor alignment but also regaining lost fusion ability, as documented before decompensation, or strongly inferred from the history of an acquired deviation. In keeping with the supposed pathogenesis of this condition, MR weakening procedures are the preferred approach except when there is severe visual deficit in one eye, in which case unilateral recession/resection surgery is an effective substitute. Because most strabismologists agree that the portion of the total deviation representing a continuing accommodative anomaly is not appropriate for surgical correction, the goal is to reestablish any postoperative residual deviation as purely accommodative. The quantity of surgery needed for optimal alignment has been the subject of different surgical strategies.

A long-standing practice was to employ the individual surgeon’s usual scheme for a given angle of nonaccommodative esotropia. Based on the impressive frequency of undercorrection in decompensated esotropia, various authors have suggested using the near deviation as the basis for calcula-
Monofixational strabismus refers to a binocular sensory state seen in some strabismus patients able to maintain straight eyes in ordinary visual circumstances. Semantically, all manifest strabismus of any angle is monofixational, i.e., one eye, habitually or because of amblyopia, is preferred for fixation. In larger angles of deviation of any direction, conflicting sensory informational input from the deviated eye most often is suppressed. What distinguishes the monofixation syndrome is that the manifest deviation, if present, is sufficiently small (up to 8 PD) to permit binocular peripheral fusion, despite monocular central suppression. Hence, a reference to monofixational strabismus or syndrome implies retention of peripheral fusion.

Monofixation syndrome typically is seen when a preexisting strabismus is controlled nonoperatively (as in compensated accommodative esotropia) or after surgery. In this setting, the monofixation state is described as secondary or "adaptive," on the assumption that it is the sensory outcome of the prior uncorrected strabismus state. Monofixational fusion also may be detected even in nonstrabismic individuals, suggesting that it may be an inherent primary sensory condition and raising the conjecture that even when it occurs in strabismus, it could be an independent condition and not an adaptation. Some authors have explained this paradox by resorting to an explanation based on a "microtropia" that is not detectable by ordinary examination methods. There may be an inheritable predisposition to this anomaly, which some observers think is distinct from that due to a patient's refractive or strabismic state. This, however, remains debatable.

The reader attempting to explore this subject will be confounded by earlier literature that almost interchangeably referred to "microtropia" in two different senses, as well as "monofixational phoria," "small angle esotropia with fixation disparity," and similar designations. It is important to realize that, whatever term one employs, monofixational strabismus is not a discrete ocular motility abnormality but rather a particular sensory state common to many types of strabismus.

**Clinical Characteristics and Diagnosis**

Contrary to its original description, monofixational fusion requires no measurable manifest or latent misalignment at any fixation distance. In such instances, the diagnosis is indicated entirely by sensory testing for both central and peripheral fusion. When deviations are detectable, particularly when the angle increases under conditions of disrupted fusion, cover testing alone is sufficient. Most cases of monofixational esodeviation will show the classic feature of an
Table 9–3. Cover Test Responses as Indicators of Fusion

<table>
<thead>
<tr>
<th>Cover/Uncover</th>
<th>Alternate Cover</th>
</tr>
</thead>
<tbody>
<tr>
<td>No shift</td>
<td>No shift</td>
</tr>
<tr>
<td>No shift</td>
<td>Shift</td>
</tr>
<tr>
<td>Shift*</td>
<td>Larger shift</td>
</tr>
<tr>
<td>Shift*</td>
<td>Equal shift</td>
</tr>
<tr>
<td>Shift*</td>
<td>Smaller shift</td>
</tr>
<tr>
<td>Fusion Interpretation</td>
<td>Peripheral Fusion</td>
</tr>
<tr>
<td>No information</td>
<td>No information</td>
</tr>
<tr>
<td>Present</td>
<td>No information</td>
</tr>
<tr>
<td>Present</td>
<td>No information</td>
</tr>
<tr>
<td>No information</td>
<td>Not possible</td>
</tr>
</tbody>
</table>

*Up to 8 PD of shift, measured by simultaneous prism and cover.


8 PD or smaller manifest esotropia by the cover/uncover test (measured using simultaneous prism and cover), with a larger esophoria as the alternate cover test disrupts even peripheral fusion. However, absence of the following still are compatible with the monofixation syndrome: (1) re fixation movement on the cover/uncover test, (2) build-up from the manifest small deviation, and (3) misalignment on either form of cover testing (Table 9–3).

Although a debate exists as to whether fusion under these conditions is based on normal or anomalous retinal correspondence, the ophthalmologist who manages these patients need not be constrained by this controversy, because current strabismus management decisions do not emphasize such a distinction.

**Treatment**

Whether monofixation, as an outcome in a strabismus patient, is satisfactory or unsatisfactory depends on the observer’s attitude. For those who attach primary importance to the lack of central (bifoveal or bimacular) fusion, a monofixational result is both disappointing and frustrating. For the ophthalmologist who instead emphasizes the retention of peripheral fusion, especially when restored by treatment, monofixational syndrome is a useful status. It is more practical, clinically, to emphasize the stability of peripheral fusion, and to determine whether fusional efforts produce unwanted symptoms. These findings dictate case management. Determining whether central fusion is present is a secondary consideration.

In general, identifying the monofixation syndrome is less important than understanding how to manage the strabismus it accompanies. No specific treatment will convert a monofixational to a bifixational fusion status, that is, where there is both central and peripheral binocular cooperation, although instances of spontaneous improvement after correction of amblyopia have been reported. Efforts should be directed at minimizing amblyopia of the nonfixing eye.

Decompensation occurs in monofixational strabismus when the misalignment, previously controlled by peripheral fusion, changes from a latent or occasionally manifest deviation to a substantially constant one. From a binocular sensory standpoint, the consequences are manifest in the peripheral fusion system. The central monofixational pattern existed before the alteration and will persist even after decompensation has been corrected. The treatment of this form of strabismus aims to optimize whatever fusion ability is present.

The presence of this condition in itself does not interfere with other treatment efforts. It is an acceptable outcome, and the most realistic goal in the majority of cases. The capacity for monofixational fusion is not lost except when some complicating event, such as a poor surgical result or a spontaneous increase in the basic angle, supervenes.

**UNDERCORRECTED ESOTROPIA**

Surgical undercorrections of esotropia may be divided into two classes: those that are acceptable and those that are not. Acceptable undercorrections are those in which further realignment is unlikely to improve function and is not essential to enhance a patient’s personal appearance. Unacceptable undercorrections are those in which binocular cooperation is an achievable goal through further treatment and those giving cosmetic dissatisfaction.

By definition, undercorrections are residual misalignments in the same direction as the original condition. They should be distinguished from successful results that later recur, not for purposes of treatment selection but as a means of estimating the likelihood of functional success. This may depend on whether the patient has had the benefit of an intervening period of binocular cooperation.

**Clinical Characteristics and Diagnosis**

The diagnosis of undercorrection should be withheld until a full healing period of about 6 weeks has passed. Initial indications of the result can change dramatically over this interval. There is hardly ever a reason to proceed more rapidly. At the end of this period, additional observation is warranted if earlier postoperative visits have indicated a progressive tendency to improve.

Once convincingly observed, determining the ultimate functional prognosis requires the ophthalmologist to consider the length of time esotropia was present before surgery or decompensation. A shorter interval tends to favor restoration
146

•

CLINICAL STRABISMUS MANAGEMENT

of binocular function. Cases known to have originated as

purely accommodative esotropia, even when decompensation
has been present over considerable time, deserve the pre
sumption of innate binocularity. In contrast, in infantile (con
genital) esotropia, if it is known that the eyes were not
acceptably straight for at least several months by about age
2 years, the chance of obtaining binocularity is poor.
Rather than relying on these inferences, the practitioner
can seek evidence of potential binocularity from sensory
evaluation tests such as the Worth four-dot test, Bagolini
striated glasses test, and the major amblyoscope. In all these
tests the deviation should be neutralized with prisms.

Two variations may be considered. When previous MR
weakening placed the new insertion no more than 10 mm
from the limbus (by actual exploration at the time of the
second procedure, not merely calculated from the operative

report), further recession of either both muscles or of one in
combination with repeat or first-time resection of the antago
nist LR is possible. The additional recession should place
the reinserted MR no farther than 12 mm from the limbus.

Even if this amounts to only an additional 2.0 mm of
recession, approximately 10 to 15 PD of additional correc
tion is a reasonable expectation. If there is a substantial
difference in acuity between the eyes, it is best to avoid or
limit surgery on the better-seeing eye even when the eye
with poorer acuity has already been operated on.

Treatment
Essentially, the reasons to consider reoperating are the
same as those prompting surgical treatment in the first place.
Residual esotropia may be modest enough in terms of
angle size to allow the temporary use of corrective prisms if
there is reasonable hope of additional improvement. How
ever, undercorrections most often persist, especially when
initial surgery consisted only of MR weakening, where it is
not simply a question of waiting until a resected muscle can
function at full strength once it has healed.
Several surgical options are available. Initial MR weaken
ing is followed by resection procedures on the lateral rectus
(LR) muscles. If the initial procedure was unilateral reces
sion/resection, the same approach on the unoperated eye is
in order. Both of these choices follow Cooper's" rule of
approaching reoperation as a new case. In repeat procedures,
it is less clear whether adding 1 mm to the usual surgical
dose is appropriate for a residual deviation that retains the
features of decompensated accommodative esotropia.

When the MR muscles have been maximally recessed,
double marginal myotomy of these muscles may be per
formed. The results, however, are more variable. This option
should be reserved for patients in whom the MR muscles
not only are already maximally recessed but are incorporated

in enough scar tissue to suggest difficult isolation and a
likelihood that scar will re-form. This especially is the case
when there has been previous surgery and scar formation on

the LR muscles as well. Other authors have advocated poste
rior fixation of the MR in this situation,” but this may be
technically difficult to perform.
Other than failure to restore proper binocular function
and cosmetic dissatisfaction, the principal difficulty from
undercorrection is that, if strabismic amblyopia was present
and required treatment, a possibly smaller angle of esotropia
continues to favor a relapse that will call for reinstitution of
the appropriate measures. Vigilance to this possibility is
necessary. If the undercorrection is small enough to allow
for exercise of monofixational fusion, declaring the result an

undercorrection is technically correct but is of minor clinical
importance.

OVERCORRECTED ESOTROPIA

Eyes that diverge soon or remotely after operation for any
type of esotropia constitute overcorrected esotropia. Unless
rotations are obviously deficient, it is common to observe
satisfactory alignment interspersed with brief intervals of
intermittent exotropia over several office visits before over
correction becomes definitely manifest. Such a fluctuating
state may continue for several years.
As is the case for undercorrections, management of over
corrected esotropia exemplifies the range of factors and
solutions the ophthalmologist also encounters in dealing with

Clinical Characteristics

and Diagnosis
The key finding when planning to remedy overcorrected

esotropia is the presence of limited rotation in the field of
action of a previously weakened MR muscle. The choice of
the term limited is deliberate. This is because there can be
no conclusion that the cause is restriction or muscle weak

ness without further analysis, especially when the antagonist

overcorrected exotropia and vertical misalignments (see also

LR has been resected or otherwise tightened. In adult pa

Chapters 12 and 38).
Overcorrection of decompensated accommodative esotro
pia has been mentioned as a complication and the frequent
consequence of operating for purely accommodative esotro
pia. Another important entity prone to overcorrection is
infantile (congenital) esotropia (discussed in Chapter 8). In
this setting, the most important risk factor for overcorrection
is difficulty in accurately determining the size of the original

tients, passive forced duction and active force generation
testing usually is possible in the office and should be per
formed to differentiate muscle paresis from restriction as a

deviation.

cause of limited rotations.” ". " The information obtained

from these tests is especially important in repeat surgery.
Ancillary tests such as saccadic velocity determination
(not always available)" and measurement of differential in
traocular pressure reinforce these “gold standard” proce
dures." ** A rise of several millimeters of mercury when


actively attempting to rotate the eye into the field of limitation, as strong muscular and other forces are applied externally to the globe, suggests intact extraocular muscle power and thus no overweakening effect.

Although application of forceps to the eye and repeated determinations of intraocular pressure typically are not feasible in a child, careful observations allow reasonable inferences to be made. If adduction can be characterized as a “slow float” rather than a “brisk saccade” with a sudden stop, weakness rather than restriction is strongly suggested. This is the counterpart to testing active force generation and determining saccadic velocity.

Classic teaching states that a lesser deviation when the uninvolved eye fixates (“primary deviation”) than when the involved eye does so (“secondary deviation”) is characteristic of paretic deviations. This dictum is true but incomplete. Hering’s law of simultaneous innervation to yoke muscles applies equally well to the extrainnervation employed in attempting to overcome a restricted rotation. This phenomenon does not differentiate weakness from restriction.

**Treatment**

Nonsurgical measures often are merely stopgaps, as the consecutive exodeviation progresses to a degree warranting operative correction. In the susceptible age group, attention to amblyopia will still be necessary.

Active intervention by orthoptics is not applicable in the younger child. When the overcorrection still is within the range where at least peripheral (monofixational) fusion is possible, cooperative older children and adults may benefit from attempts to expand compensatory fusional amplitudes, but these will not affect the overall motor angle. Prism wear similarly may help maintain binocularity but will not improve motor alignment.

The practice of reducing a hyperopic prescription or supplying an optically unnecessary myopic correction is based on the hope that, by exercising accommodation and therefore accommodative convergence, the overcorrection will be adequately controlled. This approach is not reliable. Whether sustained accommodation actually is used in everyday conditions is questionable, because there is no demand for precise focus under all conditions. Adults managed in this way, especially those approaching the presbyopic state, may find the remedy to be more onerous than the overcorrection.

Once the necessity for surgery is clear, the operative plan should distinguish between full and compromised rotations. If there are no limited rotations, Cooper’s rule again applies. The surgeon should approach unoperated muscles by planning the usual surgical dose (number of millimeters) for the size of the deviation.

If the initial procedure was recession of both MR muscles, recession of both LR muscles is indicated. When the first procedure was MR recession–LR resection on one eye, the best choice for reoperation would be LR recession–MR resection on the other eye. Unlike the formula in the initial procedure, it is not clear whether adding or subtracting a millimeter to compensate for an accommodative influence is advantageous.

Kushner has suggested that, despite apparently normal adduction, a secondary convergence insufficiency pattern (near exodeviation exceeds distance exodeviation) indicates MR underaction and requires advancement of the previously recessed MR or recession of the LR muscles above the usual amount. Because the number of millimeters of advancement cannot be stated with assurance, the alternate choice is more attractive, provided that direct exploration reveals the MR reattachment sites to be no more than 11 to 12 mm from the limbus.

As advised for undercorrections, major visual loss in one eye dictates a different approach. In this instance, recessing the previously resected LR combined with recession of the as yet untouched LR in the better eye is a compromise between the soundness of Cooper’s rule and the need to minimize risk to the remaining sound eye. If the surgeon prefers to confine the second procedure to the poorer eye, prior resection of the LR would not preclude doing the usual amount of surgery for a given angle size. However, advancement of the recessed MR is difficult to quantify and should be done on an adjustable suture if possible.

**Figure 9–2.** Thirteen-year-old child with ptosis caused by residual hemangioma of the right upper eyelid and exotropia. There are limited adduction and inferior oblique overaction, each greater in the right eye. Two of several prior operations had included recession of the right medial rectus muscle; the second procedure had moved the insertion from 9.0 to 11.0 mm from the limbus. Because adduction was not restricted preoperatively at the last procedure, any further surgery should include exploration of the medial rectus reattachment sites.
Limited adduction may be due to weakness or restriction of this rotation. With the information provided by the diagnostic investigation described earlier, it is best to defer selection of a plan pending exploration of the suspected offending muscle. If a restriction improves when any surrounding adhesions are lysed, conjunctival recession should be included in the second operation. If the restriction does not release until disinsertion of the muscle, adjustable recession should be employed whenever possible (see also Chapter 38).

Unless overcorrected esotropia associated with deficient adduction has resulted from failure to respect the conventional safe limit of MR recession, the most likely cause is slippage of this muscle and its capsule owing to loosening of its reattachment, or recession within the capsule because of failure to incorporate the tendon itself in the locked reattachment suture (Fig. 9–2) (see Chapter 40). The surgeon should not omit direct exploration merely because an operative report speaks of quantities of prior recession that are less than maximum.

Minor slippage revealed by exploration may be ignored when substantial adduction remains. With severe limitation of MR rotation, despite the difficulty in quantifying the surgical dosage, the procedure should include advancing the MR to or near the original insertion, on an adjustable suture if possible, with minimal if any resection. This is best combined with recession of the resected LR.

Overcorrections that cannot be held latent by fusional compensation lead to loss of binocular vision and, for a patient in the susceptible age range, amblyopia. When the consecutive exodeviation occurs in the potentially fusing patient, symptoms reflecting visual annoyance (e.g., diplopia, confusion of overlapping dissimilar images) may be disabling.

**CYCLIC (PERIODIC) ESOTROPIA**

Cyclic esotropia is a rare and curious form of strabismus. It has been described as “alternate-day esotropia,” although 2-day and longer periodic cycles have been reported. The condition has no particular hereditary tendency or gender predilection. It differs from essential intermittent esotropia in following rather strictly repetitive cyclic intervals of straight and crossed eyes and from the intermittency of accommodative esotropia, which is driven by the accommodative stimulus. It is unlike esotropia associated with neurologic disorders again because of its strict periodicity and the reproducible angle. However, acquired cyclic esotropia has been reported to occur in children and adults with central nervous system disease. Some authors consider the esotropia as the underlying state, with cyclic intervals of remission.

**Clinical Characteristics and Diagnosis**

The classic patient with cyclic esotropia lacks the features of even latent strabismus (esophoria) during straight intervals. These are replaced abruptly with a fully manifest esotropia, which is not influenced by suppressing accommodative demand. Most patients have, in time, developed a constant deviation for no obvious reason. Associated features such as inferior oblique overaction, dissociated vertical deviation, and motor nystagmus are not major accompaniments of this condition.

The history is characteristic and should suggest the diagnosis. The ophthalmologist can easily obtain confirmation by examining the patient during both phases of the cycle, repeated as long as necessary to establish its periodicity. There are no differences in ocular rotations in either phase, tending to eliminate periodic paresis and restriction as etiologic factors. Measurements made with and without hyperopic correction do not alter the strabismus angle. Moreover, the condition is not even peculiar to hyperopic individuals. Children usually are asymptomatic in the esotropic phase. Adults may experience diplopia.

**Treatment**

There are no specific nonsurgical ways to abolish this condition or keep it from evolving into a constant deviation. The most peculiar feature is that surgery for cyclic esotropia while it is still in the periodic state is as effective as when it has become constant and does not result in what might be expected: a substantial exodeviations. This observation is similar to what is seen in intermittent esotropia. Accordingly, the surgeon can plan correction, using the procedure dictated by personal experience, any time after the diagnosis is established.

Because of the relatively brief duration of the esotropic phase before the deviation becomes constant, amblyopia is not present without an independent cause such as anisotropia. Because this is an acquired deviation, it does not severely threaten binocular capacity, especially because early surgery is perfectly acceptable.
Consecutive esotropia is the result of surgical overcorrection of an original exotropia. Whereas several decades ago it was common for the immediate surgical goal to be straight eyes or even a small residual exodeviation, this was replaced by the realization that, in most patients, the eyes tend to diverge again almost at once, and that an early postoperative esodeviation of up to 20 PD is preferable to counteract this tendency. Exotropia correction is best done while at least part-time fusion ability is still present. These observations were made when exotropia was corrected by bilateral LR recession, but they can be considered generally valid for recession/resection procedures as well.

Recurrent exotropia occurs far more often without initial overcorrection than does persistence of the initial esotropia. The young patient, however, during an exposure of possibly several weeks, is capable of developing the unfavorable attributes of esotropia, including amblyopia and monocular suppression. More visually mature children and adults experience diplopia, which can be very disabling.

Clinical Characteristics and Diagnosis

In consecutive esotropia, the initial postoperative esotropic angle typically diminishes to some extent but not enough to bring the surgical result within the satisfactory range. There is no consistent pattern to the reduction. It may occur over the entire postoperative period and merely be insufficient or may begin but not continue to improve after the first several days. If no improvement at all has occurred in the first 2 weeks, this is a reliable indicator that overcorrection will be permanent.

Particularly just after recession of the LR muscles, abduction of one or both eyes will not be full. This is not necessarily related to overcorrection, because esotropia persists even as abduction returns to normal. Lateral-gaze esotropia measurements are greater than for straight-ahead gaze, reflecting the usually temporary decrease in LR function until further healing has occurred. The primary-position distant esodeviation is greater than at near, with the latter possibly even insignificant. This difference may become smaller as the distant esotropia decreases in time.

An anomalous situation can vary this typical sequence. An unusual case of consecutive esotropia will appear as a persisting high-AC/A-ratio accommodative deviation and respond to treatment for that entity. Preoperatively, the accommodative component may have been mistaken for typical attempts to compensate for the basic divergent eye position. The true nature of the independent accommodative abnormality emerges only after surgical correction of the exodeviation. Because such an event does not constitute a surgical overcorrection, the strained but logical conclusion must be that the patient originally suffered from two separate forms of strabismus, in which the accommodative anomaly was completely subordinated to and masked by the exotropic presentation.

Kushner has commented extensively on this observation. He believes that the finding of a high AC/A ratio determined by the gradient method, after monocular occlusion to disperse the confounding influence of “tenacious proximal fusion,” will identify these patients, allowing appropriate counseling about the likelihood of unmasked accommodative esotropia after operation for exotropia. Knowledge of this possibility, however, does not allow the surgeon to avoid it through modification of the operative scheme.

Treatment

NONSURGICAL MANAGEMENT

Most ophthalmologists share the view that immediate postoperative overcorrection of exotropia is advantageous, but many are uncomfortable with a deviation exceeding 10 PD, despite the demonstration that satisfactory alignment occurs with equal frequency for esodeviations up to 20 PD. At one extreme are ophthalmologists who will merely observe an immediate overcorrection, in expectation of the trend toward redivergence that such patients display. At the opposite pole are those who intervene with occlusion, compensating prisms, and hyperopic correction and/or anticholinesterase miotics (even when there is no demonstrable accommodative influence). It is not always clear from the published advocacy of these remedies whether they are designed to counter possible unfavorable sensory sequelae or to influence the motor progress of the overcorrection.

My treatment program is to institute daily alternate occlusion for most of the day if esotropia above 20 PD is present at the first postoperative visit, 2 to 4 days after surgery. If there is amblyopia, alternation is modified to enhance the visual experience of the disadvantaged eye. Without evidence of grossly impaired abduction, repeat visits are scheduled at intervals of 10 to 14 days. If the overcorrection responds favorably, this program is maintained, even beyond the 6-week healing period, until improvement ceases. At this point, the case is reassessed to decide on the next appropriate step.

No intervention is employed for initial overcorrections of up to 20 PD until about 2 weeks have passed, and only for those whose esodeviation is still greater than 10 PD whether or not it has improved since the first postoperative visit. The regimen thereafter is as described earlier. When the initial esotropia is 10 PD or less, there is no consistent benefit to be expected from occlusion. Because such cases are already close to an alignment at which fusion will be possible, occlusion actually might be a sensorial disadvantage. The exception is amblyopia of a degree otherwise sufficient, in the examiner’s judgment, to warrant therapeutic occlusion (Table 9–4).

For all these groups, accommodation-lowering measures are not employed except in the unusual case in which they unequivocally restore proper alignment, in which instance they are therapeutic and not merely expectant. I have not used compensating prisms in these patients.
even peripheral binocularity and persistence of a deviation characterized as disappointing but within the range of exotropia prolongs conditions that favor both the erosion of best long-term alignment, visually immature patients are at risk. Even when satisfactory or an excessively tight reinsertion. Demonstrating restricted resection procedures resulting in overcorrection, restriction is not to be expected. However, after recession—resection procedures resulting in overcorrection, restriction is a distinct possibility, either from the quantity of resection or an excessively tight reinsertion. Demonstrating restricted abduction does not eliminate the possibility of coexisting weakness through overrecession or slippage of the LR; therefore, it is important to estimate active force as well as passive restriction in cooperative patients.

Although consecutive esotropia is itself a complication, any undercorrection or overcorrection of strabismus is better characterized as disappointing but within the range of expected outcomes. The dilemma here is that, to achieve the best long-term alignment, visually immature patients are necessarily exposed to unfavorable sensory consequences of the initial postoperative esotropia. Even when satisfactory alignment is eventually achieved, a monofixational fusion pattern may result. At least in some patients, this represents a regression from the bifoveal fixation present before surgery during the straight phase of intermittent esotropia.

Whether this risk is acceptable depends on one’s attitude toward the monofixation syndrome—a state that involves usually stable peripheral fusion and minimal, if any, amblyopia. If attempted correction is delayed or there is reluctance to create the initial esotropia, persistence of intermittent esotropia prolongs conditions that favor both the erosion of even peripheral binocularity and persistence of a deviation that approaches constancy. Realignment efforts at this stage are less reliable and probably court the same consequences.

REFERENCES


33. Parks MM: Ocular Motility and Strabismus, pp 144 and 150. Hagers-


medical rectus recessions in normal and delayed children. J Pediatr


in the surgical management of acquired esotropia. Arch Ophthalmol


38. Raab EL: Interpretation of the cover test in small angle deviations. J


39. Raab EL: Etiologic factors in accommodative esodeviations. Trans Am


40. Raab EL: Cycloplegic refraction after echothiophate iodide. J Pediatr


42. Raab EL: Accommodative esotropia: A reassessment. Am Orthopt


44. Raab EL: Outcome of deteriorated accommodative esotropia. Trans


Strabismus and Ocular Motility Disorders: Proceedings of the Sixth

Meeting of the International Strabismological Association, Surfers


46. Raab EL, Parks MM: Recession of the lateral recti. Early and late


47. Raab EL, Parks MM: Recession of the lateral recti. Effect of preopera-

tive fusion and distance-near relationship. Arch Ophthalmol 1975;

93:584.

48. Raab EL, Spierer A: Persisting accommodative esotropia. Arch Oph-

thalmol 1986;104:1777.


1976;69:1588.

50. Rosenbaum AL, Jampolsky A, Scott AB: Bimedial recession in high


51. Rosenbaum AL, Ure ra PT: Investigation of limited ocular rotations:


52. Saunders RA, Helveston EM, Ellis FD: Differential intraocular pressure


53. Scott AB: Active force tests in lateral rectus paralysis. Arch Ophthalmol


54. Simon AL, Borchert M: Etiology and prognosis of acute, late-onset


56. von Helmholtz H: Uber die accommodation dus auges. Albrecht Von


57. von Noorden GK: Indications for the posterior fixation operation in


58. von Noorden GK: An alternative to marginal myotomy. Am J Ophthal-


59. von Noorden GK: A reassessment of infantile esotropia. XLIV Edward


60. von Noorden GK: Etiology of heterophoria and heterotropia. In Binocu-

lar Vision and Ocular Motility: Theory and Management of Strabismus,


61. von Noorden GK: Examination of Patient—III: Sensory signs, symp-
toms, and adaptations in strabismus. In Binocular Vision and Ocular


62. von Noorden GK: Esodeviations. In Binocular Vision and Ocular Motil-

ity: Theory and Management of Strabismus, 5th ed, p 302. St. Louis,


63. von Noorden GK: Esodeviations. In Binocular Vision and Ocular Motil-

ity: Theory and Management of Strabismus, 5th ed, p 326. St. Louis,


64. von Noorden GK: Esodeviations. In Binocular Vision and Ocular Motil-


65. von Noorden GK: Special forms of strabismus. In Binocular Vision

and Ocular Motility: Theory and Management of Strabismus, 5th ed, p


66. von Noorden GK: Principles of nonsurgical treatment. In Binocular

Vision and Ocular Motility: Theory and Management of Strabismus,


67. von Noorden GK, Avilla CW: Accommodative convergence in hyper-


68. von Noorden GK, Morris J, Edelman P: Efficacy of bifocals in the

The onset of comitant strabismus in most patients occurs during early infancy or childhood. The two major categories of early-onset comitant esotropia are congenital (infantile) and accommodative. Although there is some disagreement as to the precise underlying pathophysiology and appropriate treatment of these disorders, there is general agreement that they are not causally related to any serious underlying central nervous system pathologic process. This, of course, is not to imply that these early-onset forms of esotropia lack neurophysiologic correlates.

In contrast, an older patient who presents with acute esotropia and diplopia should prompt careful consideration of whether the strabismus might be a sign of central nervous system disease. First, one needs to establish whether the deviation is comitant or incomitant. An acute-onset esotropia with incomitance must be considered to be a result of lateral rectus muscle paresis until proved otherwise. There is no doubt that the vast majority of neuropathic and/or myopathic cases of strabismus present as an incomitant deviation.

On the other hand, comitance in acute-onset esotropia does not always ensure that the underlying cause is benign. Until recently, most authorities would probably have agreed that “acute-onset esotropia that is comitant without divergence insufficiency is benign in nature, with no risk that it might be associated with intracranial pathologic processes.” Regrettably, it is now apparent that this simple algorithmic way of viewing acute-onset esotropia, although valid in the vast majority of cases, is not inevitably appropriate, because many exceptions exist. Comitance in acute-onset esotropia does not rule out the possibility of a serious underlying neurologic condition.

How then is the clinician to evaluate the patient with acute-onset comitant esotropia? Does every patient with this form of strabismus deserve neuroradiologic and/or neurologic evaluation? If not, what features or findings would indicate that a given patient with acute-onset comitant esotropia is at particular risk for neurologic problems?

Types of Acute Comitant Esotropia

Although scattered reports discussing cases of acute-onset esotropia can be found in the ophthalmologic literature dating back over the past century, it is the paper of Burian that is generally accepted as the pivotal work in this area. In reviewing the literature and his own experience, this author presented the thesis that acute-onset comitant esotropia may be separated into three distinct categories based on the clinical features and underlying etiology. Common to all three groups are an acute onset, comitance, a relatively large angle of deviation, good binocular potential, and the lack of any serious underlying neurologic disease.

TYPE I: ACUTE-ONSET COMITANT STRABISMUS AFTER OCCLUSION (SWAN TYPE)

This form of strabismus may occur after therapeutic patching or as the result of monocular or asymmetric visual loss. The resulting strabismus in children and young adults is usually an esotropia, whereas exotropia predominates in adults. Although most children who develop comitant esotropia after occlusion or asymmetric visual loss are hyperopic, several reports emphasize that this may occur in the absence of significant refractive error. It would appear, therefore, that occlusion of one eye or unilateral loss of vision, even in the absence of significant hypermetropic refractive error, may precipitate acute-onset comitant esotropia in some children or young adults.

TYPE II: COMITANT CONVERGENCE STRABISMUS (FRANCESCHETTI TYPE)

These patients develop an acute-onset esotropia that at first may be intermittent but quickly becomes constant. The
refractive error is usually minimally hyperopic, with a normal accommodative convergence/accommodation (AC/A) ratio. There are reports that this type of esotropia may occur in multiple siblings as well as in monozygotic twins. Surgery is necessary to reestablish ocular alignment, but the prognosis for regaining normal binocularity is good. Some patients in this category undoubtedly have had previously unrecognized microstrabismus that decompensated. In these patients, completely normal binocularity may not return.

**TYPE III: COMITANT CONVERGENT STRABISMUS ASSOCIATED WITH MYOPIA (BIELSCHOWSKY TYPE)**

In 1922, Bielschowsky reported a series of patients with acute-onset comitant esotropia associated with myopia. Similar cases had been described previously by von Graefe. The consistent features of this group were myopia of 5 D or less, esotropia at distance but maintained fusion at near, and no evidence of lateral rectus paralysis. Since Bielschowsky's original report, the characteristics have been redefined to include higher levels of myopia and constant deviations at both near and distance fixation. Subsequent reports have emphasized that good binocular function may be maintained with prisms. However, these patients often present with a reasonably small angle of esotropia (10 PD or less) but gradually develop an increasingly large angle that may preclude the use of prisms. Fortunately, surgery is usually effective in reestablishing normal binocular function.

Bielschowsky had no doubt that the myopia and the resulting accommodative convergence induced by near fixation play a central role in the origin of this form of acute-onset comitant esotropia. Recently, this theory was challenged by a report suggesting that direct damage to the lateral rectus muscle may be the primary pathologic problem. Neuroimaging studies suggest that displacement of the lateral rectus muscle may be important in the pathogenesis of this syndrome.

**Clinical Features of Comitant Esotropia Associated with Neurologic Disorders**

Of the three categories of acute-onset comitant esotropia described by Burian and Miller, only type II is likely to perplex the clinician with regard to its appropriate evaluation. Patients with type I will invariably have a history of monocular occlusion or visual loss, and those with type III will have a myopic refractive error. Patients with type II, on the other hand, are the real worry. How does one distinguish them from the uncommon but worrisome patients whose acute-onset comitant esotropia is associated with serious neurologic disease?

**HYDROCEPHALUS**

The association of comitant esotropia and hydrocephalus has been discussed in great detail. Harcourt should be credited for calling our attention to the frequent association of both comitant and incomitant esotropia with hydrocephalic disorders. Moreover, he challenged the notion that the comitant esotropia frequently seen in this setting is merely an expression of abducens nerve dysfunction. We believe it is useful to divide patients with comitant esotropia and hydrocephalus into two distinct categories.

First, a significant number of patients with hydrocephalus are seen with an early-onset large-angle comitant esotropia, resembling infantile esotropia. An A pattern is seen in the vast majority of these patients. These patients may demonstrate a comitant esotropia with nystagmus in abduction (Ciancia syndrome). Most of the patients with this form of comitant esotropia and hydrocephalus have a meningomyelocele or encephalocele. These patients rarely pose a diagnostic dilemma. They present in early childhood with a large-angle esotropia, usually an A pattern, and obvious neurologic problems associated with hydrocephalus or other congenital central nervous system anomalies. An A pattern often seen in these patients clearly speaks against the notion that sixth nerve paresis is the underlying pathologic process responsible for the esodeviation, because a small V pattern is expected in sixth nerve palsies.

In contrast, a second and smaller group of patients with hydrocephalus may present with the acute onset of comitant esotropia as a sign of dramatic elevation in intracranial pressure and/or ventricular shunt failure (Fig. 10–1). The comitant esotropia seen in these patients is usually a large-angle one, although they may initially present with a divergence paralysis that only later becomes constant at both near and distance fixation. An A pattern is distinctly unusual in this group. Associated ocular motor signs may include bilateral supranuclear upgaze palsy and nystagmus in primary gaze. Most of these patients will experience realignment of the ocular axes after restoration of normal intracranial pressure. Indeed, the acute onset of comitant esotropia with diplopia is now well recognized by neurologists and neurosurgeons as a sign of shunt failure. Several reports have documented recurrent comitant esotropia with repeated attacks of raised intracranial pressure, with a complete return of normal binocularity between attacks when intracranial pressure is normal.

The nature of the onset, the presence or absence of an A pattern, and the prognosis for binocularity are distinctly different in these two subgroups of patients with hydrocephalus. Moreover, the clinical findings imply that the underlying pathophysiology is not the same. Only in the second group of patients with an acute onset of comitant esotropia resulting from a sudden elevation in intracranial pressure is it likely that a diagnostic dilemma may occasionally occur. However, our experience even in these cases suggests that esotropia is rarely the presenting sign in an otherwise healthy child. In the vast majority of patients, hydrocephalus has been previously diagnosed, usually in infancy, and the esotropia is a sign of shunt failure.

**CHIARI TYPE I MALFORMATION**

Chiari malformation is the eponym given to a hindbrain anomaly first described by Chiari in 1891. In 1894, Arnold added a more detailed description of the disorder. Four types have now been described. The Chiari type I malformation involves displacement of the cerebellar tonsils into the upper
cervical canal (Fig. 10–2). Unlike many congenital malformations of the central nervous system, patients with this condition usually remain asymptomatic until late childhood or the early adult years. The most frequent symptoms are neck pain and headache. Other neurologic findings may include ataxia, dysarthria, dysphagia, diplopia, dizziness, and, on occasion, limb weakness and numbness.

Ophthalmic abnormalities may be the sole manifestation of Chiari type I malformation. Symptoms may include diplopia and oscillopia secondary to various forms of nystagmus. Increasingly, it has been recognized that acute acquired esotropia may be an early hallmark of this disorder. It may present initially as divergence paralysis; and as in long-standing comitant esotropia associated with hydrocephalus, an A pattern is frequent. However, not all patients with comitant esotropia and the Chiari type I malformation have associated hydrocephalus. Other ocular motor signs are frequent and may help the clinician establish the correct diagnosis.

Nystagmus is present in up to 60% of affected patients; it may be horizontal, rotatory, downbeating, or even upbeating. Sixth nerve paresis, skew deviation, internuclear ophthalmoplegia, spasm of the near reflex, Horner syndrome, ocular dysmetria, and loss of optokinetic nystagmus all have been described in association with this syndrome. Downbeating nystagmus is especially diagnostic and helps to localize the site of pathology to the area of the craniocervical junction. Nystagmus was lacking initially in several reports of patients with Chiari malformation and comitant esotropia. Comitant esotropia may be the presenting sign of a Chiari type I malformation, even in the absence of nystagmus or other obvious brain stem signs.

**BRAIN TUMORS**

That acute-onset comitant esotropia may be associated with a brain tumor has been increasingly recognized. Even those whose patients with acute-onset comitant esotropia have no obvious neurologic disease may agonize about when neurologic evaluation should be undertaken. No single type or site of brain tumor can account for all cases described thus far. Undoubtedly, more than one etiologic mechanism is at play. Nevertheless, patients with comitant esotropia and intracranial tumors do have some common clinical features.

By and large, these are pediatric patients. To our knowledge, only two adults with acquired comitant esotropia and intracranial brain tumors have been described. The onset of esotropia may be sudden, with a constant comitant esotropia with diplopia being noted. In some cases, the esotropia is initially intermittent and a constant comitant strabismus develops over a few days or weeks. The angle of deviation is often small or even intermittent initially, but over time tends to increase. It is essential to note that an A pattern is distinctly unusual in this group of patients. This is in sharp contrast to patients having long-standing hydrocephalus and a Chiari type I malformation. We are aware of only one patient with a brain tumor and comitant esotropia in whom an A pattern was described. This patient also had hydro-
Figure 10–3. A 6-year-old with a cerebellar astrocytoma had no repetitive signs other than comitant esotropia.

cephalus, but whether this was primarily responsible for the A-pattern esotropia is not clear. A small V pattern is common in patients having comitant esotropia and brain tumor and should suggest the possibility that the primary problem is subclinical paresis of the abducens nerve.19

Most unnerving about these patients is that the comitant esotropia is often the initial and only sign of intracranial pathology (Fig. 10–3). Indeed, in the most disturbing case described by Zweifach,20 a 9-year-old boy had comitant esotropia persisting for 28 months without any other neurologic symptoms or signs. Ultimately, other signs of posterior fossa dysfunction became evident, and a diagnosis of cerebellar medulloblastoma was established. At no time did the esotropia become incomitant, nor did other signs of abducens paresis become evident. It seems clear that paresis of the sixth nerve cannot explain all cases of comitant esotropia associated with brain tumors. In fact, in all but two such patients reported, a lack of motor and/or sensory fusion has been evident.4, 33 This suggests that a more central part of the ocular motor control system is involved in development of the strabismus. One should be concerned about a patient with sudden-onset comitant esotropia who does not demonstrate motor fusion with the appropriate prism correction or after strabismus surgery. A significant proportion of patients with brain tumors and comitant esotropia will exhibit nystagmus, particularly nystagmus in abduction (Ciancia syndrome).

Tumors of the cerebellum,11, 35, 48 brain stem (Fig. 10–4),48 sellar region (Taylor D, personal communication, 1997), and corpus callosum3 all have been associated with acute-onset comitant esotropia. However, the cerebellum is by far the most common tumor site (Fig. 10–5).4, 11, 33, 48 Some of these

Figure 10–4. A 4-year-old boy with diffuse pontine glioma had a 30–PD esotropia with a small V pattern.
patients present with papilledema and other obvious signs of raised intracranial pressure. In many cases, however, comitant esotropia is the presenting feature and remains the only sign of neurologic dysfunction for a considerable period. In contrast, brain stem tumors presenting with comitant esotropia usually produce other neurologic signs (e.g., sixth or seventh nerve paresis, gaze palsy, hemiparesis) within a short time.

**THALAMIC DISEASE**

An acute acquired comitant esotropia may be a prominent sign of pathology involving the thalamus. This is particularly true for adults with hypertensive bleeding within the thalamus. The comitant esotropia may be associated with forced downward deviation of the eyes and miosis. Both computed tomographic analysis and autopsy cases suggest that only patients whose thalamic hemorrhage extends into the dorsal midbrain are likely to develop these signs. This has led some authorities to suggest that the esotropia associated with thalamic dysfunction is a direct result of interference with vergence neurons in the midbrain. These adult patients rarely present a diagnostic dilemma, because associated neurologic signs invariably point to this life-threatening condition. These cases are of special interest because they may offer a model for understanding the esotropia frequently seen in premature infants with intraventricular hemorrhage. The recent finding of neurons within the dorsal midbrain of monkeys that respond to both convergence and divergence signals lends weight to the argument that this form of esotropia is caused by interference with vergence/fusional mechanisms.

**MYASTHENIA GRAVIS**

Whereas most patients with myasthenia gravis and diplopia will have evidence of incomitant strabismus with restricted ocular movements, occasionally in children this is not necessarily the case. Comitant esotropia and exotropia have been described as the initial presenting sign of infantile myasthenia gravis. These comitant deviations may continue to be the only sign of myasthenia gravis for several months before the variable nature of the disorder and associated ptosis become apparent. We have even seen cases of myasthenia gravis in which comitant strabismus resolved spontaneously, only to recur months later. Normal motor and sensory fusion has been documented during remission, even after months of diplopia with a comitant deviation.

**SEIZURES**

In at least two patients a constant comitant esotropia has been associated with epileptic activity. Of interest is the fact that, with medical control of the seizure disorder, esotropia resolved in both cases. The mechanism whereby esotropia is produced by cortical dysfunction in epilepsy is not known. It is a rare occurrence in a common disorder and may represent nothing more than decompensation of a previously unrecognized esophoria.

**Diagnosis**

Acute-onset comitant esotropia is an uncommon form of strabismus, even in practices taking a special interest in neurologic disorders of children. The vast majority of cases of acquired comitant esotropia will not be associated with serious underlying neurologic disease. When there is a history of previous strabismus, occlusion therapy, monocular visual loss, or myopia, acquired comitant esotropia need cause little worry. However, if a patient has no apparent cause for acute-onset comitant esotropia, the possibility of an underlying neurologic disorder should at least be considered. Attention to the clinical features of the ocular motor disturbance will help determine whether a neurologic problem is likely to be present.

The size of the deviation and the presence or absence of an A or V pattern are especially important in assessing these patients. An A pattern in an unequivocally acquired comitant esotropia almost invariably indicates that the patient has an underlying neurologic problem, either hydrocephalus or the Chiari type I malformation (or both). However, the patient

![Figure 10-5. A 7-year-old girl with a medulloblastoma and no hydrocephalus presented with an acute-onset comitant esotropia of 25 PD.](https://example.com/figure10-5.png)
is not likely to have a brain tumor. A V pattern may be present in patients with or without neurologic disease, and careful reassessment is essential to be certain that it is not an early sign of sixth nerve palsy. Now that it is apparent that not all divergence paralysis represent subclinical sixth nerve palsies,12 we need to pay special attention to patients who present with this form of comitant esotropia, particularly children. These patients are very likely to have an underlying neurologic problem, especially hydrocephalus or the Chiari type I deformity.

Forms of manifest and latent nystagmus are frequently seen in patients with infantile esotropia. Nystagmus is conspicuous by its absence in all benign forms of acquired comitant esotropia. Nystagmus therefore is an important sign suggesting underlying neurologic disease. Downbeating nystagmus localizes the pathologic process to the area of the cranio-cervical junction, and any form of nystagmus seen in a patient with acquired comitant esotropia should suggest an underlying neurologic process. Nystagmus may be seen in patients with hydrocephalus, the Chiari type I malformation, or a brain tumor.

Although further studies are needed to evaluate fusion potential in patients with comitant esotropia and a brain tumor, it appears at present that a lack of significant motor and sensory fusion, even after appropriate optical and/or surgical treatment, is found in a considerable number of patients. Indeed, we are struck by the fact that, in patients with recurrent obstructive hydrocephalus who have several episodes of comitant esotropia after resolution of the intracranial hypertension, normal motor and sensory fusion is almost invariably present. This stands in sharp contrast to patients with brain tumors, in whom fusion is often lacking even after multiple strabismus surgeries. We believe that a patient who presents with an acquired comitant esotropia and whose eyes cannot be made to fuse with hand-held prisms or on synoptophore examination is very likely to have an underlying neurologic problem, especially a brain tumor.

The patient with acquired comitant esotropia needs to be carefully examined to detect other neuro-ophthalmic signs of neurologic disease. Because the vast majority of reported patients with brain tumors and comitant esotropia have posterior fossa lesions, a careful examination for papilledema is of the utmost importance. However, a lack of papilledema does not exclude posterior fossa dysfunction. Not all ophthalmologists will be comfortable in assessing motor function in a patient who presents with acute comitant esotropia. However, most patients with significant cerebellar dysfunction can be identified by simply asking the patient and/or relatives about problems with motor function and coordination. Any recent evidence of clumsiness or loss of motor skills should prompt thorough investigation for a cerebellar tumor. It should be emphasized once again, however, that a number of patients with posterior fossa tumor have presented with comitant esotropia and no other signs of cerebellar dysfunction, even when examined by a competent neurologist.

**Treatment**

**NONSURGICAL MANAGEMENT**

By far the most essential issue in evaluating the patient with acquired comitant esotropia is establishing that there is no underlying neurologic problem. That does not mean that all patients with this form of strabismus require neurologic or neuroradiographic investigation. Certainly, the patient with nystagmus, poor fusional potential, and/or other ocular motor abnormalities demands an immediate neurologic and/or neuroradiographic workup. Even when the clinician believes that no serious underlying neurologic pathology exists, frequent reevaluation of patients with acquired comitant esotropia is mandatory. It should be recalled that a number of patients ultimately found to have the Chiari type I malformation were at first thought to be cured of their strabismus by surgery and/or hyperopic lenses, only to be subsequently reassessed when the esotropia recurred or it became obvious that a stable small-angle esotropia had not been established.

We want to caution against the partial response of acquired comitant esotropia to hyperopic spectacles. In reviewing all reported patients with neurologic disease and comitant esotropia, a significant number had delayed diagnosis because the angle of esotropia was partially reduced when the appropriate hyperopic correction was prescribed. Because the vast majority of patients with neurologic disease and acute comitant esotropia are children, the presence of a hyperopic refractive error is to be expected. Only if fusion can be reestablished with the hyperopic prescription should underlying neurologic disease be ruled out.

In most instances, the appropriate neuroradiologic approach to a patient with acquired comitant esotropia who is suspected of having neurologic disease is magnetic resonance imaging. In patients with hydrocephalus or a brain tumor, no special instructions are generally needed to identify the underlying neurologic process. The Chiari type I malformation, however, is often not apparent unless special attention is paid to the area of the cranio-cervical junction and a contrast magnetic resonance imaging study is performed.

**SURGICAL MANAGEMENT**

The esotropia associated with hydrocephalus and the Chiari type I malformation typically exhibits an A pattern. Surgical treatment will therefore require a determination of whether superior oblique overaction is present and accounts for the A pattern. If not, it may be necessary to transpose the medial rectus muscles to eliminate the A pattern (see also Chapter 14).

In general, we have found that patients with acquired comitant esotropia and underlying neurologic disease respond reasonably predictably to the usual amounts of surgical correction, even if fusion is not reestablished—as is often the case in patients with brain tumors. We want to emphasize that the patient who presents with acute comitant esotropia in association with shunt failure or acutely elevated intracranial pressure will probably not require strabismus surgery as long as the shunt is revised or the intracranial pressure returns to normal.

**REFERENCES**


Clinical Strabismus Management

3. Arnold J: Myelocyste, Transposition von Gewebskeimen und Sym- 


5. Biglan AW: Ophthalmologic complications of meningomyelocele: A 

6. Bixenman WW, Laguna JF: Acquired esotropia as initial manifestation 

7. Boering KP, Lorenz L: Normosesoriches Spatschielen OperationsZeit-

8. Burian HM: Motility clinic: Sudden onset of comitant convergent 

9. Chiari A: Uber Veranderungendes Kleinhirns inforte von Hydro-


15. Davidson JL, Rosenbaum AL, McCa LC: Strabismus surgery in pa-

16. Demer JL, von Noorden GK: High myopia as an unusual cause of 

17. Faria MA, Spector RH, Tindall GT: Downbeat nystagmus as the salient 
manifestation of the Arnold-Chiari malformation. Surg Neurol 
1986;36:165.

Trans Am Neurol Assoc 1959;84:56.

Orleans Academy of Ophthalmology, p. 449. New York, Raven Press, 
1986.

20. France TD: The association of “A” pattern strabismus with hydroceph-
alus. In: Moore S, Mein J, Stockbridge L (eds): Orthoptics: Past, Present, 

ology 1988;38:1759.

22. Harcourt RB: Ophthalmologic complications of meningomyelocele and 

23. Hertle RW, Bienfang DC: Oculographic analysis of acute esotropia 
secondary to a thalamic hemorrhage. J Clin Neuroophthalmol 

24. Hoyt CS, Good WV: Acute onset comitant esotropia: When is it a 

25. Hoyt WF, Daroff RB: Supranuclear disorders of oculocerebral control 

related to divergence eye movements and accommodation. J Neuro-

thalamic hemmorhages and their clinical implications. Neurology 
1986;36:165.

28. Kirkham TH, Bird AC, Sanders MD: Divergence paralysis with raised 
1972;56:776.

29. Krizioth TH, Kaufmann H, Traupe H: Elucidation of restrictive motility 
in high myopia by magnetic resonance imaging. Arch Ophthalmol 
1997;115:1019.

30. Lemmersand B, Gallo FE, Samuelson N: Neuro-ophthalmologic find-
ings in relation to CNS lesions in patients with meningomyelocele. 

31. Lewis AR, Kline LB, Sharpe JA: Acquired esotropia due to Arnold-

32. Lim L, Rosenbaum AL, Demer JL: Saccadic velocity analysis in 
patients with divergence paralysis. J Pediatr Ophthalmol Strabismus 
1995;32:76.

33. MacPherson H, DeBecker I, MacNeil JR: Beware: Armed and danger-

lateral rectus muscle in myopia with esotropia: An ultrastructural study. 

35. Micketavage RC: Ophthalmologic disease presenting as orthoptic prob-

36. Moore S, Welter P: Ophthalmologic diagnosis and evaluation of prism 

37. Norbis AL, Malbran AE: Comitant esotropia of late onset— 
pathologic report in four cases in siblings. Br J Ophthalmol 
1956;40:373.

of normal stereoacuity in acute-onset comitant esotropia. Am J Ophthal-


Orleans Academy of Ophthalmology, p 456. New York, Raven Press, 
1986.

41. Stolz SE, Chatrian GE, Spence AM: Epileptic nystagmus. Epilepsia 

42. Tamura EE, Hoyt CS: Ocular motor consequences of intraventricular 

43. Tischler AM, Rees MC, Dunn HG, et al: Esotropia and epileptic eye 

44. Vollrath-Junger C, Lang J: Akuten Strabismus konvergens bei erhöhtem 

45. Von Graefe A: Uber di bon Myopie abhangige Forme Conviergerenden 

46. Watson AP, Fielder AR: Sudden-onset squint. Dev Med Child Neurol 
1987;29:207.

47. Weber E: Mit Diplopie einhergehende Convergenschielen Myoper 
(Bielschowsky). Ophthalmologica 1947;1114:320.

48. Williams AS, Hoyt CS: Acute comitant esotropia in children with brain 

49. Wybar K: The significance of squint in certain forms of malignant 

50. Zweifach PH: Childhood esotropia with delayed appearance of cerebel-
Divergence paralysis is a relatively uncommon motility disorder seen infrequently in clinical practice. Typically, an elderly patient complains of the sudden onset of horizontal double vision. Diplopia is present for distance vision but disappears when reading.

Whether divergence is an active process remains controversial. Electromyographic studies show it to be associated with active innervation of the lateral rectus (LR) muscles. The existence and site of a divergence center are still under investigation, but it is postulated to be somewhere in the brain stem, most likely in the midbrain and pons, which also contains the oculomotor, trochlear, and abducens nuclei. Studies of rhesus monkeys have identified divergence burst cells located in an area of the mesencephalic reticular formation just dorsal and lateral to the oculomotor nucleus.

Jampolsky argues against the existence of a divergence center and does not recognize divergence paralysis as a clinical entity. He asserts that if the divergence center is paralytic, patients would be expected to have no divergence function. And yet, fusion/divergence function can be demonstrated in every patient with so-called divergence paralysis by stimulating unused function after restoring fusion by full prism correction.

### Clinical Characteristics

The clinical signs and symptoms of divergence paralysis were first described by Parinaud in 1883 and were elaborated by Duane in 1899. Bielschowsky summarized the diagnostic criteria: (1) homonymous diplopia at distance (beyond 10–20 inches); (2) comitance in lateral gaze; (3) orthotropia and fusion at near (10–15 inches); and (4) full ocular ductions and versions.

Increased intracranial pressure, intracranial tumors, head trauma, and vascular lesions of the brain stem are among the many neurologic disorders that may be associated with divergence paralysis. A case associated with ingestion of diazepam has been reported. In some instances the diploic symptoms disappeared when the underlying disease resolved. Divergence paralysis was the initial finding in a patient having severe ophthalmoplegia associated with Miller-Fisher syndrome. It may also present in isolation and remain the only neurologic deficit in an otherwise healthy adult. In a long-term study by Krohel and coworkers, no further neurologic sequelae developed in eight patients with divergence paralysis, although their esotropia persisted.

### Diagnosis

#### Clinical Examination

Strabismus measurements should be obtained, especially in primary and lateral gazes, to check for incomitance. Patients exhibit a small comitant esotropia, usually in the range of 12 to 15 PD at distance, but it may occasionally be as large as 30 PD. There is hardly any deviation or, at most, a small esotropia at near. The analysis of eye muscle movements should emphasize horizontal rotations. Ductions and versions are normal in divergence paralysis but are slightly limited in bilateral sixth nerve palsy, which may be confused with this condition.

Careful assessment of sixth nerve function is required to make an accurate diagnosis. Bedrossian recommended plotting accurate fixation fields to unmask minimal abduction weakness and seeking nystagmus on extreme lateral gaze to identify subclinical sixth nerve paresis.

The most important concern is whether divergence paralysis of new onset is truly an isolated occurrence. Such patients should undergo neurologic evaluation to rule out increased intracranial pressure and neuroradiologic examination to detect any lesion in the central nervous system. If the workup is negative, the patient should be observed at regular intervals. A patient who presents initially with associated focal neurologic signs or develops such signs requires a complete neurologic workup.

#### Laboratory Evaluation

Horizontal saccadic velocity testing using electro-oculography has served to measure LR muscle function (Fig. 11–
Reduced abduction velocities in a patient with elevated intracranial pressure who develops classic features of divergence paralysis are considered by some authors to reflect minimal sixth nerve palsy. Normal peak abduction velocities, indicating unimpaired LR muscle function, have been documented in patients with isolated divergence paralysis.

Differential Diagnosis

Most authors recognize divergence paralysis as a clinical entity distinct from sixth nerve palsy (Table 11–1). Jampolsky, however, believes that divergence paralysis is synonymous with bilateral sixth nerve palsy and that careful evaluation will demonstrate LR muscle dysfunction and the presence of divergent fusional amplitudes. Many of the same underlying neurologic disorders that are sometimes associated with divergence paralysis are known to cause LR palsy. The most common is increased intracranial pressure from any cause. Abducens nerve paresis may mimic divergence paralysis. Bielschowsky observed numerous patients with symptoms typical of unilateral or bilateral LR palsy who subsequently developed features indistinguishable from divergence paralysis. The incomitant deviation observed initially became comitant over time. Cases of benign sixth nerve palsy presenting initially with symptoms consistent with divergence paralysis also have been reported.

Two other forms of acquired comitant esodeviation in adults should be considered in the differential diagnosis of divergence paralysis: decompensated esophoria and decompensated monofixational esotropia. In decompensated esophoria, the fusional mechanism that initially controlled the deviation has deteriorated so that the latent deviation has become manifest. Careful evaluation will reveal that the distance and near deviations are equal, distinguishing this entity from divergence paralysis. Particular care should be taken to control accommodation at near fixation to ensure accurate measurements. In patients with decompensated monofixation esotropia, a search for the specific sensory and motor abnormalities found consistently in monofixational syndrome will distinguish this condition from divergence paralysis.

Treatment

Nonsurgical Management

Diploic symptoms that persist are treated with base-out prisms before strabismus surgery is considered. Prisms are
particular benefit to patients having smaller deviations. However, because of the difference in the size of the distance and near deviations, separate spectacles are required for distance and near. Prisms are usually needed only for the distance deviation. Alternatively, Fresnel (add-on) prisms may be trimmed to fit only the upper (distance) segment of a spectacle lens with a bifocal segment (Fig. 11–2).

Orthoptic training is an alternative approach to treating divergence dysfunction. The goal of divergence orthoptic exercises is to enhance fusional divergence amplitudes so that the patient can fuse a manifest deviation and obtain symptomatic relief. Repeat training sessions are necessary when symptoms recur. Because of the excellent results gained from prism therapy and strabismus surgery, orthoptic exercises are rarely used and little is known of their long-term effectiveness.

**SURGICAL MANAGEMENT**

Strabismus surgery is indicated in patients who do not respond adequately to prism therapy. Surgical intervention is highly successful in the treatment of this form of strabismus. Tenotomy of one or both medial rectus muscles was initially performed, but the appropriateness of weakening the muscles of convergence to treat divergence paralysis was questioned. It was argued that it is more rational to advance or resect the LR muscles. The logical operative principle in treating divergence paralysis is to strengthen the muscles of divergence. LR resection also has greater effect in the distance than at near.

Both unilateral and bilateral LR resection reportedly are effective in treating 8 to 30 PD of distance esotropia. Satisfactory results were obtained by performing a 5.0- to 6.0-mm resection to correct 8 to 18 PD of distance esotropia. Some patients may require a small prism to maintain single binocular vision postoperatively.

My colleagues and I performed a study in which a 4.0- to 6.0-mm resection of both LR muscles on adjustable sutures corrected 16 to 30 PD of distance esotropia in five patients. Addition of the adjustable suture technique provides an opportunity to fine-tune ocular alignment in the immediate postoperative period. The goal is to produce orthotropia or a small exophoria (5–7 PD) that the patient can easily fuse after adjustment. The tendency is for LR resection procedures to relax over time. Thus, a small overcorrection at distance will dissipate in the long term. Rarely, a small overcorrection or undercorrection present postoperatively may be managed with Fresnel prisms until the patient is able to fuse the residual.

Hoover and colleagues corrected 12 to 20 PD of distance esotropia by performing 6.0 to 8.0 mm of unilateral LR muscle resection under peribulbar or retrobulbar anesthesia in six patients with divergence paralysis. Local anesthesia is a viable alternative to general anesthesia and may be the preferred technique for elderly patients with high-risk medical conditions.

**Conclusions**

Divergence paralysis is one form of strabismus most successfully treated surgically. The strabismus surgeon may be reluctant to proceed with definitive management, because of the patient's age, higher medical risk, and the "small" (12–15 PD) amount of esodeviation. The surgical procedure (LR resection) is, however, relatively simple and requires only a brief anesthetic. Local anesthetic delivery systems preclude the need for general anesthesia. The peribulbar and sub-Tenon routes avoid the risks connected with bilateral retrobulbar injection. Using an adjustable suture technique reduces the possibility of overcorrection and undercorrection. After realignment, these patients often are among the happiest to be encountered in clinical practice.

**REFERENCES**

Historical Perspective

Duane theorized that exodeviations are caused by innervational imbalance between convergence and divergence mechanisms. Whether divergence is an active or passive process remains a subject of debate. Neuroanatomic substrates, including the area of the tegmentum of the brain stem in humans, and divergence burst cells in the area of the mesencephalic reticular formation (located dorsolateral to the oculomotor nucleus in rhesus monkeys) have been suggested as possible sites of a divergence center. No true divergence nucleus, however, has yet been found. That divergence is a passive process, conditioned by relaxation of accommodation and the absence of simultaneous contraction of both lateral rectus (LR) muscles, is a view shared by other authors. The deviating eye demonstrates increased innervation on electromyographic recordings of the LR muscle at the onset of exodeviation, whereas the fixing eye fails to show any increase in motor activity.

Bielschowsky argued that there is an abnormal position of rest in exodeviations, related to anatomic factors, interpupillary distance, the shape and size of the globe and orbits, and properties of the extrabulbar tissues. Orbital depth and growth, as well as horizontal muscle length and insertion, influence the equilibrium of passive forces between the medial and LR muscles.

Burian believed that exodeviations are governed by both static (mechanical and anatomic) and dynamic (innervational) factors. The static position of rest is the relative position of the visual axes when there is no stimulus for fusion. Refractive errors are corrected and the dominant eye is fixing at a distant object in primary gaze. There exists a dynamic interplay between ocular convergence and divergence mechanisms. Any imbalance favoring divergence may result in the development of exodeviations.

Epidemiology and Risk Factors

Exodeviations occur less frequently than esodeviations in a ratio of 1:3 and are more common in females. They occur more frequently in the Middle East, subequatorial Africa, and the Orient than in the United States, and less frequently in central Europe. Exodeviation is found more frequently at latitudes with higher levels of sunlight. Thirty-five percent to 40% of cases are seen before the second year of life.

Facial asymmetry associated with exodeviations has been described. The deviated eye is invariably on the side with a slanted posterior orbit and is usually accompanied by antimongoloid lid fissures. There may be a history of birth trauma, less facial development on the affected side, or anisometropic amblyopia resulting from unequal sensory input (such as that occurring with Descemet’s tear due to forceps extraction).

Children born with craniofacial anomalies are more likely to exhibit exotropia (XT) (see also Chapter 30). Exotropia is also more common in patients with neurologic deficits. Both maternal smoking during pregnancy and low birth weight are significant and independent risk factors for the development of horizontal deviations.

Genetics

Heredity plays a significant role in exodeviations, but the genetics of the disorder probably are multifactorial. A positive family history is often elicited. The chance of XT developing if one sibling from a multiple birth is affected is increased 17-fold. No such association is found for siblings from separate births. An autosomal dominant pattern of constant XT has been documented in a single case report.

Clinical Characteristics

Classic Presentation

Exodeviation is characterized by visual axes that form a divergent angle. It usually begins as an exophoria (X). During this phase, patients are assumed to have bifixation
and normal retinal correspondence. When XT develops in a patient with a mature visual system, diplopia occurs during periods of exodeviation because suppression is absent. In younger children with an immature visual system, the development of bitemporal suppression precludes the awareness of diplopia. Exophoria may later progress to intermittent exotropia (X[T]), in which case the deviation vacillates between phoric and tropic phases (Fig. 12–1) and finally to constant XT.

Characteristically, many patients—especially children—will close one eye in bright light or direct illumination (Fig. 12–2). This may precede the actual observation of an exodeviation. It has been assumed that bright light dazzles the retina, somehow disrupting fusion and causing the deviation to become manifest. This implies that one eye is closed to avoid diplopia and visual confusion, despite lack of awareness of diplopia.

Present evidence suggests that eye closure is due to a binocular summation of photalgia associated with exposure to high-intensity light. The threshold for monocular photophobia is significantly higher than that for binocular photophobia. It represents an adaptive response intended to reduce discomfort and decrease photophobia rather than diplopia. This finding is also seen in patients with other forms of strabismus and in people without strabismus.

Bright light has been shown to adversely affect the amplitude of fusional convergence in patients who are in a delicate state of balance between X and X(T). This effect is not seen in patients with orthotropia and constant XT.

Figure 12–1. Child with intermittent exotropia. Top photograph shows orthotropia. Bottom photograph shows exotropia.

Figure 12–2. Monocular eye closure in bright lights may precede the appearance of exotropia in some patients.

Children with X(T) and hemiretinal suppression have very few symptoms. Without suppression, older children and adults complain of asthenopia, blurred vision, headaches, diplopia or visual confusion, and reading difficulties—especially after prolonged periods of near work. Patients commonly complain of losing their place while reading and needing to repeatedly start on the same line. Rarely, micropsia has been reported. Accommodative convergence is used to control exodeviation at distance so that objects appear smaller and closer.

There have been few studies documenting the natural history of untreated XT. In a series of 48 patients 6 to 22 years of age with unoperated X(T) who were observed for an average of 11.7 years, 65% showed improvement. A majority of these patients were exophoric rather than exotropic, with deviations less than 20 PD. In a personal communication, the authors reported that 80% of those who improved received some type of orthoptic treatment. Because the study does not represent a random sample of exotropic patients, the only conclusion that can be drawn is that not all patients with X(T) experience a deterioration.

In a separate study of 51 patients 5 to 10 years of age, observed for 3.5 years on average, 75% showed progression of X(T), 9% remained unchanged, and 16% improved without intervention.

The progressive nature of the disease has therapeutic implications. Factors that affect progression include decreasing tonic convergence with advancing age, the development of suppression, a gradual reduction in accommodative power, and increased divergence of the orbits with age. Progression takes several forms. Fusional control may decline so that an X develops into an X(T). The phoric phase may be more prominent than the tropic component. Later, the tropia manifests more often and may progress to a constant XT. The tropic phase often is aggravated by daydreaming, fatigue, illness, and distance viewing. Progression may also take the form of an increase in magnitude of the deviation. Deteriorating control may be monitored by both distance and near stereoacuity tests (see later discussion).

CLASSIFICATION

The foundations of the clinical classification of exodeviations were established by Duane, Scobee, and Burian.
and associates. Today, we incorporate the strength of fusional mechanisms into the functional classification scheme. (A comparison of the Burian and Kushner classification is shown in Table 12–1.)

**Basic Exodeviation**

Basic distance deviation is within 10 PD of the near deviation. These patients have a normal accommodative convergence/accommodation (AC/A) ratio.

**Convergence Insufficiency**

Near deviation measures at least 10 PD more than distance deviation. Patients have either a low AC/A ratio or fusional convergence insufficiency (decreased fusional convergence amplitudes). Monocular occlusion does not increase the deviation at distance. This condition needs to be distinguished from a subgroup of patients having a pseudoconvergence insufficiency, in whom monocular occlusion or fixation at infinity increases the amount of the deviation.

In the early stages of convergence insufficiency, patients complain of asthenopia, visual fatigue, blurred vision, and intermittent diplopia at near. Monocular occlusion commonly relieves patients of symptoms but no X at either distance or near is present. Patients may present in their teens, when reading demands are greater. As the process progresses, an X or frank X(T) with a convergence insufficiency pattern develops. The early stages are most responsive to orthoptic exercises when the deviation is less than 8 PD. In the latter stages, exercises should be tried, but surgical management may be required for definitive therapy.

**True Divergence Excess**

Exodeviation measures at least 10 PD more at distance than at near. The AC/A ratio is normal. Measurements at near do not increase with +3.00-D lenses nor with prolonged occlusion.

**Pseudodivergence Excess**

The initial measurements reveal a distance exodeviation more than 10 PD of that at near. Several types are recognized:

1. Normal AC/A ratio with pseudo high AC/A ratio and tenacious proximal fusion. These patients may initially appear to have a high AC/A ratio following an increase in the deviation at near with +3.00-D lenses. With prolonged monocular occlusion, the tenacious proximal fusion is eliminated and the near deviation increases. When +3.00-D lenses are used after monocular occlusion, the deviation will be the same at both distance and near. This stresses the need for prolonged occlusion before using +3.00-D lenses.

2. Normal AC/A ratio with tenacious proximal fusion but without pseudo-high AC/A. Only monocular occlusion will increase the size of the near deviation, and +3.00-D lenses will not affect measurements at near.

3. High AC/A ratio. Monocular occlusion does not increase the deviation at near because of the absence of tenacious proximal fusion. A high AC/A ratio can be documented with either the gradient method or +3.00-D lenses. A small esodeviation can be documented at distances closer than ½ m. The distance deviation may or may not be affected by fixation at more than 6 m (20 ft) by monocular occlusion.

**Diagnosis**

**CLINICAL EVALUATION**

**Assessing Control of X(T)**

Evaluation of control is necessary to obtain a baseline assessment as well as to monitor deterioration and progression of X(T). It allows the ophthalmologist to be sensitive to early signs of deterioration and institute timely intervention.

**Subjective Methods.** Traditionally, most methods of assessing control in X(T) have been subjective. They include observing control in the office and at home and determining the frequency and duration of the deviation.

We have devised guidelines to provide semiquantitative criteria for evaluating control, but they remain subjective and yield variable results among different observers.

**Home Control.** The parents or a caregiver may be asked to categorize control of the deviation at home. Patients who manifest the deviation rarely and only at distance when fatigued, daydreaming, or inattentive may be considered to have excellent control. When the deviation is manifest fewer than five times a day and only at distance, control is considered good. When the patient exhibits the deviation more than five times a day but only at distance, while maintaining alignment at near, control is considered fair. With poor control of the deviation, the patient “breaks” frequently at both distance and near. Only occasionally can one observe orthotropia.

**Office Control.** In the office, the ophthalmologist can confirm good control when the patient “breaks” only after cover testing and resumes fusion rapidly without need for a blink or refixation. Patients who blink or refixate to control the deviation after disruption with cover testing have fair control. The patient who “breaks” spontaneously without any form of fusion disruption has poor control.

**Table 12–1. Burian Versus Kushner Classification of Exotropia**

<table>
<thead>
<tr>
<th>Burian’s Classification</th>
<th>Kushner’s Classification</th>
</tr>
</thead>
<tbody>
<tr>
<td>Divergence excess</td>
<td>Proximal convergence</td>
</tr>
<tr>
<td>Simulated divergence</td>
<td>High AC/A ratio</td>
</tr>
<tr>
<td>Simulated divergence</td>
<td>Tenacious proximal fusion</td>
</tr>
<tr>
<td>excess (based on monocular</td>
<td>(pseudo-high AC/A ratio)</td>
</tr>
<tr>
<td>occlusion)</td>
<td></td>
</tr>
<tr>
<td>Simulated divergence</td>
<td>Basic</td>
</tr>
<tr>
<td>Simulated divergence</td>
<td>Low AC/A ratio</td>
</tr>
<tr>
<td>excess (based on +3.00-D</td>
<td>Fusional convergence</td>
</tr>
<tr>
<td>lenses)</td>
<td>insufficiency</td>
</tr>
<tr>
<td>Basic</td>
<td>Pseudo-convergence</td>
</tr>
<tr>
<td>Convergence insufficiency</td>
<td>insufficiency</td>
</tr>
</tbody>
</table>
Objective Methods

**Distance Stereacuity.** Distance stereacuity measurements provide an objective assessment of both control of the deviation and the deterioration of fusion that occurs early in this disorder. Normal distance stereacuity probably indicates good control with little or no suppression. The Mentor B-Vat II BTS assesses distance stereacuity using both contour circles and the random dot E test from 240 to 15 seconds of arc disparity (Fig. 12-3). With contour circles, median distance stereacuity in patients with X(T) was 60 seconds of arc compared with 30 seconds of arc in nonstrabismic individuals. This difference was accentuated with the random dot E test. The median value for exotropic patients exceeded 240 seconds of arc, compared with 120 seconds of arc for the control group. Poor control of the distance deviation correlated well with changes in office control.

**Near Stereacuity.** Near stereacuity does not correlate well with the degree of control in X(T). In our series, the normal control group and patients with X(T) performed equally well in the near stereacuity test. Performance in this test is only minimally affected by surgery.

**Alternate-Letter Suppression Testing.** Some authors contend that central suppression precedes a recordable loss of distance stereacuity. Alternate-letter suppression testing may be a sensitive early test. It is available with the Mentor BVAT II systems and the Reichert or Stereo Optical vectographs.

**Detecting Amblyopia and Assessing Refractive Error**

Amblyopia occurs in 9% to 13% of patients with X(T). It is not as common in patients with X(T) as in those with esotropia. When it occurs, it is usually associated with anisometropia. Patching in unilateral XT may improve control of the deviation, presumably because of improvement in amblyopia.

### Measuring the Deviation

The usual distance deviation is measured with an accommodative target (e.g., 20/70 or smaller Snellen optotype) at 6 m. In X(T), where control of the distance deviation is compromised first, the full deviation may not be apparent at distances less than 6 m. In some instances, we have found it useful to take the patient out of the examining lane to fixate at a more distant target 50 to 100 ft away to obtain the full exotropic angle. The exodeviation may increase by 16 to 30 PD at distances beyond 6 m. In a prospective randomized trial, 86% of patients who underwent surgery for the largest angle had a satisfactory outcome, compared with 62% who were operated on for the "standard" distance (of 6 m) deviation.

Because fusional control may be tenacious, prolonged occlusion may be required to eliminate all fusional vergence. An orthoptic patch is applied over one eye for 30 minutes to an hour, or sometimes as long as 24 hours, before repeating measurements. The patient is not permitted to resume fusion before the alternate cover test is performed. Both eyes should remain closed until the examiner has removed the patch and put a cover in front of one eye. The alternate cover test is done for both distance and near in this manner (see also Chapter 1 for details of performing the test).

Monocular occlusion should be used before +3.00-D lenses to measure near deviation, to avoid misdiagnosing a high AC/A ratio. The +3.00-D lenses suspend normal accommodative convergence, whereas monocular occlusion relaxes fusional convergence mechanisms.

### Measuring the AC/A Ratio

Any AC/A determination in X(T) will be contaminated by tenacious proximal fusion (postocclusion vergence aftereffect). This effect from sustained vergence masks the true magnitude of the near deviation and will require more than a brief cover test to break. The AC/A ratio may still be measured with the gradient method or the heterophoria method in these patients as long as prolonged monocular occlusion has been performed before its determination (see Chapter 5 for measuring AC/A ratio). Most patients who show an increase in the near deviation with both occlusion and +3.00-D lenses will be found to have normal AC/A ratio when monocular occlusion is performed before testing with +3.00-D lenses.

### Lateral Incomitance and Ocular Rotations

Lateral incomitance was first described by Moore and was defined as a difference in size of the deviation in primary position and lateral gaze. To be considered significant, there must be a reduction of 20% or greater in lateral gaze from the primary position. Others use a difference of 5 to 10 PD. Horizontal incomitance has been reported in as few as 5% and as many as 24% to 60% of exotropic patients. It may occur primarily or as a result of surgical intervention. Recognizing lateral incomitance is important in determining whether to modify the amount of surgery on the horizontal rectus muscles. Measurement artifacts can be avoided by maintaining the proper position of the prism.

---

*Figure 12-3. Bar graph of distance and near stereacuity testing in normal and exotropic patients. Preoperative and postoperative values are shown. Notice that preoperative contour circle stereacuity is poorer in exotropic and normal patients. Some improvement is observed after surgery. Comparison with near stereacuity testing with Titmus vectograph demonstrates relatively excellent near stereacuity despite deterioration in distance stereacuity.*
when measuring the deviation in lateral gazes \(^{92}\) (see also later discussion under Common Pitfalls in Management).

Particular attention must be paid to ocular rotations, especially in the fields of the medial rectus (MR) and LR muscles. Abduction deficit may be due to either a tight MR muscle \(^{22}\) or an underacting LR muscle. \(^{31, 122}\)

**Measuring Convergence**

Convergence is a binocular vergence movement that increases the angle formed by the visual axes, usually through simultaneous adduction of both eyes. \(^{119}\) Any deficiency in convergence amplitudes (near amplitudes less than 20 PD) or a remote near point of convergence (NPC) (10–30 cm or more) constitutes convergence insufficiency. \(^{126}\)

Convergence amplitudes are commonly measured with rotary prisms or prism bars for both distance and near. Base-out prisms are gradually added, with both eyes open, until the blur point is reached. At this point, images remain single but blurry. Additional base-out prisms then are added until the patient reports diplopia, which corresponds to the break point. In patients who do not recognize diplopia because of suppression, the break point may be determined by observing when one eye starts to deviate outward. \(^{120}\) Normal convergence amplitudes are 20 PD for distance \(^{120}\) and 30 to 35 PD for near. \(^{129}\) Base-out prisms are then gradually reduced until the patient regains single vision to determine the recovery point. The recovery point is normally 2 to 4 PD less than the actual break point. In some patients with intermittent deviations such as X(T), this difference may be marked. This indicates that once disrupted, it is difficult to regain fusion. \(^{120}\)

Starting with a fixation object at 30 to 40 cm, the patient is instructed to maintain fixation as the object is brought closer to the eyes. The near point of convergence is defined as the distance at which the eye starts to lose fixation and turns out. It normally is 5 to 10 cm from the bridge of the nose. \(^{120, 129}\)

**Prism Adaptation Test**

The prism adaptation test (PAT) was first described for acquired esotropia \(^{64}\) and its efficacy confirmed by later studies. \(^{53, 105}\) Fresnel add-on prisms are applied on the patient’s spectacle correction preoperatively. The surgical procedure is directed at the total prism-adapted deviation.

The high incidence of long-term surgical undercorrections for X(T) patients \(^{96, 108}\) prompted some authors to use the same principles of PAT for esotropia in preoperatively determining the magnitude of the total exodeviations. \(^{96, 115}\) Improved surgical results were obtained when the preoperative PAT was used to measure the deviation. \(^{96, 115}\) These studies were, however, confounded by several factors, including differing surgical procedures such as recess-resect or bilateral LR recessions, different ages between groups at the time of surgery, and varying preoperative sensory status.

**Treatment**

**NONSURGICAL MANAGEMENT**

Nonsurgical management is indicated in patients with excellent control as measured by normal distance stereoaucity and in young children who are at risk of developing monofixational esotropia from persistent surgical overcorrection. At the present time, fusional ability is considered intact if distance stereoaucity is normal. These patients should be periodically monitored for motor control and sensory status. In children, nonsurgical techniques such as minus lenses and prisms can prevent or reverse early sensory anomalies by maintaining the potential for equal vision in each eye and preserving binocular fusion status. \(^{62, 99}\)

**Refractive Errors**

Unequal clarity in vision represents an obstacle to fusion and can facilitate suppression, contributing to progressive loss of control in X(T). Significant refractive errors, especially astigmatism and anisometropia, need to be corrected. \(^{63}\)

The patient with X(T) and hyperopia of more than 3.00 D poses a unique dilemma for the strabismus surgeon. Some children with moderate to severe hyperopia have been shown to actually improve their ocular alignment after hyperopic spectacle correction. \(^{94}\) This probably occurs because of the influence of improved visual acuity on ocular alignment.

Most patients with X(T), however, have a smaller exotropic deviation without the hyperopic correction. If the patient is able to control the deviation in this manner, then only the minimal hyperopic correction providing comfort should be prescribed. In the presence of manifest hyperopia, the goal is to provide the best acuity with the smallest amount of hyperopic correction. Patients should be informed of the possibility that hyperopic spectacles may lessen control and augment the size of the exotropic deviation. When necessary, small amounts of base-in prisms may be used if the hyperopic correction makes control of the deviation more tenuous. Similarly, beginning presbyopic correction can interfere with the control of an exotropic deviation at near. Only the minimum amount of reading addition should be prescribed. \(^{98}\)

If control deteriorates and surgery is required, it is impossible to obtain consistent measurements without the full hyperopic correction in place. Children aged 7 years or older and adults may not tolerate the full correction. A compromise will be required; the maximum tolerated hyperopic correction will suffice. Patients should wear the correction for at least a month, and an attempt made to increase the hyperopic correction at each follow-up visit. Of course, this will cause the X(T) to worsen and will require that hyperopic spectacles be worn postoperatively. \(^{98}\) The total exotropic angle obtained with the maximum tolerated hyperopic correction is the target angle for surgery.

**Minus Lenses**

Caltrider and Jampolsky \(^{39}\) have advocated minus lenses for the primary treatment of X(T) in children. They described results of treating 35 patients who did not receive surgery for X(T) but were treated by 2.00 to 4.00 D of overcorrecting minus lenses for a median duration of 18 months. The authors reported that approximately half of the patients had improved quality of fusion while undergoing therapy. A fourth had both improved quality of fusion and a quantitative decrease in the angle of deviation. Conversely, therefore,
the strabismus angle was not decreased in the majority of the patients.

Overcorrection can result from wearing minus lenses. Two patients in the series just mentioned developed esotropia while wearing the minus lenses. Although most children with high AC/A ratios would readjust their initial near deviation, a few would remain esotropic wearing minus lenses. All patients in minus lenses should be seen within 3 to 4 weeks after starting the therapy. Minus lenses should be discontinued if esotropia develops. Wearing minus lenses improves the quality of fusion and occasionally even decreases the angle of the exodeviation so that surgery may be deferred.

In our experience, the use of minus lenses (2.00–4.00 D) may be helpful in younger patients with exodeviations of 5 to 15 PD. Using over-minus lenses stimulates accommodative convergence. Lenses may be tried when surgery is contraindicated for various reasons. Success with minus lenses, both as a primary treatment and for postoperative undercorrection, approaches 50% in some reports. The effect may last for up to 1 year after discontinuation of therapy in 70% of those who improved initially. As the child grows older, asthenopic symptoms with over-minus lenses become prominent as the amount of near work increases.

**Prisms and Orthoptics**

For small comitant deviations of up to 20 PD, base-in prisms may be used to assist control and relieve asthenopic symptoms. Patients do become dependent on prisms, however, reducing the need for convergence effort. In time, patients may "eat up" the prisms and gradually develop an increasing exotropic angle.

Convergence exercises are indicated for symptomatic patients with convergence insufficiency because of the high risk of overcorrection with surgery. The goal is to increase the ranges of fusional convergence and divergence. They include near point exercises (e.g., pencil push-ups), prism convergence exercises, and red glass convergence exercises to name but a few. In pencil push-ups, the fixation target is presented at a remote distance where it is easily fused and gradually brought toward the nose until the break point is reached. In prism convergence exercises, a prism bar oriented base-out is gradually increased as the amount of near work increases.

Some ophthalmologists use occlusion therapy for preoperative antisuppression, but the efficacy of this treatment remains debatable. Alternate patching has also been tried in an effort to improve control and reduce suppression.

**Botulinum Toxin**

Botulinum toxin has been tried for the management of exodeviations but remains a reasonable alternative in patients with small-angle X(T) and to achieve a better cosmetic outcome in patients who cannot undergo anesthesia or surgery. A small dose of the toxin (1 to 1.5 units) may be given for small overcorrection or undercorrection especially in adults in whom the procedure can be performed in the office (see also Chapter 32).

**SURGICAL MANAGEMENT**

**Indications for Surgery**

Opinions vary widely with regard to the appropriate timing of surgical intervention in a child or adult with X(T). For example, the mere existence of X(T) may be considered an indication. The finding that as many as 75% of patients with X(T) show progression alerts us to the need for timely intervention. However, determining the best time to operate may be a difficult question to answer definitively. Current data suggest that surgery be performed once deterioration is documented. Waiting until deterioration has relentlessly progressed may reduce the chance of obtaining an excellent surgical outcome.

**Poor Control of X(T)**. Poor control can be judged both by home control and office control. X(T) occurring at least 50% of the time may be a manifestation of poor control calling for surgical intervention.

**Deterioration of Control of X(T)**. Before deterioration can be assessed, serial observations are required. These include monitoring for any increase in the size of the deviation; progressive deterioration in stereoacuity, especially at distance; loss of control; a more frequent manifest or tropic phase of the exodeviation; and progressive inability to refuse once the deviation has become manifest.

**Deterioration in Stereoacuity**. Earlier studies have recognized sensory decompensation as indicating loss of control, but this was measured in terms of near stereoacuity. Loss of control of the distance deviation precedes loss of control at near. Rosenbaum and coworkers provided a way of assessing control of the distance deviation using distance stereoacuity techniques. We recommend surgery for patients who show poor distance stereoacuity or progressively deteriorating distance stereoacuity.

**Increase in the Size of Exodeviation**. Increase in the size of exodeviation indicates progression of the exodeviation.

**Development of Suppression**. Development of suppression occurs especially in children.

**Severe Asthenopia**. Most patients with X(T) are usually asymptomatic because of the development of hemiretinal suppression. In some cases, however, such as in the convergence insufficiency type of exodeviation, severe asthenopic symptoms are especially bothersome at near. Asthenopia may be a consequence of deteriorating control. If a trial of vigorous orthoptic exercises is unsuccessful, definitive surgery will be required.

**Visual Confusion and Diplopia**. Bothersome diplopia is usually an indication for surgical correction. However, visual confusion is often overlooked because patients usually do not volunteer this information. The ophthalmologist therefore has to inquire about this symptom. Visual confusion may occur even in the setting of poor vision or amblyopia.
Factors Affecting Response to Surgery

Age. Advocates of early surgery for X(T) believe that surgical correction before age 4 years yields better results than later surgery. 16, 80, 90 In another study, no relationship between age and success was apparent. 99 Reoperation rates, risk of developing amblyopia, and loss of fusion were, however, greater in the younger age group. 62

In younger patients, a small overcorrection may lead to a monofixational type of esodeviation. This precludes the development of full near stereocuity and increases the risk of mild amblyopia 19, 33, 62, 95 (see also the discussions on monofixational esotropia in Chapter 9 and monofixational exotropia in Chapter 13). 99 This has led authors to recommend exercising caution before attempting surgery in children younger than age 4. 122 We reserve surgery in this age group for patients in whom rapid loss of control is documented. 99 In the interim, minus lenses may be used to encourage fusion, base-in prisms tried, or antisuppression therapy attempted. 20, 59, 62, 122

Degree of Control. Patients with better fusion control before intervention fare better than those whose control has deteriorated. In cases in which X(T) has deteriorated to constant XT, binocular function with stereopsis may not return. 84, 122 It is important to recognize that even adults who have deteriorated constant XT will have a chance to improve their fusion status. 62, 82

Sensory Destabilizing Factors. It is very tempting to discontinue optical correction of moderate postoperative refractive error once the X(T) has been corrected surgically. This seemingly trivial sensory destabilizing factor may predispose to recurrences of the exodeviation. 52, 68

The practice of monovision should be discouraged in strabismic patients who are able to fuse. Unequal binocular input, with one eye blurry at distance and the other blurry at near, predisposes to decompensation of control of the strabismic deviation. The same principle applies to patients who undergo refractive surgical procedures. Overcorrection of the myope by refractive correction will reduce the accommodative convergence effort and compromise control of the exotropic deviation.

Tenacious Fusion at Near. Tenacious proximal fusion, as manifested by the need to use prolonged occlusion preoperatively to bring out the true magnitude of the near deviation, may predict better results after surgery. 67 Patients with good fusion control had a surgical success rate of 83%, compared with 40% in those without evidence of tenacious fusional ability and a normal AC/A ratio (basic exodeviation). In addition, patients with tenacious distance fusion and increased deviation beyond 6 m had an undercorrection rate of 35%, compared with 18% for patients who had neither. 67

Criteria for Surgical Success

Just as there are no consistent guidelines for the timing of surgical intervention, there is no agreement between authors in how to assess the surgical outcome.

There was a time when strabismologists were content to achieve good motor alignment. Success was defined as alignment to within 8 to 10 PD of orthotropia. With this criterion alone, success rates were estimated at 42% to 81% with one surgical procedure. 26, 48, 49, 89, 95, 124 and 82% to 90% after a second surgery. 49, 95

Table 12-2. Sensory Criteria for Success of Surgery in Intermittent Exotropia

<table>
<thead>
<tr>
<th>Authors</th>
<th>Criteria for Success</th>
<th>Success Rate (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hardesty et al 49</td>
<td>Some degree of stereopsis at near (Titmus)</td>
<td>78</td>
</tr>
<tr>
<td>Haase and de Decker 114</td>
<td>40 seconds of arc at near</td>
<td>17</td>
</tr>
<tr>
<td>Pratt-Johnson et al 88</td>
<td>40 seconds of arc at near</td>
<td>41</td>
</tr>
<tr>
<td>Burian and Spivey 48</td>
<td>Stereopsis of 50% to 100% at near</td>
<td>55</td>
</tr>
<tr>
<td>Beneish and Flanders 4</td>
<td>40 to 60 seconds of arc at near</td>
<td>61</td>
</tr>
</tbody>
</table>

Hardesty and associates 49 highlighted the importance of including the presence of some degree of stereopsis when judging surgical success. Using more stringent sensory criteria, the results become more sobering (Table 12–2). 3, 18, 40, 89, 114 Pratt-Johnson and colleagues 40 achieved motor alignment in 81%, but sensory fusion of 40 seconds of arc at near was achieved in only 41% of cases. Similarly, Folk 40 achieved “straight” eyes in 50%, but a functional cure in only a fourth of his patients. It is disconcerting to note the low success rates when near stereocuity is used as a sensory parameter.

Seventy-five percent of patients show an improvement in distance stereocuity with contour circle testing after surgery and 45% by random dot E testing. 84 Preoperative distance stereocuity of 240 seconds of arc improved to 60 seconds of arc after surgery on contour circle testing. 84 However, distance stereocuity is not restored to normal population values, possibly reflecting relatively late intervention. 64, 99

Motor alignment alone is not sufficient to gauge the success of surgery for X(T). Sensory functions should be measured using both distance and near stereocuity tests.

Postoperative Alignment

The desired surgical goal in the first 1 to 2 weeks after surgery is to create a small angle of 5 to 10 PD of esotropia. 98, 100 Postoperative diplopia is used to stimulate the development of fusional vergences and stabilize postoperative alignment. 82, 122 This intentional overcorrection should be avoided in children with an immature visual system. In the immediate postoperative period, base-out prisms will prevent monofixational exotropia. If a patient has some amount of hyperopia, this correction may be given.

In older children and adults who develop X(T) after age 10 years, diplopia is usually present with little or no suppression. In these patients, the surgical goal should be orthotropia on the first postoperative day, not intentional overcorrection. 62, 98 If bothersome diplopia develops, prisms may be used temporarily to allow patients to function. 46, 77 Nonsurgical management of overcorrection should be tried for at least a month rather than reoperating because of the high likelihood of spontaneous resolution. 65

Surgical Procedures

Lateral Rectus Recession. Bilateral LR recessions are preferred for patients with true divergence excess. 49, 98 MR
resection should be avoided because of its effect at near. Patients may have bothersome symptoms from overcorrection at near. Some authors believe that LR resections are more effective in reducing distance deviation than near deviation. For smaller deviations of 14 to 16 PD, large unilateral LR resections of up to 12 mm on the nondominant eye may be effective with minimal abduction deficiency.

**Recess-Resect Procedures.** Patients with basic exodeviation do not do as well as those with true divergence excess after bilateral LR recession. A prospective randomized clinical trial revealed only a 52% success rate in patients with basic X(T) if managed with LR recession compared with 82% if surgery consisted of the recess-resect procedure. This occurs despite the same reduction in the distance-near disparity with either surgery (contradicting proponents of more effect on distance deviation if LR surgery was performed).

The recess-resect procedure involves LR recession and MR resection. Some strabismologists believe that this is probably the ideal procedure for obtaining stable results, at least for some patterns such as basic X(T) and convergence insufficiency. Kushner theorizes that patients with tenacious proximal fusion may be managed equally well with either bilateral LR recession or the recess-resect procedure. Patients without tenacious fusion at near may benefit from the latter procedure because of the tether against abduction caused by the MR resection.

MR resection may produce lid fissure narrowing and overcorrection in the abducted field of the eye undergoing the recess-resect procedure. This slight overcorrection may be advantageous in eliminating suppression because of the esotropia caused in the field of the recessed LR. Overcorrection in left gaze, however, can be especially bothersome because of problems with driving. The amount of MR resection should probably be limited to 4.5 mm to avoid horizontal incomitance, especially in adult patients.

In cases in which one eye is amblyopic, the surgeon often chooses unilateral surgery. Depending on the size of the deviation, a recess-resect procedure is done if surgery on two muscles is required. Large-angle X(T) of more than 50 PD should probably be managed with bilateral LR resections combined with resection of one or both MR muscles. A conjunctival recession enhances the recession effect of an LR weakening procedure and may augment the correction.

Adjustable suture techniques are helpful in cooperative patients.

**Medial Rectus Resection.** Bilateral MR resections are useful for the convergence insufficiency type of X(T) because of the effect of this surgery at near. We prefer doing at least one on an adjustable suture. Overcorrection at distance is expected up to 6 weeks after surgery. Fresnel add-on prisms are used for distance until the esotropia at distance is overcome. Because of the risk of overcorrection at distance, surgery should be reserved for highly motivated patients.

**Common Pitfalls in Management**

**Lateral Incomitance.** If significant lateral incomitance is documented, the usual surgical numbers for LR recession should be reduced to avoid overcorrection. In cases in which the LR is underacting, MR resection strategies have given satisfactory results. When the lateral incomitance is due to a tight MR muscle, MR resections will make it worse. The recommended procedure is recession of the tight MR with enhanced LR recession to compensate for the effect of MR recession. The adjustable suture technique improves the postoperative results.

**Tight LR Syndrome.** This is usually seen in patients with long-standing constant monocular XT. Mildly reduced adduction may be observed in association with pseudo-overaction of both the superior and inferior oblique muscles, because of the leash effect caused by a tight LR. The overaction disappears after LR recession. Recognition of this pattern will suggest LR recession and the avoidance of oblique muscle surgery.

**Concomitant Vertical Deviations.** Concomitant vertical deviations may be documented in as many as 50% of patients with X(T). Small vertical deviations less than 10 PD can be corrected by horizontal transposition of the muscles undergoing recession or resection procedure for X(T). Large vertical deviations should be addressed at the time of X(T) surgery.

**COMPLICATIONS**

**Undercorrection**

Undercorrection after an initial bilateral LR recession is common, with 21% to 38% requiring a second surgical procedure. Several studies have concluded that the age at the time of initial surgery and the amount of the exodeviation were not contributory factors to failure after the initial bilateral LR recession. The following make the initial surgery (usually bilateral LR recession) less likely to be successful:

1. Increasing constancy of the deviation for viewing distant objects
2. Small amounts of coexisting vertical misalignment
3. Failure to identify oblique muscle dysfunction and subsequent A or V patterns
4. Failure to reveal the total quantity of distance X(T) measurements preoperatively
5. Uncorrected hyperopia

Any vertical muscle dysfunction in any gaze field increases the total amount of misalignment. The motility examination should include measurements in vertical upward and vertical downward gaze to reveal any pattern strabismus. When present, surgery to correct the pattern should be addressed when horizontal muscle surgery is performed for the X(T) (see also Chapter 14).

To determine the full amount of deviation with which to base target surgical angles, fusional convergence must be fully relaxed as discussed earlier. If the patient has significant hyperopia, there is an increased tendency for the patient to use some accommodative convergence if corrective glasses are not worn while measurements are being made for distant targets. Patients should be wearing their full "tolerable" hyperopic correction while the prism cover test is being performed for distance and near.

**Clinical Presentation.** After initial bilateral recession of
INTERMITTENT EXOTROPIA • 171

Figure 12-4. Tight lateral rectus in long-standing right exotropia. Note apparent overaction of right inferior oblique and right superior oblique. Both oblique "overactions" manifest with significant abducting component. Forced duction testing is positive for resistance to full adduction. No oblique muscle surgery is indicated.

the LR muscles or recession of one lateral and resection of the antagonist MR, the patient often maintains orthotropia or a small angle X for near but may demonstrate residual small to moderate X(T) while viewing distant targets. This residual distance misalignment may be small at first, but, without additional therapy, tends to increase with the passage of time. The undercorrection noted while viewing distant targets is the most common form of failure after initial surgery for X(T).

Management of Undercorrection

Nonsurgical Management. Use of base-in prisms to neutralize residual deviation at distance has been advocated by Hardesty, who referred to prism neutralization of the distance misalignment as a "fusion priming device." Prisms should be worn for a minimum of 6 months before a resection of the MR muscle is performed. Ultimately, however, most of the patients in his series required secondary surgery.

Minus lens therapy was advocated as a substitute for surgery in the treatment of X(T) in children, but may also be used to treat residual X(T) (less than 12 PD) after surgical treatment. Successful treatment of small undercorrections approaches 50% in some reports.

If there is a greater residual deviation for near than distance (convergence insufficiency), occasionally convergence exercises will help control the residual deviation. However, convergence exercises for basic divergence excess X(T) may perpetuate an abnormal AC/A ratio. Consequently, preoperative convergence exercises for X(T) may make overcorrection for near targets more common after a surgical procedure and should be avoided.

Surgical Management. In the past, it was common to wait 6 months or more after the initial surgery before considering additional surgery, but a recent outcome study showed that relatively few patients respond well to secondary surgery when surgery is performed an average of 2 years after the initial surgery. Continued suppression of one eye with the persistence of an uncorrected distance deviation in these patients seems to be the major cause of the poor response to reoperation. Thus, secondary surgery should be performed earlier (e.g., 8 to 12 weeks after the initial surgery), if there is residual X(T) for distance. This has increased the overall percentage of successful final alignment.

Secondary surgery for residual X(T) becomes problematic if maximum or large bilateral LR recession has been performed as the initial procedure and patients present with residual X(T) for distance with relatively straight eyes for near. The surgeon has a choice of (1) re-recessing the LR to 7.5 to 10 mm from the original insertion or (2) resecting one or both MR muscles. Re-recession of both LR muscles has not been routinely successful. We recommend resection of one or both MR muscles for undercorrection. This procedure may be used for both early and late undercorrection. The following case illustrates successful management of a small overcorrection with one MR resection.

Case 1. A 3-year-old patient who had 30 PD of distance exotropia but was orthotropic at near underwent bilateral LR recession of 5.5 mm. Despite a small esotropia in the early postoperative period, she subsequently developed 15 PD of exotropia for distance and remained orthotropic at near. Patching was unsuccessful. After undergoing a left MR resection of 3.0 mm 3 months after the initial surgery, the patient maintained orthotropia at distance and 4 PD of esotropia at near at the 6-month postoperative visit.

Patients with secondary surgery must, of course, also be watched for overcorrection, and, if the initial esotropia from the resection of the MR does not decrease to zero within 3 weeks, base-out membrane prisms may be prescribed to maintain fusion. The quantity of the prisms can usually be reduced gradually.

Overcorrection

Overcorrection after initial surgical therapy is less common than undercorrection. Even when patients were operated upon for the largest angle of exodeviation, no increase in
overcorrection was observed. In the series by Hardesty and Richard and Parks, overcorrection persisting beyond several months was found in only 6% after bilateral LR recession. More recently, however, in an unpublished consecutive series by one of us (MRI), overcorrections were as common as undercorrections at the 6-month postoperative visit. Rarely, large overcorrection after a slipped or lost LR muscle may occur (Fig. 12–5).

An initial rather large esotropic response during the first postoperative week is not necessarily a poor response to surgery. Long-term success in some series was highest in the group of patients who demonstrated up to 20 PD of esotropia in the first 10 days postoperatively after bilateral recession of the LR muscles. However, a persisting esotropia for near targets beyond 3 postoperative weeks becomes worrisome, and therapy should be initiated to prevent suppression and deterioration of the fusional status.

**Risk Factors.** Lateral gaze incompatance (reduced X(T) on lateral gaze measurements compared with primary gaze) has been identified as a risk factor for overcorrection after bilateral LR recession. It seems prudent to reduce the surgical dosage for bilateral LR recession by 1 mm for patients in whom measurements show 20% less deviation on lateral gazes compared with primary position.

Pratt-Johnson and associates concluded that surgical results were more successful for patients aligned before age 4 years but that risk of overcorrection and a monofixation syndrome result was also greater. In one of the authors' series, success could not be correlated with the age at surgery. Furthermore, the mean age of patients with monofixation syndrome result was, in fact older than the mean age of the group as a whole (5 years, 1 month vs. 4 years, 8 months). Delaying initial surgery until after the age of 4 years did not appear to provide protection against developing the monofixation syndrome.

Adults tolerate overcorrection for X(T) very poorly; and for this reason, deliberate undercorrection of divergent strabismus has been recommended by others. We try to avoid sensory diplopia postoperatively by selecting a surgical dosage determined by measurements made by base-in prisms up to, but not at the point of, subjective diplopia. If a patient reports diplopia with base-in prisms that totally neutralizes X(T) on alternate prism cover test, the base-in prisms that are held over one eye should be reduced to a point at which

the patient does not report any diplopia. The surgeon should then use the smaller amount of prisms as the appropriate surgical target angle.

Although many X(T) patients have distance-near disparity in the amount of deviation, most will respond to surgery well without maintaining this disparity after surgery. However, infrequently patients develop a high AC/A ratio after surgery, manifesting as esotropia at near. If AC/A ratio was determined after prolonged occlusion, these patients can be identified preoperatively, minimizing postoperative “overcorrection” at near.

**Clinical Presentation.** Older patients will complain of double vision for near or distant targets. Closing one eye will be seen as the patient avoids diplopia in this manner. The patient will tilt the head back or drop the chin down to avoid diplopia if an A or V pattern is present. Decreased abduction of the LR may be observed on side gazes. A difference in the activity of the LR muscles suggests a slipped muscle on the weaker side.

A transient esotropia for near targets is commonly found in the first few postoperative weeks. Thereafter, any esotropia for distant targets can be examined more thoroughly by using prism cover test measurements in side gazes and examining versions. Slippage or excessive recession of the LR will be revealed by deficient abduction. Diagnosis and management of the latter is discussed in Chapter 40.

**Management of Overcorrection**

**Nonsurgical Management.** Alternating occlusion for any esotropia persisting beyond 3 weeks postoperatively may reduce or eliminate overcorrection. For patients whose esotropia persists beyond 3 weeks, other nonsurgical measures should also be employed.

Echothiophate iodide (Phospholine Iodide) 0.125% eye drop solution instilled in both eyes daily often dramatically reduces esotropia at near that persists postoperatively, especially if there was a large X(T) for distance and orthotropia for near targets preoperatively (abnormal AC/A ratio). The drops are used daily until sufficient improvement is noted and subsequently tapered to a dose that relieves esotropia. Echothiophate iodide may be very difficult to use in some patients because of headaches and blurred vision induced by miosis.

The use of plus lenses (+2.50 D or +3.00 D) at near in

**Figure 12–5.** Overcorrection after bilateral lateral rectus recession for intermittent exotropia. Patient developed 25 PD of right esotropia, with limitation of abduction of the right eye evident on right lateral gazes. Differential diagnosis should include slipped or lost right lateral rectus muscle. A lost muscle, however, is less likely, because of the relatively good right lateral rectus rotation, albeit reduced.
the form of bifocals will preserve fusion in patients with distance phoria but esotropia for near targets. Eventually, however, surgical treatment with bilateral MR recession should be considered if the esotropia for near does not dissipate after 6 months.

Base-out prisms can also serve to preserve fusional status and can often be reduced gradually. This is especially important in preventing development of permanent monofixational esotropia. If prisms are still necessary after 6 months, surgery to reduce or eliminate the esotropia should be considered.

The following case illustrates successful nonsurgical management of overcorrection.

**Case 2.** A 4-year-old boy with X(T) of 35 PD at distance and orthotropia for near underwent bilateral 6-mm LR recession. Postoperatively, he developed 15 PD of esotropia for distance and 35 PD of esotropia for near. Alternating occlusion and 0.125% Phospholine Iodide were used for 10 days, after which Phospholine Iodide was maintained for 3 months. The esotropia was reduced to 4 PD for both distance and near. On discontinuing the miotics, however, the esotropia increased to 8 PD for distance and 12 PD for near. Base-out membrane prisms were given and tapered over 8 months. At the 6-year postoperative visit, the patient had remained orthotropic for distance and near, with 20 seconds of arc near stereoaucity.

**Surgical Management of Overcorrection.** A small-angle esotropia persisting for distance with X or orthotropia for near targets is probably best left untreated by secondary surgery because this result is usually very stable.

The persistence of a larger angle of esotropia for distance with complaints about diplopia can be surgically treated by advancing the previously recessed LR, especially if there is underaction of the LR on versions.

A more common presentation of an overcorrection, however, is the persistence of esotropia for near targets that has not decreased despite bifocals or base-out prisms. These patients respond rather well to unilateral or bilateral MR recession with or without a posterior fixation suture. The following case illustrates this type of patient:

**Case 3.** A 3-year-old girl who had 25 PD of esotropia at distance and orthotropia at near underwent 4.5 mm recession of both LR muscles. Postoperative deviation drifted to 10 PD esotropia for distance and 12 PD esotropia for near despite good alignment of only 2 to 4 PD for near for 3 weeks after surgery. Base-out prisms were used for 2 years until the esotropia for near increased to 20 PD. At age 6, she received a 4 mm left MR recession with a posterior fixation suture. At the time of her last examination (2 years after her last surgery), the patient was orthotropic for both distance and near and the Randot stereoaucity test indicated 40 seconds of arc.

**Conclusions**

The relatively high rate of overcorrections and undercorrections despite what is considered appropriate intervention may be a reflection of our attempts to control a defective central supranuclear signal with extraocular muscle surgery. The strabismus surgeon must clarify the mechanism for a distance-near disparity. Prolonged monocular occlusion (before testing with +3.00 D lenses) is necessary to distinguish between patients with tenacious proximal fusion (who may be treated with either bilateral LR recession or unilateral recess-resect procedure) and those without. In the latter, the AC/A ratio needs to be investigated. Minus lens therapy may be attempted in patients with high AC/A ratio because surgery may result in overcorrection at near. If surgery is performed, bifocal correction may be required to manage residual esotropia at near. Patients with basic X(T) (and assumed to have normal AC/A ratio) do better with unilateral recess-resect procedure.

The possibility of failure of the initial surgical procedure in the treatment of X(T) should be anticipated in some patients and should be discussed thoroughly with the patients or parents in the preoperative period. Any informed surgical consent should include a signed acknowledgment that more than one surgical procedure may be necessary to align the eyes and that patching, miotic drops, bifocals, or prism glasses may be necessary to supplement the result after the initial surgery.

The operating surgeon should also recognize that a monofixation syndrome result may be found in 5% to 10% of surgically treated cases. These patients, however, usually have a very satisfactory motor alignment, and parents are usually happy with the result despite the ophthalmologist’s finding of mildly reduced stereocuity.

With secondary surgery and other techniques described in this chapter, the vast majority of patients with X(T) can have their eyes successfully aligned and binocular function preserved or even improved.

**REFERENCES**


Infantile (or congenital) exotropia is a rare motility disorder characterized by divergent strabismus that is apparent in infancy and persists beyond age 6 months. It is generally agreed that the misalignment must begin before age 6 months to be considered infantile strabismus. 14, 79, 103, 144 Infantile exotropia may be a primary or a secondary disorder (Table 13–1).

Table 13–1. Classification of Infantile Exotropia

<table>
<thead>
<tr>
<th>Primary Infantile Exotropia</th>
</tr>
</thead>
<tbody>
<tr>
<td>Constant (congenital) exotropia</td>
</tr>
<tr>
<td>Idiopathic</td>
</tr>
<tr>
<td>Hereditary</td>
</tr>
<tr>
<td>Early-onset intermittent exotropia</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Secondary Infantile Exotropia</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ocular disorders</td>
</tr>
<tr>
<td>Diseases causing vision loss (e.g., cataract, retinoblastoma, ptosis)</td>
</tr>
<tr>
<td>Miscellaneous disorders (e.g., albinism, nystagmus)</td>
</tr>
<tr>
<td>Other strabismus conditions</td>
</tr>
<tr>
<td>Innervational (e.g., third nerve palsy, Duane syndrome)</td>
</tr>
<tr>
<td>Mechanical (e.g., Brown syndrome, strabismus fixus, orbit tumor)</td>
</tr>
<tr>
<td>Systemic disorders</td>
</tr>
<tr>
<td>Neurologic disease (e.g., cerebral palsy, hydrocephalus)</td>
</tr>
<tr>
<td>Nonspecific developmental delay (e.g., prematurity)</td>
</tr>
<tr>
<td>Craniofacial syndromes</td>
</tr>
<tr>
<td>Genetic syndromes (e.g., Prader-Willi syndrome)</td>
</tr>
</tbody>
</table>

Primary infantile exotropia is seen in patients who are otherwise healthy and have no evidence of systemic or ocular disease (Fig. 13–1). 14, 52, 90, 92, 119, 147 This disorder is almost always sporadic, 13, 14 although a familial form has been reported. 16 Intermittent exotropia, the most common form of childhood exodeviation, may rarely occur in the first few months of life 14, 56, 133 (see Chapter 12).

Secondary infantile exotropia is seen in patients who have ocular or systemic abnormalities. An early exotropia may develop along with any condition that causes congenital loss of vision, such as cataracts, retinoblastoma, or optic nerve anomalies. 35, 79, 90 It may be associated with ptosis 2, 3 and can accompany other ocular disorders such as albinism. 32 Secondary infantile exotropia may be a feature of various strabismus syndromes, including innervational disorders such as congenital third nerve palsy and Duane syndrome, as well as mechanical disorders such as Brown syndrome, strabismus fixus, 5, 10, 90 or congenital orbital tumors. Many of these entities are discussed elsewhere in this book.

Infantile exotropia may be associated with neurologic diseases such as cerebral palsy and hydrocephalus and also with general developmental delay. 16, 79, 112 This exotropia also occurs in various forms of craniofacial syndrome 16, 16, 21, 92, 93 and has been described in patients with chromosomal anomalies and other systemic syndromes (Fig. 13–2) 14, 23, 50, 85, 114, 120 (see also Chapter 30).

Incidence

In contrast to infantile esotropia, the incidence of primary infantile exotropia is very low. Most reports in the literature include very small numbers of cases accumulated over a
number of years. Infantile exotropia occurs in 1 in 30,000 births in the general population, compared with 0.5% to 1.0% in large population studies of infantile esotropia. By extrapolation, cases of infantile strabismus include 150 to 300 infants with esotropia for every case of exotropia. This estimate may, in fact, be too high, as suggested by studies of Archer and colleagues. Follow-up of more than 3000 infants first examined as neonates showed that 3 developed esotropia, whereas 2 developed exotropia that persisted beyond 6 months of age.

**Causes of Infantile Exotropia**

**GENETIC CONSIDERATIONS**

The cause of primary infantile exotropia is unknown. Its low prevalence and sporadic occurrence preclude detailed epidemiologic and genetic studies. Brodsky and Fritz document the occurrence of exodeviation in three consecutive generations of one family: two children in the third generation and their maternal grandfather had infantile exotropia and their mother had an exophoria. These findings suggest autosomal dominant inheritance with variable penetrance or else multiple genetic and intrauterine factors causing the strabismus. Two sources suggest that infantile exotropia may be more common in persons of Asian and African origin than in those of Caucasian lineage.

**NEUROPHYSIOLOGIC STUDIES**

Examination of neonates revealed that exodeviations are very common in the first few days of life: they occur in one
third of otherwise healthy children, whereas a smaller number have esodeviation.\textsuperscript{97} This suggests that a large number of normal infants have an anomalous binocular sensory experience during the first few weeks of life. During normal visual maturation, the eyes straighten in the first 2 to 4 months of life as the vergence system matures.\textsuperscript{4, 8} Primary infantile exotropia may, therefore, reflect deficient development of the convergence system during this vulnerable early period.

Studies in primates show that monococular defocusing or total deprivation from birth yields a high rate of exotropia in the first weeks of life. This suggests that early pattern vision is necessary for the development of normal ocular alignment and normal vergence reflexes.\textsuperscript{109, 135} This is borne out by findings in infants with congenital ptosis who develop early deprivation amblyopia, because their predominant associated misalignment is exotropia.\textsuperscript{2, 3} Disruption of binocular vision shortly after birth is a potent factor that may disrupt the development of normal vergence reflexes and cause strabismus.

These findings do not, however, explain how exotropia, in particular, develops when binocular vision is disrupted. There is a difference in the efficiency of monococular temporal and nasal smooth pursuit that is characteristic of infantile strabismus and is related to the early disruption of normal binocular processing in the visual cortex.\textsuperscript{98, 136} Targets followed monocularly from the temporal to the nasal field are tracked more efficiently than the reverse. Data from primate studies suggest that this deficit is associated with a deficit in cortical control of the vergence reflex.\textsuperscript{136} This temporal-nasal disparity occurs whether the animal has early-onset exotropia or esotropia. It is also demonstrable in humans with infantile strabismus, whether esotropia or exotropia.

Despite the common pursuit defects in these two states, esotropia is much more common than exotropia in humans with infantile strabismus. This may result from the greater efficiency of the nasally directed pursuit system. If this is the case, there must be some other factor that predisposes a small group of patients to develop exotropia rather than esotropia. There may be an innate asymmetry in muscle structure between the lateral rectus (LR) and medial rectus (MR) in these patients. Rosenbaum and colleagues\textsuperscript{117} have shown differences in the length-tension relationship between the LR and MR in intermittent exotropia compared with that in esotropia. Another study documented an increased diameter of the LR compared with that of the MR in a case of infantile exotropia, and the reverse in cases of esotropia.\textsuperscript{53} Unfortunately, these studies do not prove whether the anatomic differences cause the exotropia or are a result of the misalignment. Congenitally abnormal insertions of the MR can lead to infantile exotropia that simulates congenital third nerve palsy.\textsuperscript{99} for intermittent exotropia to be manifest by age 6 months, but it has a course and prognosis more consistent with the typical forms of intermittent exotropia seen in older children. It should probably not be grouped with primary infantile strabismus.\textsuperscript{112, 133}

The deviations in the primary form of infantile exotropia are almost always over 30 PD and may be as large as 90 PD.\textsuperscript{13, 14} Amblyopia is seen in a minority of cases, ranging from zero to 25\% in reported series; it is usually due to strabismus rather than anisometropia. The amblyopia associated with this form of exotropia seems to be as responsive to traditional therapy as that combined with infantile esotropia.\textsuperscript{13, 14}

Both dissociated vertical deviation (DVD) and inferior oblique overaction may be associated with infantile exotropia, just as they are with infantile esotropia. The combined incidence of these two anomalies in infantile exotropia appears to be lower than in infantile esotropia, in the range of 50\% overall.\textsuperscript{13, 14, 48, 52, 92, 119, 147} Superior oblique overaction has also been reported.\textsuperscript{13, 14, 147} Unlike infantile esotropia, latent nystagmus is uncommon,\textsuperscript{14} although it has been noted by some observers.\textsuperscript{90}

Pattern strabismus may occur: V patterns are much more frequent than A patterns.\textsuperscript{13, 14, 16} Amblyopia is seen in a minority of cases, with long-standing large exotropia. These patients may exhibit pseudo-overaction of the four oblique muscles (see section entitled Sensory Esotropia).

\begin{center}
\textbf{Clinical Characteristics}
\end{center}

Primary infantile exotropia is defined by the onset of exotropia before age 6 months and confirmation by an ophthalmologist before age 1 year (see Fig. 13–1). Because the angle of strabismus is usually large and constant, it is often reliably documented by a parent or guardian or by the child’s primary physician in the first few weeks of life. It is possible to determine whether the infant alternates fixation or prefers one eye, whether the deviation is constant or intermittent, and whether movement is obviously limited in one or both eyes, either in the horizontal or the vertical plane. The parent or guardian should be questioned about whether the infant makes eye contact or follows targets at near. Any history of local ocular disease or trauma should be noted in addition to any family history of ocular problems or strabismus.

\begin{center}
\textbf{Diagnosis}
\end{center}

\begin{center}
\textbf{HISTORY}
\end{center}

The physician must take a careful history to determine the time of onset of the exotropia. To diagnose infantile strabismus, there must be evidence of an onset before age 6 months. Family photographs are valuable for confirmation. Notations from the family physician or pediatrician are also helpful, especially if measures such as the Hirschberg (corneal light reflex) test or cover test were taken to confirm exotropia.

Ocular, genetic, and general systemic conditions should be considered (e.g., cerebral palsy, craniofacial disorder). One should inquire about the prenatal and birth history, as well as developmental milestones. The examiner must determine whether the infant alternates fixation or prefers one eye, whether the deviation is constant or intermittent, and whether movement is obviously limited in one or both eyes, either in the horizontal or the vertical plane. The parent or guardian should be questioned about whether the infant makes eye contact or follows targets at near. Any history of local ocular disease or trauma should be noted in addition to any family history of ocular problems or strabismus.
EXAMINATION

Systemic Evaluation

At the outset, the infant’s general behavior should be noted. Features to take note of include alertness, eye contact, and muscle tone (is it appropriate to the child’s age?). Obvious external deformities should be sought, including facial dysmorphism, abnormal skin color, limb anomalies, and abnormal head size or shape. Abnormalities in any of these areas are grounds for pediatric, neurologic, or genetic evaluation.

Ocular Assessment

An ophthalmologic examination is performed to determine whether the exotropia is primary or associated with other ocular or strabismus conditions noted earlier (see also section entitled Sensory Exotropia). Note should be taken of any abnormal head posture, because it may suggest an incomitant deviation or ocular fixation preference. Depending on the child’s age, vision or fixation should be determined. The red reflexes and pupillary reactions to light are observed. Any orbital or eyelid abnormalities such as ptosis or proptosis should be ruled out, as well as any external disease or congenital anomaly of the anterior segment. Portable slit-lamp evaluation may be required if there is an abnormality of the anterior segment.

The motility examination should include assessment of the positions of the light reflexes in both eyes by looking at the angles kappa. This distinguishes true exotropia from pseudoexotropia with positive angles kappa (Fig. 13–3). Ideally, exotropia should be measured at distance with the alternate prism cover test in primary and secondary positions (up, down, left, and right gaze) to detect incomitance at distance. The test is repeated in the primary position at near. If the child is younger than age 1 year, measurements may be possible only at near using the Krimsky test; measurement of the angle in secondary positions at near should be attempted as well.

If the Krimsky test is used, the angle kappa of the preferred eye must be noted, because the angle of deviation is correctly measured when the light reflex in the nonpreferred eye is symmetric with the reflex in the pupil of the preferred eye. For angles over 50 PD, the measurement should be done with prisms of approximately equal strength, placed before each eye, to minimize the inaccuracies that occur when prisms of differing sizes are used. Stacking prisms together to measure large deviations leads to inaccurate results (see Chapter 5). Careful scrutiny is needed to detect coincident vertical deviations, including DVD and oblique muscle overaction.

Ocular movements are assessed in the cardinal positions of gaze to observe any overaction or underaction of the ocular muscles and any other anomalies such as globe retraction, pupillary changes, and nystagmus. If the child is old enough, sensory testing should be done to detect suppression or anomalous retinal correspondence (ARC).

Posterior segment examination and refraction are important to rule out any media opacity, significant refractive error, and posterior segment disease or tumor. If the child has any systemic condition known to have ocular associations, such as hydrocephalus, signs of such associations must be sought.

It is worthwhile to briefly check alignment in close relatives to see if any other family members have strabismus. The presence of strabismus in first-degree relatives suggests a familial tendency.

Optokinetic testing can show whether there is a difference in the nasal and the temporal smooth-pursuit responses, which is highly suggestive of infantile strabismus. This can be done during the routine examination with an optokinetic drum or formally in an eye movement laboratory using more sensitive devices. Any ocular or orbital disease that is detected may require further imaging studies. Examination under sedation may be needed to complete the assessment. Electrophysiologic studies are considered for infants with nystagmus and infantile exotropia if subnormal vision is suspected.

DIFFERENTIAL DIAGNOSIS

A positive angle kappa (see Fig. 13–3) can cause pseudoexotropia when the eyes are in fact straight. Confirmation that the eyes are normally aligned with foveal fixation is provided by a lack of movement on cover testing and good vision in both eyes, as well as visuscopic analysis showing the fovea to be the retinal locus used for the primary line of fixation in each eye.

An ectopic macula that is dragged temporally by diseases such as retinopathy of prematurity or familial exudative vitreoretinopathy may force the patient to fix with the eye in an abducted position, making the eyes appear to be exotropic. Finally, early-onset intermittent exotropia is occasionally diagnosed in a child younger than 6 months of age. This type of exotropia does not exhibit the typical nasal-temporal smooth-pursuit asymmetry seen in primary infantile exotropia. Sensory testing shows a high degree of binocularity and normal retinal correspondence (NRC) when the eyes are straight, findings not typically seen in infantile exotropia.
Treatment

NONSURGICAL MANAGEMENT

Most patients with primary infantile exotropia have stable alignment from the time of diagnosis. The amblyopia seen in this condition almost always responds to conventional therapy. Patching and correcting refractive errors can stabilize the deviations but in general do not lead to resolution of the strabismus. Two patients reportedly gained an improved angle of deviation after patching of the dominant eye, but they may represent cases of early-onset intermittent exotropia rather than true primary infantile exotropia.

Injection of botulinum toxin into the ocular muscles was introduced in 1978 as an alternative to surgery for treating strabismus in adults. In recent years this measure has been used to treat infantile strabismus, mainly esotropia. There are no large studies on the treatment of infantile exotropia because of its rarity. Botulinum injection of one LR muscle in young children with constant exotropia has led to successful alignment in fewer than 50% of cases. Because most infantile exotropia cases are large, often exceeding 40 PD, it is unlikely that botulinum toxin injections of one LR muscle will match the results achieved with surgery. As in infantile esotropia, simultaneous bilateral injections of both LR muscles may lead to improved success rates in the long term; the results of this approach are yet to be reported.

SURGICAL MANAGEMENT

Timing

Surgery is required in almost all cases of infantile exotropia to align the eyes within the first few months of life and maximize the chance for binocular single vision to develop and persist. For healthy infants with primary infantile exotropia, published reports suggest that the optimal motor and sensory results are obtained in children whose eyes are aligned successfully before age 24 months. This is consistent with the experience in patients having infantile esotropia. Once the diagnosis is made, the patient should be followed for a few weeks to be sure that the angle is stable and to treat any coincident amblyopia. If the angle does not change over two or three consecutive visits, surgery should be performed with minimal delay.

Procedures

The aim of surgery is to realign the eyes to a stable angle of less than 8 PD to allow the development of binocular responses, both motor vergences and sensory fusion. These goals are similar to recommendations for infantile esotropia. Long-term success is likeliest in patients who achieve small-angle esotropia or whose eyes are straight the first week after surgery and remain straight after 8 weeks. Despite concern that small-angle esotropia after surgery for infantile exotropia may create a risk of amblyopia with monofixation syndrome, this outcome has not been common in children whose alignment was slightly overcorrected immediately after surgery. Because of the high rate of undercorrections reported for this condition, it is probably best to aim for slight overcorrection in the immediate postoperative period, as is commonly recommended for intermittent exotropia in older children.

By far the most common initial procedure has been bilateral symmetric resections of the LR muscles. The amount of recession has varied according to the measured angles, ranging from 5.0 mm for deviations of 30 PD up to 12.0 mm for deviations greater than 70 PD. Alignment can also be achieved by monocular recession and resect procedures for exotropia less than 40 PD. Large angles of exotropia (over 40 PD) can also be safely treated by three- or four-muscle surgery, recessing both LR muscles, and resecting one or both MR muscles. Inclusion of an MR muscle resection should be considered if the deviation at near matches or exceeds that at distance.

Surgery for DVD and oblique overactions may be planned at the same time as the horizontal repair. However, in most cases of infantile strabismus (both esotropia and exotropia), if the horizontal repair is done at an early age, the vertical strabismus is often not evident or is not within the surgical range. The horizontal realignment is usually done first, followed later by any correction of vertical misalignment if it develops or worsens after the first surgery. Options for correcting DVD and oblique overactions are dealt with elsewhere in this book (see Chapter 17).

Outcomes

The reported long-term alignment results after surgery for infantile exotropia have been disappointing. Reoperations are needed by up to 50% of patients. Most reoperations are done for undercorrection, but patients also have had second or third operations to eliminate a coexisting DVD or oblique muscle overaction.

If LR recessions were done initially, repeat surgery for residual exotropia should include a resection of the MR muscle to prevent recurrent exotropia. It is rare to see chronic overcorrections of more than 10 PD at distance or at near after primary repairs.

In healthy children with infantile exotropia, successful surgical alignment before age 2 years can lead to the recovery of peripheral fusion in up to 50% of patients. Gross stereopsis may be recorded in these patients. It appears, however, that bifoveal fixation, reflected by central fusion and high levels of stereopsis, is not regained, even with successful early realignment. Monofixation syndrome may be the optimal result in these cases (see section on decompensated monofixational exotropia). Realigning patients for the first time after age 2 years can lead to long-term stable alignments but rarely is accompanied by sensory fusion.

COMPPLICATIONS

Primary infantile exotropia, if left untreated, can lead to amblyopia, even though the risk appears to be less than for infantile esotropia. Failure to diagnose amblyopia during the crucial period of visual development in the first few years of life can lead to permanently impaired vision in the affected eye. Suppression, although plastic early on, appears to be deeply set within the first few months of life. Realigning surgery should be carried out within the first few
months of life so that the initial surgery and any necessary reoperation can straighten the eyes before the age of 2 years. Research on animals has documented the effects of artificially induced strabismus (including exotropia) in infancy on the developing visual system. Cats reared with surgically induced exotropia from shortly after birth exhibit deteriorating spatial frequency and contrast sensitivity at the level of the cortex, especially under photopic conditions. In primates, artificially induced exotropia reduces sensitivity to flicker responses, especially at high temporal frequencies. It also leads to a loss of contrast sensitivity that is less severe than that in animals reared with infantile esotropia but worse than that experienced by nonstrabismic animals reared with monocular cycloplegia to degrade the image in one eye.

Finally, asymmetry of the nasal versus temporal smooth-pursuit system characterizes patients with infantile exotropia, just as it does those with infantile esotropia. Although long-term alternating occlusion reportedly reverses this deficit in infants with esotropia, it is not known whether this holds true for infants having exotropia. For this reason it may be prudent to consider alternating occlusion, at least for a few hours per day, for infants with infantile exotropia until surgery can be performed.

Other complications include the development of muscle contractures, as may occur in any form of long-standing strabismus. Contractures are especially common in infantile exotropia because of the large angles of the strabismus. They may lead secondarily to a tight LR syndrome with X patterns and pseudo-over-attachments of the oblique muscles.

**DECOMPENSATED MONOFIXATIONAL EXOTROPIA**

**Definitions**

The term monofixational exotropia refers to a divergent strabismus characterized by an underlying defect in the ability to fix with both foveas. It is a subcategory of the disease complex termed monofixation syndrome, in which a combination of characteristic sensory and motor features is detected under binocular conditions. The sensory findings include monofoveal fixation with a scotoma in the macula of the nonfixing eye, peripheral fusion, preserved gross stereopsis, and, frequently, amblyopia of the nonfixing eye. The motor features are a small manifest angle of misalignment (a tropia under 8 PD), a superimposed latent component (heterophoria), and preserved fusional divergence and convergence amplitudes. A monofixation syndrome that demonstrates no shift on cover testing may also exist; these cases have been termed monofixational phorias. Despite the slightly different criteria that various authors have used to define these conditions, there are consistent features: suppression of the fovea of the nonfixing eye under binocular conditions, the presence of binocular vision involving parafoveal and peripheral retinal areas, and the frequent presence of amblyopia.

The monofixation syndrome is subdivided into primary and secondary forms. If the syndrome is associated with anisometropia or a macular lesion, or occurs after surgery for congenital strabismus, it is termed a secondary monofixation syndrome. In the absence of any of these conditions it is called primary monofixation syndrome.

A heterophoria, or latent component, is frequently observed along with the monofixation syndrome. If fusion control is disrupted or lost, the latent deviation may become the predominant clinical feature. This condition is termed a decompensated monofixational tropia; if the deviation is an exodeviation, it is designated a decompensated monofixational exotropia.

**Incidence**

Although there are numerous studies dealing with monofixation syndrome, the overall incidence of this condition and the rate of deterioration remain uncertain. Part of the difficulty arises because young children may not be diagnosed lacking confirmation by sensory tests, which may be too advanced for young children to perform. In a review of a large clinic population in Europe, Lang found microtropia in 2.8% of 33,600 patients. In North America, the rate of monofixation syndrome may be as high as 1%. Among patients with this diagnosis, exotropia is much less common than esotropia and vertical monofixation syndrome is even rarer. The reported frequency of exotropia among diagnosed cases of monofixation syndrome ranges from 4% to 40% and averages 15% to 20%. Several authors believe that these estimates may be too low, especially because many cases of intermittent exotropia may in fact be monofixational exotropia that goes undetected before surgical correction.

Secondary forms of monofixation syndrome, both exotropia and esotropia, occur more frequently than the primary forms. Galloway-Smith and colleagues found that 11% of their cases of monofixational exotropia were primary, the rest being secondary to optical or surgical treatment.

It is difficult to determine the rate of decompensation in monofixational exotropia. Because fusional vergences are intact, monofixation remains stable in a majority of cases. The reported incidence of deteriorating alignment in monofixational exotropia is less than 25% over many years. Many cases of monofixational exotropia, however, are diagnosed only after surgical correction of intermittent exotropia, suggesting that some cases of intermittent exotropia may...
represent undiagnosed decompensated monofixation syndrome.\textsuperscript{12, 38}

**Causes and Physiologic Factors**

**PRIMARY MONOFIXATION SYNDROME**

Patients with primary monofixation syndrome have an inherent defect that precludes central fusion even if the eyes are straight.\textsuperscript{100, 101} This defect may be hereditary in some patients.\textsuperscript{81, 91, 123} It has been proposed that there may be a small error in the fovea-to-fovea correspondence in both eyes, sometimes termed fixation or fusion disparity, that would normally be handled by the sensory flexibility of Panum’s fusional space. This error may be convergent or divergent in orientation and is defined by the degree to which the visual axes miss the intersection of the fixation point under binocular conditions.\textsuperscript{69} If the amount of the error exceeds the extent of Panum’s fusional space at the fovea, diplopia or retinal rivalry results. These conditions can be accommodated by an expansion of Panum’s area or, alternatively, by suppression in the macula of the eye whose fovea is slightly off axis. Persistence of this state could lead to continuing dominance of one eye and robust suppression in the fovea of the fellow eye that may not resolve spontaneously.\textsuperscript{24, 100, 101}

This process may operate in the esotropic form of the syndrome, but much less often in the exotropic form. This presumably is due to differences in mechanisms of suppression between esodeviations and exodeviations resulting from the differing sensitivities of the nasal and temporal retinas.\textsuperscript{61, 96, 145} Because the temporal retina is physiologically inferior to the nasal retina, exodeviations exhibit an “all-or-none” suppression pattern as control wanes, leading from exophoria to intermittent exotropia and on to constant exotropia. Esodeviations, in contrast, may assume intermediate states with partial or regional sensory adaptations in the nasal hemiretina.\textsuperscript{61, 62} These findings suggest that monofixational exotropia develops when divergent foveal disparity is combined with a rare alteration in the normal nasal-versus-temporal retinal sensitivity. Activation of the suppression mechanism may produce the unusual result of a facultative scotoma in the temporal macula.

Causes of decompensation in the exotropia form include stress factors such as family crisis, acute illness, and chronic fatigue (similar to esotropia)\textsuperscript{81, 84} that lead to loss of the normally stable sensory situation. This form may also be affected by factors that affect vergence control, such as alteration in the accommodative convergence/accommodation ratio.\textsuperscript{24}

There may be a greater chance of the exotropia form decompensating than the esotropia form. Analysis shows that the size of the heterophoria is nonlinearly related to the amount of fixation disparity for the divergent form. The relationship is much more proportional for the convergent form, especially for targets at distance. This relationship is thought to reflect differences in the mechanisms of foveal suppression seen in esodeviations and exodeviations. In esodeviations there are various possible intermediate patterns of suppression and retinal correspondence, depending on the size and level of control of the deviation. Exodeviations do not exhibit such flexibility in sensorial adaptations; instead, there is more of an “all-or-none” suppression adaptation.\textsuperscript{24, 59, 61, 62, 70, 108} Because the suppression mechanism may be less flexible, the strongest suppression adaptation may tend to take over under stresses that draw out the latent component. Moreover, any mechanical factors that favor the LR over the MR muscles can lead to decompensation.

**SECONDARY MONOFIXATION SYNDROME**

Most cases of monofixation syndrome are secondary forms that are seen after the correction of strabismus in childhood or are associated with anisometropia. However, reports of the exotropia form suggest that anisometropia is a much less important factor than in the esotropia form; most secondary cases result from surgery for constant or intermittent exotropia.\textsuperscript{38, 80}

Monofixation syndrome is described as an outcome of surgical treatment for infantile exotropia, intermittent exotropia, and constant exotropia. In any of these situations, a monofixation syndrome is probably the result of a preexisting deficit in bifoveal fusion.\textsuperscript{12, 24, 38, 101}

Anisometropia is a strong obstacle to bifoveal fusion and may initiate a chain of events that include ocular dominance, loss of retinal rivalry, and suppression of the disadvantaged fovea.\textsuperscript{27, 49, 61} If this process develops early in life, a monofixation syndrome may develop despite correction of the difference in refractive errors. Spherical refractive-error differences as small as 1.0 D may produce the syndrome.\textsuperscript{38, 128}

A destructive unilateral macular lesion that causes an organic scotoma may lead to findings matching those seen with any of the preceding causes. In both adults and children, unioocular loss of foveal vision often leads to a divergent sensory strabismus with the severe facultative scotoma that typically develops in cases of exotropia. If the macular lesion is small, the rarer alternative of monofixational exotropia may develop.

In any of these scenarios there is a small chance of decompensation because of an abnormal binocular state associated with the underlying disorder. Recurrent strabismus after surgery is not uncommon, and it may be more likely when the initial surgery leads to monofixation rather than bifoveal fixation. Any stimulus that disrupts the vergence reflex may lead to a loss of control and allow a heterophoria to manifest as an intermittent deviation. Untreated anisometropia leaves a defocused image on one retina and, in time, allows divergence forces to destabilize the alignment, even with a previously stable monofixation phoria.\textsuperscript{65}

**Clinical Characteristics**

**SYMPTOMS**

It is unusual for patients with stable monofixation syndrome to complain of any symptoms. The combination of motor fusion, with its vergence amplitudes and sensory fusion comprising a facultative suppression scotoma of the nonfixing fovea and peripheral fusional reflexes, eliminates any diplopic symptoms, even in the presence of a small angle of deviation.
In contrast to patients with intermittent exotropia or uncomplicated exophorias, patients with monofixation rarely shut one eye in sunlight. Nevertheless, some patients with monofixation exotropia do complain of asthenopic symptoms, which are rare in cases of esotropia. Asthenopia has been reported in as many as 40% of cases. This contrasts to a frequency of up to 80% in patients with pure exophorias, even though the latent deviations are smaller in pure exophoria than in monofixational exotropia.

When exotropia decompensates, patients lose the protective monofixation adaptation and may begin to close one eye in sunlight. They may also complain of diplopia, because their retinal suppression mechanism may not be sufficiently strong. Over time, however, they may develop the suppression pattern of the typical patient with intermittent exotropia to avoid diplopia.

**MOTOR ALIGNMENT**

Monofixation syndrome, whether it is esotropia or exotropia, may exhibit either no shift or a small tropia of less than 8 PD in the nonfixing eye when the fixing eye is covered. Up to one third of patients show no movement on cover testing—a finding associated more frequently with anisometropia than with any of the other causes. A lack of shift on cover testing may be noted in cases in which eccentric fixation has developed with a small angle of strabismus. It is also possible that a small tropia is too small to detect clinically on cover testing, leading to the erroneous conclusion that there is no tropic component. Ocular shifts with angles of less than 2 PD may be difficult to discern, even by experienced observers.

Characteristics of all subtypes of monofixation syndrome is the frequent presence of a superimposed heterophoria, noted in at least 40% of cases. This phoria may coexist with tropia on cover testing. On average, the exophoria component in patients with monofixational exotropia tends to be twice as large as the exophoria in bifoveal patients. The total deviation rarely exceeds 25 PD. The most common clinical finding is intermittent exotropia, but patients may present with an exophoria or, if the deviation becomes more frequent over a long period of time, with a constant exotropia. There may also be a difference in the type of exodeviation at distance and at near, as is commonly found in bifoveal patients with intermittent exotropia.

The fusional vergences in cases of monofixation syndrome are usually close to normal. This differentiates the syndrome from cases of constant small-angle exotropia in which the amplitudes are very low. Vergences are preserved because of intact peripheral fusion and are not compromised by the loss of bifoveal fixation.

**SENSORY CHARACTERISTICS**

**Sensory Fusion**

When the exophoria decompensates in a monofixational exotropia, the suppression pattern resembles that of intermittent exotropia. This typically includes a temporal suppression scotoma extending from the deviating fovea to the diplopia point and may include ARC peripheral to the fovea of the deviating eye. The patient may exhibit differing motor and sensory findings at distance and near. For example, if the patient can maintain alignment at near, the sensory pattern may be characteristic of a monofixation syndrome, with reduced stereopsis and a unilateral macular scotoma.

There is controversy about whether the type of sensory correspondence present in the monofixation state is NRC or ARC. This uncertainty has arisen because of the varying diagnostic criteria used by different authors, as well as the particular sensory tests used. Some authors have found that ARC is a primary feature of the syndrome, especially when a coexisting tropia is present. Others suggest that NRC is possible if the deviation is not measurable or is extremely small—so small that Panum’s area is elastic enough to accommodate it, even in the presence of a disparity in the foveal directions. Whether the results of sensory testing indicate NRC or ARC depends on the size of the targets used, how closely the stimuli resemble real-life visual conditions, the patient’s age and level of understanding, and the dissociating ability of the test apparatus. In addition to sensory adaptations, there may be eccentric fixation by the eye with a foveal scotoma when it is forced to fixate. This condition is confirmed by using a visuscope to detect the parafoveal area used for fixation.

**Stereopsis**

Stereopsis is almost always subnormal in monofixation syndrome. It is detectable in at least two thirds of cases of monofixational exotropia. A lack of high degrees of stereovision is viewed as a diagnostic feature of the disorder. It has been used by many authors to distinguish patients who fix bifoveally, such as the typical patient with intermittent exotropia, from those with monofixation syndrome who lose the function of one fovea under binocular conditions. This is predicated on the assumption that the finest degrees of stereovision, 40 seconds of arc or better, are only possible with bifoveal fixation. Stereocuities are usually higher for the primary than for the secondary forms of monofixational exotropia, as is reported for esotropia.

The reliability of stereopsis testing has been questioned because of evidence that stereopsis of 40 seconds of arc may occur without bifoveal fixation and that poor stereocuity can be found in otherwise normal patients with bifoveal fixation. Stereocuity tests that suggest subnormal binocular vision therefore must be interpreted in the context of other sensory tests confirming the absence of bifoveal fixation.

**AMBLYOPIA**

Amblyopia, defined as a difference of at least two lines of linear Snellen acuity, frequently accompanies monofixation syndrome. The incidence in the exotropia form appears to be less than in the esotropia form. Figures for amblyopia in monofixational exotropia range from 28% to 65%, whereas those for esotropia range from 35% to more than 90%. Parks found that amblyopia risk depends on the underlying cause: postsurgical forms have the lowest rate, anisometropia has the highest rate, and primary monofixation syndromes have an intermediate rate.
Diagnosis

**MOTOR TESTS**

The cover test must be done carefully to detect a small tropia under binocular conditions. If one is found, the angle should be measured by a simultaneous prism cover test to freeze the binocular alignment (Fig. 13–4). This angle should be less than 8 PD. The total deviation may be measured by an alternating prism cover test to uncover the superimposed latent deviation. In many cases the phoric component may amount to several prism diopters. Measurements of convergence and divergence amplitudes with prisms or the major amblyoscope should confirm normal values.

In the presence of decompensated monofixational exotropia, the total deviation may already be manifest and can be measured with alternate prism cover test. Only if the patient has intervals of minimal or normal alignment can the presence of a monofixational tropia be detected. This may be possible in patients with decompensated tropias who still have control of alignment at near, similar to patients with typical intermittent exotropia and a divergence excess pattern.

Measurements should be done in secondary positions of gaze to detect any lateral incomitance or pattern strabismus. The extraocular movements are examined for evidence of any muscle underactions or overactions.

**SENSORY TESTS**

To diagnose monofixation syndrome, the physician must show that the patient is not able to fix with both foveas. This can be demonstrated by a number of sensory tests, including the Worth four-dot, polarized four-dot, and Bagnolini striated lenses tests and the major amblyoscope. When done correctly with appropriate-sized targets, these tests can show that the patient functions with peripheral sensory fusion but fixes monocularly when fusible targets are projected onto the macula of both eyes.

A macular scotoma that is present under binocular conditions is a more convincing clue to monofixation. It may be detected with the 4-PD base-out prism test, the vectograph test, or binocular perimetry.

Stereopsis testing may be done using any of the available tests, including the Wirt, Titmus, and Randot. In monofixation syndromes, stereacuity is typically reduced. Work by Rosenbaum and Stathacopoulos has shown that the distance stereotest may be a useful gauge of the degree of control of intermittent exotropia. This test may be adapted to diagnose the presence of a monofixation syndrome before surgical correction: the examiner can offset the deviation with prisms and measure distance stereacuity. Reduced distance stereacuity despite prism neutralization suggests monofixation. This procedure may be more sensitive than testing at near with traditional tests or with the major amblyoscope.

![Figure 13-4. Simultaneous prism cover test to measure small-angle tropia of the left eye. A, The cover and prism are ready to be moved into position. B, The cover is brought in front of the fixing right eye at the same time as the prism is placed in front of the deviated left eye.](image)
Differential Diagnosis

Several conditions must be differentiated from monofixational exotropia, especially once the strabismus has decompensated into a larger deviation. The most common are the typical or pure exophorias and intermittent exotropias. In both disorders there is clear evidence of bifoveal fixation during times when the eyes are aligned. In addition, there is no discrepancy between binocular alignment (tropia) and the fully dissociated deviation (tropia plus phoria)—in contrast to monofixational exotropia. The eyes are straight (orthotropic) when fusion operates; the full deviation is measured only when the dissociation occurs.

Another condition to distinguish from monofixation exotropia is dissociated horizontal deviation, which is most commonly an exodeviation. This form of exotropia is quite variable and does not obey Hering’s law of equal forces to yoke muscles: the angle measured with one eye fixing differs from that measured when the fellow eye fixes. This occurs in infantile strabismus with its associated anomalies, including latent nystagmus and even DVDs. In contrast, monofixational exotropia that has decompensated does not exhibit dissociated deviation but rather displays about the same degree of deviation, no matter which eye is fixing.

Treatment

Nonsurgical Management

Amblyopia

The most important goal of treating decompensated monofixation syndrome is to reverse any existing amblyopia. Conventional therapy is used as indicated and includes spectacles for significant refractive error and anisometropia and also patching or penalization. Treatment continues until maximum vision is obtained in the amblyopic eye.

In the past, physicians were admonished not to improve vision beyond 20/40 (6/12) or 20/30 (6/9) so as to avoid reducing the size of the facultative suppression scotoma and thereby increase the risk of diplopia if the deviation decompensates. This is not borne out by experience in recent years. Treatment of amblyopia may, in fact, lead to improved control of the deviation, as occurs in typical intermittent exotropia.

Alignment

Asymptomatic patients with small angles of exophoria and good vision in both eyes whose amblyopia is controlled well during most of the day may safely be observed. If control is poor and symptoms are present, several treatment options exist. No treatment, however, will reverse the monofixation pattern, which seems to nearly always be innate and irreversible. Attempts to convert a patient from a monofixator to a bifoveal fixator have proved unsuccessful and impractical.

Occlusion therapy may be tried for symptomatic patients to improve control of the deviation or reduce its size, as happens in some patients with typical intermittent exotropia. Occlusion must be used daily on the fixing eye for at least half the waking hours. It often must continue for several months to achieve optimal control, followed by gradual tapering.

Orthoptic exercises are not indicated, because the fusional vergences are typically good. Exercises may, however, be of use if there is a problem of control at near fixation as well as diplopia or asthenopic symptoms. In addition, because there is an innate inability to fix bifoveally, any antisuppression methods that attempt to reduce the depth of suppression are contraindicated, just as they are in monofixation exotropia.

Prisms may be used to relieve symptoms of diplopia or asthenopia caused by deteriorating control of the exotropia. They can be used as Fresnel-type prisms and tapered, or may be ground into spectacles once the minimum amount needed to control the symptoms is determined. Treatment of typical intermittent exotropia with prisms reportedly improves control of the deviation in more than half of patients, especially at distance fixation, although the size of the tropia does not lessen significantly. Rarely does the angle increase when prisms are worn, in contrast to experience with prism adaptation for exotropia. Minus lenses to stimulate accommodation are effective in managing intermittent exotropia and should benefit some patients.

Pharmacologic Denervation

Pharmacologic denervation with botulinum toxin injection of extraocular muscles is an alternative to surgery for patients with a variety of strabismus problems, including intermittent exotropia. The reported success rate in reducing the deviation to less than 10 PD in intermittent exotropia is about 45%. The results are better for angles smaller than 20 to 25 PD than for larger ones; success rates exceeding 70% have been reported. Because most cases of decompensated monofixational exotropia are less than 25 PD, botulinum treatment may be a good alternative for a majority of patients. Its use specifically in cases of decompensated monofixational exotropia has not been investigated.

Surgical Management

Preoperative Considerations

Surgery is done in symptomatic patients when nonsurgical measures are unsuccessful or thought to be ineffective. No therapy, even surgery that restores good alignment, can alter the monofixation pattern. The best achievable result is restoration of an angle of deviation small enough to allow the macular scotoma to redevelop and permit peripheral fusion and vergence reflexes to regain their ability to control the deviation.

Preoperative evaluation for monofixational exotropia resembles that for a typical intermittent or constant exotropia. The distance and near deviations should be measured with an accommodation-controlling target and measurements repeated at extreme distance (60 m). Alignment should be checked in all positions of gaze to detect any concurrent pattern strabismus and lateral incomitance. A 45-minute patch test should be done to dissociate the eyes and detect the maximum possible deviation.

There are descriptions of the preoperative prism adaptation used to define the maximum angle of deviation, as in
esotropia. However, prism adaptation does not yield any information that cannot be gained from prism cover testing at 6 or 60 meters on accommodative targets.12,38

**Procedures and Outcomes**

The most common procedure described for correcting decompensated monofixational exotropia is to recess both LR muscles.12 The success rate in achieving a stable angle less than 8 PD on cover testing is well above 80% when patients are followed for at least 1 year. Preoperative near stereoaucuity is retained in more than 80% of patients.12 These excellent results have been achieved when small-angle esotropia is attained in the first week after surgery for decompensated monofixational exotropia.

**EXOTROPIA WITH HEMIANOPIC VISUAL FIELD DEFECTS**

Exotropia can occur when vision is poor in one eye, or if both eyes develop significant visual field loss. A divergent strabismus that manifests with unilateral visual loss is termed *sensory exotropia* (see section entitled Sensory Exotropia). This section deals with exotropia associated with bilateral hemianopic visual field defects, which is rarely described in the literature.

**EXOTROPIA WITH HEMIANOPIC VISUAL FIELD DEFECTS**

**Bilateral Homonymous Visual Field Defects**

**Incidence**

Exotropia may occur with homonymous visual field loss secondary to congenital or acquired intracranial lesions.16 The combination of congenital homonymous deficits with exotropia is believed to be rare,41,51 but some authors think it may be more common than previously reported.40,55,84 Several of the neurologic disorders such as cerebral palsy and hydrocephalus that may be associated with infantile exotropia (see section on infantile exotropia) can also be seen with congenital brain malformations—a small number of which are known to cause hemianopic defects.

**Physiologic Considerations**

The binocular field of vision in a normal patient with straight eyes extends well beyond 80 degrees to either side of primary fixation. This is termed the normal *panorama* of binocular vision.51,84 A homonymous field defect reduces the extent of this panorama. The rare presence of exotropia in the direction of the field defect enlarges the visual field. This phenomenon has different features depending on whether the visual deficit is congenital or develops early in life during the age of visual immaturity or whether the onset is beyond this age.

**CONGENITAL AND EARLY-ONSET DEFECTS**

Children who have congenital structural brain disorders or who acquire early lesions that lead to hemianopic defects are able to adapt surprisingly well to their seemingly major visual impairment.40,41,55 In fact, they often are completely unaware of their deficit.13 This adaptation is postulated to reflect rewiring of the plastic neural system in the developing brain, taking advantage of extrageniculostriate pathways.55 There is evidence that such a process takes place in infant kittens rendered hemianopic but not in adult animals subjected to acquired hemianopia.42

Infants who have congenital or early-onset hemianopic deficits are noted to spontaneously develop two other changes that may improve their performance: (1) a face turn in the direction of the missing field may be an attempt by the brain to more efficiently center the intact field in the visual space,40,54 and (2) the development of exotropia, seen much less often than a face turn, benefits the child when the eye preferred for fixation is the one contralateral to the visual field defect (Fig. 13–5). If the deficits are to the left in both eyes and the patient uses the right eye for fixation, exotropic deviation of the left eye would move its intact visual field toward the left. This enlarges the binocular field by several degrees to the left side in proportion to the size of the exotropia.41,51,84

The patient also may develop ARC, as well as suppression in the still-overlapping portions of the visual fields, both of which can be found on binocular sensory testing.41,51,84 Some authors believe that ARC develops only in patients whose exodeviation, like the visual deficits, arises early in life, generally before age 2 years.41,51 Others believe that exotropia may develop later and that ARC can still arise even if the exotropia is first noted after age 2.84 This process is also thought to improve the child’s ability to localize two objects in space, even if each is seen independently by either eye.51
Figure 13–5. Sample monocular and binocular visual fields in a patient with a congenital left complete homonymous deficit and exotropia. Upper left, Monocular visual field of the left eye. Upper right, Monocular visual field of the right eye. Lower left, Binocular visual field with the left eye fixing. Note the expansion of the total extent of the field to the right. The shaded area represents the overlap of the monocular fields. Lower right, Binocular visual field with the right eye fixing. Note the expansion of the field to the left in the direction of the monocular deficits. The shaded area represents the overlap of the monocular fields. (Redrawn from Gote H, Gregersen E, Rindziunski E: Exotropia and panoramic vision compensating for an occult congenital homonymous hemianopia: A case report. Binocular Vision Eye Muscle Surg Q 1993;8:129, with permission from Binoculus Publishing.)
Whether the exotropia that develops in such cases is truly a compensatory phenomenon or a coincidental finding remains controversial. If exotropia is helpful by enlarging the total visual field, it must represent an active process driven by the visual cortex. Other authors disagree with this concept, citing the fact that in some cases exotropia takes years to develop. If exotropia were in fact an active compensatory process, strabismus would regularly be apparent at a much earlier age. Various visuomotor strategies may develop to mitigate problems with such tasks as reading. They preclude the need for enlargement of binocular fields by active divergence over time. In addition, the development of exotropia would come at the expense of bifoveal fixation, a seemingly retrogressive adaptation.

**LATER ACQUIRED DEFECTS**

Older children and adults who acquire neurologic disorders that produce homonymous field defects have much more difficulty adapting to their visual disability than do young children. Many of them may also be unaware of the hemianopia but complain of problems tracking into the blind field and trouble localizing targets in space. Young children with congenital deficits, in contrast, are able to localize targets using adaptive saccadic strategies. Older children and adults may use a ruler or finger to track lines of text. Like younger children, they may adopt a face turn in the direction of the field defect.

Some patients have been noted to have exodeviation combined with a visual field deficit. The four adult patients reported by Roper-Hall likely had exophorias or intermittent exotropia but did not complain of diplopia, presumably because they had NRC and enough fusional reserve to maintain alignment. Orthoptic evaluation of these adult patients using the synoptophore demonstrated retained central fusion but reduced fusional amplitudes. The patient reported by Robertson and Williams had complete homonymous hemianopia after brain surgery and constant exotropia with diplopia. The diplopia was relieved by prism neutralization, indicating that ARC had not developed. It is unclear whether exotropia predated the surgery.

In summary, no adult patient is known to have developed exotropia in response to a hemianopia as intended compensation for the loss of visual field. Such cases are believed nonexistent. However, Levy and associates reported a boy with a presumed congenital homonymous deficit who developed exotropia after age 6 years. The phenomenon may not be limited to patients whose lesions occur before age 2 years. It conceivably could occur beyond the early years when the visual system is most plastic.

**Clinical Characteristics**

**SYMPTOMS**

In the congenital or early-onset forms of bilateral homonymous visual field defect, the patients—although they do not generally notice their visual deficit—develop various oculomotor strategies to help them cope with their field loss. One such adaptation is to use saccades into the blind field, followed by a smooth pursuit recovery to fix on the target. Even in the presence of exotropia, patients do not complain of diplopia, probably because of their ability to develop ARC. These patients may also exhibit a face turn in the direction of the field deficit.

In contrast, the later acquired forms lead to significant visual difficulties. Patients may not be aware of their field defects but do experience problems caused by loss of the hemifield. They frequently bump into objects not seen on one side and find reading difficult because of problems tracking into and out of the blind field. If they develop exotropia, they may complain of diplopia because they are generally bifoveal fixators who have NRC. ARC does not develop later in life; it is an adaptation seen only in infants and young children with early-onset strabismus. Adults may, however, be aware of an enlarged visual field secondary to the exotropia.

**MOTOR ALIGNMENT**

In congenital and early-onset cases of exotropia with a homonymous visual field defect, the magnitude of the deviation is uniformly constant and large, ranging from 40 to 70 PD. The exotropia may be associated with vertical tropias and pattern strabismus. Deviation at near may differ from that at distance by several prism diopters.

The deviations in later-onset cases are small, generally less than 20 PD. They may take the form of exophoria, intermittent exotropia, or constant exotropia.

**Visual Fields and Sensory Anomalies**

Depending on the underlying condition, homonymous visual field deficits can vary in their extent, depth, and congruity. The difficulty an adult or older child has adapting to an acquired deficit is proportional to the severity of the deficit. A young child can adapt better to any level of deficit from a partial one up to complete dense deficits in both eyes.

Patients with exotropia associated with congenital homonymous defects have all exhibited complete congruous hemifield losses. No reports have described the development of exotropia as compensation for a partial or incongruous defect, presumably because these patients can still develop normal binocular vision with vergence reflexes and have a visual panorama that is not compromised enough to induce exotropia.

The expansion of the total visual field in the direction of the missing field ranges from 20 to 45 degrees and, as expected, is proportional to the size of the exotropia. This feature is only present when the patient fixes with the eye contralateral to the hemifield defect (see Fig. 13–5). These patients all show suppression within the overlapping portions of the fields, as well as ARC on sensory testing. This ARC can be either a harmonious or a nonharmonious adaptation.

The ARC adaptation is not found in late-onset cases of homonymous field deficit with exodeviation. The exotropia leads to diplopia if the angle decompensates, overriding any benefit from expansion of the total visual field.
Diagnosis

VISUAL FIELDS

Both monocular and binocular visual fields must be documented in patients with homonymous field deficits. Monocular fields are plotted to diagnose the homonymous defect; to assess its congruity, density, and extent; and to determine whether the maculae are spared. The binocular field is plotted to determine the extent of the panorama of the visual field. With homonymous defects, the fixing eye must be noted: in the presence of exotropia, the overall field would be enlarged in the direction of the missing field, as long as the fixing eye is contralateral to the field deficit.

MOTOR ALIGNMENT

Initial observation of the patient should reveal whether a compensatory head posture is adopted in response to the field defect. This feature can occur with hemianopias caused by either early- or late-onset disorders. The exotropias seen in congenital hemianopia are large and constant, whereas exotropias in the late acquired forms may be phorias or intermittent or constant exotropias. The deviation can be measured in the primary position, both at distance and at near, by prism and alternate cover testing in older children and adults, and by the Krimsky test in young children. The deviation should be measured in the secondary fields as well, to detect pattern strabismus or lateral incomittance. In cases with macular sparing it is possible to determine an endpoint in the prism measurements.

Eye movements must be observed to detect any underaction or overaction of the muscles. The eye movements are often normal in both the early-onset and late-onset forms. Vergence amplitudes may be assessed by measuring prism vergences, both base-in and base-out, or by using the synoptophore. They may be decreased in some patients with acquired homonymous deficits.

SENSORY TESTING

In the congenital forms of homonymous deficits, suppression in the nonfixing eye is detected by fovea-to-periphery tests such as the Worth four-dot and Bagolini striated lenses. When the eye ipsilateral to the field defect is fixing, the image in the exotropic fellow eye falls on its nonseeing temporal retina. When the fellow eye fixes, the image in the now exotropic first eye falls on the functioning temporal retina and is suppressed.

The presence of ARC in an early-onset form can be demonstrated by offsetting the angle of deviation with prisms and noting if diplopia develops. The synoptophore may serve to confirm whether ARC is harmonious or otherwise. Binocular perimetry may disclose suppression in the overlapping visual field and show evidence of ARC in a portion of the overlap.

In the later-onset forms of homonymous hemianopia, patients usually exhibit NRC and diplopia when an exotropia is present. It is unlikely that such a patient can develop suppression unless the exotropia predates the field defect and begins in childhood.

Treatment

It is unclear whether exotropia with a congenital or early-onset homonymous hemianopia develops as a compensatory mechanism. Proponents of either view strongly caution against any attempt to realign the eyes of such patients. Two reasons are cited to support this admonition: (1) straightening the eye with exotropia may reduce the panorama of total visual field, upsetting a patient who has adapted to an enlarged field; and (2) preoperative sensory testing in the few patients reported revealed ARC, and thus realigning the eyes then carries a significant risk of inducing paradoxical diplopia.

If a patient in this situation wants to have the eyes aligned, several maneuvers should be considered. The patient should be patched monocularly for a time to see whether he or she is able to adapt to the smaller panorama of binocular visual field that will result from realigning the eyes. In addition, prisms used to offset the full angle of exotropia, when placed before the eye with exotropia, allow the patient to experience realignment of the visual axes. This maneuver also allows the surgeon to predict postoperative paradoxical diplopia. Finally, the surgeon can temporarily realign the eye by injecting lidocaine into the LR muscle of the exotropic eye under electromyographic control. For a short time while the effect is wearing off, the eyes should be close to straight, giving the patient an opportunity to experience the effect of realignment surgery.

In the case of an acquired defect, the patient may not be symptomatic because the exodeviation is controlled. If the deviation becomes constant, diplopia usually develops. It is therefore unlikely that such a patient would wish to continue having exotropia, even with an expanded binocular visual field. If a patient wishes to have the eyes straightened to eliminate diplopia, the options include nonsurgical methods such as patching, prisms, or botulinum injections as well as surgical realignment. Robertson and Williams reported one patient with constant exotropia, diplopia, and NRC whose diplopia responded to prisms. Gradual tapering of the prisms then aided recovery of control to a stable exophoria.
Bilateral Temporal Visual Field Defects

Incidence

Bitemporal hemianopia is a sequel to lesions such as pituitary tumors and aneurysms that are near the optic chiasm. It is rare for strabismus to develop unless there is significant field loss in both eyes that includes the macula. In such cases there is loss of binocular fusion; in the absence of fusion lock, the eyes may dissociate to exotropia or esotropia. There are no studies documenting the exact prevalence of this phenomenon, but it probably is a frequent finding in large field losses involving the macula.

Physiologic Considerations

Whereas homonymous bilateral field defects may not interrupt binocular vision, a bitemporal deficit almost always interferes with fusion. Normal functioning of the fusion mechanism depends on intact sensory and motor components. These include normal decussation of the retinal fibers in the optic chiasm and an intact vergence system that tracks targets in depth and responds to stimulation of disparate retinal points proximal and distal to the point of fixation.25, 29, 116 This process depends on overlap of the visual fields from both eyes.

The loss of extensive areas of the temporal visual field in both eyes disrupts this system by preventing stimulation of corresponding retinal points, leading to several problems.25, 29, 37, 116, 129 It results in failure to generate corrective fusional vergences, and this may promote the decompensation of a preexisting heterophoria. If the deviation is an exophoria, an intermittent exotropia may develop.25, 37, 116 Patients also describe a hemiretinal slide, because the visual fields of the two eyes are segregated from one another and cannot be synchronized by the normal fusional vergence reflex. This creates significant reading problems.25, 37, 116, 129 Exotropia will duplicate features of the target or elongate an object, whereas esotropia will lead to a loss of certain features or truncate the target (Fig. 13–6).25, 37, 116, 129

Patients also have a blind area distal to the fixation point, termed postfixation blindness (Fig. 13–7).73, 116 If they fixate bifoveally, they do not respond to targets located beyond the fixation target and will not respond to uncrossed physiologic diplopia or base-in prisms because of the loss of divergence disparity (see Fig. 13–7, left).116, 129 If one eye diverges, the area of postfixation blindness becomes more remote. Objects proximal to this area, including the target, may then be seen as diplopic unless suppression develops (see Fig. 13–7, right).

Clinical Characteristics

SYMPTOMS

As noted, patients have disruption of the vergence reflex because of the loss of corresponding points in the retina of both eyes. If they develop exotropia, they complain of retinal sliding: the independent partial images of the target seen by the temporal retinas of both eyes overlap.37, 73, 116, 129 This overlap leads to the subjective sensation of an elongated target, or duplication of some features of the target. Serious reading difficulties result when words or letters are duplicated on the page. These patients also may see split-screen images.25, 29, 129

Loss of fixation beyond the fixation target, or postfixation blindness, may lead to several complaints. The patient may not see items beyond the target, leading to mishaps while moving about in a room or doing routine visuomotor tasks such as cutting food or pouring liquids.129 This can occur whether the patient fixes bifoveally or develops exotropia in one eye (see Fig. 13–7).

Despite the disordered binocular vision, many patients may not be aware of many of the changes in their vision.37

MOTOR ALIGNMENT

The angles of the exodeviations associated with bitemporal hemianopic visual field defects are typically only a few prism diopters. Strabismus creates symptoms because the patients generally have NRC and are unable to suppress the image from either eye when their visual fields overlap.25, 29, 37, 129

VISUAL FIELDS AND SENSORY ANOMALIES

Fusion problems occur with this class of visual field defects when there is very little or no overlap of the visual fields of the two eyes. This promotes decompensation of a preexisting phoria because motor-vergence reflexes are absent.116, 129

When the eyes are straight, the total panorama of the visual field is normal, with each half of the total being provided by the contralateral eye. Once the eyes diverge, the total field actually contracts and the patient notes a duplication of features in the center of the target object (see Fig. 13–6).

Diagnosis

VISUAL FIELDS

As for homonymous field defects, both monocular and binocular visual fields must be documented in patients with heteronymous field defects.41, 51, 84, 116 The monocular fields are plotted to diagnose heteronymous defects; to assess their congruity, density, and extent; and to determine whether the maculae are spared. In the presence of bitemporal defects, the temporal limits of the binocular field may contract if exotropia becomes manifest because of overlap or may widen if an esotropia is present.
A. Straight Eyes

B. Left Esotropia

C. Left Exotropia

Figure 13-6. Hemifield sliding and abnormal binocular phenomena observed in a case of complete bitemporal hemianopia. A. In the orthotropic state, the two separate monocular visual fields juxtapose to form a complete image of the target. B. In esotropia, portions of the target are deleted. C. In exotropia, portions of the target are duplicated because of the redundant reception by the functioning temporal retinas of both eyes. (Redrawn from Fritz KJ, Brodsky MC: Elusive neuro-ophthalmic reading impairment. Am Orthopt J 1992;42:160, with permission of the University of Wisconsin Press.)

MOTOR ALIGNMENT

Patients with complete bitemporal deficits tend to have variable exotropia because of poor or nonexistent fusion vergences. The deviation should be measured by prism tests, but the endpoints may be difficult to determine. These patients show no divergence responses to base-in prisms when fusion amplitudes are tested because a blind area is present distal to the fixation point. Extraocular movements typically are normal.

Patients with an incomplete deficit and preserved macular fixation may have very good fusional vergence responses. They are much less likely to manifest an intermittent exotropia or exophoria.

SENSORY TESTING

When the eyes are fixing bifoveally on the target, the blind area present distal to fixation causes patients with bitemporal deficits to show physiologic diplopia only in the proximal position relative to fixation, not in the distal position. Stereopsis is detected if the targets are held just proximal to fixation. When an exotropia is present, the patient may note diplopia unless suppression develops in the deviating eye.

Sensory tests confirm the loss of fusion and may reveal either diplopic responses or suppression of one eye. These patients should be tested with reading material and asked to draw or describe what they see. This can demonstrate orthotropia, exotropia, or esotropia (see Fig. 13–6).

Treatment

Bitemporal field defects, especially if accompanied by exotropia or esotropia, can be very upsetting to a patient because of their effects on such activities as driving, reading, and simple tasks like cutting food and pouring liquids.
Figure 13-7. Postfixation blind area in a case of complete bitemporal hemianopia. Left, While both foveas (F) fix on a target (T), any object (O) within the shaded area would have its image fall on the nonfunctioning nasal retina in each eye, causing a lack of perception of the object. Right, Left exotropia with the right eye fixing. Target (T) may be seen as diplopic because its image falls on the functioning temporal retina of the left eye. The postfixation blind area, containing a nonvisualized object (O), is more remote than when eyes are orthotropic. (Left redrawn from Roper-Hall G: Effect of visual field defects on binocular single vision. Am Orthopt J 1976;26:74, with permission of the University of Wisconsin Press.)

Prisms are not helpful in many cases because of the variable alignment of the ocular deviation. In the case of exotropia, however, prisms may increase separation of the images and so avoid diplopia without sacrificing the total visual field.129

Patients should be encouraged not to view objects on lateral gaze, because the visual axes might remain misaligned on refixation in the primary position. Occlusion of one eye is rarely helpful because the patient will have to function with only the nasal half of one monocular field.

Surgery also is fraught with difficulty because of the varying nature of the alignment. If a constant exotropia is causing distress because of the hemifield-sliding phenomenon, surgery may be offered in an attempt to eliminate the distortions and diplopia experienced with binocular viewing. Adjustable sutures are recommended in such cases, because accurate postoperative alignment is crucial. Nevertheless, even with optimal alignment on the day of surgery, the results are unstable because of the absence of binocular vision and vergence reflexes.25
Definition and Incidence

*Sensory exotropia* is the term used for a unilateral divergent misalignment that is a sequel to loss of vision or long-standing poor vision in an eye. The exotropia can develop within days to weeks of the onset of an acquired visual deficit such as a cataract, or it may evolve over a longer period when a chronic visual problem such as amblyopia exists.

Sensory exotropia may result from a monocular vision problem at any age. In children younger than 5 years old, the rates of exotropia and esotropia in strabismus occurring in poorly sighted eyes are about equal. There appears to be no correlation between the severity of visual impairment and the type of deviation (esotropia or exotropia) that will develop. Among children older than age 5 years and adults, an eye with poor vision tends to develop exotropia. In adults, sensory exotropia accounts for 20% to 25% of all cases of exotropia.

Causes of Sensory Exotropia

The causes of sensory exotropia are numerous; they include all pathologic states that reduce visual acuity in one eye, whether congenital or acquired. A classification of causes is listed in Table 13–2.

**EYE PROBLEMS IN CHILDHOOD**

Any unilateral media opacity present from birth, such as a cataract or corneal scar, may degrade the retinal image sufficiently to cause strabismus. Eyelid disorders such as ptosis and hemangioma can lead to amblyopia through deprivation and/or astigmatism. Posterior segment anomalies include structural defects involving the macula, such as coloboma, optic atrophy, and retinal lesions such as hemangioma.

Various acquired problems may also lead to visual loss and exotropia. Cataract can occur in a previously clear lens after trauma or in lens disorders such as posterior lenticous. Posterior segment lesions can impair vision. Most worrisome is a tumor such as retinoblastoma, but more common retinal diseases like cicatricial retinopathy of prematurity may also be responsible.

One common cause is anisometric amblyopia that is unresponsive to therapy or is diagnosed beyond the critical period within which amblyopia can be reversed. It is principally associated with anisometropia—especially high myopia—and anisoastigmatism, both of which degrade distance acuity. The effects are most noticeable to the child after the first few months of life, when interest and gaze are increasingly directed away from the immediate surroundings. Exotropia also may occur in amblyopia because of anisohyperopia, although esotropia is much more common in this condition.

**EYE PROBLEMS IN ADULTS**

Both traumatic and nontraumatic conditions can lead to reduced vision. As in children, any opacity in the cornea or lens can degrade the image seen by the eye. Various inflammatory conditions may involve either the anterior or posterior segment. Disorders in the posterior segment include chronic retinal detachment and acute detachment that was not successfully repaired or in which the macula is scarred and not fully functional. As with children, a tumor, though rare, must be considered in any case of acquired strabismus.

Late-onset cataract, either traumatic or nontraumatic, commonly causes sensory exotropia in adults. Exotropia may develop within weeks of onset of the visual deficit from an advancing cataract, usually when a preexisting exophoria decompensates (Fig. 13–8). It may also become mani-

---

**Table 13-2. Classification of Causes of Sensory Exotropia**

### In Children

<table>
<thead>
<tr>
<th>Category</th>
<th>Causes</th>
</tr>
</thead>
<tbody>
<tr>
<td>Congenital</td>
<td>Eyelid lesions (e.g., hemangioma, ptosis)</td>
</tr>
<tr>
<td></td>
<td>Media opacity (e.g., cataract, corneal leukemia)</td>
</tr>
<tr>
<td></td>
<td>Developmental defects (e.g., uveal coloboma, optic atrophy)</td>
</tr>
<tr>
<td></td>
<td>Tumor (e.g., retinoblastoma, hemangioma)</td>
</tr>
<tr>
<td>Acquired</td>
<td>Trauma, penetrating or nonpenetrating</td>
</tr>
<tr>
<td></td>
<td>Refractive (e.g., anisometropic amblyopia)</td>
</tr>
<tr>
<td></td>
<td>Media opacity (e.g., developmental cataract)</td>
</tr>
<tr>
<td></td>
<td>Intraocular tumor (e.g., retinoblastoma)</td>
</tr>
<tr>
<td></td>
<td>Posterior segment disease (e.g., cicatricial retinopathy of prematurity)</td>
</tr>
</tbody>
</table>

### In Adults

<table>
<thead>
<tr>
<th>Category</th>
<th>Causes</th>
</tr>
</thead>
<tbody>
<tr>
<td>Trauma, penetrating or nonpenetrating</td>
<td>Inflammatory (e.g., chronic uveitis)</td>
</tr>
<tr>
<td></td>
<td>Media opacity (e.g., cataract, corneal scars)</td>
</tr>
<tr>
<td></td>
<td>Posterior segment disease (e.g., macular degeneration, retinal detachment)</td>
</tr>
<tr>
<td></td>
<td>Intraocular tumor (e.g., uveal melanoma)</td>
</tr>
</tbody>
</table>

---

*Figure 13-8. Patient with sensory exotropia of the left eye caused by a cataract.*
Physiologic Considerations

STIMULI FOR DIVERGENCE

Several theories about why loss of vision predisposes to exotropia have been proposed. Jampolsky emphasizes that the primary stimulus common to all forms of sensory exotropia is the diffusion of light that results in blurred contours of the images transmitted to the retina of one eye while clearly focused images are received by the retina of the normal fellow eye. This sets up a constant rivalry between the two eyes under binocular conditions: signals from the normal eye lead to inhibitory signals to the fellow eye.

The tendency of older children and adults to develop sensory exotropia is ascribed to a difference in efficiency between the nasal and temporal hemiretinas. The nasal retina (directed to the temporal field) is more sensitive in localizing bright-light stimuli and in sensing the brightness of targets than is the temporal retina. Blurred contours and competitive interaction between the sound and abnormal eyes, due to the asymmetric sensory input, augment the difference in efficiency between the two hemiretinas. As a result, the physiologic superiority of the nasal retina in the affected eye is enhanced while the response of the temporal retina (nasal field) is diminished. This leads to loss of fusional reflexes, because the overlapping visual fields essential to the vergence control system are disrupted. The result is stimulation of an active retinomotor-divergence reflex that makes the eye diverge at an increasing angle over time.

Jampolsky further qualifies his hypothesis by indicating that diffusion or degradation of the image transmitted to that eye must be present simultaneously in both the central and peripheral fields. This implies that a lesion involving only the central field (such as a focal chorioretinal scar), or only the peripheral field (such as early glaucomatous field loss), is not an adequate retinomotor stimulus. In addition, the loss of vision must be partial and not total; complete blindness does not cause nearly as powerful a binocular interaction as that seen with blurred contours in one eye.

A formerly well-controlled exophoria may decompensate when a media opacity or other visual deficit develops in one eye. The loss of fusion lock caused by degradation of peripheral visual field responses in the abnormal eye may allow the eye to settle into a divergent rest position. This phenomenon differs from the active divergence tone induced by blurred contours. Dissociation of a phoria is not considered to be an active process. Rather, it is a settling of the eye into a position of rest determined by the relative tone of the LR and MR under dissociated conditions—which in the case of exophoria is a divergent position. The size of the exotropia should not increase significantly over time, unless active divergence occurs secondarily as a result of ongoing blurring of image contours.

Other theories implicate altered refractive, mechanical, and tonic vergence states. In the past, it was thought that the eyes tend to develop exotropia if the fixing eye is hyperopic or exotropia if the error is myopic. This has not, however, proved to be a consistent trend. Increasing horizontal separation of the orbits through adolescence may mechanically predispose the eyes to reach a divergent rest position when binocularity is upset. There may be a loss of tonic convergence during late childhood and into adulthood, disposing to exotropia if the motor and sensory fusion reflexes are disrupted by loss of vision in one eye.

Studies in infant primates have shown that deprivations shortly after birth, such as monocular defocusing or optically corrected aphakia, lead predominantly to exotropic deviation in the early weeks of life. In addition to exotropia, some of these animals develop nystagmus whose intensity is greater in the deprived eye. Temporal modulation and contrast sensitivity are markedly reduced in the deprived eye compared with the nondeprived eye.

SENSORY ADAPTATIONS

The development of an exotropia may lead to several possible sensory compensations. Various authors have shown that there may be a deep regional hemiretinal suppression in the temporal retina or that the area of suppression may be more extensive and extend into the nasal retina as well. The results depend on the nature of the binocular test used and on the extent of binocular dissociation created by the test.

The type of retinal correspondence that develops in sensory exotropia depends on the quality of vision in the poorer eye and the cause of visual loss. If there is useful vision in the deviated eye, a rudimentary binocular reflex may allow the development of ARC, seen in a significant minority of patients. Some cases exhibit both ARC and NRC, depending on the dissociating properties of the test, the viewing conditions (dark versus illuminated surroundings), the target position (distance or near), and the angle of misalignment. When vision is very poor in the deviated eye, there may be no evidence of binocularity at all, even with tests that minimally dissociate the eyes.

The extent of sensory adaptations may influence the results of realignment surgery for exotropia. For example, cases of sensory exotropia caused by long-standing cataract in adults, or associated with congenital cataract, may fail to develop any evidence of binocularity despite alignment of the eyes to within less than 10 PD. Some adult patients with exotropia caused by cataract or corneal scar may have intractable diplopia after correction of the media opacity and restoration of normal alignment. Such cases have been termed central fusion disruption or horror fusionis.
Clinical Characteristics

Ocular Abnormalities

Sensory exotropia develops in response to a loss of formed vision in one eye. Therefore, in each case there must be a detectable abnormality in the eye or optic nerve that could diminish vision. Table 13–2 lists these causes in both children and adults.

It is important at any age to exclude an intraocular cause of strabismus, whether esotropia or exotropia. Cataract or an intraocular tumor such as childhood retinoblastoma may first manifest as exotropia. A complete ocular examination, including refraction and detailed fundus assessment, is mandatory at the outset when evaluating unilateral strabismus with poor vision.

Motility Problems

The size of exotropia is variable, even within the same class of origin. The angles are characteristically large, ranging from 30 to 100 PD. The angle of exotropia usually increases gradually over time, as long as the cause of visual deficit remains active.

An eye with long-standing sensory exotropia often develops any of several mechanical and innervational abnormalities, especially if the angle is more than 40 PD. The LR often becomes shortened and contractured, causing a mild to moderate limitation of adduction. Its intermuscular attachments and overlying conjunctiva and Tenon’s fascia also are shortened and tightened. The MR muscle may become inelastic or may become thinned and atrophic because of chronic stretching of its fibers.

In long-standing exotropia there is a characteristic upshoot and downshoot of the adducted eye. This phenomenon has been attributed to three different mechanisms. First, when the eye initially adopts a markedly exotropic position, the superior and inferior oblique muscles become slack. Over time they become shortened, leading to overaction of both muscles in their respective fields of action. A second explanation is sideslipping of the short, tight LR along the sclera as the eye rises above a falls below the midline. This “leash” effect is also seen in some cases of Duane syndrome and causes an abrupt upshoot and downshoot in the adducted position. A third possibility is that the abnormal vertical movements are, in fact, pseudo-overactions of the oblique muscles and are not caused by true shortening of the muscles or tendons. Rather, the eye may have more room to elevate or depress because it does not reach the medial wall of the orbit when it adducts in tandem with the fixing fellow eye.

Clinically a pattern of limited adduction is evident, with excess elevation and depression of the eye in the adducted position. The superior and inferior rectus muscles may show underaction in their fields of action as well. These features may also be noted in the normal fellow eye. Bilateral anomalies may create an X pattern. This constellation of findings is termed the tight LR syndrome and is characteristic of long-standing large-angle exotropia (Fig. 13–9). Often, a vertical tropia accompanies long-standing exotropia; hypertropias are more common than hypotropias.

In addition to the tight LR muscle, the overlying conjunctiva and Tenon’s fascia may be shortened. The stiffened orbital tissues may lead to an exotropic bias of the globe. This can be demonstrated in some cases of large-angle exotropia at the time of surgery: even after the MR and LR muscles have been detached from the globe, a spring-back balance bias favors exotropia. There appears to be a positive correlation between the size of the angle of exotropia in the alert patient and the amount of exo bias as determined at surgery. Patients with reduced vision because of unsuccessful surgery for retinal detachment may have additional restrictive forces because of the position of the scleral buckle or postsurgical fibrosis.

Exotropias less than 40 PD may not exhibit these anomalies but can develop oblique muscle dysfunction. Superior oblique overactions leading to A patterns are much more common than inferior oblique overactions. In addition, patients may have different-sized angles at distance and at near. These result in recognizable patterns typical of nonsensory exotropia, including divergence excess and convergence insufficiency.

Figure 13–9. Nine gaze positions of a patient with a large sensory exotropia of the left eye and tight lateral rectus syndrome. Note the limited adduction and updrift and down-drift of both eyes in adduction. An X pattern is present.
SENSORY FINDINGS

As noted earlier, several sensory adaptations may occur in sensory exotropia. If vision is very poor, there may be no binocular response on any sensory test. If vision is less severely compromised, some binocular response may be preserved. Young children may develop ARC in response to a stable exotropia as long as the angle is not excessive. They may sometimes retain NRC, as in sensory exotropia caused by anisometropic amblyopia. ARC adaptation is unlikely to develop in angles greater than 40 PD. In older children or adults with a later onset of visual loss, there may be diplopia because ARC cannot be established, unless vision is so poor that the image in the deviating eye can be ignored.

If binocular responses are present, the findings on various sensory tests may conflict. The results depend on the dissociating ability of the sensory test used and on the depth of suppression that has developed. A patient may show NRC on one or more tests and ARC on others. Stereopsis rarely is preserved in sensory exotropia caused by the combination of reduced vision in one eye and a large angle of deviation.

Diagnosis

A complete ocular examination including evaluation of the adnexal tissues is mandatory in any case of unilateral exotropia, at any age, to identify an organic lesion. Because sensory exotropia develops in response to reduced vision in one eye, a reduction in best-corrected visual acuity must be documented in the deviated eye. This is evaluated by conventional tests appropriate for the patient’s age. An accurate refraction is needed to diagnose anisometropia or high refractive errors that may be factors in the development of sensory exotropia (see also Chapter 1).

MOTILITY EVALUATION

The method used to measure the angle of exotropia depends on the level of vision in the misaligned eye and the patient’s age. If vision is good enough to permit fixation on an accommodative target, the prism cover test can yield an accurate result. This test is valid for both adults and children who are able to cooperate. If vision is too poor to allow reliable fixation, or if the child is young and cannot cooperate for a prism cover test, the angle can be determined with the prism and light reflex (Krimsky) test.

When the Krimsky test is used, the preferred method is to place the prism base-in over the fixing (better) eye. First the angle kappa is noted in the fixing eye. Then a series of increasingly strong prisms are placed over that eye, drawing the poorly seeing fellow eye toward the primary position. The true angle is measured when the light reflex in the pupil of the fellow eye reaches the same angle kappa as that originally noted in the fixing eye (Fig. 13–10).

If the prism cover test can be performed, the angle should be measured in the primary position at distance and near. The angles in the secondary positions are also measured: side gaze alignments will detect lateral incomitance; up and down positions will reveal any pattern (A, V, or X). If vision in the exotropic eye is too poor to allow a prism cover test at distance, the primary and secondary position measurements may be done using the Krimsky test. The examiner must also be alert to the possibility of concurrent vertical deviations, because these occasionally occur with sensory exotropia.

The eye movements must be documented, paying careful attention to limitations of adduction, oblique muscle overactions, and vertical rectus muscle underactions. Any or all of these features may occur in long-standing exotropia, especially if the angle is large. These anomalies also may occur in the normal eye.

The presence of a tight LR muscle is confirmed by a forcedduction test. Restriction is due to tightness of the lateral orbital tissues, both muscle and soft tissues.

SENSORY TESTING

Depending on the patient’s age, the length of time that exotropia has existed, and the cause of vision loss, different sensory adaptations may be present. Several tests are avail-
able, rated according to their dissociating ability and whether they are fovea-to-periphery or fovea-to-fovea tests. The most common clinical tests used to confirm the presence of total suppression, ARC, or NRC include the Worth four-dot or polarized four-dot tests, Bagolini striated lenses, the synoptophore (amblyoscope), the red glass test, and the afterimage test. An adult or older child with an acquired disorder may have diplopia, which may be confirmed by measures such as the red glass test.

Suppression in cases of sensory exotropia may be further studied with binocular perimetry, by dissociating tests such as the Lees screen with red-green dissociation, or with haploscopic devices. The suppression scotoma size, its location, and the depth of suppression can be ascertained in older children and adults.

Once patients have had any underlying disorders identified and treated, they should be tested to see if they have central fusional disruption before realignment is considered. This phenomenon can result from several forms of visual loss, including long-standing cataract in adults, congenital cataract, macula-off retinal detachment, and eye injury associated with blunt head trauma. It may be identified with prisms that neutralize the deviation in free space, or with a synoptophore (amblyoscope).

**Differential Diagnosis**

Sensory exotropia must be distinguished from the common forms of exotropia, both constant and intermittent. In the typical intermittent exotropia the eyes are straight at times, either at distance or near. A person with long-standing constant exotropia not due to a sensory cause will have good vision in both eyes or mild amblyopia and may have alternating suppression on sensory tests.

Sensory exotropia may be confused with consecutive exotropia, defined as one that arises spontaneously in a patient who was originally esotropic. It is commonly associated with anisohyperopia and frequently is complicated by amblyopia. Sensory exotropia also can be confused with postsurgical exotropia occurring after strabismus surgery for esotropia. In both entities there is a clear history of preceding esotropia.

Various innervational and mechanical disorders that can cause congenital or acquired exotropia may produce concurrent amblyopia or organic visual loss. The innervational disorders include third nerve palsy (both congenital and acquired), internuclear ophthalmoplegia, and exotropic Duane syndrome. Mechanical causes include orbital diseases such as thyroid and fibrosis syndromes and orbital tumors that deviate the eye into a divergent position. These disorders have their own characteristic features in addition to the exotropia.

**Treatment**

### General Ocular Conditions

The most important aspect in managing sensory exotropia is to eliminate or reverse a treatable underlying condition such as amblyopia, cataract, ptosis, retinal detachment, or intraocular tumor. It is possible for sensory exotropia to improve if a reversible disorder is treated in a timely manner.

There are two general principles in the treatment of sensory exotropia. First, even with an irreversible organic cause such as uveal coloboma or a macular lesion, functional amblyopia may be superimposed on an organic visual loss. This element of the visual deficit may be reversible by patching. Second, any patient whose best-corrected vision is less than 20/40 (6/12) should receive a prescription for safety lenses. A patient already in spectacles should be advised to check with the optician to ensure that glasses and frames meet safety specifications. Three key design features include the following: (1) the frame should be a solid one-piece design that is more resistant to impact than thin or wire frames; (2) the lenses must meet safety specifications for the thickness or type of plastic; and (3) lenses should be beveled so that they pop out of the frame rather than inward when stressed.

### Strabismus Management

#### Nonsurgical Management

**Treating the Underlying Pathology.** Once an underlying ocular disorder is successfully treated, sensory exotropia may resolve. This is not uncommon in surgery for adult cataract or retinal detachment that is caught early, when the eye regains good vision. For children with an early-onset media opacity such as corneal leukemia or cataract, the problem must be corrected within the first few weeks of life to prevent severe permanent visual loss.

If the cause is anisometropic amblyopia, correcting refractive errors may eliminate exotropia if vision improves significantly. Patching or penalization may be needed to maximize vision. Amblyopia therapy is an integral part of visual rehabilitation for infants having congenital media opacities.

**Prisms.** Some adults who complain of diplopia or asthenopia because of exotropia despite poor vision in the eye may be helped by prisms if the angles are not large (usually less than 30 PD). The angle of misalignment is often large, however, making it impractical to prescribe prisms, even in a Fresnel (paste-on) form. In addition, these patients are often bothered by a combination of blurring from the prisms and a defocused image from the underlying disease. It is unusual for patients with sensory exotropia to achieve comfortable binocular vision with prisms.

In some cases the patient has central disruption of fusion that may preclude comfortable binocular vision, even if the deviated eye were realigned. These patients might best be left without realignment. Alternatively, the eye may be realigned, followed by intentional blurring of the poorer eye using optical measures.

**Pharmacologic.** Chemodenervation of the LR muscle of the involved eye with agents such as botulinum toxin may serve both diagnostic and therapeutic purposes. If the surgeon is considering realigning the eye but is concerned about resultant diplopia or fusion problems, temporary pharmacologic realignment may allow the patient to experience what it would be like to have the eye straight. A diagnostic injection of lidocaine will give the patient a few minutes of good alignment, whereas botulinum toxin can produce a straight eye for several days.

Botulinum injections are an alternative to surgery if the
angle of exotropia is not excessive. The overall rate of realignment to a deviation less than 10 PD after injecting one LR muscle is approximately 45%, even with multiple injections. The results approach 70% when the angle is less than 25 PD.\textsuperscript{1, 12} However, the sensory exotropia angle often exceeds 40 PD, making this therapy much less effective overall than surgery. Nevertheless, if the goal is to improve alignment for cosmetic reasons rather than precisely, botulinum injection is a good alternative to surgery. Periodic injections are often required for large angles to maintain acceptable alignment.\textsuperscript{83}

**Surgical Management**

**Preoperative Considerations.** The goals for the patients' vision should be defined on a case-by-case basis. The surgeon must make an effort to determine whether there is a reasonable chance of fusion. Even in adults with reduced vision in one eye, it is possible to gain peripheral fusion or stereopsis in many cases; cases can benefit by more than just motor alignment.\textsuperscript{99, 94, 127} This possibility can be examined using various sensory tests and diagnostic pharmacologic realignment.

If there is a chance of regaining fusion, an effort should be made to straighten the eyes to within a few prism diopeters of orthotropia. Where vision is very poor, the eye should be realigned to an optimal motor position. If preliminary tests show a strong probability of postoperative diplopia or if there is central fusional disruption, surgery may be contraindicated. Alternatively, the surgeon can test the patient for the subjective response to a partially corrected angle. Some patients can adapt with partial realignment, which improves the appearance but leaves the blurred image far enough out of conscious view so that it does not interfere with the clear image perceived by the normal eye.

Preoperative forced duction testing will help confirm the presence of mechanical restriction. The operation must free lateral restrictions to achieve successful long-term alignment. The surgeon may choose to address concurrent A or V patterns or a vertical tropia at the time of exotropia repair.

Finally, the overall health of the eye must be taken into consideration. The evaluation must assess the intraocular pressure, the extent of conjunctival scarring, the state of the cornea, and the posterior segment. The surgical plan may have to be modified if any of these abnormalities are present.

**Surgical Procedure.** Surgery for sensory exotropia should, if possible, be confined to the eye with the visual deficit, even with a large-angle deviation.\textsuperscript{110, 111, 138, 143} The procedure must include a weakening procedure on the LR muscle. Attention must be given to contractured soft tissues on the lateral side, a frequent finding in this condition. The best approach is a large LR recession, usually of 8 mm or more, combined with recession of the tight conjunctiva and Tenon's fascia.\textsuperscript{111, 121, 138} To stabilize the result and prevent a recurrence, the MR muscle should be resected in nearly all cases.

The forced duction test is repeated after each muscle undergoes surgery to see if the lateral restriction is substantially or completely reduced and to learn whether resection of the MR muscle has created a mild to moderate restriction of abduction. Spring-back balance testing usually shows an exotropic bias in the early stages of surgery, even without using a depolarizing muscle relaxant.\textsuperscript{99} This should be rechecked once the MR muscle is resected. The resection should create enough tension in favor of adduction so that the spring-back balance is biased slightly in the esotropic direction at the end of surgery. This bias, along with a limitation on attempted forced abduction, is the best security against recurrence.

Despite adhering to these principles, the immediate postoperative alignment may not be optimal. It is helpful to use an adjustable suture technique in the LR recession.\textsuperscript{75}

Almost always an exotropic drift occurs after surgery for most forms of exotropia. This drift usually is in the range of 5 to 7 PD.\textsuperscript{126, 146} To ensure long-term stability, it is probably best to align the deviated eye to within 5 to 7 PD of esotropia. Agreement with this goal is not, however, unanimous. In cases of a large exotropia associated with amblyopia, Rayner and Jampolsky\textsuperscript{111} caution against intentional overcorrection soon after surgery, especially in adults. Leaving a small angle of exotropia is better accepted. Patients' eyes do not look "too crossed," but these individuals should be warned of postoperative drift. If vision is very poor, surgery may be designed to align the eye so that the light reflex is centered, rather than making the angles kappa symmetric. For example, if the patient has a positive angle kappa in the fixing eye, perfect realignment may leave the eyes appearing pseudoexotropic.

Some authors employ a unilateral four-muscle procedure to correct extremely large angles (more than 50 PD) of exotropia.\textsuperscript{110, 138} Both oblique muscles are weakened in addition to the horizontal rectus muscle surgery. The principle is to release all abducting forces, including those from the oblique muscles. The inferior oblique is recessed, and the superior oblique tendon is tenotomized. The LR is recessed, and the MR muscle resected, both in the range of 6 to 8 mm. Both horizontal muscles are secured to sclera a few millimeters inferior to the original insertion so as to prevent hypertropia after weakening both oblique muscles simultaneously.

**Results of Surgery.** The long-term postsurgical alignment in sensory exotropia is expected to be poor because the chance of regaining stable fusion is low. However, a stable small deviation has been reported in as many as 75% of patients when only horizontal surgery is used. The postoperative goal is a small-angle esotropia, especially for angles less than 45 PD.\textsuperscript{127, 138} With larger angles, only 40% of eyes remain aligned.\textsuperscript{138} Patients lacking preoperative evidence of central fusional disruption have less than a 10% risk of permanent postoperative diplopia.\textsuperscript{127}

Once the eye is realigned by horizontal rectus muscle surgery, it is common for any oblique overactions, upshoots or downshoots in adduction, and resulting X patterns to resolve within a few weeks after surgery. Similar improvement is noted in the normal eye if previously affected.\textsuperscript{143}

For patients with more than 50 PD of exotropia, the addition of superior and inferior oblique weakening to the horizontal recess-resect procedure reduced the deviation to less than 20 PD in 80% of cases.\textsuperscript{110, 138} No overcorrection has been reported as a result of a four-muscle procedure.

**Complications**

The development of sensory exotropia indicates loss of fusional control. The misalignment has detrimental effects on both sensory and motor function.
In children, a chronic deviation may cause amblyopia to worsen. The resulting deficit in vision, if not treated during the crucial stage of visual immaturity, may be permanent. Chronic strabismus may have a negative effect on the quality of achievable binocularity. In adults, loss of fusion may be accompanied by diplopia or asthenopia. The longer the visual deficit (e.g., cataract) is left untreated, the higher is the risk of central fusional disruption developing.

In both children and adults the angle of exotropia may increase over time, especially if a blurred image is constantly presented to the retina of the deviated eye. When the angle becomes large, a tight LR syndrome may develop secondarily, with accompanying features of oblique overaction or pseudo-overaction, limited abduction, and X patterns. The orbital tissues may become contractured. Once the nonmuscular tissues become tight, the chance of long-term operative success is reduced.

REFERENCES


CLINICAL STRABISMUS MANAGEMENT


Definition

A significant difference in size of the horizontal strabismic deviations in defined positions of upgaze and downgaze is termed A- or V-pattern strabismus. Magee suggested that a difference of 20 PD be used to denote a pattern. Knapp defined the V pattern as a difference equal to or greater than 15 PD from 25 degrees upgaze to 25 degrees downgaze. When the eyes diverge more than 10 PD from upgaze to downgaze, an A pattern is diagnosed; when they converge more than 15 PD from upgaze to downgaze, a V pattern is diagnosed. Downgaze generally is used for near tasks. Because of the need for convergence when viewing objects in downgaze at near, a greater degree of esodeviation is accepted as satisfying the definition for a V pattern. Other authors have used similar values to define these patterns but specify 25 degrees upgaze and 30 degrees downgaze. Parks examines the horizontal deviation in 30 degrees upgaze and 30 degrees downgaze.

A precise nomenclature for pattern strabismus has evolved. Jampolsky suggested using the term tent or "tepee" syndrome. Albert used the terms A and V pattern, which have become the most commonly used notations.

The recognition and treatment of A- and V-pattern strabismus will increase the effectiveness of treating horizontal strabismus.

Historical Perspective

The earliest description of V-pattern strabismus was given by Duane in 1897. He described a patient with bilateral superior oblique (SO) palsy and esotropia in downgaze. Despite this observation, the importance of measuring ocular alignment in upgaze and downgaze was not recognized and acted on until recently. Lancaster, in a personal communication to Costenbader in 1944, recommended measuring the deviation in upgaze and downgaze, whereas Scobee in 1947, emphasized using versions to detect oblique muscle overaction. Costenbader, Berke, McLean, and Urrets-Zavalia underscored the importance of recognizing pattern strabismus with horizontal deviations in the midline vertical positions of gaze.

Knapp wrote an elaborate essay on the causes and treatment of A and V patterns in 1959. He recommended that the oblique muscle dysfunction that causes A and V patterns be treated by operating on the dysfunctional muscle. A- and V-pattern strabismus without oblique dysfunction is treated by vertically displacing the horizontal rectus muscles. The stated principles are similar to those currently used to manage these patients.

Classification

A and V patterns are classified on the basis of the deviation in primary gaze. A-pattern esotropia refers to an esodeviation in primary gaze in which the eyes have increased convergence in upgaze and relative divergence in downgaze (Fig. 14–1). A-pattern exotropia is an exodeviation in primary gaze that increases in downgaze and is reduced in upgaze (Fig. 14–2). V-pattern esotropia is an esodeviation...
with greater convergence in downgaze than in upgaze (Fig. 14–3). V-pattern exotropia is one featuring greater divergence in upgaze than downgaze (Fig. 14–4). It is possible to have an A or V pattern with orthotropia in primary gaze.

Some patients have divergence of the eyes in both upgaze and downgaze when compared with ocular alignment in the primary gaze position, a condition described as an X pattern (Fig. 14–5). This condition is common in patients with combined SO and inferior oblique (IO) overaction and is frequently associated with exotropia. A lambda (λ) pattern is a variant of the A pattern in which the eyes diverge in downgaze but remain horizontally comitant in upgaze and primary gaze (Fig. 14–6). This infrequent pattern may be seen in patients with bilateral SO overaction. A Y pattern is present when the eyes diverge in upgaze but remain horizontally comitant in primary gaze and downgaze (Fig. 14–7). This pattern, also encountered infrequently, has been associated with co-contraction syndrome. A T pattern can also be seen as a variation of the Y pattern. The eyes remain orthotropic until they reach the extreme upgaze and abduct in this position (Saunders RA, personal communication, 1996).

**Prevalence**

The coexistence of an A or V pattern with horizontal strabismus is seen in 12.5% to 50% of cases. However, a study of vertical incomitance in a South American population revealed an A or V pattern in only 0.28% of children with any oculomotor disturbance. In a population of patients with abnormal head posture, 9% had a significant A or V pattern. An A pattern was found in 31% of patients with strabismus and spina bifida. An estimated one in five patients with strabismus may be expected to have an A or V pattern. The prevalence of the various patterns observed in several populations of strabismus patients is shown in Table 14–1.
Etiology

HORIZONTAL MUSCLE OVERACTION

Horizontal rectus muscle overaction was initially believed to be responsible for vertical gaze inconstancy in patients with A and V patterns. In V-pattern esotropia, overaction of the medial rectus (MR) muscles was suggested as the cause of increased convergence in downgaze. Overaction of the lateral rectus (LR) muscles was held responsible for increased divergence in upgaze. Recession without displacement of the rectus muscles was accordingly recommended for treating A and V patterns.

Electromyography has shown that the electrical activity of the horizontal rectus is similar in patients with pattern strabismus and those with comitant horizontal deviations. Innervation of the horizontal extraocular muscles was altered only in the deviating eye during vertical eye movements; the fixing eye showed no change. Breinin concluded that the change or inconstancy in horizontal alignment on vertical gaze is not caused by abnormal innervation of the horizontal rectus muscles. At this time, there is no convincing evidence supporting dysfunctional innervation of the horizontal rectus muscles as an exclusive cause of A or V patterns.

VERTICAL RECTUS MUSCLES

Weakness of the inferior rectus (IR) and superior rectus (SR) muscles was considered a possible cause of the inconstancy seen in pattern strabismus. An A pattern is noted with an underacting IR and compensatory overaction of the yoke SO. A V pattern occurs with an underacting SR and overacting IO. This hypothesis was applied when nasal and temporal displacement of the vertical rectus muscles was used to treat A and V patterns. For example, transposition of the insertion of the IR temporally is used for V-pattern esotropia. Although these recommendations have some limited practical value in managing select forms of A- and V-pattern strabismus, most practitioners have abandoned the theory that vertical rectus muscle dysfunction is the principal cause of A- or V-pattern strabismus.

OBLIQUE MUSCLE OVERACTION OR UNDERACTION

Knapp was one of the first authors to suggest dysfunction of the oblique muscles as the principal cause for A and V patterns. These patterns may exist without overaction of the oblique muscles, although they are less common. A patterns generally are associated with overaction of the SO muscles and underaction of the IO muscles, and V patterns are associated with IO muscle overaction or relative weakness of the SO muscles. Not all cases, however, follow this rule. Paradoxical IO muscle overaction may occur with A-pattern esotropia. A role for the oblique

Figure 14–3. Patient with V-pattern esotropia. A, Preoperative esotropia is 20 PD in primary gaze that increases in downgaze. Note inferior oblique overaction when patient looks to right and left, right and up, and left and up gazes. B, Postoperative series after bilateral medial rectus recession and bilateral inferior oblique myectomy. Note improved alignment in vertical gazes and primary gaze and improved rotations of the inferior oblique muscles.

Figure 14–4. Patient with V-pattern exotropia and overacting inferior oblique muscles. Exotropia in primary gaze increases in upgaze and decreases in downgaze. Overacting inferior oblique muscles also show abducting component and increased elevation in adduction. Failure to identify the abducting component may mask the diagnosis of inferior oblique overaction.
Figure 14-5. Patient with X-pattern exotropia. The eyes are relatively well aligned in primary gaze with small exotropia of 10 PD. The eyes diverge both in upgaze and downgaze from the primary position.

Figure 14-6. Patient with lambda (λ) pattern. No deviation is seen in primary and upgaze. In downgaze, the eyes diverge with overaction of both superior oblique muscles. (Courtesy of Joseph L. Demer, MD, PhD, and Robert Clark, MD.)

Figure 14-7. Patient with Y-pattern exotropia. The eyes are orthotropic in primary gaze and downgaze. In upgaze, the eyes diverge. In up and lateral gaze right and left, the inferior oblique muscles are overacting.
Table 14–1. Prevalence of A- and V-Pattern Strabismus (Expressed as Percent)

<table>
<thead>
<tr>
<th>Pattern</th>
<th>Deviation</th>
<th>Costenbader'</th>
<th>Knapp&quot;</th>
<th>Scott'</th>
<th>von Noorden and Olsen&quot;</th>
</tr>
</thead>
<tbody>
<tr>
<td>V</td>
<td>ET</td>
<td>41</td>
<td>37</td>
<td>5</td>
<td>24</td>
</tr>
<tr>
<td></td>
<td>XT</td>
<td>23</td>
<td>30</td>
<td>25</td>
<td>30</td>
</tr>
<tr>
<td>Total V</td>
<td></td>
<td>64</td>
<td>67</td>
<td>30</td>
<td>54</td>
</tr>
<tr>
<td>A</td>
<td>ET</td>
<td>25</td>
<td>17</td>
<td>48</td>
<td>19</td>
</tr>
<tr>
<td></td>
<td>XT</td>
<td>11</td>
<td>9</td>
<td>22</td>
<td>27</td>
</tr>
<tr>
<td>Total A</td>
<td></td>
<td>36</td>
<td>26</td>
<td>70</td>
<td>46</td>
</tr>
</tbody>
</table>

ET, esotropia; XT, exotropia.

*Knapp's series does not add up to 100%. Ten patients (7%) had an A or V pattern but were orthotropic in the primary gaze position.

muscles in causing A- and V-pattern strabismus is also supported by the observation that surgical correction of the appropriate oblique muscle dysfunction is effective in correcting these patterns.

ANATOMIC FACTORS

The configuration and rotation of the orbit appear to play some role in producing A and V patterns. Patients with the syndromes of Crouzon, Apert, and Pfeiffer have shallow orbits, so that the angle between the visual axis and the insertion of the IO and SO tendon is increased. This decreases the abducting ability of the SO and IO muscles in downgaze and upgaze, respectively. Outward or exocyclorotation of the bones of the orbit may cause excyclorotation of the globe, which in turn causes the MR insertions to be displaced superiorly. This gives the MR muscles a slight elevating effect in addition to their adducting force. Similarly, the LR will serve not only as an abductor but also as a depressor. These abnormal force vectors have been suggested as possible causes of A and V patterns (see also Chapter 30).

Coronal computed tomography was used to study the relationship of horizontal and vertical rectus muscles in patients with and without a V pattern. Rotations of the globe and rectus muscles and a displaced orientation of the position of the rectus muscle insertions were related more to age, and not significantly associated with V syndromes. Clark and colleagues proposed that upward displacement of pulley systems around the LR may cause a V pattern and downward displacement an A pattern.

Anomalies of the external facial configuration have long been associated with cyclovertical muscle dysfunction and A- and V-pattern strabismus. IO overaction was correlated with downward slanting (antimongoloid) palpebral fissures, malar hypoplasia, and an S-shaped contour of the lower eyelid (Fig. 14–8). SO overaction is associated with mongoloid obliquity of the palpebral fissure, well-developed cheek bones, and upward displacement of the lateral canthus. In a Bolivian-based population study, esotropia combined with mongoloid-oriented eyelid fissures was associated with overacting SO muscles, whereas exotropia correlated with IO overaction. With orthotropia in primary gaze, IO overaction and a V pattern were more common. Malar hypoplasia was associated with overacting IO muscles and exotropia. Malar hypoplasia and esotropia suggest a tendency toward underaction of the IO muscles.

On the basis of orbital and facial anthropometric data, some authors regard facial types to be unreliable predictors of the expected pattern. Others who studied acquired conditions such as hydrocephalus thought that the anterior displacement of the trochlea noted with bossing of the frontal bones could cause the commonly associated A pattern. In a study of patients with spina bifida in which some had normal ocular motility and others a large A-pattern strabismus, computed orbital tomography showed no difference in orbital bone structure, the length of the SO muscle, or the position of the trochlea. The dorsum of the midbrain exhibited a "beaking" defect in its rostral section that correlated with an acquired A pattern. This deficit is in an area of the brain stem associated with upgaze. It was speculated that the absence of tissue in this area causes a relative reduction of inhibitory fibers to the upgaze center, resulting in downgaze with overaction of the SO muscles and an A pattern.

SAGITTALIZATION OF THE OBLIQUE MUSCLE INSERTIONS

Gobin hypothesized that sagittalization of the oblique muscles might be a plausible cause of A and V patterns. Alterations of the angle of the insertion of the oblique muscles with the visual axis can reduce cyclorotating and abducting forces but will increase their vertical function (Fig. 14–9).
Normal sup. oblique position

Figure 14-9. The globe is viewed from a superior position. The angle of the reflection of the superior oblique tendon with the visual axis will increase or decrease its abducting force. A more acute angle (α) will reduce abduction and incyclorotation but increase depression.

SENSORY DEPRIVATION

Guyton and Weingarten hypothesized that poor binocular function may cause some forms of A and V syndromes. This is supported by the parallel observation that many patients with an A or V pattern exhibit poor binocular function. Deficient or absent fusion is commonly associated with exocyclotorsion of the globe. This effect is usually bilateral and often asymmetric. Kushner also discussed the effect that torsion of the globe has on horizontal rectus function in upgaze and downgaze. With excyclorotation, the MR becomes a partial elevator whereas the SR has a reduced elevating function. This rotation produces the clinical appearance of IO muscle overaction. With time, sarcomeres of the IO muscle become reduced secondary to shortening of this muscle. The muscles may then overact because of their shortened or contracted fibers.

Diagnosis

Most A and V patterns are not evident at birth. This may be due to difficulty in observing the relatively small changes from upgaze to downgaze in a patient with large-angle esotropia or to incompletely developed following movements that do not permit smooth pursuit of targets into far upgaze and downgaze positions.

Those at high risk for having an A- or V-pattern strabismus include patients with craniofacial anomalies, in particular, the craniosynostoses (A or V), spina bifida (A), fourth cranial nerve palsy resulting in underaction of the SO muscle (V), and infantile esotropia (V). Patients with mongoloid and antimongoloid lid fissures, patients with thyroid disease who have unilateral or bilateral IR recessions (A), and those with aberrant regeneration of the third cranial nerve are also at risk. Patients with thyroid disease are especially susceptible after large recession procedures of the IR muscle. This will weaken the contribution of the muscle to adduction and increase innervation to the yoke SO in downgaze. In third cranial nerve palsy, innervation of the ipsilateral IO muscle directed to the MR muscle causes convergence of the affected eye in upgaze (Fig. 14–10). It can also produce a V pattern if regrowing fibers to the IR innervate the MR, producing esotropia on attempted downgaze.

EVALUATION

The age of the patient at the time the A or V pattern is recognized usually correlates with the severity of vertical incomitance. Some patients may have their strabismus recognized before 1 year of age. Of 421 patients with A and V patterns reported by Costenbader, 58% had an age at onset of 12 months or younger. Supporting Guyton’s hypothesis, 26% of these patients had a visual acuity of 20/200 (6/60) or less in one eye. Eleven percent of patients may have an associated abnormal head posture. A pattern may also be acquired after age 2 years. V patterns may be related to acquired fourth cranial nerve palsy. If the patterns are small in magnitude, they may not be recognized until the early school years, when a head posture becomes apparent or reading difficulties are noted. Children with craniofacial anomalies, abnormally oriented eyelid fissures, and severe degrees of oblique muscle overaction or underaction usually are referred for evaluation for other reasons at a young age.

The history should include inquiries about diplopia, asthenopia, head posturing, lid position abnormalities, and abnormal movements of the eyes, especially in extreme positions of gaze. In adults, an occupational history includes an estimate of the occupational need for binocular vision and the potential impact of diplopia.

Visual acuity should be measured and recorded with the best optical correction in place. Anisometropia should be corrected even if the patient does not wear glasses. The preferred eye for fixation should be identified. Amblyopia should be considered and, if present, treated.

Motor Testing

Alternate prism cover testing should be performed with the head held erect in primary gaze using an accommodative target at 20 ft (6 m). The head is tilted downward so that the eyes are positioned in 25 degrees of upgaze and tilted back to place the eyes in 25 to 35 degrees of downgaze. The 35-degree position is used to bring out any deviation in the reading or extreme downgaze position. I prefer obtaining measurements at 20 ft, but they may be taken at near. Measurements may be made either with the head held stable and the fixation target moving or by tilting the head up, down, and into side gazes with the fixation target held constant. The effect of input from the otoliths in quantitating measurements is probably insignificant. Measurements are taken in right and left gaze to document horizontal as well
as vertical deviations. Over several office visits, measurements are repeated in upgaze and downgaze until consistent and stable values are recorded.

In young or uncooperative children, quantitation of the A or V pattern may be difficult; it may require estimation using either the Krimsky or Hirschberg technique. The examiner moves the eyes from upgaze to downgaze and estimates the amount of deviation. When possible, quantitation with the alternate prism cover test is desirable and recommended in different gaze positions. In some uncommon situations the pattern is large and accompanied by a head posture. This may be observed in young children with a V pattern associated with fourth cranial nerve palsy. The pattern may need to be quantified because surgical correction, if necessary, should be performed early and can be effective.

**Pseudo A and V Patterns.** A pseudo V pattern may be seen in patients with accommodative esotropia. This occurs if the correction for the hypermetropic refractive error is not used when assessing a patient with a small degree of hyperopia. With uncorrected hyperopia there is a natural tendency to accommodate in primary gaze and downgaze, as opposed to upgaze, simulating a V pattern.

Similarly, V-pattern strabismus may simulate an increased accommodative convergence/accommodation (AC/A) ratio. In patients with a V pattern, measurements at distance and near will be the same in primary gaze. If the fixation target is permitted to drop below primary gaze at near, an increased AC/A may be simulated. The AC/A ratio in patients with A and V patterns does not change in upgaze or downgaze. In patients who have an increased AC/A ratio and are treated with bifocal correction, the bifocals may interfere with accurate measurements in downgaze at distance. The bifocals should be removed and the distance prescription placed in a trial lens carrier. Using a more practical method, glasses are moved down on the bridge of the patient’s nose when measuring downgaze and moved up when measuring upgaze, making sure that the distance prescription is used to view the fixation target. The change in vertex distance will be insignificant, unless there is a large amount of hyperopia or myopia.

**Examination of Ocular Rotations**

Examination of versions in extreme positions of gaze is recommended, but it is not necessary to measure the deviation in these extreme fields. Versions are tested beyond the recommended limits into the extreme positions of gaze: in the up, down, up and lateral, up and medial, down and lateral, and down and medial gaze positions. This will reveal any overaction or underaction of the oblique muscles.

The interaction between SO and IO dysfunction contributes to the observed pattern. In A patterns, SO overaction is responsible for divergence (abduction) in downgaze whereas IO underaction contributes to decreased abduction in upgaze. In V patterns, IO overaction opens up the deviation in upgaze and SO underaction contributes to the weakened abducting force in downgaze, causing a relative eso-shift. Y patterns are probably more common in patients with IO overaction but no corresponding SO underaction. The normal SO provides adequate abducting force to keep the eyes aligned in downgaze. Lambda (λ) patterns may represent SO overaction without corresponding IO underaction. The normal IO provides adequate abducting force to maintain orthotropia in upgaze.

In the presence of pattern strabismus, overaction of the oblique muscles may have an abducting component that can mask the overaction in the vertical direction, especially in exotropia (Figs. 14–4 and 14–11). In V-pattern exotropia, for example, overacting IO muscles will produce a curvilinear...
counterclockwise movement in the right eye and clockwise motion in the left eye before finally settling in position in upward medial gaze (see Fig. 14–4). In A-pattern exotropia, the same curvilinear movement is seen with overacting SO muscles—clockwise movement of the SO is seen in the right eye and counterclockwise motion in the left. Less commonly, the same phenomenon may be observed in V-pattern deviations (see Fig. 14–11).

**Torsion**

In older children and adults, torsion of the globes should be assessed using the double Maddox rod test and by viewing the fundus with an indirect ophthalmoscope (Fig. 14–12). Detection of torsion with other signs of oblique muscle overaction and underaction has implications when selecting the appropriate surgical procedure. Torsion may also be documented using binocular perimetry, noting displacement in the relationship of the blind spot to the fovea. When available, photographic documentation of fundus torsion is recommended (see also Chapter 4).

**Sensory Testing**

Binocular function is usually decreased in patients with an A or V pattern. Helveston and colleagues hypothesized that these patients have sliding anomalous retinal correspondence when changing from upgaze to downgaze. Worth four-dot testing will sometimes reveal a fusion response at near but seldom at distance. High levels of stereoaucity are usually not expected. Patients with incomplete patterns such as Y and λ patterns tend to have better levels of stereoaucity. Patients with anomalous head posture such as a chin-back or chin-down position may exhibit sensory fusion with their preferred posture but not in the forced primary position or other gaze positions. Patients with a normal posture may have fusion in the primary position but not in downgaze or upgaze, owing to the larger angle of horizontal deviation in these positions. These findings may influence the decision on whether to operate to enlarge the field of binocular vision.

**Treatment**

The management of A and V patterns is principally surgical. Oblique prisms have been tried, but their prolonged use has proved unsuccessful. Small horizontal deviations accompanied by a small A or V pattern may not require any treatment other than correction of the refractive error and treatment of amblyopia.

**INDICATIONS AND TIMING OF SURGERY**

The measurements in young children may be variable owing to lack of cooperation and a short attention span. Patients should be evaluated on several occasions to document the magnitude and nature of the deviation in upgaze, downgaze, and the primary and lateral gaze positions. Refractive errors should be corrected and amblyopia treated with the goal of achieving equal vision. Deviations greater than 15 PD and those considered to interfere with the development of binocular function should be corrected.

Surgery is performed when accurate, consistent measurements have been obtained. If the pattern is detected early in life, correction is suggested to promote the development of some degree of binocular function. Surgery after age 8 years may be associated with troublesome postoperative vertical, horizontal, or torsional diplopia. After this age, children lose the ability to suppress a second image. Careful patient selection is important, especially when treatment for A-pattern exotropia is being considered. Surgery on markedly overacting SO muscles with an A pattern may produce torsion and disturbing cyclo-diplopia, which may be difficult to remedy. This effect can be minimized by selectively weakening only the posterior fibers of the SO.

Treatment of young patients with spina bifida and an A pattern has not produced diplopia as long as surgery is done before age 8 years. With surgery, these patients are able to regain the previous eye alignment and, in some cases, binocularity.

If a significant anomalous head posture is present, correction is indicated to preserve fusion. If there is a small pattern but the eyes are well aligned in primary gaze and only slightly out of alignment in downgaze, correction may be postponed. Early correction is suggested for patients who appear to have a chance of establishing binocular fusion in the functional positions of gaze (primary and reading position).

It is reasonable to recommend surgery to improve motor alignment. In patients with marked overaction of the oblique
muscles who exhibit a large disparity in the deviation from upgaze to downgaze, the appearance may be disturbing. Cautious surgical correction is indicated to preserve the patient’s self-image, foster self-esteem, and permit comfortable, secure eye contact when speaking with others.

Surgical goals must be realistic: (1) correct horizontal and vertical alignment in useful positions of gaze, (2) achieve or regain comfortable single binocular vision, and (3) correct or eliminate any head posture. In the past, staging surgical procedures to correct A or V patterns was recommended. Simultaneous surgery on the horizontal and oblique muscles corrects the horizontal and vertical defects in one procedure. However, additional surgery may be expected, especially in more complex cases.

**SURGICAL STRATEGY**

A strategy must be carefully developed when deciding on corrective surgery for A- and V-pattern strabismus. Because of the complex actions of the cyclovertical muscles in the extreme positions of gaze or when there are coexisting defects in orbital anatomy, muscle function may not be normal. Defects in eyelid position, such as pseudoptosis or true ptosis, and the effect of recessing the vertical rectus muscles on the lid position should be considered. If oblique muscles are significantly overacting or underacting, they should be the target of surgery. If there is no oblique muscle dysfunction, treatment of the pattern by suprplacation or infraplacement of the recessed or resected horizontal rectus muscle can be effective. The MR is always transposed toward the apex of the V (infraplaced) or the A (suprplaced). The LR is suprplaced to correct V patterns and infraplaced to correct A patterns (Fig. 14–13).

The degree and asymmetry of oblique muscle overaction or underaction should be noted. Oblique muscles that are underacting frequently do not respond well to strengthening procedures (Fig. 14–14). In general, it is better to identify an overacting muscle and weaken it than to attempt to strengthen an underacting muscle.

The size of the pattern from upgaze to downgaze determines the number of muscles that require surgery. The effect of surgery on the vascular supply of the anterior segment of the eye should be weighed. Oblique muscles do not contribute to the anterior segment circulation. Patients with 30 to 40 PD of A or V pattern frequently will require surgery on 4 to 6 muscles to correct the misalignment. Patterns of smaller magnitude may be corrected by symmetric surgery on the oblique muscles or by repositioning the horizontal rectus with appropriate vertical displacement of the insertion. Horizontal transposition of the vertical rectus muscles has been advocated for some forms of A- and V-pattern strabismus. Results after this type of surgery are difficult to predict. If fusion is present, the horizontal, vertical, and torsional aspects of the outcome must be considered. For example, temporal displacement of the IR to treat a V pattern will reduce its adducting component and enhance abduction but will also increase the excyclorotating component in downgaze. The SO will attempt to counter this by resisting the increased excyclorotation and possibly overact as a depressor. These procedures should be used judiciously in selected patients who do not have oblique muscle overaction. If there is a horizontal deviation that requires surgery on the horizontal rectus muscles, additional movement of vertical rectus muscles may place the eye at risk for anterior segment ischemia. Whenever possible, surgery on the SR and IR muscles to correct A- and V-pattern strabismus should be considered only after oblique or horizontal rectus surgery has been rejected.

**TECHNICAL CONSIDERATIONS**

Several methods are used to suprplacate and infraplacate horizontal rectus muscles. Weakening oblique muscles that have abducting ability may affect horizontal alignment. Understanding these technicalities will help the surgeon obtain more predictable results, especially when procedures are combined or when concomitant correction of a horizontal deviation is desirable.

**Techniques Used for Horizontal Muscle Transposition**

The location and configuration of the new insertion when offsetting horizontal rectus muscles for managing an A or V
The lax tendon of a patient with Crouzon syndrome and a very large V-pattern exotropia. Tucking of this lax tendon as an isolated procedure will produce little or no effect on correcting the compensatory overaction of the relatively unopposed inferior oblique muscle.

Pattern have been described with several variations (Fig. 14–15). The lower border of the new insertion of the MR may be advanced within infraplacement so that the new insertion remains a similar distance from, or concentric with, the limbus at its upper and lower borders (see Fig. 14–15[1]). Other recommended slanting the rectus muscle without supraplacement or infraplacement. A modification of this technique has been used to treat convergence insufficiency (see Fig. 14–15[2]). The tendon may be suprplaced or infraplaced so that the new insertion is parallel to the old one (see Fig. 14–15[3]). The upper or lower border may be slanted more posteriorly when recessing the MR or LR muscle. For example, to treat V-pattern esotropia, the MR muscle is recessed measuring from the center of the tendon to the old insertion and is shifted downward, and its lower portion is anchored more posteriorly on the sclera than the superior part (see Fig. 14–15[4]). As the anterior portion of the globe moves downward on its axis of rotation, the pull or tension generated by the inferior fibers of the recessed MR is reduced further because the distance between the origin and insertion is reduced more. This further decreases the adducting force in downgaze.

One study has shown that vertical shifting of the horizontal rectus muscle beyond one-half tendon width has no significantly increased effect when treating an A pattern. The size of the pattern correlates better with the effect achieved than the actual amount by which the insertion of the horizontal rectus muscle is vertically displaced.

Superior Oblique Weakening

The effect produced by a tenotomy of the SO tendon will vary depending on the amount of pattern present and whether the tenotomy is performed nasal (more effect) or temporal (less effect) to the lateral border of the SR muscle. Temporal tenotomy may correct more than 40 PD of A pattern, averaging 30 PD of change. Performed nasally, the reduction in pattern ranges from 25 to 45 PD.

The effect of SO tenotomy on the size of the horizontal deviation in primary gaze is controversial. A tenotomy performed temporally produces a predictable esoshift of 8 PD in primary gaze. Others failed to note any esoshift in primary gaze after recession of the SO. SO tenotomy performed nasal to the SR produces an esoshift of 0 to 3.3 PD. Knowing that a small esoshift of the horizontal alignment in primary gaze may follow SO tenotomy helps to achieve more predictable results when combining horizontal rectus surgery with SO surgery.

The weakening effect on the SO tendon can be graded. Dividing the tendon nasal to the border of the SR produces slightly greater weakening than dividing it temporal to the SR. Cutting the attachments between the SO tendon and the undersurface of the SR muscle permits greater slippage of the cut tendon nasally or toward the trochlea (Fig. 14–16). This can produce excessive weakening and may cause late iatrogenic SO palsy. Others maintain that SO tenotomy should be performed cautiously in patients who have fusion. Disturbing diplopia may result, and the procedure is
Figure 14–16. This schematic adaptation from Fink's atlas shows the superior rectus muscle reflected to demonstrate the filamentary attachment of the superior oblique tendon to the superior rectus tendon. If the tendon is cut temporal to the lateral border of the superior rectus, these attachments will persist and the superior oblique muscle will still show some function. These attachments will serve as the new insertion. The function of the superior oblique muscle will be reduced in force and excursion of movement. Severing the tendon nasal to the superior rectus is equivalent to a complete disinsertion because few, if any, attachments will be preserved.

very difficult to reverse. Alternatively, partial tenectomy of the posterior fibers of the SO tendon can be performed to collapse A-pattern strabismus with SO overaction without affecting torsion (see Chapter 35).

Inferior Oblique Weakening

In patients with overacting IO muscles, any of the weakening procedures performed temporal to the lateral border of the IR muscle will eliminate as much as 20 PD of V pattern. There is little, if any, effect on horizontal alignment in primary gaze after weakening the IO.

TREATMENT ALGORITHM

Some general treatment guidelines are outlined for two subgroups of patients with A and V patterns: (1) those without oblique muscle overaction and (2) those with oblique muscle overaction.

A-Pattern Esotropia

This pattern is frequently associated with a chin-back head posture and is often associated with SO muscle overaction. In patients without SO muscle overaction, recession and symmetric suprarelacement of the tendons of the MR muscles by one-half tendon width (5–6 mm) is suggested. This corrects up to 15 PD of A pattern. The amount of collapse of the pattern will depend on its magnitude more than the amount of transposition. Urist performed this procedure with discouraging results. After recession and supravelacement, several patients developed V-pattern exotropia. This early report had insufficient information to make a critical judgment on this procedure because oblique muscle dysfunction and measurements in lateral gazes were not noted.

Patients with A-pattern esotropia and SO overaction require bilateral SO tenotomy and horizontal rectus recessions to correct esotropia in primary gaze. In a patient with fusion and an A-pattern esotropia or exotropia, SO weakening may be achieved by posterior tenectomy in order not to induce unwarranted torsion effects. Patients with A-pattern esotropia and SO overaction are best treated by recession of the LR muscles and infravelacement of the new insertion. I usually infravelace the new insertion one-half tendon width for 12 to 15 PD of A pattern and three-fourths to a full tendon width, combined with posterior slanting of the inferior fibers of the LR, for patients with 18 to 20 PD of A pattern (see Fig. 14–15) Excellent long-term results have been reported using infravelacement of the LR to treat A-pattern exotropia. All patients had their A pattern collapsed to within 10 PD after at least a year.

For patients with A-pattern esotropia and overacting SO muscles, bilateral SO tenotomies will correct 35 to 50 PD of A pattern. SO tenotomy may be effectively combined with symmetric surgery on the horizontal rectus muscles in most cases (see Fig. 14–2). Advancement of the insertion of the SO fibers to a position more anterior on the globe or recession of the fibers has been reported to reduce an A pattern. I have had no experience with either of these techniques.

V-Pattern Exotropia

Patients with V-pattern esotropia usually demonstrate a depressed chin position to maintain parallel alignment of the eyes in upgaze. Recession of the MR muscles without infravelacement will correct approximately 10 PD of V pattern. If combined with infravelacement of the insertions, an additional 15 PD of V pattern may be corrected. This form of V pattern is common in patients with infantile esotropia and patients with bilateral SO palsy. With bilateral SO palsy, weakening of the direct antagonist IO muscles combined with MR recession produces satisfactory results. Patients with IO overaction and infantile esotropia require combined IO weakening with MR recession (Fig. 14–3). If there is profound weakness of the SO muscles on version testing and a V pattern of 40 PD or more, symmetric tucking of the SO tendons may be added to the procedure. The Harada-Ito procedure may be used if there is significant fundus excyclotorsion in the absence of meaningful SO underaction. Weakening of the IO muscles, unlike weakening of the SO muscles, rarely changes the horizontal deviation in primary gaze.

V-Pattern Exotropia

In patients with V-pattern exotropia without IO muscle overaction, recession of the LR muscles with supravelacement
of the superior portion of the transposed tendon, positioning it more posterior to the insertion than the inferior fibers of the tendon, will correct 10 to 15 PD of pattern. This will reduce the pull of the uppermost fibers of the new insertion when the patient is looking up.

In patients with V-pattern exotropia and IO overaction, the IO muscles should be weakened symmetrically and appropriate recession of the LR muscles performed to correct the exodeviation in primary gaze. The amount of horizontal muscle surgery may be based on the size of the exodeviation in primary gaze because an esoshift of the ocular alignment in this gaze is not expected. Horizontal rectus surgery should be performed at the same time as the corrective oblique procedure.

Monocular Surgery

If there is decreased vision in one eye with or without oblique muscle overaction, A and V patterns may be corrected by performing surgery on one eye. Appropriate vertical displacement of the horizontal rectus (medial down, lateral up) for V pattern and (medial up and lateral down) for A pattern is effective. This can potentially cause up to 7 degrees of torsion of the globe. Metz, however, observed only 1.6 degrees mean cyclorotation of the globe.

Y Pattern

Patients with Y esotropia or exotropia usually have IO overaction with evidence of fundus excyclotorsion. Bilateral weakening of the IO muscles will reduce or eliminate the pattern in upgaze. If there is a horizontal deviation of more than 10 PD in primary gaze, recession of the appropriate horizontal rectus muscles should be combined with this procedure. For Y-pattern esotropia with underacting SO muscles, an alternative procedure is graded tucking of the SO tendons, which should reduce esotropia in downgaze. The incyclotorsion induced by the tuck helps to correct excyclotorsion. Iatrogenic Brown syndrome is a concern but usually improves with time.

Lambda (λ) Pattern

Patients with a λ deviation and exotropia usually present with a chin-down head posture to achieve fusion in a slight upgaze position. Bilateral SO tenotomies will reduce the λ pattern if SO overaction is present. However, care must be taken to ensure that surgical weakening is complete on each tendon to produce symmetric results. Asymmetric weakening may cause problematic vertical and torsional diplopia.

Theoretically, it is possible to have a diamond-shaped pattern, with the eyes converging in upgaze and downgaze. This suggests bilateral SO and IO underaction or restriction of the LR muscles. The situation is rarely encountered in clinical practice.

Complications

The results of surgery designed to simultaneously correct A or V pattern and horizontal deviations in primary gaze may be difficult to predict. Planning must take into consider-

DIPLOPIA

Most A and V patterns are treated before the age of visual maturation at about 8 years. It is during this period that fusion develops. If there are impediments to fusion, suppression is easily learned. Diplopia is infrequent. On the other hand, diplopia may be a problem if correction is undertaken after the first decade of life. The patient should be made aware of this risk. The ocular alignment problem must be severe enough to warrant surgical correction if diplopia is a possibility. Vertical and torsional diplopia are of more concern in patients with A-pattern exotropia. Tenotomy of the SO tendon may produce an asymmetric effect and lead to excyclotorsion of the globe. Vertical and torsional fusional amplitudes are able to compensate for small vertical and torsional alignment defects. Corrective strabismus surgery would be difficult to plan and technically difficult to execute.

ANTERIOR SEGMENT ISCHEMIA

If surgery on the horizontal rectus muscles has been performed in patients without oblique muscle dysfunction but a pattern persists, care must be taken to avoid operating on both vertical rectus muscles in the same eye. This places the eye at risk for anterior segment ischemia.

EMERGING OVERACTION OF OBLIQUE MUSCLES

Scott has reported that weakening the IO muscles to treat a V pattern may lead to an A pattern with overaction of the SO muscles many years later. I have seen at least four such cases and have had to weaken the IO tendons to correct this problem. A similar problem may occur after SO weakening procedures that can produce iatrogenic SO palsy with a V pattern.

FAILURE TO DIAGNOSE OR TREAT OBLIQUE MUSCLE OVERACTION

Probably the most difficult complication to correct follows the treatment of an A or V pattern with vertical displacement of the horizontal rectus muscles in the presence of unrecognized oblique muscle overaction. The IO overaction will persist and will sometimes be exacerbated.

HEAD POSTURE

Head postures may be created by asymmetric weakening of the oblique muscles, which can produce hypertropia or ocular torsion. Asymmetric weakening may result from the failure or inability to identify all the fibers of the SO tendon, or missing a segment of muscle when weakening the IO.

Fine-tuning the horizontal alignment in primary gaze and the reading position with prisms is usually unsuccessful, because patients rarely develop enough fusion to maintain the alignment. The appearance and optical distortion caused by prism glasses may be undesirable for small deviations in primary position or downgaze.
Conclusions

The variable response of weakening procedures, coupled with supraveloc readjustment or infraveloc readjustment of the force vector of the horizontal rectus muscles, yields results that are less predictable than those of horizontal rectus surgery alone. Accurate tables are not available to guide the surgeon to the proper amounts of surgery. These procedures are performed infrequently, using variable techniques and for varying causes of pattern. Each case should be assessed individually. Care must be taken to provide for an alternative surgical plan if there is distorted anatomy or absence of the muscle or if technical difficulties are encountered.

Familiarity with the primary, secondary, and tertiary actions of the oblique muscles and facility with techniques for weakening them or altering their direction of action are essential for a successful outcome. Even then, overcorrection and undercorrection of the horizontal deviation in primary gaze and the creation of a persistent vertical muscle imbalance occur more often when correcting A or V patterns than with the management of comitant horizontal strabismus.

Nevertheless, alignment of the eyes can be improved in patients with A- and V-pattern strabismus. The goal should be to reduce the pattern and at the same time correct the horizontal deviation so that the patient can develop some binocular function and, one hopes, achieve fusion with amplitudes. A high level of sensory function is uncommon with A and V patterns. Patients with Y and A patterns seem to have a better opportunity for achieving fusion, as do those with acquired SO palsy causing a V pattern.57

ACKNOWLEDGMENTS

The author would like to thank Professor Ewy Meyer, MD, from the Rambam Medical Center in Haifa, Israel, for her constructive thoughts during the preparation of this manuscript. This chapter was supported in part by a research grant from the Department of Ophthalmology, Children’s Hospital of Pittsburgh, Pittsburgh, Pennsylvania.

REFERENCES

VERTICAL DEVIATIONS
SUPERIOR OBLIQUE PALSY

Superior oblique (SO) palsy is a common motility disorder—the most common cyclovertical muscle palsy encountered by the strabismologist. Most SO palsies seen by the pediatric ophthalmologist are congenital, but ophthalmic practitioners with larger numbers of adult patients encounter many acquired palsies resulting from trauma, inflammation, infection, vascular malformation, infarct, tumor, or myasthenia gravis, as well as iatrogenic cases after sinus, orbital, or neurologic surgery. Although there is usually no genetic predisposition to develop SO palsy, familial cases have been reported.1, 14

Widely varying reports on the etiology of SO palsy have been published (Table 15–1).10, 23, 30, 45, 48, 49, 54 This variation in etiology is related to referral patterns of a particular practice and the observer’s propensity to classify long-standing palsies of no known cause as congenital.

Classification

CONGENITAL

Approximately three fourths of cases of SO palsy are congenital, although not all of them present in childhood. In fact, many cases do not present until adulthood when the patient no longer can sustain a long-standing effort to maintain fusional vergence control of the hyperphoria. Some of these patients may slowly develop large vertical fusional amplitudes of 30 PD or more. Such fusional amplitudes are not specific for congenital palsy, however; many clearly acquired cases with large amplitudes have been reported.

Although cases are categorized as having historical and/or photographic evidence of SO palsy dating back to childhood as congenital, the precise cause of congenital SO palsy is not well understood. Birth, which can be a traumatic event, is not associated with the kind of severe head injury needed to cause SO palsy. Is congenital SO palsy caused by agenesis of the cranial nerve nucleus, damage to the peripheral nerve, or some other neurogenic factor? The answer to this question is unknown.

One of the most interesting recent findings is that, in congenital SO palsy, the tendon is usually lax and abnormally long.21, 32 In some cases it may be misdirected or even absent.16, 17, 38, 55 This finding begs the question of whether the tendon abnormality in congenital SO palsy is secondary to denervation atrophy or is a primary anatomic defect. Certainly, other anatomic defects of the SO are well known:

<table>
<thead>
<tr>
<th></th>
<th></th>
<th></th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>No. of Patients</td>
<td>190</td>
<td>147</td>
<td>657</td>
<td>270</td>
</tr>
<tr>
<td>Congenital</td>
<td>137 (72%)</td>
<td>106 (72%)</td>
<td>79 (12%)</td>
<td>107 (40%)</td>
</tr>
<tr>
<td>Acquired</td>
<td>53 (28%)</td>
<td>41 (28%)</td>
<td>578 (88%)</td>
<td>163 (60%)</td>
</tr>
<tr>
<td>Trauma*</td>
<td>55%</td>
<td>88%</td>
<td>29%</td>
<td>56%</td>
</tr>
<tr>
<td>Tumor*</td>
<td>9%</td>
<td></td>
<td>5%</td>
<td></td>
</tr>
<tr>
<td>Aneurysm*</td>
<td></td>
<td></td>
<td>1%</td>
<td></td>
</tr>
<tr>
<td>Iatrogenic*</td>
<td>23%</td>
<td>12%</td>
<td>33%</td>
<td>39%</td>
</tr>
</tbody>
</table>

*Percent of acquired palsy
they range from the short restricted tendon found in a majority of Brown syndrome patients to the ultimate abnormality—an absent SO tendon.

Not all patients with congenital SO palsy have significant laxity of the affected tendon, but most do. Conversely, although the rule for acquired palsies is symmetric tendon tightness even in unilateral cases, there are exceptions to this rule. Some of these “exceptions” may reflect inaccurate diagnosis of the cause of palsy. We have seen one middle-aged patient who presented with bilateral SO palsy after significant head trauma but at surgery was found to have anomalous insertion of the SO tendons into Tenon’s capsule rather than the sclera—clearly a congenital defect.

If the sole cause of “palsy” in these congenital cases is an anatomic tendon abnormality, then one might expect that the skeletal muscle portion of the SO complex would be normal; this, however, is not the case. Magnetic resonance imaging (MRI) data on patients with both congenital and acquired palsies show small cross-sectional areas of the belly of the involved SO. 

Surgical exploration in two patients with preoperative MRI evidence of muscle atrophy demonstrated lax tendons.

These findings conform to our observations. Figure 15–1 shows the MRI findings in a 35-year-old woman with congenital left SO palsy. Marked asymmetry in muscle size between the affected and normal sides is apparent. Significant left SO tendon laxity, seen at surgery, is shown in the intraoperative photograph in Figure 15–2. Another example of unilateral congenital palsy in an adult is shown in Figure 15–3. The MRI image of a 38-year-old man who sustained bilateral SO palsy after severe head trauma, demonstrating bilateral muscle atrophy 1 year after injury, is shown in Figure 15–4. This patient did not have tendon laxity at the time of his surgery.

In an unpublished series, my colleagues and I performed quantitative MRI analysis in nine patients having unilateral congenital SO palsy. Five adults showed marked asymmetry in SO muscle size, ranging from 33% to 51% of that on the uninvolved side. Four children aged 5 to 13 years showed great variability in the size of affected muscle, ranging from 15% to 149% of those on the normal side. Observing these children many years later should help answer the question of whether the observed “atrophy” of the involved muscle worsens over time. Unfortunately, this will not indicate whether the observed differences in muscle size are due to denervation atrophy or whether some other mechanism such as atrophy secondary to lack of isometric resistance from the lax tendon is responsible. Regardless of the pathophysiology of tendon laxity, it is evident that clinical assessment of...
tendon laxity is helpful in formulating an effective surgical plan.

**ACQUIRED**

Insult to the trochlear nerve may occur anywhere in its long course from its exit in the dorsal midbrain, along the brain stem close to the tentorium cerebelli, through the cavernous sinus, and across the superior orbital fissure into the orbit where it innervates the skeletal muscle portion of the SO. This extended course makes the trochlear nerve especially vulnerable to injury from trauma. Motor vehicle accidents are the most common cause of traumatic SO palsy, but any severe frontal head trauma may injure the nerve(s). A direct blow to the midbrain will likely cause bilateral palsy due to the crossing fibers in the anterior medullary velum emanating from the trochlear nuclei on each side. A contrecoup blow may compress the peripheral nerve against the tentorium, causing unilateral or bilateral palsy. A substantial blow to the head, usually accompanied by loss of consciousness, is required to cause SO palsy in most cases. Other possible causes should be considered when a patient reports only trivial trauma.

**Clinical Characteristics**

**SYMPTOMS**

The SO muscle depresses, intorts, and abducts the eye. Therefore, paresis of this muscle will cause the involved eye to have some combination of hypertropia, exycyclotropia, and, to a lesser extent, esotropia. As a practical matter, the tertiary abducting function of the SO is so minor that the induced esotropia from paresis is clinically significant only in bilateral palsies and mostly in downgaze.

Patients may complain of diplopia, which can be vertical, diagonal, or torsional. If the principal complaint is torsion, bilateral palsy should be suspected. Other common complaints are asthenopia and cervical discomfort (if a compensatory head posture is adopted).

**SIGNS**

A compensatory head posture is the most common presenting sign of SO palsy. Typically, this is a torticollis with the head tilted away from the affected side. Rarely, a paradoxical head tilt toward the affected side may be preferred by a patient who is trying to maximize separation of the annoying second image from the primary image. In bilateral acquired palsy, the typical head posture is chin down to compensate for the induced V pattern in downgaze.

Another very important and commonly overlooked physical finding is the facial asymmetry characteristic of congenital SO palsy. This was first noted by Parks and consists of shortening of the midfacial region between the lateral canthus and the edge of the mouth (Fig. 15–5). In two recent studies, facial asymmetry was found in more than three fourths of patients with congenital palsy. This finding is not specific for SO palsy and is clinically indistinguishable from facial asymmetry seen in children with torticollis dating from infancy secondary to causes such as congenital muscular torticollis or the Klippel-Feil anomaly. This type of facial asymmetry should not be confused with frontal plagiocephaly, an anatomical skull abnormality caused by coronal synostosis, which may also be associated with vertical strabismus—including SO palsy. Children who develop torticollis late in childhood because of dissociated vertical deviation or other disorders do not develop such facial asymmetry. Facial asymmetry in SO palsy is very strong evidence that torticollis dates back to infancy, and it may preclude the need for an expensive and unnecessary neurologic workup.

The cause of facial asymmetry is controversial. Observations in children with muscular torticollis have led some to suggest that asymmetry develops because these children...
habitually sleep with the same side of the face against the pillow. This mechanism seems particularly unlikely in children with torticollis due to SO palsy. Unlike children with muscular torticollis, the tendency for patients with SO palsy to maintain a head tilt disappears when supine. Parental observations of infants with SO palsy confirm that they sleep in any and all possible head positions. Although the exact cause remains uncertain, the midfacial hypoplasia that develops on the dependent side in patients having torticollis from a young age appears to result from the head tilt.

**Diagnosis**

**Clinical Diagnosis**

**The Three-Step Test**

The key to diagnosing an isolated cyclovertical muscle palsy is the three-step test, requiring motility measurements in the primary position, right and left side gazes, and right and left head tilts. The first step excludes half of the eight cyclovertical muscles by incriminating the two depressors of the hypertropic eye and the two elevators of the hypotropic eye. The second step reduces the four remaining muscles to the two whose strongest action is in the field of the greatest side gaze deviation. The third step selects the remaining muscle whose elevating weakness is exposed when its torsional power is called on by the Bielschowsky head-tilt test. (See Chapter 1 for details of the three-step test.)

In addition to the five gaze position measurements required for the three-step test, measurements should be obtained in the upgaze and downgaze positions. Kushner pointed out that measurements in the oblique fields can help identify masked bilateral SO palsy. For surgical planning, the primary, up, down, and both side gazes will suffice.

**Torsion**

Whenever possible, torsion should be subjectively assessed with a double Maddox rod test and objectively assessed by ophthalmoscopy or photography (see also Chapters 1 and 4). A few helpful generalizations regarding torsion: (1) congenital SO palsy frequently has very little or no measurable subjective torsion; (2) acquired palsy invariably has a measurable degree of torsion; and (3) a spontaneous complaint of torsional diplopia or measurable excyclotorsion greater than 10 degrees suggests bilateral SO palsy. In addition, measuring cyclotorsion in downgaze helps magnify the difference between the normal state and SO palsy, as well as between unilateral and bilateral SO palsy, but no inviolable boundaries exist that will distinguish normal from unilateral or unilateral from bilateral SO palsy based on torsion alone.

**Versions**

Three abnormalities in version movements may be observed (Fig. 15–6): ipsilateral inferior oblique (IO) overaction, ipsilateral SO underaction, and contralateral SO "overaction." IO overaction of some degree will be present in most cases. It is usually moderate to marked but, particularly in older patients with small deviations, it may be minimal to absent.

SO underaction is observed only in cases with marked neurogenic paresis or patients with very lax SO tendons. This finding is much less common than ipsilateral IO overaction and is not required to diagnose SO palsy.

Contralateral SO "overaction" is really a misnomer and generally is underappreciated as an important clinical finding. This seems to be due to failure of the paretic eye to infraduct fully in abduction, giving the sound adducting eye the appearance of overdepression. Traction testing at the time of surgery will confirm that the problem is caused by superior rectus (SR) restriction in the paretic eye. This condition is most apparent in long-standing, large hyperdeviations. It is not needed to diagnose SO palsy but its recognition is critical in formulating an appropriate treatment plan when it is present.

**Bilateral SO Palsy**

Bilateral SO palsy should be suspected in a patient with any combination of the following: (1) SO palsy after closed-head trauma; (2) subjective complaints of torsion; (3) objective torsion exceeding 10 degrees; (4) alternating hypertropia on head tilt (e.g., right hypertropia on right head tilt and left hypertropia on left head tilt); (5) V-pattern esotropia; and (6) a chin-down head posture.

**Laboratory Diagnosis**

Most patients with isolated SO palsy do not require an extensive neurologic workup. Because most palsies are congenital, documenting the characteristic facial asymmetry, a review of old family photographs showing a long-standing head tilt (so-called family album tomography [FAT scan]) or simply eliciting a history of symptoms of many years' duration is sufficient to rule out an acute acquired process. Neuroimaging or neurologic consultation may be indicated when a patient has an acquired palsy that cannot be directly linked to trauma or a palsy associated with other neurologic findings.

Brain stem lesions affecting the trochlear nerve are expected to be accompanied by other neurologic signs such as Parinaud syndrome, ataxia, internuclear ophthalmoplegia, Horner syndrome, or other cranial nerve palsies. Damage to the nerve within the cavernous sinus will usually affect other cranial nerves in the sinus and may be painful. An isolated, painless, unilateral fourth nerve palsy may be caused by an aneurysm of the internal carotid or superior cerebellar artery.

Myasthenia gravis should be considered in any acquired motility disorder, although it is a very rare cause of isolated SO palsy. A positive edrophonium (Tensilon) test can confirm this diagnosis.

**Differential Diagnosis**

A complete eye examination emphasizing motility measurements in the appropriate positions of gaze, measurement of torsion when possible, assessment of versions, and the three-step test should provide the correct diagnosis of SO palsy in most cases. It is crucial to remember that the three-step test is reliable only in isolated cyclovertical muscle
palsy. It cannot be applied to restrictive processes such as thyroid ophthalmopathy, blow-out fracture, Brown syndrome, fibrosis of a muscle, or other vertical misalignments such as dissociated vertical deviation and skew deviation. Prior strabismus surgery may also negate the results of a three-step test.

Distinguishing factors of other vertical deviations include the following:

1. **Skew deviation.** Skew deviation is an acquired, usually acute hyperdeviation that may be, but is not necessarily, comitant. It may be confused with acquired SO palsy but is always associated with other neurologic signs referable to the brain stem or cerebellum. Measurable torsion is invariably absent in skew. This may help differentiate it from SO palsy, but some congenital cases of SO palsy may have little or no torsion.

2. **Thyroid-related ophthalmopathy (TRO).** Chronic TRO can usually be identified as a restrictive process in one or both eyes. The inferior rectus (IR) is the most commonly involved muscle; its restriction may lead to an erroneous diagnosis of SO palsy in the fellow eye. Proptosis, lid retraction, or lagophthalmos may be present. Active TRO is associated with chemosis, proptosis, and other signs of orbital congestion. Neuroimaging will show enlargement of the affected muscle belly.

3. **Brown syndrome.** Brown syndrome causes underaction of the ipsilateral IO owing to restriction of the SO—in contrast to IO overaction, which is more typical of SO palsy. The combination of Brown syndrome and SO palsy in the same eye was considered by Knapp to be a class VII SO palsy. This condition is also called the “canine tooth” syndrome in reference to its most common cause—a dog bite in the area of the trochlea.

4. **Primary IO overaction.** Primary IO overaction is frequently seen in association with congenital esotropia, but it may be an isolated finding either bilaterally or unilaterally. It is distinguished from SO palsy by the absence of a primary-position hyperdeviation, a lack of subjective torsion, and a negative Bielschowsky head-tilt test.
Step 1
With the surgeon seated above the patient's head, the eye is grasped diagonally, i.e., at the 2 and 8 o'clock positions on the left eye, and the 4 and 10 o'clock positions on the right eye.

Step 2
In one move the eye is rotated up and in while simultaneously pushing the eye toward the floor. This puts the SO tendon on maximum stretch. The normal left SO tendon shows no change from 7C to 7E as the tendon is maximally stretched. The laxity of the right SO tendon is demonstrated by the further stretching out of that tendon from figure 7D to 7F.

Figure 15-7. A through H, Traction test.
Treatment

NONSURGICAL MANAGEMENT

SO palsy, seen as an incidental finding in a patient without torticollis or symptoms, does not require treatment. Patients with very small, relatively comitant deviations and those who are not surgical candidates for other reasons may gain some relief from prisms. However, the usefulness of prisms is limited by incomitance of the hyperdeviation, seen in most cases, or the V pattern present in bilateral cases. The vast majority of patients presenting to the ophthalmologist with SO palsy do so because of troubling symptoms and usually are surgical candidates.

SURGICAL MANAGEMENT

Knapp, in his landmark paper on the classification of SO palsy, emphasized the importance of the relative magnitude of hyperdeviation in the various fields of gaze. Although some of the specific treatment recommendations in Knapp's original classification are no longer considered optimal, the importance of his contribution to diagnosing and treating SO palsy remains. The message of the Knapp classification may be summarized in the generalization that one should match the fields of greatest deviation to the muscles that exert their strongest action in those fields. To this framework we may add more recent observations regarding the significance of SR contracture, SO laxity, quantification of the SO tuck, and management of torsion. A tailored treatment plan thereby can be drawn up for each patient using a few basic tenets. Formulating the plan assumes that the correct diagnosis has been made and requires information from both the clinical and intraoperative examination.

For unilateral palsy in patients having motility measurements in all fields of gaze, the preoperative information needed includes knowledge of (1) the magnitude of deviation in primary position; (2) relative deviations in up, down, and side gazes; (3) versions, with emphasis on the presence or absence of IO overaction; and (4) SR contracture if clinical signs are present. Intraoperatively, SO tendon laxity is assessed by oblique traction testing and suspected SR restriction is confirmed by traditional traction testing.

Oblique Traction Testing

Intraoperative traction testing of the SO has become an indispensable part of evaluating patients with SO palsy. It is especially relevant for young children in whom precise motility measurements cannot be obtained. Traction testing identifies very lax tendons that should be tucked but, just as important, identifies tendons of normal length that should not undergo this procedure because of the risk of causing Brown syndrome. Even though the majority of SO tendons in congenital palsy have some degree of excess laxity, most do not have the marked laxity that requires a tuck.

Technique. The technique for assessing laxity of the SO tendon is based on the method of "exaggerated forced ductions" of the obliques, described by Guyton for evaluating patients with SO overaction. It has been modified to help emphasize the laxity in these redundant tendons (Fig. 15-7).

Step 1: With the surgeon seated above the head of the supine, anesthetized patient, two toothed forceps are used to grasp the limbus diagonally, that is, at the 2- and 8-o'clock positions on the left eye and the 4- and 10-o'clock positions on the right eye.

Step 2: The eye is rotated up into an elevated, adducted position in the superior nasal quadrant while simultane-
ously pushing the globe down toward the orbital apex. This maneuver puts the reflected tendon on maximal stretch and thereby shows the degree of tendon laxity between its firm attachments to the trochlea and its insertion onto the globe.

**Step 3:** Once the tendon is put on stretch, the eye is moved back and forth (temporally and nasally) while maintaining the tendon taut. This back and forth movement allows the surgeon to "feel" the tendon as a band across which the globe is rocked.

**Step 4:** Steps 1 to 3 are repeated on the fellow eye.

### Important Points Regarding the Traction Test

All normal SO tendons can be easily felt using this technique. Becoming comfortable with the method on normal patients will allow the surgeon to be more aware of what an abnormal tendon feels like. Because there is a range of normal tendon laxity, it is the relative laxity of the tendon compared with its fellow eye that is the determining factor. A subjective grading scale is used to compare the paretic and normal tendons:

- **Grade 1**—Tendon only mildly lax
- **Grade 2**—Tendon obviously more lax than normal
- **Grade 3**—Tendon markedly lax but definitely present
- **Grade 4**—Tendon cannot be felt with certainty. The tendon may be absent, but some tendons are so lax that the end point of the test comes from resistance of other tissues attached to the globe, not the outstretched tendon itself.

In general, grade 4 tendons should always be tucked; grade 3 tendons usually are tucked; grade 2 tendons are rarely tucked; and grade 1 tendons are almost never tucked. The gain from tucking grade 1 or 2 tendons usually is not outweighed by the potential risk of causing Brown syndrome.

This vigorous maneuver will easily tear the fragile conjunctiva of adults. It is generally more valuable in children; and, if done in older adults, locking toothed forceps should be used on the episclera/sclera, not just the conjunctiva.

### Treatment Algorithm

*If the patient has less than a 15-PD deviation in the primary position, one-muscle surgery is probably sufficient (Fig. 15–8). The choice of which muscle to operate on depends on IO muscle evaluation.*

1. **If the IO overacts,** it is weakened by either myectomy or graded recession, depending on the surgeon’s preference. Although good results have been reported with anterior transposition of the IO, this procedure may cause incomitance from the relative restriction to elevation it frequently induces in patients with SO palsy and excellent fusion potential. It is rare to overcorrect a patient with SO palsy and IO overaction by weakening the ipsilateral IO. The only question is whether that procedure alone is sufficient to correct the deviation.

2. **If there is no IO overaction,** the following alternatives exist: (a) If the ipsilateral SR is restricted, recess the SR. As with any form of strabismus, it is generally appropriate and necessary to relieve any restriction that is present. Significant superior restriction, however, is unlikely in patients with small deviations. (b) If there is marked laxity of the SO, tuck the muscle. Marked SO laxity is rarely encountered in patients without IO overaction. (c) Recess the contralateral IR.

*If the primary position deviation is greater than 15 PD, surgery on two or more muscles will be necessary to correct the deviation. Typically, the ipsilateral IO would be weakened, with the second muscle chosen based on the following: (1) ipsilateral SR recession if this muscle is restricted, (2) ipsilateral SO tuck if there is marked laxity of the tendon,*
(3) contralateral IR recession, or (4) a combination of these techniques if multiple procedures are appropriate.

In cases of contralateral SO “overaction,” a tenotomy or other weakening procedure should never be performed on the offending SO. This was originally recommended by Knapp for treating class IV and V SO palsy but has not withstood the test of time; recession of the ipsilateral SR should be done instead. Weakening the contralateral SO in a case of unilateral SO palsy can result in bilateral SO palsy that is very hard to correct.

In infants and toddlers with torticollis due to SO palsy, it is not possible to obtain motility measurements in different gaze fields. This has caused some ophthalmologists to delay surgery for years until precise measurements can be made. I believe delay is unnecessary. There is the possibility of progressive loss of fusion in certain gaze fields, as well as potential adverse effects on the child’s facial development.

Theoretically, developmental facial asymmetry should be prevented by early surgery to correct the torticollis. This supposition has been borne out in many children with congenital SO palsy who are operated on at a young age. If head posture is surgically corrected before facial asymmetry is apparent, it does not develop later.

The failure rate when correcting torticollis in infants with SO palsy is high after IO weakening alone. This is where use of the intraoperative traction test of SO laxity is most helpful. If the tendon is markedly lax, as it usually is in infants with torticollis, an SO tuck should be performed in addition to the IO weakening. If the SO is not significantly lax, only the IO should be operated on.

**How Much to Tuck?** One cannot determine preoperatively how many millimeters of tendon should be tucked. An intraoperative assessment of tendon laxity is required. The endpoint of the tuck is a feel on traction testing that is the same as on the normal side. It is not necessary to create a Brown syndrome to achieve an effect if the ipsilateral IO is weakened at the same time.

**Absent Superior Oblique.** The presence of a horizontal deviation and/or amblyopia in patients with congenital SO palsy should alert the surgeon that the SO tendon may be absent. The traction test can noninvasively prove that the tendon is present. However, if the test suggests that the tendon may be absent, surgical exploration is necessary to confirm the diagnosis. There are times when the SO tendon is so lax that it cannot be appreciated by the traction test. If the tendon is absent, I empirically recess the ipsilateral SR by a moderate amount in addition to IO weakening as the first procedure.

**Bilateral Superior Oblique Palsy.** Bilateral acquired SO palsy is typified by a large degree of excyclotorsion, a small primary-position deviation, and V pattern with increasing esotropia in downgaze.

Large amounts of excyclotorsion (more than 10 degrees) require a procedure aimed specifically at reducing it. Bilateral IO weakening is not sufficient. Harada and Ito described splitting the SO and advancing the anterior half of the tendon. The anterior fibers are primarily responsible for the torsional action of the SO. The primarily vertical-acting posterior fibers are left undisturbed. A modification of the Harada-Ito procedure advances the anterior SO fibers to the superior border of the lateral rectus 8 mm posterior to the lateral rectus insertion. Use of an adjustable procedure also has been described. In addition to the torsional procedure, recession of the yoke muscle—the contralateral IR of each eye—is helpful. If residual torsional diplopia persists in downgaze, nasal transposition of one or both IR muscles may be curative.

If the amount of excyclotorsion is less than 10 degrees, a Harada-Ito procedure need not be performed. Surgery should be undertaken to correct the V pattern and/or diplopia on side gazes. It includes IO weakening on one or both sides and bilateral IR recessions. Recessing both IRs will help the V pattern in downgaze and may be done asymmetrically to correct any primary-position deviation. For example, for 5 PD of right hypertropia in primary position, one might recess the left IR 5 mm and the right IR 3 mm. If there is no significant vertical deviation in primary position, the medial rectus muscles can be transposed one-half to a full tendon width to correct the V pattern, and this may be combined with a recession if there is more than 15 PD of esotropia in primary gaze.

**RESULTS**

Using the treatment algorithm just described has significantly altered the frequency of various procedures used to treat SO palsy. A 1976 report from Indiana University described a series of 106 patients with SO palsy, 65% of whom had at least an SO tuck as part of the surgical plan. This high rate of SO tuck, coupled with less selective criteria for performing it, resulted in 17% of patients requiring takedown of the tuck because of troublesome Brown syndrome.

In a 1996 series of 190 patients from the same institution using the treatment scheme described in this chapter, 171 patients (90%) had IO weakening as part of their operative procedure. Only 26 patients (13%), all with unilateral SO palsy, had enough tendon laxity to warrant an SO tuck. Another 15 patients (7%) had a Harada-Ito procedure for excyclotorsion. This management resulted in a 92% success rate, with success defined as the resolution of symptoms that caused the patients to seek treatment. Seventy-six percent had no deviation in the primary position. There were no cases of postoperative Brown syndrome requiring reoperation.

**Complications**

**OVERCORRECTION**

Overcorrection, converting a hypertropia into even a very small hypotropia in the affected eye, should be avoided. Patients who have built up fusional vergence amplitudes in the direction of their preoperative deviation are usually comfortable with an acceptably small residual deviation (undercorrection). They generally do not have much fusional amplitude in the opposite direction and may be miserably diplopic if the affected eye is made hypotropic.

**BROWN SYNDROME**

Excessive SO tuck will produce a Brown syndrome and diplopia, especially in upgaze. This complication may be avoided by carefully selecting patients based on traction.
testing. Only those with marked laxity of the SO tendon should have it tucked. Even among these patients, the endpoint of the tuck should be to make it feel like the normal tendon of the fellow eye. Tendons of normal length, such as those in acquired palsies, are never tucked because even a small tuck can cause Brown syndrome in these cases. Although it is true that some excessive tightness of a tucked tendon will relax over time, this is a bothersome and unnecessary problem when alternative muscles may be operated on with no such risk (see also Chapter 41).

**SUPERIOR OBLIQUE MYOKYMI A**

A clinical entity characterized by uniocular paroxysms of small-amplitude, high-frequency rotary nystagmus was first described by Duane in 1906 and termed unilateral rotary nystagmus. Hoyt and Keane gave the condition its present name, SO myokymia.

The etiology of isolated SO myokymia is unknown. There is some evidence that it is a response to the regeneration of damaged trochlear nerve axons. Lee reported SO myokymia in two patients after recovery from SO palsy and inferred that aberrant regeneration may be the cause. Electromyographic studies showed an independently firing motor unit under the control of the trochlear nerve nucleus. Two patients with SO myokymia whose affected SO muscle was smaller than the fellow normal SO have also been reported. One of these patients had coexisting traumatic SO palsy, which may have caused denervation atrophy.

These findings suggest, but certainly do not prove, that relatively minor damage to the trochlear nerve may be enough to cause regeneration and consequently SO myokymia, as well as some atrophy of the SO muscle. The more marked damage needed to cause SO palsy and marked muscle atrophy may or may not be accompanied by pathologic regeneration. This is supported by reports of SO myokymia occurring after relatively minor head trauma. 

A variation of the Harada-Ito procedure, in which the anterior half of the SO tendon is transposed nasally to the globe. A standard tenotomy of the SO is not sufficient in the long term because the cut tendon will invariably reattach to itself or somewhere else on the globe through the periorcular tissues. For similar reasons, a silicone spacer placed in the tendon to weaken it would likely be ineffective.

**Clinical Features and Diagnosis**

Patients with SO myokymia complain of intermittent bursts of torsional and/or vertical diplopia, or a “shimmering” of vision. The episodes last from several seconds to several minutes. Specific stimuli of these episodes cannot usually be identified, although fatigue and stress seem to be exacerbating factors.

The diagnosis is made from the characteristic history of uniocular shimmering or intermittent rapid torsional or vertical diplopia and by documenting the characteristic eye movements. A rapid, small-amplitude torsional nystagmus may be observed grossly, but the movements may be so subtle that slit-lamp observation is necessary. The eye movements are not discernible during periods of symptom-free remission.

In the absence of any other neurologic complaints or findings, neuroimaging is generally unrewarding and therefore not indicated. If such findings are present, however, including contralateral SO palsy, neuroimaging is indicated with emphasis on the midbrain.

**TREATMENT**

**Nonsurgical Management**

No treatment is necessary if the patients’ symptoms are not bothersome. In the absence of any other neurologic signs or symptoms, a reassuring explanation of the cause of the symptoms and the benign nature of the problem may suffice. If the patient is significantly troubled by the symptoms, both medical and surgical options are available.

Numerous systemic medications including carbamazepine, baclofen, benzodiazepines, and beta-blockers have been tried. Although some patients have gained some benefit from one or more of these drugs, systemic treatment is generally disappointing. In addition, significant side effects frequently limit their prolonged use. Topical beta-blockers including betaxolol have had short-term success in anecdotal reports. They cause minimal systemic side effects, but their long-term usefulness has yet to be determined.

**Surgical Management**

If medical treatment fails to control symptoms or is not tolerated, surgery may be indicated. Successful surgery requires removing any attachment of the affected SO to the globe. A standard tenotomy of the SO is not sufficient in the long term because the cut tendon will invariably reattach to itself or somewhere else on the globe through the periorcular tissues. For similar reasons, a silicone spacer placed in the tendon to weaken it would likely be ineffective.

A large portion of the SO tendon needs to be removed (tenectomy) to lower the chance of reattachment to the globe. This procedure invariably causes an SO palsy, making prophylactic IO weakening necessary at the same time. A lasting symptom-free postoperative course has been reported after this procedure in a small number of patients.

Some patients required a second procedure to treat residual diplopia.

A variation of the Harada-Ito procedure, in which the anterior half of the SO tendon is transposed nasally to effectively weaken the torsional action of the muscle, has shown promise. In 1 year of follow-up the patient who underwent this procedure remained free of symptoms.
Complications

The principal complication of surgery for SO myokymia is residual SO palsy. The principles of SO palsy management described earlier in this chapter are not applicable, because further manipulation of the tenectomized SO is usually not beneficial. Assuming that the ipsilateral IO has already been weakened, further surgery should be directed at recessing the contralateral IR, with a nasal shift of one or both IRs if residual excyclotorsion is significant. The contralateral IR muscle is the yoke to a parietal SO.

REFERENCES

Historical Perspective

Inferior oblique (IO) palsy is the least common isolated muscle palsy seen in ophthalmology\textsuperscript{7, 9, 18}; few cases have been described in the literature. White and Brown,\textsuperscript{19} in a 1939 study of 1955 patients with ocular muscle anomalies, described 20 patients with IO underaction, but this group undoubtedly included cases of the superior oblique (SO) tendon sheath syndrome that Brown\textsuperscript{3} described in 1950. In 1965, McNeer and Jampolsky\textsuperscript{8} reported patients having underaction of the IO with overaction of the ipsilateral SO and free forced ductions. In 1977, Scott and Nankin\textsuperscript{16} reported a series of patients with IO palsy as strictly defined by the criteria popularized by Parks\textsuperscript{10} for cyclovertical muscle palsy: free forced ductions in adduction and improvement of elevation in adduction on duction testing compared with versions. Subsequently, Olivier and von Noorden,\textsuperscript{9} Frey,\textsuperscript{4} and Pollard\textsuperscript{11, 12} all reported series of patients having IO palsy according to the same criteria.

Epidemiology and Risk Factors

The prevalence and incidence of IO palsy are unknown. Pollard\textsuperscript{12} estimates that, on average, 1.4 patients with IO palsy are seen in his referral clinic each year. Risk factors have not been identified. IO palsy may affect any age group, and there is no sex or racial predilection.

This palsy is usually an idiopathic condition, although congenital, vascular, and traumatic causes have been proposed.\textsuperscript{12} In a 1986 series of patients having IO palsy, the benign nature of this entity was stressed.\textsuperscript{11} However, treatable illnesses such as syphilis, stroke, and myasthenia gravis can potentially cause IO palsy.\textsuperscript{11} No genetic cause has been demonstrated, but presumed congenital cases have been described.\textsuperscript{12, 14}

Clinical Characteristics

CLASSIC APPEARANCE

Patients present with a vertical deviation. Associated features include diplopia, a head tilt to the side of the palsy, and/or a face turn away from the side of palsy. The vertical deviation is a hypotropia on the side of the palsy if the patient fixes with the uninvolved eye (Fig. 16–1A) or a hypertropia on the opposite side when fixing with the involved eye (secondary deviation) (Fig. 16–2A). In either case the deviation worsens with horizontal gaze into the field of action of the affected IO, or with tilt toward the side opposite the paretic eye, thereby fulfilling the three-step criteria for IO palsy (see Fig. 16–2A and B). Elevation in adduction is impaired on version testing but improves on monocular duction testing (see Fig. 16–2C). There is no restriction to elevation in the adducted position on forced duction testing. SO overaction is usually present as well.

CLINICAL PRESENTATIONS

Patients with IO palsy may present with a hypertropia on the side opposite the palsied oblique if they prefer to fix with the palsied eye. Fixation with the palsied eye requires greater innervation to the palsied IO and, according to Hering's law, more innervation of the contralateral superior rectus (SR) as well. Therefore, secondary deviations tend to be larger than the primary ones. Although a secondary deviation may be confusing, three-step testing may be relied upon to isolate true palsy to the involved IO.

Two subtleties in the diagnosis of IO palsy deserve clari-
Figure 16-1. Group 1. A. Right inferior oblique palsy with right hypotropia. The left eye is used for fixation. Significant right superior oblique overaction and right inferior oblique underaction cause the hypertropia to increase in left gaze. However, there is little deviation in right gaze, that is, out of the field of action of the affected inferior oblique. B. Postoperative results after superior oblique tenotomy alone reduced the hypertropia in primary and left gazes and improved oblique dysfunction.
Figure 16–2. Group 2. A and B, Right inferior oblique palsy with left hypertropia. The right eye is used for fixation. The three-step test is fulfilled for inferior oblique palsy. Significant right superior oblique overaction and right inferior oblique underaction causes the hypertropia to increase in left gaze. Also, there is significant hypertropia in right gaze, that is, out of the field of action of the palsied inferior oblique. C, Duction better than version. Left photograph shows underaction of the right inferior oblique on version. Right photograph shows improved action on duction testing. D, Postoperative results: right superior oblique tenotomy combined with left superior rectus recession was required to improve the hypertropia in primary, right, and left gazes. Note oblique dysfunction is improved.

Diagnosis

CLINICAL EXAMINATION

There are three key features on clinical examination that confirm the diagnosis of IO palsy. Three-step testing for isolated cyclovertical muscle palsy must be positive for IO
palsy. The vertical deviation worsens with gaze and tilt away from the affected eye. Gazing away from the affected side places the IO in position for maximal vertical action. The underaction of a palsied IO becomes more prominent, resulting in a greater vertical deviation. Tilting away from the side of the affected palsy places the affected eye in exocyclotorsion. Because attempted exocyclotorsion is achieved primarily by the IR when the IO is palsied, this muscle has greater action and causes more vertical deviation that is unopposed by the usual depressing action of the IO.

The second key feature is impaired elevation of the affected eye in adduction on version testing, which improves on duction testing (see Fig. 16–2). Finally, free forced duction to elevation in adduction must be documented. Laboratory testing of IO muscle function, such as active force generation and saccadic velocity testing, is not helpful.

The fusional status of patients with IO palsy falls into a spectrum comprising diplopia, gross stereopsis, and bifoveal fixation. The majority of patients exhibit some degree of fusion with an abnormal head position, and 20% to 40% have bifoveal fixation.\(^2\),\(^3\),\(^7\) Few patients complain of torsional diplopia, but exocyclotorsion is found in a majority of cases using double Maddox rod testing or indirect ophthalmoscopy.\(^2\),\(^3\),\(^7\)

Patients whose IO palsy is believed to be caused by stroke, myasthenia gravis, or tertiary syphilis should undergo appropriate diagnostic testing. Magnetic resonance imaging, edrophonium (Tensilon) testing, or serum testing for syphilis may be indicated.

### DIFFERENTIAL DIAGNOSIS

The differential diagnosis includes primarily SO palsy, Brown SO syndrome, monocular elevation deficiency, and primary bilateral SO overaction (Table 16–1). Other possibilities include ocular myasthenia, Graves disease, progressive external ophthalmoplegia, orbital blow-out fracture, and other restrictive conditions.

IO palsy is differentiated from SO palsy using the criteria established by Parks.\(^16\) In both IO and SO palsy the vertical deviation worsens in the horizontal field of gaze where the primary action of the obliques is vertical, that is, in gaze away from the paretic eye. Overaction of the unaffected oblique muscle is often noted. SO overaction occurs in IO palsy, whereas IO overaction is seen with SO palsy. In either case, head tilting confirms the diagnosis. With IO palsy the deviation worsens with tilting away from the paretic eye, because extorsion is impaired. In SO palsy, the deviation worsens with tilt toward the paretic eye because intorsion is impaired.

Differentiating IO palsy from Brown SO syndrome relies on comparing the action of the IO on version versus duction testing, as well as forced duction testing. An ability to elevate the paretic eye in adduction effectively rules out Brown syndrome, whereas restricted elevation in adduction—both voluntarily and on forced duction testing—confirms it. An A-pattern strabismus is seen with IO palsy, whereas a V pattern is seen in Brown syndrome, especially with bilateral involvement. SO overaction is associated with IO palsy but usually is not found in Brown syndrome. A history consistent with congenital or acquired Brown syndrome is supportive; it may include deviation since birth, orbital trauma, previous strabismus surgery, and inflammatory diseases.

Double elevator palsy is the inability to elevate the eye in both adduction and abduction due to concomitant paresis of the SR and IO muscles, with or without restriction of the IR. Hypotropia on the affected side may be associated with ptosis or pseudoptosis. A chin-up head position may be adopted to maintain fusion and binocular vision. If there is no anomalous head posture, amblyopia has probably developed. The cause is thought to be a supranuclear lesion affecting both the IO and the SR, restriction of the IR, or both (see Chapter 20).

Bilateral primary SO overaction may also be confused with IO palsy. An A-pattern strabismus with SO overaction may mimic IO palsy. Head tilting will not cause any difference in the amount of deviation in primary SO overaction, whereas IO palsy exhibits vertical deviation with increased head tilt away from the affected eye and reversal of the vertical deviation if the IO palsy is bilateral.

Ocular myasthenia may present as an isolated extraocular muscle palsy but is distinguished from IO palsy by the variable course of diplopia and by prism measurements.

| Table 16–1. Differential Diagnosis of Inferior Oblique Palsy |
|---------------------------------|------------------|------------------|------------------|------------------|
| Inferior Oblique Palsy          | Brown Syndrome   | Superior Oblique Palsy | Monocular Elevation Deficiency* |
| Inferior oblique action         | Underaction      | Apparent underaction: same on duction and version | Underaction: same on duction and version |
|                                 | on version improves on duction | Inferior oblique overaction | Inferior oblique overaction |
| Elevation                       | Limited in adduction | Limited in adduction | Variable |
| Forced duction                  | No restriction   | Restriction         | Limited in primary gaze, abduction, and adduction |
| Superior oblique action         | Overacting       | Normal              | No restriction unless secondary inferior rectus restriction |
| Three-step test                 | Fulfilled        | Not fulfilled       | Usually normal |

*Also known as double elevator palsy.
Edrophonium testing confirms the diagnosis. In Graves’ disease, restriction of the IR causes limited elevation in both adduction and abduction. Forcedduction testing is positive. Additional clinical findings distinguishing Graves’ disease from IO palsy include lid retraction, lid lag, proptosis, and enlarged rectus muscle bellies on echography or computed tomography without involvement of the tendons. An orbital blow-out fracture also causes limited elevation that may be confused with IO palsy. A typical history of blunt trauma or surgical repair of the orbital floor is usually elicited. Secondary strabismus after a blow-out fracture is caused by either restriction or muscle palsy, usually affecting the IR. However, IO palsy has been reported after orbital injury, and three-step testing confirms the diagnosis. (See respective chapters for a more thorough discussion.)

**Treatment**

**NONSURGICAL MANAGEMENT**

The decision to treat hinges on the presence of diplopia, a noticeable vertical deviation in primary gaze, an abnormal head position, and the needs and expectations of the patient and family. As with SO palsy, it is wise to document the stability of the palsy for approximately 6 months. Treatable illnesses that may cause IO palsy should be considered and addressed.11 Prism management may be helpful as primary treatment11 or as an adjunct to surgery.5 Prisms may be only marginally useful in certain positions of gaze if the vertical deviation is incomitant.

**SURGICAL MANAGEMENT**

The aim of surgical treatment is to eliminate an abnormal head position, a noticeable vertical deviation, and diplopia. Options for achieving these goals include surgical weakening of the ipsilateral antagonist (the SO muscle); weakening of the contralateral yoke muscle (the contralateral SR); and strengthening the contralateral antagonist (the contralateral IR). The decision on which muscles and how many of them to operate on to correct the deviation is not a simple one. The surgeon must individualize the surgical strategy depending on each patient’s findings. Patients must be aware preoperatively that complications such as undercorrection or overcorrection, loss of fusion, and iatrogenic SO palsy may occur. A second surgery is always a possibility.

Three clinical findings help the surgeon decide what type of procedure to perform. These are the magnitude of the hypertropia; the spread of comitance, in particular the degree of hypertropia in gaze ipsilateral to the palsied SO; and the degree of dysfunction of the oblique muscles causing incomitance (overaction of the SO and underaction of the IO). Patients may be grouped into three categories based on these findings. A fourth group with bilateral IO palsy is discussed as well.

**Group 1—Incomitant Deviation with Significant Oblique Dysfunction and Little or No Deviation in Gaze Out of the Field of Action of the Palsied Inferior Oblique**

Patients having mild to moderate hypertropia in primary gaze, significant oblique dysfunction (SO overaction and IO underaction) causing increased hypertropia in gaze contralateral to the palsied IO, and little or no hypertropia in gaze ipsilateral to the palsied muscle (little or no spread of comitance) will benefit from SO weakening procedures alone (see Fig. 16–1A and B). Surgery on an oblique muscle is needed to reduce both the hypertropia in primary gaze and overaction of the ipsilateral SO with the resultant incomitance. Surgical options for weakening the SO include SO tenotomy, tenectomy, tendon Z-plasty, and tendon recession.2, 5, 9, 12, 14

Generally, for SO surgery alone to be successful, the deviation in primary gaze must be no larger than 15 to 20 PD, oblique dysfunction must be present, and the deviation may not worsen to more than 10 additional PD on ipsilateral gaze. Success is defined by resolution of the abnormal head position, relief of diplopia, and improved vertical deviation without any of the following untoward events: undercorrection or overcorrection, deterioration of binocular fusion, or iatrogenic SO palsy.

In several series, “intrasheath” SO tenotomy5, 12 and SO recession2 are viable options for group 1 patients having more than 6 PD of vertical deviation in primary gaze. A theoretical advantage of recessing the SO tendon, as opposed to a tenotomy or tenectomy, is that recession is graded, reversible, and adjustable. However, a recession may be technically more difficult. SO tenectomy causes a high rate of SO palsy and overcorrection and should be avoided.9

If the deviation in primary gaze is less than 5 PD, a complete SO tenotomy may lead to iatrogenic SO palsy. Instead of a full-thickness tenotomy, a Z-tendonectomy may be performed, resulting in a less abnormal head position without overcorrection.5 Patients with small deviations in primary gaze but significant head tilt benefit from minimal weakening of the SO.5

Weakening procedures on the ipsilateral SO are not the only options for group 1 patients. Success has been reported with contralateral SR recession alone.4, 12

Undercorrection after weakening of the SO usually results from improper patient selection. In the following discussion of group 2 patients, those with greater than 20 PD of deviation in primary gaze and more than 10 PD of deviation out of the field of action of the palsied muscle respond poorly to weakening of the SO alone.

**Group 2—Incomitant Deviation with Significant Oblique Dysfunction and Significant Deviation Out of the Field of Action of the Palsied Inferior Oblique**

Patients with moderate to severe hypertropia in primary gaze, significant oblique dysfunction causing increased hypertropia in gaze contralateral to the palsied IO, and significant hypertropia in ipsilateral gaze (spread of comitance) require weakening of both the SO and the contralateral SR (see Fig. 16–2A through D).1 Vertical deviation of more than 10 PD in ipsilateral gaze is significant.

Weakening of the overacting SO achieves only a partial reduction of hypertropia in both primary position and contralateral gaze. It does not affect the deviation out of the field of action of the SO. Surgery on a second vertical muscle therefore is required. This achieves a greater reduction of hypertropia in primary gaze and also reduces hypertropia out of the field of action of the palsied IO that is unaffected.
by SO weakening alone. An adjustable suture technique on
the vertical rectus muscle may be beneficial.

In our early experience, ipsilateral SO tenotomy alone
resulted in undercorrection in patients meeting group 2 crite-
ria. All undercorrected patients responded well to contralat-
eral SR recession. Since then, all group 2 patients have
been treated by SO tenotomy combined with SR recession,
producing excellent results. Good results were also reported
using 12 to 13 mm of SO recession to correct 8 to 12 PD
of hyperdeviation in primary gaze and SO tenectomy to
correct 7 to 12 PD of hyperdeviation.

Three-muscle surgery, adding resection of the contralateral
IR, may be indicated if the hypertropia measures 30 PD or
more. In these cases the adjustable suture technique is advis-
able because overcorrection is more likely.

Group 3—Comitant Deviation Without
Significant Oblique Dysfunction

Patients with comitant deviations in primary position and
side gazes without significant oblique dysfunction will ben-
fit from surgery on the vertical rectus muscles alone (Fig.
16–3). Contralateral SR recession and/or ipsilateral IR
recession are preferred; they usually are effective in reducing
the deviation in all fields of gaze. Surgery on the SO is not
needed, because oblique dysfunction is not significant
enough to increase the vertical deviation in the field of action
of the affected obliques. Group 3 patients are relatively easy
to treat. Undercorrections and overcorrections are less likely,
because patients have no oblique dysfunction and do not
require SO surgery.

Group 4—Bilateral Inferior Oblique Palsy

Patients with bilateral IO palsy have been treated in a
variety of ways (Fig. 16–4). Bilateral, asymmetric SO reces-
sions have been used successfully. Pollard treated a patient
having symmetric IO palsies by elevating only the medial
rectus insertions. The main problems in this case were eso-
tropia in gaze above midline with diplopia and use of a chin-
up head position to fuse. Postoperatively, fusion was restored
up to 20 degrees above midline. Masked bilateral IO palsy
may be treated by a second SO tenotomy on the unoper-
atated eye.

Complications

Residual IO palsy, inadvertent severing of the SR, iatro-
genic SO palsy, and loss of fusion are complications that
may occur after surgery for IO palsy. Emergence of a masked
bilateral IO palsy after surgery for presumed unilateral palsy
has been reported. Patients should be counseled about these
possibilities.

Residual IO palsy may be due to incomplete tenotomy of
the SO or an insufficient amount of surgery. Inadvertent
severing of the SR seems unlikely but has been reported. A
stretched SR could be mistaken for the tendon of the SO.
Full visualization of the SO tendon, good exposure, and
identification of the SR are essential for avoiding these
complications.

Iatrogenic SO palsy is a complication of surgery on the
SO and is much more frequent after SO tenectomy (approxi-
approximately 50%). It occurs in approximately 9% of patients having SO tenotomy, with or without contralateral IR recession. Loss of fusion after SO tenectomy also has been reported but has not occurred with SO tenotomy despite iatrogenic SO palsy. Subjective and objective excyclotorsion occurs in these patients and may result in torsional diplopia.

REFERENCES

### Historical Perspective

Dissociated vertical deviation (DVD) has been an enigma for longer than a century. Stevens first reported on double vertical strabismus in 1895. Several decades later prominent ophthalmologists described DVD, albeit under various names (Table 17–1). These different terms reflect the confusion that surrounds DVD.

**Table 17–1. Synonyms for Dissociated Vertical Deviation**

<table>
<thead>
<tr>
<th>Name</th>
<th>Author</th>
</tr>
</thead>
<tbody>
<tr>
<td>Alternating hyperphoria</td>
<td>Crone, 1954&lt;sup&gt;22&lt;/sup&gt;, Bielschowsky, 1938&lt;sup&gt;7&lt;/sup&gt;</td>
</tr>
<tr>
<td>Alternating hypertropia</td>
<td>Duane, 1896&lt;sup&gt;25&lt;/sup&gt;</td>
</tr>
<tr>
<td>Alternating supproduction</td>
<td>Duane, 1896&lt;sup&gt;25&lt;/sup&gt;</td>
</tr>
<tr>
<td>Alternating sursunmuduction</td>
<td>Lancaster, 1958&lt;sup&gt;11&lt;/sup&gt;, Scobee, 1947&lt;sup&gt;7&lt;/sup&gt;</td>
</tr>
<tr>
<td>Alternating vertical deviation</td>
<td>Ohm, 1928&lt;sup&gt;92&lt;/sup&gt;</td>
</tr>
<tr>
<td>Anaphoria/anatropia</td>
<td>Stevens, 1995&lt;sup&gt;16&lt;/sup&gt;</td>
</tr>
<tr>
<td>Augenwaage</td>
<td>Wilson and McClatchey, 1991&lt;sup&gt;96&lt;/sup&gt;</td>
</tr>
<tr>
<td>Dissociated hyperphoria</td>
<td>Bielschowsky, 1938&lt;sup&gt;7&lt;/sup&gt;</td>
</tr>
<tr>
<td>Dissociated strabismus complex</td>
<td>Burian, 1944&lt;sup&gt;19&lt;/sup&gt;</td>
</tr>
<tr>
<td>Dissociated vertical deviation</td>
<td>Brown, 1966&lt;sup&gt;13&lt;/sup&gt;</td>
</tr>
<tr>
<td>Dissociated vertical divergence</td>
<td>Good and Hoyt, 1996&lt;sup&gt;38&lt;/sup&gt;</td>
</tr>
<tr>
<td>Dissociated vertical anomaly</td>
<td>Cords, 1922&lt;sup&gt;22&lt;/sup&gt;</td>
</tr>
<tr>
<td>Divergent vertical deviation</td>
<td>Stevens, 1895&lt;sup&gt;25&lt;/sup&gt;</td>
</tr>
<tr>
<td>Double dissociated hypertropia</td>
<td>Verhoeff, 1941&lt;sup&gt;19&lt;/sup&gt;</td>
</tr>
<tr>
<td>Double hypertropia</td>
<td>Anderson, 1959&lt;sup&gt;5&lt;/sup&gt;</td>
</tr>
<tr>
<td>Double vertical strabismus</td>
<td>Verhoeff, 1941&lt;sup&gt;19&lt;/sup&gt;</td>
</tr>
<tr>
<td>Occlusion hyperphoria</td>
<td>Cords, 1922&lt;sup&gt;22&lt;/sup&gt;</td>
</tr>
<tr>
<td>Occlusion hypertropia</td>
<td>Good and Hoyt, 1996&lt;sup&gt;38&lt;/sup&gt;</td>
</tr>
<tr>
<td>Periodic vertical squint</td>
<td>Stevens, 1895&lt;sup&gt;25&lt;/sup&gt;</td>
</tr>
<tr>
<td>Presumptive hyperphoria</td>
<td>Cords, 1922&lt;sup&gt;22&lt;/sup&gt;</td>
</tr>
<tr>
<td>Strabismus sursoabductorius</td>
<td>Cords, 1922&lt;sup&gt;22&lt;/sup&gt;</td>
</tr>
</tbody>
</table>

**Definition**

Dissociated vertical deviation is commonly defined as an intermittent anomaly of the nonfixing eye consisting of upward excursion, excyclotorsion, and lateral deviation. DVD violates Hering's law of simultaneous innervation of yoke muscles. In contrast to a true vertical deviation, the fellow eye does not exhibit refixation movement in the opposite direction, whether in the vertical, torsional, or horizontal planes.

The upward drift is traditionally referred to as DVD, whereas the excyclotorsion is termed dissociated torsional deviation (DTD) and the lateral deviation, dissociated horizontal deviation (DHD). The term dissociated strabismus complex was introduced to call our attention to the presence of all three components in a patient traditionally regarded as having DVD. Torsion associated with DVD is not a new observation. Lang described excyclotorsion in 60% of patients with DVD. Although recognized earlier, DHD as a clinical finding regained prominence after being described in the 1990s. If DVD is usually intermittent, but it may have a phoric or tropic phase. The phoric phase is seen only under cover, whereas the tropic phase manifests itself when a patient is daydreaming, inattentive, fatigued, or in poor health.

**Theories**

The exact cause of DVD remains unknown. Although several theories have been proposed (Table 17–2), current concepts focus on either a supranuclear disorder or a superior oblique (SO) malfunction.

Bielschowsky postulated that aberrant impulses from a vertical divergence center in the brain stem are responsible for DVD. Alternating hyperphoria or hypertropia was ex-
Table 17–2. Etiologic Theories of Dissociated Vertical Deviation

<table>
<thead>
<tr>
<th>Author</th>
<th>Theory</th>
</tr>
</thead>
<tbody>
<tr>
<td>Stevens, 1895&lt;sup&gt;a&lt;/sup&gt; and Duane, 1896&lt;sup&gt;b&lt;/sup&gt;</td>
<td>Imbalance caused by stronger elevators and weaker depressors</td>
</tr>
<tr>
<td>Ohm, 1928&lt;sup&gt;c&lt;/sup&gt;</td>
<td>Vestibular imbalance</td>
</tr>
<tr>
<td>White, 1933&lt;sup&gt;d&lt;/sup&gt;</td>
<td>Hyperfunction of the superior rectus</td>
</tr>
<tr>
<td>Bielschowsky, 1938&lt;sup&gt;e&lt;/sup&gt;</td>
<td>Aberrant impulses from a vertical divergence center</td>
</tr>
<tr>
<td>Verhoeff, 1941&lt;sup&gt;f&lt;/sup&gt;</td>
<td>Hypofunction of superior oblique or inferior rectus</td>
</tr>
<tr>
<td>Posner, 1944&lt;sup&gt;g&lt;/sup&gt;</td>
<td>Nuclear hypoplasia of superior oblique and inferior rectus</td>
</tr>
<tr>
<td>Scobee, 1947&lt;sup&gt;h&lt;/sup&gt;</td>
<td>Abnormal tone of the extraciliary muscles</td>
</tr>
<tr>
<td>Brown, 1966&lt;sup&gt;i&lt;/sup&gt;</td>
<td>Upper motor neuron anomaly in uncomplicated DVD; upper and lower motor neuron in complicated DVD</td>
</tr>
<tr>
<td>Anderson, 1954&lt;sup&gt;j&lt;/sup&gt;</td>
<td>Deficient impulses from inferonasal retina</td>
</tr>
<tr>
<td>Guyton and Kirkman, 1956&lt;sup&gt;k&lt;/sup&gt;</td>
<td>Fundamental torsional deviation</td>
</tr>
<tr>
<td>Helveston, 1980&lt;sup&gt;l&lt;/sup&gt;</td>
<td>Birth difficulties lead to arrested development of binocular fixation</td>
</tr>
<tr>
<td>Lyle, 1964&lt;sup&gt;m&lt;/sup&gt;</td>
<td>Supranuclear disturbance of a vertical vergence center</td>
</tr>
<tr>
<td>Stegall, 1981&lt;sup&gt;n&lt;/sup&gt;</td>
<td>Neuromuscular block at synaptic junction of the superior oblique</td>
</tr>
<tr>
<td>Spielmann, 1987&lt;sup&gt;o&lt;/sup&gt;</td>
<td>Imbalance of binocular stimulation</td>
</tr>
</tbody>
</table>

The superior oblique (SO) muscles formerly were implicated as a possible cause of DVD.<sup>13, 75, 89</sup> However, the report of an A-pattern exotropia, SO overaction, and DVD made this hypothesis unpopular.<sup>40</sup> Recent studies have resurrected this theory based on the fact that the eye elevates and undergoes excyclotorsion in DVD. A hypofunctioning SO muscle could explain both findings. Either a chemical defect or interruption of neuromuscular transmission in the SO could result in intermittent DVD. This theory is plausible even in the presence of an overacting SO—the overaction occurs only when the stimulus passes through the synapse, whereas if transmission is interrupted, DVD results.<sup>84</sup>

Recent sclerosearch coil eye movement recordings by Guyton in patients with DVD have shown that vertical versions are predominantly mediated by the oblique muscles in the fixing eye and by the vertical rectus muscles in the nonfixing eye. Vertical vergences, as in normals, are mediated predominately by the oblique muscles in both eyes.<sup>306</sup>

Latent nystagmus is strongly associated with DVD and is likely recordable in every patient with DVD. The slow phase of the latent nystagmus is always conjugate for the two eyes, with the fixing eye drifting in, down, and intorting. The nonfixing eye drifts in, down, and extorts. The latent nystagmus thus has both horizontal and cyclovertical components. Convergence is often used to damp the horizontal component. The cyclovertical eye movements in DVD appear to damp the cyclovertical component of the latent nystagmus. The damping occurs via a vertical vergence involving activation of the superior oblique muscle in the fixing eye, which tends to cause depression, intorsion, and abduction. In the nonfixing eye the inferior oblique muscle causes elevation, extorsion, and abduction. Simultaneously, vertical and horizontal versions occur to neutralize the tendency toward abduction and depression of the fixing eye, in order to maintain fixation. The upward version is produced by the inferior oblique muscle of the fixing eye and by the superior rectus muscle of the nonfixing eye, elevating the nonfixing eye still farther. The horizontal version occurs toward the nonfixing eye, causing the nonfixing eye to abduct still farther.<sup>306</sup>

The end result is that the fixing eye continues to fix, although slightly intorted, and the nonfixing eye is elevated, extorted, and abducted. Convergence compensation for the horizontal component of the latent nystagmus, if present, can negate the abduction of the nonfixing eye and often produces an adducted posture of the upward deviated eye. This convergence compensation, in combination with the net adducting effect of the DVD mechanism itself, is the basis of “dissociated horizontal deviation.”<sup>306</sup>

**Epidemiology and Risk Factors**

DVD is frequently associated with a history of infantile esotropia but may occur as an isolated finding.<sup>41, 42</sup> From 14% to 90% of patients with infantile esotropia will eventually develop DVD.<sup>16, 17, 41, 42, 47</sup> For this reason, some authors consider DVD to be a marker for congenital esotropia.<sup>24, 63</sup>

Patients with sensory deviations may acquire DVD as well.<sup>5, 7, 80</sup> The deviation has been described after cataract extraction<sup>86</sup>; after prolonged occlusion<sup>3</sup>; and in association with amblyopia, hyperopia, exotropia, and vertical deviations.<sup>41</sup>

**Genetics**

The genetic background of DVD remains uncertain. Suggestions that genetic factors contribute to DVD are based on anecdotal information. DVD alone has been reported in the sibling of a patient with esotropia and DVD.<sup>36</sup> Identical twins with DVD have been documented, as have three patients with affected siblings.<sup>41</sup>

**Clinical Characteristics**

**CLINICAL PRESENTATION**

DVD is usually asymptomatic because of poor fusion and suppression.<sup>36</sup> Patients may have evidence of peripheral
Dissociated vertical deviation

It usually is latent (phoric phase) but may be manifest (tropic phase). Friends and relatives may notice the cosmetic deformity, particularly during periods of inattention, lack of concentration, stress, or illness. Occasionally, diplopia and visual confusion may be reported when the DVD is manifest, simulating a true vertical deviation.

DVD generally presents before 12 months of age, but a lower mean age of 9 months was documented in one study. Its occurrence is not related to either the control of horizontal deviation or the timing of surgery.

DVD is commonly a bilateral but asymmetric condition. The signs are more prominent in the nonfixing, nondominant eye. Unilateral DVD is seen with profound amblyopia and sensory deviations.

Nystagmus is a frequent associated finding, most commonly in the form of latent nystagmus or manifest latent nystagmus, conditions also associated with infantile esotropia. Less commonly, patients with manifest nystagmus may exhibit DVD.

The literature is confusing with regard to the association of DVD with anomalous head posture. Crone described a syndrome consisting of esotropia, hypertropia in lateral gaze, nystagmus, DVD, and head tilt. A description of a similar syndrome noted that the head tilt was variable and unrelated to a null zone for the nystagmus. Both ipsilateral and contralateral head tilts have been observed (Figs. 17–1 and 17–2). These head postures may be adopted in an attempt to control nystagmus, or establish peripheral fusion. In many cases, however, the reason for the anomalous head posture remains obscure.

CLASSIFICATION

Dissociated vertical deviation that measures roughly the same (i.e., within ± 7 PD) in abduction, the primary position, and horizontal gaze is called comitant DVD. This is the most common presentation. Incomitant DVD refers to a measurable disparity in the magnitude of DVD in different gaze fields, commonly in adduction, primary gaze, and abduction. It is important to distinguish between these patterns because of the differing surgical strategies required to control the incomitance as well as the DVD. The different patterns will be discussed further in connection with their individualized surgical management.

Diagnosis

Clinical Evaluation

Measuring DVD

Measuring DVD poses a challenge even to the astute clinician. Accurate measurements are difficult to obtain. DVD is often more marked at home—or when the patient is fatigued, daydreaming, under stress, or sick—than when looking at a fixation target in an ophthalmologist’s office. Casual observation may reveal a larger deviation than when one attempts to quantify it. DVD is also influenced by the patient’s attention span and concentration—factors that are often variable, especially in a child. The magnitude of the DVD may vary from time to time and may increase as the examination proceeds.

Hirschberg’s corneal light reflex test may be used to estimate the amount of deviation. For cooperative patients, the prism under cover test is more precise. A cover is placed to dissociate the eye in question, and the size of DVD under cover is estimated. A translucent occluder may be used to disrupt fusion while at the same time observing DVD under cover (see Fig. 17–2). Base-down prisms and cover are placed in front of the dissociated eye. As the cover is switched to the fixing eye, only the movement from the uncovered (DVD) eye is observed. The point of neutralization is reached when no further downward movement is seen.
Some investigators use a semiquantitative grading scale: a small DVD is 0 to 9 PD; a moderate DVD, 10 to 19 PD; and a large DVD, more than 20 PD. A three- or four-point grading scale may be used. The different components of the dissociated strabismus may also be graded separately: for example, DVD +3, DTD +1, and DHD +1.

**Bielschowsky’s Phenomenon**

When the fixing eye is presented with light of decreasing intensity, the eye with DVD falls. Conversely, increasing light in the eye with DVD will cause an increase in upward vertical drift. Light intensity may be varied by using graded neutral density filters or the Bagolini red filter bar, or by rotating two polarized filters (the same ones used with most near stereotests) on one another to create a darkening filter effect. This is termed Bielschowsky’s phenomenon after his description in 1938 (Fig. 17-3). It is found in at least 50% of patients with DVD and has been demonstrated in patients with DHD as well. The phenomenon is regarded as strong evidence that DVD is a sensory anomaly, because altering the sensory input to the fixing eye produces the characteristic findings.

**Red Glass Test**

Red glass testing yields peculiar results in DVD. Regardless of whether the red filter is placed before the right or left eye, the patient describes a red image below a white image. This contrasts to the findings in patients with a true vertical deviation. In true hypertropia, the second (red) image is seen above or below the primary image, depending on whether the red filter is placed in front of the hyperdeviated or the hypodeviated eye.

**Anomalous Head Posture**

Anomalous head posture is a frequent correlate of DVD. The deviation is first measured in the preferred posture and again with forced head tilt in the opposite direction (see Fig. 17-1). If DVD is increased or control breaks down with the latter maneuver, it is contributing to the anomalous posture.

**Incomitant DVD**

Horizontal incomitance with DVD should be sought. The size of the DVD is measured in abduction, adduction, and the primary position, and the results are correlated with careful analysis of inferior oblique (IO) and SO function as well as superior rectus (SR) underaction if SR recessions have been performed. The findings will allow the surgeon to plan a suitable operative strategy.

**LABORATORY EVALUATION**

The diagnosis of DVD does not require laboratory confirmation, but the methods described contribute to our understanding of the mechanisms of DVD. These studies will become increasingly important as the mystery of the DVD phenomenon continues to unfold.
Eye Movement Recordings

Electromyography (EMG). Scott pioneered many studies on motility using EMG and saccadic velocity analysis. He demonstrated increased firing of elevator muscles in DVD and also observed that the velocity of elevation and of the return or refixation movements was variable. The variability depended on the patient’s attentiveness, the visual environment, and placement of the cover in front of the fixing or the nonfixing eye. Increased IO innervation was reported in a case of DVD.

Electro-oculography (EOG) and Magnetic Scleral Search Coil Techniques. Helveston demonstrated that the upward movement in DVD was slow, less than 2 to 40 degrees/sec. Downward refixation movements were slightly faster at 10 to 200 degrees/sec. These figures contrast to normal saccadic velocities of 200 to 400 degrees/sec. DVD recordings were the same in the supine and upright positions. These EOG findings later were confirmed using scleral search coil techniques.

Magnetic Resonance Imaging

In a recent update, 7 of 18 patients who underwent magnetic resonance imaging showed disorganized architecture and altered size of the pars compacta of the substantia nigra. However, because of the small sample size, further studies are required to substantiate these findings.

Visual Evoked Potentials

Earlier studies reported an albino-type of misrouting of the temporal retinal fibers in DVD, crossing at the chiasm. These findings have not been replicated by more sophisticated studies using flash visual evoked potentials and pattern reversal paradigms.

DIFFERENTIAL DIAGNOSIS

It is critical to distinguish isolated IO overaction from DVD, because either may cause elevation of the eye in the primary and adducted positions. The common association of these entities adds to the confusion. Important differences between DVD and IO overaction are summarized in Table 17–3.

Table 17–3. Dissociated Vertical Deviation Versus Inferior Oblique Overaction

<table>
<thead>
<tr>
<th>Features</th>
<th>DVD</th>
<th>IO Overaction</th>
</tr>
</thead>
<tbody>
<tr>
<td>Elevation</td>
<td>Seen in adduction, primary gaze, and abduction</td>
<td>Adduction ± primary gaze; none in abduction</td>
</tr>
<tr>
<td>V pattern</td>
<td>Not seen</td>
<td>Often present</td>
</tr>
<tr>
<td>Superior oblique function</td>
<td>May see operation</td>
<td>Usually underacting superior oblique</td>
</tr>
<tr>
<td>Hypertropia</td>
<td>Usually not seen</td>
<td>Seen in adduction</td>
</tr>
<tr>
<td>Hering's law</td>
<td>Violated</td>
<td>Obeyed</td>
</tr>
<tr>
<td>Speed of vertical deviation</td>
<td>Slow (2–200 degrees/sec)</td>
<td>Rapid (200–400 degrees/sec)</td>
</tr>
<tr>
<td>Incycloduction on refixation</td>
<td>+</td>
<td>–</td>
</tr>
<tr>
<td>Latent nystagmus</td>
<td>Often +</td>
<td>Usually –</td>
</tr>
<tr>
<td>Pseudoparesis of contralateral superor rectus</td>
<td>–</td>
<td>+</td>
</tr>
<tr>
<td>Bielschowsky’s phenomenon</td>
<td>±</td>
<td>–</td>
</tr>
</tbody>
</table>

+ = present; – = absent; ± = may or may not be present.

Combined DVD and Inferior Oblique Overaction

This is a more difficult situation to appreciate, and it is not infrequent. True hypertropia may occur when both DVD and IO overaction occur simultaneously. DVD with hypertropia may also result from secondary SR contracture in a patient with DVD.

Measuring the hypotropic refixation movement in the contralateral eye by the prism cover test reveals the true hypertropic component. The total upward vertical drift is then measured by the prism under cover test. The difference between the two measurements constitutes the component that is caused by DVD. Refixation movements observed in the contralateral eye in true vertical deviation (by Hering’s law) are not observed in DVD.

Treatment

NONSURGICAL MANAGEMENT

Observation

Some investigators believe that DVD improves with time. However, in 100 patients followed for a mean of 7.3 years, no significant decrease in the magnitude of DVD was observed. A nonsurgical approach remains preferable for DVD that is controlled as a phoria and for smaller deviations that occur infrequently.

Encourage Fusion or Bifixation

The logical first step in management is to strengthen fusional mechanisms. Optimal spectacle correction should be provided. Blurred vision may cause a latent DVD to become manifest more frequently. Providing clear images...
to both eyes encourages fusion.\textsuperscript{57} Correcting an associated horizontal deviation, either surgically or with prisms, promotes the possibility of peripheral fusion.

### Switching Fixation

Oclusion of the fixing or dominant eye, either as a means of treating amblyopia or encouraging fixation, may be tried in an attempt to improve DVD in the nonpreferred eye. Foggng the good eye with plus lenses may be more cosmetically acceptable than an orthoptic patch. Progressively stronger hyperopic lenses that are tolerated without cyclopia until the patient switches fixation in the examining room may be tried.\textsuperscript{39} Permanent alteration of fixation is infrequent, however, and occurs only in very young patients.

### SURGICAL MANAGEMENT

#### Indications

Indications for surgery in patients with DVD vary widely among strabismologists. Surgery is frequently performed in the following situations:

**Increasing Frequency of Manifest DVD in a Patient with Peripheral Fusion.** When control of DVD deteriorates and patients are still at risk of developing amblyopia, a more aggressive approach to surgery should be taken. Unfortunately, because of the variable behavior of DVD, experts may disagree on their recommendations depending on the size and frequency of DVD as measured in the office. Prolonged DVD may cause secondary contracture of the SR and may deteriorate into a true hyperdeviation.\textsuperscript{57}

**Patients Who Adopt an Anomalous Head Posture to Control the Magnitude of the DVD.** With the preferred head tilt, a significant subset of patients control DVD better from a motor standpoint (smaller DVD) or gain peripheral fusion. Forced head tilt testing to the opposite side will reveal poorer control of DVD or a larger deviation. Successful surgery may reduce the degree of head tilt.\textsuperscript{72}

**DVD That is Large and Occurring Frequently.** Often surgery is undertaken electively to improve a patient’s psychosocial well-being.

#### Issues Related to Surgical Management

**Doing Surgery for an Imprecise Preoperative Measurement.** Experts recognize that DVD is difficult to measure. In a setting where reliable strabismus measurements dictate surgical strategy, some believe that DVD is probably better left alone.

**Unilateral Versus Bilateral Surgery.** Unilateral surgery was advocated by Scott\textsuperscript{40, 74} for unilateral manifest DVD. He differentiated DVD from occlusion hyperphoria, in which the deviation shows up only under cover. His results from unilateral graded SR recessions were better than with bilateral SR recessions.\textsuperscript{90} Unfortunately this comparison may be biased by retrospective review, because only 10 patients were in the bilateral group compared with 57 in the unilateral group.

Most authors perform bilateral surgery for DVD.\textsuperscript{57, 61, 73, 91} Unilateral surgery runs the risk of unmasking a DVD in the unoperated eye when the patient is capable of switching fixation.\textsuperscript{57, 61, 73, 91} In one report, 83% required surgery on the second eye.\textsuperscript{79} Fixation preference played no role in determining which patients would need a second procedure unless visual acuity was worse than 20/80 in one eye.\textsuperscript{79}

Monocular surgery limits the amount of superior rectus recession without inducing a true hypotropia. When the patient switches fixation, the unoperated eye may develop a large hypertropia.\textsuperscript{57} After unilateral surgery on one SR, a larger innervational input is required to keep this eye balanced in primary gaze. By Hering’s law, the opposite SR and/or IO receive the same innervation. Jampolsky\textsuperscript{43} calls this “fixation duress.” Theoretically, surgery in one eye can result in asymmetric lid changes.\textsuperscript{74} Hence, some practitioners reserve unilateral surgery for patients having dense unilateral amblyopia.\textsuperscript{57, 61, 73, 91} We prefer bilateral surgery unless there is reason to believe that the nonfixing eye will never be preferred for fixation. In the latter situation, unilateral surgery may be acceptable. If the patient is able to alternate fixation freely, bilateral surgery is performed.

**Symmetric Versus Asymmetric Surgeries for Bilateral Involvement.** Bilateral symmetric operations are indicated for cases of bilaterally symmetric DVD (within ±7 PD). Asymmetric DVD is more common, however, with the larger DVD in the nonfixing eye. The larger recession is done on the eye with a larger DVD.\textsuperscript{57}

#### Surgical Procedures

Different surgical procedures have been advocated for DVD. The sum of forces in any DVD surgery should favor depression.\textsuperscript{46, 73} Realistic goals are to minimize the dissociation or improve control. DVD is never actually totally eradicated.\textsuperscript{41, 91} Both preoperative measurements of the size of the DVD and quantitation of the amount of surgery remain relatively imprecise. Recurrences are common. The deviation tends to be difficult to overcorrect even with extensive surgery such as large SR recessions.\textsuperscript{73, 57} The many alternative procedures may confuse the novice surgeon. To aid our readers, we have outlined our treatment scheme for DVD in Table 17–4. It is imperative that the pattern of incomitance be recognized and the appropriate surgery performed.

**Surgeries for Comitant DVD.** Vertical rectus procedures work best for horizontally comitant DVD.

**SR Recession.** For symmetric DVD, we do large bilateral SR recessions of up to 10 mm (see Appendix: Surgical Dose Tables). Whereas a conjunctival incision made near the muscle insertion has become unpopular, we recommend its use for patients who are to undergo large SR recessions. The incision works well for this type of surgery. It is located superiorly and hidden by the upper lid, and the patient’s comfort level is improved. To maximize the recession effect, we prefer to suture the SR muscle to bare sclera. Beyond 10 mm, the insertion of the SO tendon interferes with scleral fixation.\textsuperscript{53} At times, we have found it helpful to remove the lid speculum and use a Desmarres retractor to gain better exposure of the posterior aspect of the globe.

Larger recessions may be achieved using the hang-back technique in case the surgeon experiences technical difficulty.\textsuperscript{34, 57} The attachment of muscle to sclera may, however, be compromised by the presence of the SO in this area.

For comitant but asymmetric DVD, we perform large bilateral asymmetric SR recessions. The larger recession of
### Table 17-4. Surgery for Dissociated Vertical Deviation (DVD)

<table>
<thead>
<tr>
<th>DVD Features</th>
<th>Surgery</th>
</tr>
</thead>
<tbody>
<tr>
<td>Comitant DVD</td>
<td>Large BSR of 7–10 mm; IR resection (up to 4 mm) after maximal BSR</td>
</tr>
<tr>
<td>DVD greatest in field of IO in nonfixing eye</td>
<td>IO weakening procedure (usually recession or anterior transposition)</td>
</tr>
<tr>
<td>DVD greatest in abduction</td>
<td>Vertical rectus surgery (BSR) and IO weakening</td>
</tr>
<tr>
<td>DVD greatest in abduction SO overaction; A pattern</td>
<td>Moderate IO weakening (i.e., recession 8–10 mm)</td>
</tr>
<tr>
<td>DHD Exodeviation</td>
<td>Vertical rectus surgery for DVD, posterior three-fourths tenectomy of SO for collapse of A pattern</td>
</tr>
<tr>
<td>DHD Esodeviation</td>
<td>LR recession ± fadenoperation</td>
</tr>
<tr>
<td>DHD Large BSR of 7–10 mm; IR resection (up to 4 mm) after maximal BSR</td>
<td>Fadenoperation on MR; ± MR recession</td>
</tr>
</tbody>
</table>

10 mm is carried out in the eye with a larger vertical dissociation—usually the nondominant, nonfixing eye. The eye with a smaller deviation receives a recession of 6 to 7 mm. If comitant DVD recurs, further SR recessions up to 10 mm may be done.

In the earlier literature, several combinations of SR recessions with or without the fadenoperation were recommended for DVD. SR recession often was combined with the fadenoperation when only a small recession (3–5 mm) was done. Some authors advocated small recessions alone, but these patients have proved to have frequent recurrences. Large bilateral SR recessions are now generally preferred. Graded recessions based on the magnitude of the DVD also are done. The recommendations vary from 5 to 9 mm and 7 to 9 mm to 8 to 14 mm. A small SR recession of 4 to 5 mm combined with a 6-mm inferior rectus (IR) resection has been described for unilateral large DVD.

**IR Resection.** Although earlier reports advocated this as a primary procedure, we now reserve it for use as a second procedure in recurrent comitant DVD after large (10 mm) SR recessions have failed, or when the surgeon is concerned about causing “double-elevator weakness.” We limit the amount of resection to 4 to 5 mm to minimize lid fissure changes. Larger resections of the IR not only restrict elevation but also cause noticeable narrowing of the palpebral fissure.

**Patterns of Incomitance and Recommended Surgical Procedures**

**DVD Greatest in the Field of the IO of the Nonfixing Eye** (Fig. 17–4). This type is associated with DVD in abduction.

![Figure 17-4. Incomitant DVD due to inferior oblique overaction. A, Preoperative photographs show DVD of right eye greatest in adduction (>10 PD), less in primary gaze (6 PD), and absent in abduction. B, Photographs taken after right inferior oblique recession showing improvement of DVD across the horizontal gazes and control of inferior oblique overaction.]
tion of less than 5 PD. A V-pattern horizontal deviation may be observed. In extreme adduction, a true hypertropia may be recorded in addition to a true DVD. In primary gaze, only DVD is seen. These cases do well with IO recession or anterior transposition.

It was formerly believed that IO weakening procedures (e.g., disinsertion, myectomy, and recession) do not affect the DVD. In patients in whom DVD and IO overaction coexist, eliminating the IO overaction does reduce the cosmetic defect in lateral gaze. Anterior transposition of the IO, a stronger weakening procedure, was reintroduced for moderate to severe IO overaction. Later, several studies have found that anterior transposition helps control DVD in the primary position.

**DVD Greatest in the Field of the IO of the Nonfixing Eye with Significant DVD in Abduction (Fig. 17–5).** The findings in this type are similar to the first type with the exception of DVD exceeding 5 PD in abduction. Compared to the first type, the DVD is larger in primary gaze, but the greatest deviation still is seen in adduction. These patients do well with vertical rectus muscle surgery for DVD in abduction, combined with an IO weakening procedure for DVD and/or hypertropia in adduction.

**DVD Greatest in the Field of IO of the Nonfixing Eye After Bilateral SR Recessions (Fig. 17–6).** These patients have had large bilateral SR surgeries for DVD that was previously horizontally comitant. Postoperatively a DVD remains that has now become incomitant, with the deviation greatest in the field of the IO of the nonfixing eye. In these cases one needs to look for moderate to severe underaction in the field of the IO. Pseudo-IO overaction develops secondary to a weakened yoke muscle, the SR. The DVD is much smaller in abduction (the field of the SR) but persists in the primary position and is even greater in adduction.

Further weakening of the SR will worsen the incomitance. We recommend moderate IO recessions of 8 to 10 mm. We

---

**Figure 17-5.** Incomitant DVD is greatest in the field of an overacting inferior oblique but with significant DVD in abduction as well. A, Preoperative photographs show esotropia and DVD of left eye in primary position. DVD of left eye persists in abduction and adduction. Top right, The left inferior oblique is seen overacting. Both superior obliques in down and medial gaze are underacting. B, Postoperative photographs taken after left inferior oblique recession and bilateral superior rectus recession. Note improvement of DVD in middle row across horizontal gaze and improvement in left inferior oblique rotation.
are reluctant to perform anterior transposition of the IO because of the significant limitation of upgaze that may ensue.8, 27, 99

Because of frustrating recurrences of DVD even after large bilateral SR recessions, some have advocated the use of combined weakening procedures on both elevator muscles—the SR and the IO.37, 87 When necessary, smaller SR recession was combined with the IO weakening procedure, usually putting the SR muscle on an adjustable suture whenever possible.57 Despite a “double-elevator weakness,” only mild duction deficiencies were observed postoperatively.53, 87

**DVD Greatest in Abduction with Associated SO Overaction** (Fig. 17–7).31, 59 The DVD is greater in abduction in the nonfixing eye than in primary gaze. There is an associated A pattern, and esotropia40 or exotropia.40 IO weakening procedures aggravate both the DVD and the A pattern.58, 70

In this pattern, overaction of the SO causes incomitance of the DVD.58 In the adducted position, the nonfixing eye is subject to opposing forces—the DVD causing elevation and the SO overaction, causing depression. This accounts for the lesser degree of DVD noted in adduction. SO overaction in the fellow eye requires increased innervation to the IO to balance the eye in a vertically neutral position. By Hering’s law, increased innervation from the yoke IO of the fixing eye increases innervation to the SR of the nonfixing eye, favoring elevating forces and worsening the DVD in abduction.

If an IO weakening procedure is done or the SO is strengthened (SO tuck),46, 68 incomitance will increase from enhanced SO overaction. This type of patient should undergo vertical rectus surgery for DVD in abduction and posterior three-fourths SO tenectomy at the insertion to collapse the A pattern without affecting torsion.58, 78

**Dissociated Horizontal Deviation (DHD).** The presence of a horizontal component, usually abduction, is usually seen in combination with DVD but may occur alone (Fig. 17–8).69, 96 DHD is frequently seen after bilateral resections of the lateral rectus muscles for infantile esotropia. Bielschowsky’s phenomenon (see earlier discussion) and violation of Hering’s law are noted in the horizontal planes. Latent nystagmus, excyclotropia, and variable horizontal deviation are observed. When present, DHD is found in all gaze positions.96 An associated SO overaction may neutralize the DVD component in primary gaze, allowing only the DHD component to become manifest. DVD may still be seen in abduction.70 DHD should be differentiated from variable intermittent exotropia69 and variable esotropia.98 In addition, patients with unilateral uncorrected hyperopia and esotropia will exhibit an adduction fixation movement unilaterally. Lateral rectus recession with or without a posterior fixation suture is usually effective in reducing the size of DHD.96, 98

Dissociated esodeviation is rarer than dissociated exodevi- ation.91 It occurs when either eye is covered with a semiopaque occluder. The eyes remain aligned when both are occluded. This finding distinguishes the deviation from esophoria.82 If surgery is contemplated, patients do well with a posterior fixation suture on the medial rectus.91

**Complications**

Perhaps the most bothersome of all the potential complications is the recurrence of DVD or the persistence of residual DVD. These problems may reflect the fact that we are trying to solve an underlying central cortical problem by extraocular muscle surgeries. Despite altering innervational forces to favor depression of the eye and improve DVD, the defective supranuclear signal remains.

Despite bilateral large recessions of the SR, DVD may recur. This has led some authors to recommend larger bilateral recessions of the SR even during initial surgery.57 Residual incomitant DVD may become manifest consequent to secondary IO overaction. In these cases, further weakening of the SR will aggravate the incomitance. IO weakening procedures are recommended.34

A true vertical deviation due to secondary contracture of the SR may develop after prolonged DVD. A vertical imbalance may also follow surgery on any of the cyclovertical muscles, especially if it is done unilaterally or asymmetri-
An ipsilateral hypotropia may follow either unilateral SR recession or asymmetric bilateral SR recession, with the hypotropia developing in the eye that received the larger recession. IR resections and anterior transposition of the IO may also be responsible.

After effective surgical correction of DVD, some deficiency of upgaze is usually present. In fact, if this does not occur, surgery is usually ineffective and undercorrection occurs. Restricted upward rotations can occur after supramaximal SR recessions, large IR resections, anterior transposition of the IO, and weakening of both the SR and IO for recurrent DVD. The latter condition results.
in a “double-elevator weakness” that is usually mild. Patients are not commonly bothered by this type of elevator deficiency.

Palpebral fissure changes may accompany vertical rectus muscle surgery. SR recessions may produce a widened palpebral fissure as well as ptosis, whereas large IR resections are accompanied by a narrowed lid fissure. The cosmetic defect may be so pronounced as to replace the original DVD, which was intermittent, by permanent disfigurement.

REFERENCES


OTHER PARALYTIC STRABISMUS
The focus in this chapter is on the diagnosis and management strategies for total, partial, and isolated muscle involvement in third cranial (oculomotor) nerve paralysis. In-depth neurologic evaluation of a patient with third cranial nerve paresis is not addressed. Specific surgical techniques used to correct the resulting misalignment are discussed in Chapter 36. These procedures are mentioned in this chapter only as appropriate surgical strategies for correcting strabismus.

### Etiology

Knowing the etiology of third nerve palsy assists in developing strategies for management. Similar causes of this type of palsy are found in children and adults, but they vary in frequency of occurrence. In adults, the most common causes are aneurysm and vascular disease (including ischemia, hypertension, diabetes, and inflammatory arteritis) (Table 18–1). In children, congenital factors, including birth trauma and neonatal hypoxia, are the leading causes of third nerve palsy (Table 18–2). Trauma is a fairly common cause of acquired third nerve palsy in both adults and children.

Migraine, which may last as long as 4 weeks, may present as transient third nerve palsy in adults and children. The palsy affects the third cranial nerve alone in as many as 83% of cases. Ptosis usually precedes ophthalmoplegia. The average age at onset is 15 years, but the palsy may occur in infancy, in which recurrent, painless ophthalmoplegia and/or ptosis may be the only manifestations of a migraine attack. In older children, ipsilateral ophthalmoplegia usually accompanies the headache.

Other less common causes in both age groups include carotid cavernous fistula, frontal sinus mucocele, and mesencephalic infarction.

### Genetics

The mode of transmission of hereditary third nerve paresis may be either dominant or recessive. A dominant hereditary type, which has passed through several successive generations, is autosomal but may show incomplete penetrance.

---

**Table 18–1. Causes of Adult-Onset Oculomotor Nerve Palsy (Expressed in Percent)**

<table>
<thead>
<tr>
<th>Cause</th>
<th>Rucker(^{26, 29}) (N = 609)</th>
<th>Green et al(^ {14}) (N = 130)</th>
<th>Goldstein and Cogan(^{12}) (N = 61)</th>
<th>Rush and Young(^{46}) (N = 290)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Aneurysms</td>
<td>19</td>
<td>30</td>
<td>18</td>
<td>14</td>
</tr>
<tr>
<td>Vascular</td>
<td>18</td>
<td>19</td>
<td>47</td>
<td>21</td>
</tr>
<tr>
<td></td>
<td>diseases</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Trauma</td>
<td>14</td>
<td>11</td>
<td>8</td>
<td>16</td>
</tr>
<tr>
<td>Neoplasm</td>
<td>14</td>
<td>4</td>
<td>10</td>
<td>12</td>
</tr>
<tr>
<td>Syphilis</td>
<td>1</td>
<td>9</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Undetermined</td>
<td>25</td>
<td>23</td>
<td>11</td>
<td>23</td>
</tr>
<tr>
<td>Miscellaneous</td>
<td>10</td>
<td>4</td>
<td>6</td>
<td>14</td>
</tr>
</tbody>
</table>

**Table 18–2. Causes of Childhood-Onset Oculomotor Palsy (Numbers Expressed in Percent)**

<table>
<thead>
<tr>
<th>Cause</th>
<th>Miller(^6) n = 30</th>
<th>Harley(^{16}) N = 32</th>
</tr>
</thead>
<tbody>
<tr>
<td>Congenital</td>
<td>43</td>
<td>47</td>
</tr>
<tr>
<td>Aneurysm</td>
<td>7</td>
<td></td>
</tr>
<tr>
<td>Neoplasm</td>
<td>10</td>
<td>9</td>
</tr>
<tr>
<td>Vascular disease</td>
<td>10</td>
<td>6</td>
</tr>
<tr>
<td>Trauma</td>
<td>20</td>
<td>13</td>
</tr>
<tr>
<td>Inflammation</td>
<td>13</td>
<td>9</td>
</tr>
<tr>
<td>Miscellaneous</td>
<td>7</td>
<td>16</td>
</tr>
</tbody>
</table>
When transmission is recessive, third nerve palsy may occur in only one generation.¹⁰

**Clinical Characteristics**

**TOTAL THIRD NERVE PARESIS**

Total third nerve paresis may be central, sparing the pupil, or peripheral with pupillary involvement. If the pupil is spared, the cause is most likely vascular.¹² When the pupil is involved, the cause is likely to be an aneurysm. This lesion may be as far anterior as the superior orbital fissure and orbit or as far posterior as the point where the third nerve exits the brain stem. The patient exhibits a large exotropia with hypotropia. A fixed, dilated pupil is observed that may be more pronounced as the ptotic lid elevates (Fig. 18–1). On attempted adduction, the eye incyclotorts if there is no concurrent superior oblique (SO) palsy. The paralysis may occur alone or may be accompanied by other cranial nerve involvement.

The pupillary fibers arise from the nucleus of Perlia (of the visceral nuclei) joining the third nerve before leaving the midbrain. A dilated, unreactive pupil with no other signs of third nerve dysfunction is not an isolated third nerve paresis but rather the result of trauma, Adie syndrome, or a drug-related event.⁴ Involvement of the pupil in third nerve palsy is always associated with some other signs of third nerve dysfunction.

In children, total paralysis is often congenital. When the neuroradiographic workup is negative and there is no neurologic deficit, it is probably caused by an intrauterine insult or birth trauma.¹⁵, ₃⁰ Childhood paralysis frequently presents with aberrant regeneration. A child with cyclical third nerve paresis experiences spasmodic episodes of total third nerve paresis occurring at regular intervals of 15 to 30 seconds.²₂, ₃⁰

Excluding birth trauma, the congenital form of external ophthalmoplegia has certain distinguishing features: (1) It is generally bilateral; (2) the extraocular muscles can vary in their degree of involvement and weakness, which is not always total; and (3) although the same biotype almost always occurs in the same family, it is not unusual to find the whole range of third nerve disorders—from simple ptosis or isolated paresis to total paresis—in a single family.

**PARTIAL PARESIS**

The third cranial nerve separates into its superior and inferior divisions in the area of the cavernous sinus. Involve-

---

*Figure 18–1. Patient with complete right third nerve palsy: A, Complete ptosis. B, Composite photographs in different gazes. Note fixed dilated pupils and that only right abduction rotation is present.*
ment of the superior division of the third cranial nerve is often associated with other cranial nerve palsies; it rarely occurs alone. A patient with cryptococcal meningitis completely recovered superior division function after antifungal therapy.6

Isolated involvement of the inferior division of the third cranial nerve occurs more frequently. Paresis of this branch presents as an internal ophthalmoplegia associated with med- ial rectus (MR), inferior rectus (IR), and/or inferior oblique (IO) palsy as well as pupillary involvement.24, 40, 49 Inferior-division palsies have been associated with trauma, neoplasm, and a viral insult. Presumed viral cases frequently resolve spontaneously.48 Recovery may follow treatment with oral corticosteroids.24

**ISOLATED MUSCLE PALSY**

Isolated paralysis of the muscles supplied by the third nerve is difficult to establish. The anatomic location in the third nerve complex of individual nuclei makes a central origin unlikely. Each of the muscles supplied by the third nerve is vulnerable to the local effects of a mass, surgery, or trauma.

**Superior Rectus Palsy**

Isolated paralysis of the superior rectus (SR) muscle may be congenital or acquired. Birth trauma may lead to isolated SR palsy and/or true ptosis. Acquired traumatic palsy can result from a direct blow or from use of either a speculum or bridle suture with cataract surgery.22 Initially, the paralyzed muscle shows the greatest deviation in elevation in the abducted position. Upward gaze remains normal in adduction. With long-standing paresis, upward rotations may be limited in both adduction and abduction and may present as hypotropia in the primary position, simulating double elevator palsy. There may be an accompanying ptosis or pseudoptosis. The distinction is made by having the patient fix on a target with the paralyzed eye and observing the change in the palpebral fissure. True ptosis persists in the paralyzed eye, whereas the lid droop disappears in pseudoptosis (see also Chapter 20).

IR restriction may follow a retrobulbar or peribulbar injection and can mimic SR paresis.

**Inferior Rectus Palsy**

When the IR is weak, IR palsy is considered. A series of such cases in which all patients recovered has been reported.37 The branch of the inferior division of the third nerve enters the IR at the junction of the middle and posterior third of the muscle. IR weakness and total paralysis may follow direct trauma to the orbit, with IR entrapment and injury of the muscle or its nerve supply.16 Interventions such as retrobulbar or peribulbar anesthetic injections may create the same weakness of depression.

**Medial Rectus Palsy**

Isolated loss of adduction, although suggesting MR paresis, may represent other neurologic deficits. Internuclear ophthalmoplegia, medial orbital wall trauma, congenital absence of the MR muscle, type II Duane syndrome, and supranuclear clear lesions all present with decreased or absent adduction of the involved eye.3

**Inferior Oblique Palsy**

Of all the extraocular muscles supplied by the third cranial nerve, the IO is the least likely to be individually affected. The diagnosis is established by the Parks three-step test. Like SO palsy, there is an anomalous head position.11, 44 These cases are almost always congenital and must be distinguished from the spread of comitance from a contralateral SO palsy, Brown syndrome, or a tight SR syndrome secondary to contralateral inhibitional palsy of an SO palsy18 (see Chapter 16).

**ABERRANT REGENERATION**

Aberrant regeneration of the third nerve may occur during its recovery. During assessment of versions and ductions, ipsilateral retraction of the upper lid may be observed on attempted adduction and/or attempted infra- duction, as well as iris sphincter (pupillary) constriction (Fig. 18–2).22 These abnormal movements are presumed to be caused by miswiring after a break in the axonal cylinders and subsequent misdirection of sprouting axons. Aberrant regeneration without an acute history of trauma may indicate an indolent space-occupying lesion, such as an aneurysm or tumor in the cavernous sinus.7, 42

**COMBINATION WITH OTHER CRANIAL NERVE INVOLVEMENT**

Although cranial nerve palsies occur most often as isolated events, they may occur in combination. Combined injuries may result from closed- and open-head trauma. Without a history of trauma, further studies are indicated to determine possible causes, which may include infection, vasculopathy, aneurysm, and neoplasm.

The most common paretic nerve found in head trauma victims is the fourth cranial (trochlear) nerve. Less frequently the sixth (abducens) nerve may be affected. Involvement of the seventh (facial) nerve has been described in association with bacterial infections.30, 40

**Diagnosis**

**CLINICAL EVALUATION**

Significant general neurologic findings can occur in association with third nerve paresis. These findings are beyond the scope of this discussion, but the ophthalmologist should abide by certain guidelines in evaluating patients with this disorder.

Patients with vasculopathy and pupillary sparing should be observed daily for 1 week, then weekly for 1 month, and finally monthly for 6 months. If the condition deteriorates, appropriate imaging studies should be performed. All patients younger than age 40 years who present with third nerve palsy should have contrast neuroimaging, cerebrospinal fluid evaluation, and possibly cerebral angiography. Pa-
Figure 18-2. Partial left third nerve palsy with aberrant regeneration. Note that ptosis, exotropia, and hypotropia are less compared with patient in Figure 18–1. Persistent deficiency in adduction, upgaze, and downgaze is observed. Pupils constrict on adduction (aberrant regeneration).

Patients older than age 40 years with pupillary involvement should undergo a similar workup. Aberrant regeneration should alert the physician to look for a possible mass lesion or aneurysm, especially if there is no history of trauma.342

The size and reaction of the pupils help localize the site of the lesion. Pupil-sparing third nerve paresis indicates a central brain stem lesion or vascular occlusive disease. Peripheral lesions, such as aneurysms and tumors, may initially spare the pupil (8% to 15% of cases) but will ultimately cause dilation by disrupting its efferent pathway.47 Patients with isolated third nerve paresis may have normal ophthalmologic findings apart from ocular rotation defects and pupillary abnormalities.

STRABISMUS EVALUATION

Prism Cover Testing

The essential element of any strabismus evaluation is defining the deviation. In a paralytic deviation, Hering’s law of equal innervation to yoke muscles plays an important role. A prism cover test in the cardinal positions of gaze (comitance test) will identify the offending eye. Determining which individual extraocular muscles are involved requires an in-depth strabismus evaluation.

Sensory Evaluation

The ultimate goal in correcting strabismus is to restore bifoveal fixation. Achieving this goal in third nerve paresis is difficult. In total paralysis, aligning the visual axes is technically challenging. Realignment may be accomplished in only one field of gaze. Even if realignment is achieved, central sensory and motor fusion of images may not be maintained. Disrupted central fusion may give rise to intractable diplopia through loss of fusional amplitudes.53–56 Synoptophore evaluation and prisms neutralization may be required for diagnosis (see also Chapter 2).

Ocular Rotations and Muscle Function

The objectives of clinically evaluating ocular rotations are to determine which muscles are affected by the third nerve palsy and to gauge the severity of paralysis of each involved muscle. The eyes are examined during version testing in the six cardinal gaze positions (see Chapter 1). An eye that exhibits limited rotation that improves on monocular version testing probably has partial paresis rather than total palsy. Surgical strategies differ for total and incomplete palsy. A resection procedure will be helpful if there is residual muscle function but ineffective if palsy is complete.

Forced duction testing is done to identify restrictions or secondary lateral rectus (LR) contracture. Force generation testing and saccadic velocity analysis identify residual muscle function (see Chapter 3). Involvement of several muscles may require laboratory quantitation of muscle function.

It is imperative that fourth nerve function be assessed. Downward rotation caused by the SO may be difficult to evaluate in the setting of complete third nerve palsy. Incycloto-
torsion, however, can be assessed. The examiner asks the patient to follow a target such as a fingertip clockwise with the right eye and counterclockwise with the left eye. Incyclotorsion of the globe should be noted if the fourth nerve is spared. At times, it may be necessary to use the magnification afforded by the biomicroscope (slit lamp). A conjunctival or limbal vessel is used as a landmark and observed for movement along the y-axis.

**Laboratory Evaluation**

Binocular fields are helpful in defining any remaining area of single binocular vision. With isolated and partial muscle paresis there is often some residual area of binocular fusion. The size and location of any remaining binocular field will influence treatment planning.

**DIFFERENTIAL DIAGNOSIS**

Individual muscle paresis results from direct trauma to the nerve or a compressive lesion in the orbit. In addition, paresis (especially of individual muscles such as the SR) should be distinguished from restrictive conditions such as thyroid eye disease, an old blow-out fracture, postoperative sinus surgery, retinal detachment repair, congenital fibrosis syndrome, and peribulbar or retrobulbar injection. The diagnosis is aided by a history of medical disease that may affect the orbit or of surgery performed in and around the orbit.

Myasthenia gravis has the potential to create an isolated weakness of any individual muscle or any combination of muscles of the third nerve. Edrophonium (Tensilon) testing is indicated when the deviation is variable.

The early stages of progressive external ophthalmoplegia may be confused with early partial third nerve paresis. As the condition progresses to its full-blown state, however, distinctive features differentiate it from third cranial nerve palsy. These include incomplete bilateral but asymmetric ptosis; inability to move either eye in any field of duction or both eyes in a version effort; and, when chronic, restricted forced duction testing in all fields of attempted passive movement. In third nerve paresis, resistance is noted only if there is secondary LR contracture.

**Treatment**

As in all cranial nerve palsies, treatment objectives are divided into those appropriate during the acute phase of the insult and those that may be required 6 months later to yield a definitive solution. The management of third nerve palsy is more complicated because up to four muscles (the SR, MR, IR, and IO) may be involved and may display different degrees of insult. Thus, the level of function of each involved muscle must be carefully evaluated and the presence of secondary ipsilateral LR contracture identified. In addition, all patients will require evaluation for possible disruption of central fusion. If this is present, the prognosis for any type of realignment will be extremely guarded. Finally, if serious progressive neurologic disease is found, treatment goals may need to be modified in light of the patient’s general condition.

**NONSURGICAL MANAGEMENT**

Nonsurgical management is indicated during the acute phase, which may last as long as 6 months, and also when definitive surgical management is contraindicated (e.g., by neurologic disease or central fusion disruption).

**Occlusion**

Patching may be necessary to eliminate bothersome diplopia in third nerve palsy. Occasionally, the images may be so far apart that the patient is unaware of diplopia. In these cases, occlusion is not required. If diplopia persists after successful surgical realignment, occluder contact lenses may be required.

**Prisms**

Aligning the visual axes with prisms can be difficult because both horizontal and vertical correction is often required. If torsional diplopia exists, realigning the visual axes in the horizontal and vertical planes will not resolve this component of diplopia.

In partial paralysis of the third nerve with residual MR function and therefore a meaningful area of single binocular vision, patients may benefit from neutralizing prisms. The goal is to restore fusion at or near the primary position. Diplopia usually persists in other fields of gaze. The deviation (both horizontal and vertical) may be corrected using add-on (Fresnel) prisms on spectacles. Prisms may be used temporarily if there is a potential for reestablishing fusion and returning neural function and muscle contractility. If recovery does not occur and surgery is contraindicated, prisms (if they are not too large) may be incorporated into the lenses. Although heavier, they provide a clearer image and help maintain fusion.

**Botulinum Toxin**

In the acute phase of partial third nerve paresis or isolated MR palsy, botulinum toxin injection of the LR will decrease and often eliminate horizontal deviation in the primary position. The injections decrease the likelihood of contracture of the MR antagonist (the LR muscle).

After recovery of the injected muscle, the remaining vertical deviation may need to be corrected by prisms or surgery. In some cases, correcting the vertical deviation may not be necessary; the patient may adopt a slight change in vertical head position to attain fusion.

Using botulinum toxin for the vertical imbalance is rarely indicated. The SR should not be injected, because ptosis occurs when toxin is placed into the levator-SR complex. It may, however, be used when vertical deviation results from a tight IR that limits upward rotation. Injection of the IR may be done alone or in conjunction with other surgical procedures.

**SURGICAL MANAGEMENT**

Before surgery, repeated measurements are needed to gauge recovery of any of the involved muscles and determine the stability of misalignment for at least 6 months.
Evidence shows that patients with traumatic partial third nerve palsy may recover after 1 year.25

**Total Third Nerve Palsy**

The goal of surgery in total third nerve involvement is realignment in primary position. The surgeon may opt to perform a very large (supramaximal) LR recession well behind the equator to totally abolish abduction. SO tendon resection and transposition to the area between the MR and SR insertion has been proposed to create a chronic adducting force that will ensure long-term stability.5, 13, 23, 29, 41, 43 Historically, a large MR resection has been performed to serve as a “stay suture,” keeping the eyes aligned in primary gaze, but it usually is ineffective. Redundant conjunctiva present medially should be partially resected for optimal cosmesis.

Operations for total third nerve palsy may align the globe in primary gaze, but little or no horizontal rotation will occur and no meaningful area of single binocular vision is achieved. When the eyes are brought closer through motor alignment, the second image is also brought closer. Patients will probably require occluder contact lenses because of an inability to develop fusional vergence, which is due to a lack of meaningful horizontal gaze in either direction.

Ptosis is a significant problem. If it is fully corrected to match the uninvolved eye, there is a high risk of corneal complications due to exposure. An adequate Bell’s phenomenon is needed before ptosis can be repaired. Partial repair to elevate the lids may be preferred rather than completely elevating the droopy lid to avoid exposure problems.

**Partial Third Nerve Palsy**

In partial third nerve palsy, the goals of surgery can be broadened to include good primary motor alignment as well as to create, move to the center, and enlarge the field of single binocular vision.

**Isolated Muscle Involvement.** If there is residual MR function, MR resection can be done in combination with antagonist LR recession—on an adjustable suture when possible. The same strategy is used for isolated vertical rectus muscle involvement. The paretic muscle with residual function is resected and the antagonist (usually with secondary contracture) is recessed, preferably with an adjustable suture.

**Multiple Muscle Involvement.** If MR function is preserved, but IR function is poor, the patient will be hypertropic and exotropic (Fig. 18–3). The degree of IR involvement can be determined by force generation testing or saccadic velocity analysis. If there is some meaningful residual function, the IR may be recessed in conjunction with LR recession and MR resection, using a ciliary vessel sparing technique on at least one of the muscles. In the absence of IR function, a horizontal recession-resection is combined with transposition of the insertions inferiorly to the IR insertion to provide a downward force (Fig. 18–4).2, 26, 27 One may also recess the contralateral uninvolved IR, with or without a posterior fixation suture, to match the deficiency in downgaze and expand the field of binocular vision. If there is poor SR function but horizontal MR function is intact, the patient may be managed using a similar plan.

Alternatively, Knapp19 suggested transposition of the SR to the MR and the LR to the IR for patients with paresis of the inferior division of the third nerve. In addition, he recommended tenectomy of the ipsilateral SO. Weakening of the SO may be included to reduce exotropia if the patient demonstrates significant in cyclotorsion. Otherwise, SO tenectomy may provoke excyclotorsion.

If there is no MR function but meaningful residual SR and IR function, both vertical rectus muscles may undergo a small resection (to enhance the transposition medially) and be transposed to the MR insertion.45 In all situations, torsion should be measured and treated as appropriate (see Chapter 4).

**Aberrant Regeneration.** Patients with aberrant regeneration and a partial return of MR function may exhibit elevation of the lid on attempted adduction. A large LR recession combined with a large MR resection may correct the exotropia and also improve ptosis.
COMPLICATIONS

Complications seen with any strabismus surgery are discussed in Chapter 41. Those unique to third nerve palsy relate to three areas of concern. First, diplopia that may be only a minor problem before surgery in patients with traumatic central disruption of fusion may become a significant problem after successful surgery because of the proximity of two images. If diplopia is detected preoperatively, surgery might be avoided. Second, in attempting to move the area of single binocular vision closer to primary position in partial paresis, binocular diplopia-free fields may contract after surgery. Lastly, torsional diplopia may develop after SO transfer operations. In this situation, symptomatic relief may be achieved by contact lens occlusion.

Conclusions

The ophthalmologist should use a logical stepwise approach in diagnosing third nerve paralysis:

1. It is necessary to ascertain whether life-threatening problems exist.
2. An evaluation to determine complete or partial third nerve palsy or isolated muscle abnormalities must be done.
3. The various conditions that may produce similar findings should be considered, identified, and excluded.

Once the diagnosis is established, strategies for treatment are addressed. Not all patients require intervention. Visual needs and rehabilitation for work and daily function must be part of the decision-making process. Management may include watchful neglect, patching, temporary or permanent prisms, and surgery. Realistic goals should be set. The patient, if capable, should participate in the decision-making process.

REFERENCES

The sixth cranial (abducens) nerve is the most frequently involved cranial nerve in ocular motor palsy. Because of its long, tortuous course, and the location of the peripheral part of the nerve near the clivus as it enters the area of the cavernous sinus, elevated intracranial pressure from any cause may tether the nerve, causing lateral rectus (LR) paresis. As such, it is regarded as a nonlocalizing nerve palsy. Other nearby anatomic structures, from the nucleus throughout its peripheral course, define several syndromes involving the sixth nerve. The reader is referred to the appropriate text for details of anatomy.

### Epidemiology and Risk Factors

#### SIXTH CRANIAL NERVE PALSY IN ADULTS

In adults, sixth nerve palsy most often occurs after age 40 to 50 years and may be due to a variety of insults (Table 19–1). Among cases in which a cause can be identified, vasculopathies due to hypertension, diabetes, or atherosclerosis are the most common. In younger adults the etiology may be more obscure, and aggressive search for a cause is imperative.

#### SIXTH CRANIAL NERVE PALSY IN CHILDREN

The causes of sixth nerve palsy in children are similar to those in adults, but vascular diseases are less often encountered, whereas trauma and neoplasia occur more often (Table 19–2). Some cases may initially masquerade as benign sixth nerve palsy or may be secondary to an infection. In the absence of trauma, a careful search should be conducted for a neoplasm, central nervous system infection, elevated intracranial pressure, or hydrocephalus. Benign idiopathic isolated sixth nerve palsy in infants and children is a diagnosis of exclusion.

In several series, viral causes were uncommon and often unilateral. Many of these palsies developed 1 to 3 weeks after a nonspecific febrile illness or were associated with immunization. The viral agent is believed to have a neurotropic effect that results in sixth nerve palsy. Spontaneous recovery within 6 months is the rule. Transient sixth nerve palsies in neonates may be caused by elevated intracranial pressure associated with forceps or vacuum extraction in assisted deliveries, overstretching of the sixth cranial nerve, hypoxia, or temporary edema caused by the birth process. Patients usually recover within 6 months.

### Genetics

Familial recurrent cranial nerve palsy, involving the sixth cranial nerve in association with the seventh (facial) nerve, may be inherited in an autosomal dominant pattern. An idiopathic hereditary vasculopathic process has been suggested. In the absence of associated congenital syndromes such as Möbius syndrome, no genetic factor for sixth nerve palsy has been discovered.

### Clinical Characteristics

#### CLINICAL FINDINGS

In acute sixth nerve palsy, the patient notices an abrupt onset of horizontal diplopia. Many will improve spontaneously. Surgical intervention is recommended only for patients who have persistent esotropia for at least 6 months after the acute onset. During this period, patients may be incapacitated by diplopia.

In mild cases, diplopia may be intermittent and manifest only as a small esophoria in primary gaze. The esophoria may be exaggerated when the eye is brought into the field of action of the paretic sixth nerve. On cover-uncover testing, the esophoria may break down to become an esotropia. The diplopia may be controlled by a small head turn that can escape detection unless suspected.
Horizontal incontinence is a prominent feature of esotropia in unilateral sixth nerve palsy. The esotropia is larger in the field of gaze of the paretic LR (Fig. 19–1). In bilateral palsies, the deviation may be fairly symmetric in all positions of gaze. A V-pattern strabismus may result from the lessened abduction and unopposed medial rectus (MR) action in downgaze.

RECOVERY

Spontaneous recovery from acute sixth nerve palsy depends on its cause.5 The majority of patients with infectious, vascular (diabetic, hypertensive, atherosclerotic), or idiopathic sixth nerve palsy will recover.5 105, 131, 146 In cases due to more serious pathology such as tumor, anatomic abnormality, trauma, or underlying neurologic disease, recovery is less likely.5 80, 118 146 Spontaneous recovery has, however, been reported with basal skull tumors.172 Overall, 20% to 50% of patients fail to recover from acute sixth nerve palsy.80, 118 Mean recovery times range from 2.4 to 5.4 months.80 131 A third of patients recover within the first 2 months, and 94% recover by 6 months.80

Table 19–1. Causes of Sixth Nerve Palsy in Adults

<table>
<thead>
<tr>
<th>Infections</th>
<th>Arachnoiditis59</th>
<th>Lyme disease62, 98</th>
<th>Psittacosis184</th>
<th>Staphylococcus aureus infection65</th>
<th>Syphilis68 182</th>
<th>Varicella zoster120</th>
</tr>
</thead>
<tbody>
<tr>
<td>Trauma</td>
<td>Cervical spine fractures6, 118, 153</td>
<td>Head injury20, 42, 97, 134, 195</td>
<td>Skull fracture112 156</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Neoplasms</td>
<td>Chondroma112</td>
<td>Chondrosarcoma65, 112, 126</td>
<td>Chordroma1 92, 126</td>
<td>Cylindroma65 126</td>
<td>Metastatic65 112, 122, 152, 163</td>
<td>Meningioma59 112, 126, 152</td>
</tr>
<tr>
<td></td>
<td>Neurinoma7</td>
<td>Nasopharyngeal carcinoma17, 126, 152</td>
<td>Pituitary adenoma48 112, 126, 152, 183</td>
<td>Plasmacytoma126</td>
<td>Pontine glioma69</td>
<td>Schwannoma38</td>
</tr>
<tr>
<td></td>
<td>Sphenoidal sinus tumor110, 112, 126</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Systemic Disorders</td>
<td>Atherosclerosis12, 146</td>
<td>Diabetes57, 73, 112, 145, 146, 152</td>
<td>Hypertension13, 112, 146, 152</td>
<td>Preeclampsia11</td>
<td>Hematologic Leukemia64, 178</td>
<td>Lymphomatous meningitis112</td>
</tr>
</tbody>
</table>

Other Vascular Causes

Aneurysm24, 44, 58, 80, 111, 131, 152, 166 | Arteriovenous malformation60, 131 | Carotidocerebral insufficiency57, 80 | Cranial (temporal or giant cell) arteritis57, 59, 73, 80, 152 | Internal carotid artery dissection89 |

Associated Neurologic Disorder

Cluster headache124 | Demyelinating disease112, 136, 152 | Elevated intracranial pressure19, 161, 166 | Intracranial hypertension15 |

Iatrogenic

Myelography12, 58, 109, 152 | Nerve blocks in the head and neck74, 78, 102 | Post lumbar puncture68, 68, 112 | Post spinal or epidural anesthesia129, 132, 176 |

Others

Idiopathic6, 112, 131, 152 | Inflammatory12 | Interferon therapy18 | Lithium toxicity18 | OKT2 therapy18 |

Table 19–2. Causes of Sixth Nerve Palsy in Children

<table>
<thead>
<tr>
<th>Infections</th>
<th>Small cell carcinoma20</th>
<th>Lymphosarcoma179</th>
<th>Rhadomyosarcoma125, 136</th>
</tr>
</thead>
<tbody>
<tr>
<td>Systemic Disorders</td>
<td>Vascular Disease</td>
<td>Atherosclerosis145</td>
<td>Diabetes145</td>
</tr>
<tr>
<td></td>
<td>Hypertension145</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Others</td>
<td>Dermatomyositis65</td>
<td>Leukemia65, 84, 130</td>
</tr>
<tr>
<td></td>
<td>Gaucher disease135</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Other Vascular Causes</td>
<td>Aneurysm60, 131, 135</td>
<td>Arteriovenous malformation80, 134, 135</td>
</tr>
<tr>
<td></td>
<td>Associated Neurologic Disorders</td>
<td>Arnold-Chiari malformation27</td>
<td>Demyelinating disease60, 84, 135</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Elevated intracranial pressure60, 175</td>
<td>Hydrocephalus62, 65, 84, 105, 169, 175</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Malignancy20</td>
<td>Pseudotumor cerebri85, 84, 133, 170</td>
</tr>
<tr>
<td></td>
<td>Others</td>
<td>Congenital13</td>
<td>Idiopathic65, 84, 127, 131, 135</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Immunization20, 83, 84, 130, 166, 175, 177, 179</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Inflammation65, 125, 136, 169</td>
<td>Lead poisoning65</td>
</tr>
</tbody>
</table>

In some cases, the actual cause may not be identified for as long as 6 months.90 Acquired sixth cranial nerve palsy may be a harbinger of serious intracranial pathology.3 133 Associated warning signs include papilledema and nystagmus. Additional neurologic signs may not become evident until several months later.3 135

RECURRANCE

Recurrences of sixth nerve palsy may occur more than once and have been reported in both children and adults.2 3, 19, 32, 104, 127, 165, 169, 171, 175 They are usually painless and occur on the same side.2 3 32 Theories include increased susceptibility of the sixth nerve once it has been exposed to inflammation.19 An aberrant anteroinferior cerebellar artery may intermittently constrict the abducens nerve.10 Childhood ophthalmoplegic migraine may also be associated with recurrent palsy.30 Intervals between episodes range from a week to 12 years.2 10, 19, 20, 127, 165, 169, 175 In some series, there is a left-sided predominance and females are affected more often than males.3, 20, 32, 169

Diagnosis

CLINICAL DIAGNOSIS

Presenting Findings

Patients usually present in the ophthalmologist’s office with a complaint of horizontal diplopia. A compensatory
Paresis Versus True Palsy

It is imperative to distinguish paresis from a true palsy in cases with sixth nerve involvement (Table 19–3). Paresis is a situation in which function of the nerve may be compromised but residual LR function remains. In true palsy, function of the nerve has been completely eradicated so that no meaningful LR contraction occurs. Simple observation of abduction is not sufficient to detect residual function of the LR. When no abduction past midline is observed, limited rotations may be due to either a tight MR or true LR palsy.141

The ophthalmologist has recourse to clinical tests such as forced duction and force generation testing and saccadic velocity analysis to evaluate muscle function.103, 108, 154, 156

In the acute stage, no resistance to full abduction will be noted with forced duction testing. However, in long-standing cases when secondary contracture of the MR muscle has occurred, resistance may be felt because of restriction by the contracture.103 A paretic muscle will have some residual force generation and will generate saccades, albeit reduced. A palsied muscle will generate neither force nor saccades. A characteristic “floating” saccade may be observed on clinical examination.103, 108, 154, 156 This distinction is paramount in determining the appropriate surgical strategy. The importance of testing muscle function has been stressed in Chapter 3.

A clinical trial with botulinum toxin during the acute phase may distinguish paresis from a true palsy.103, 135 After botulinum toxin injection of the antagonist MR muscle, residual function of the paretic LR is apparent if abduction improves. The disadvantage of this procedure is the need to delay definitive surgery by 4 to 8 weeks while waiting for the effects of the toxin to wear off.103, 141 Alternatively, injecting lidocaine (which has a shorter duration of action) into the antagonist MR may yield similar information.141

Table 19–3. Partial (Paretic) Versus Complete (Palsied) Sixth Nerve Palsy

<table>
<thead>
<tr>
<th>Parameters</th>
<th>Partial Sixth</th>
<th>Complete Sixth</th>
</tr>
</thead>
<tbody>
<tr>
<td>Saccadic velocity of LR (20 degrees)</td>
<td>More than 175 degrees/s</td>
<td>Less than 100 degrees/s</td>
</tr>
<tr>
<td>Agonist-antagonist saccadic velocity difference</td>
<td>≤40% difference</td>
<td>&gt;40% difference</td>
</tr>
<tr>
<td>Force generation</td>
<td>Reduced</td>
<td>Absent</td>
</tr>
<tr>
<td>Forced duction</td>
<td>No restriction unless MR contracted</td>
<td>No restriction unless MR contracted</td>
</tr>
<tr>
<td>Abduction</td>
<td>Usually beyond midline; if MR contracted, may see decreased abduction</td>
<td>No abduction beyond midline</td>
</tr>
</tbody>
</table>

LR, lateral rectus; MR, medial rectus; >, more than; ≤, less than or equal to; s, second.
Follow-Up

Observation should be conducted on a regular basis to document progression, resolution, or stabilization. Further evaluation of sixth nerve palsy, beyond the strabismus evaluation, is outside the scope of this chapter. Briefly, the following recommendations are proposed:

1. In children younger than 14 years of age with no other neurologic signs and symptoms, no workup is indicated, but they should be examined every 2 weeks initially and then monthly until sixth nerve palsy resolves. Persistent palsy after 3 months requires further evaluation.

2. In young adults age 15 to 40 years, risk factors should be sought.

3. Patients older than 40 years are evaluated for diabetes, hypertension, and atherosclerosis. In patients older than 55 years a workup for giant cell arteritis should be performed.

4. Patients with increasing esotropia and those developing neurologic signs and symptoms require further evaluation.

Blood studies include a fasting blood glucose determination, complete blood cell count, erythrocyte sedimentation rate, antinuclear antibody titers, rapid plasma reagin test, fluorescent treponemal antibody absorption test, thyroid function tests, and so on. Lumbar puncture studies and edrophonium testing may be required. Orbital ultrasonograms may be obtained to rule out enlarged extraocular muscles. Nasopharyngeal biopsy may be required in some cases. Radiologic evaluation includes bone scans, cerebral angiography, computed tomography, and magnetic resonance imaging.

LABORATORY DIAGNOSIS

Laboratory evaluation of muscle function is required frequently because of the clinical diagnostic techniques described earlier and in Chapter 3. In children, however, who may be less cooperative and in whom botulinum toxin may not be a reasonable diagnostic alternative, laboratory evaluation of horizontal saccadic velocities may provide the information needed to differentiate sixth nerve paresis from palsy (Fig. 19–2). A 40% or greater difference between saccadic velocities of the agonist and antagonist muscles is considered diagnostic of a true palsy. Twenty-degree saccades are sufficient to study lateral rotations (e.g., from 30 degrees adduction to 10 degrees adduction) (Fig. 19–3). Saccadic velocity analysis also serves to evaluate improvement after surgical intervention. Evaluation of binocular diplopia-free fields may be indicated before surgery for purposes of documentation. After the surgical procedure, improvement in the size and centration of diplopia-free fields may be demonstrated (Fig. 19–4). The most accessible and commonly used equipment for this purpose is the Goldmann visual field perimeter, using the size III 4e target.

DIFFERENTIAL DIAGNOSIS

Any disorder that causes an abduction deficit should be included in the differential diagnosis of sixth nerve palsy (Table 19–4, Figs. 19–5 and 19–6). Bilateral sixth nerve palsies have a uniquely different set of differentials (Table 19–5). Many can be diagnosed by a good clinical evaluation without the need for sophisticated tests. Some examples are discussed on the following pages.
In fibrosis syndromes, the loss of abduction is usually complete. Forced duction testing usually reveals a restriction. Occasionally, the diagnosis is confirmed only at the time of strabismus surgery when a fibrotic extraocular muscle is found (see Chapter 27).

Duane syndrome is associated with globe retraction and narrowed lid fissures on adduction. In type I Duane syndrome, the magnitude of the esotropia in primary gaze is small compared with what may be expected from LR palsy with a comparable abduction deficit. Contraction of the LR muscle in primary gaze in Duane syndrome, as opposed to the absence of innervation in patients with sixth nerve palsy, leads to a smaller amount of esodeviation in the former condition. The patient may show some limitation of adduction that does not occur in sixth nerve palsy. Forced duction testing may occasionally show paradoxical results. Electromyography will demonstrate co-contraction of the LR on adduction. Saccadic velocities are slowed on abduction and adduction. In children younger than 2 years, the abduction deficit may be manifest before other components of Duane syndrome have appeared (see also Chapters 3 and 24).

Inflammatory disorders involving the orbit or the globe may involve the LR muscle and cause limited abduction (see Fig. 19–6). Restricted lateral rotations may be caused by inflammatory congestion of the MR, leading to a “tight”


**Table 19–4. Differential Diagnosis for Unilateral Sixth Cranial Nerve Palsy**

<table>
<thead>
<tr>
<th>Congenital</th>
</tr>
</thead>
<tbody>
<tr>
<td>Congenital absence of lateral rectus¹⁰³</td>
</tr>
<tr>
<td>Congenital fibrosis³</td>
</tr>
<tr>
<td>Duane syndrome, types I and II²², 25, 41, 58, 103, 104, 113, 151</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Postsurgical</th>
</tr>
</thead>
<tbody>
<tr>
<td>Disinsertion or dehiscence of lateral rectus muscle¹⁰³</td>
</tr>
<tr>
<td>Excessive medial rectus resection¹⁰³</td>
</tr>
<tr>
<td>Excessive lateral rectus recession¹⁰³</td>
</tr>
<tr>
<td>Lost lateral rectus¹⁰³</td>
</tr>
<tr>
<td>Scarring and restriction on the medial aspect of globe¹⁰³</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Restrictive</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tight medial rectus¹⁰³, 112</td>
</tr>
<tr>
<td>Graves ophthalmopathy²², 59, 151</td>
</tr>
<tr>
<td>Medial orbital wall fracture²², 59, 151</td>
</tr>
<tr>
<td>High myopia with compression or restriction⁶, ³⁹</td>
</tr>
<tr>
<td>Medial rectus adherence to nasal orbital wall¹⁰¹</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Neurologic Disorders</th>
</tr>
</thead>
<tbody>
<tr>
<td>Horizontal gaze palsy¹⁰³</td>
</tr>
<tr>
<td>Myasthenia gravis²², 59, 103, 112, 161, 152</td>
</tr>
<tr>
<td>Progressive external ophthalmoplegia¹⁰³</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Miscellaneous</th>
</tr>
</thead>
<tbody>
<tr>
<td>Inflammatory orbital disease¹⁸, ²², 151</td>
</tr>
<tr>
<td>Orbital amyloidosis¹¹²</td>
</tr>
</tbody>
</table>

**Table 19–5. Differential Diagnosis for Bilateral Sixth Nerve Palsy**

| Convergence spasm²², 59, 148, 151                                         |
| Divergence palsy⁸¹, 95, 135                                                |
| Graves ophthalmopathy²², 59, 151                                           |
| Bilateral high myopia with compression or restriction⁶, ³⁹                 |
| Horizontal gaze palsy¹³                                                  |
| Infantile esotropia²², 104                                                |

MR. Graves ophthalmopathy may cause abduction deficits by the same mechanism (see Chapter 21).

Other mechanical causes of diminished abduction include excessive resection of the MR muscle, scarring and hemorrhage of the medial aspect of the globe causing a restriction, secondary contracture of the MR after LR palsy or long-standing esotropia, and congenital aberrant insertion of the MR.¹⁰³ A mechanical restriction can be confirmed by forced duction testing, whereas LR strength is validated by force generation testing.

Entrapment of the MR in the medial orbital wall after orbital trauma may be suggested by a careful history. Forced duction testing will reveal restricted abduction. Neuroimaging studies may reveal entrapment of the MR (see also Chapter 23).

LR weakness may be a result of excessive resections, traumatic disinsertion, or muscle slippage.¹⁰³ LR paresis has also been observed in association with high myopia.⁸ A progressive myopathy with fibrosis, in which muscle tissue is replaced by interstitial connective tissue and thickening of the basement membrane, has been documented. The LR muscle may be pressed against the lateral bony orbital wall by the enlarged myopic eye, causing a slow paralytic effect and development of an MR contracture.⁹ High myopia with elongated globes also may mechanically restrict full ocular rotation.³⁹

Involvelement of the LR muscle in myasthenia gravis is often associated with fluctuating ptosis and diplopia that becomes worse with fatigue. Strabismus is variable in the early stages but may become stable in time.³⁶ An edrophonium (Tensilon) test may help establish the diagnosis. Saccadic velocity analysis shows characteristic "quiver saccades," fatigue effects, and improvement with edrophonium (see also Chapter 3 and Chapter 29).

Infantile esotropia with cross fixation may be confused with bilateral sixth nerve palsy.¹⁰⁴ Full abduction may not be elicited unless an orthoptic patch is placed over one eye. Doll’s head and vestibulo-ocular maneuvers elicit abduction. Saccadic velocity analysis will show normally rapid abducting saccades. In long-standing cases the MR may tighten secondarily, causing an abduction deficit. After MR recession, abduction usually improves.

---

**Figure 19–5. Patient with limited abduction after developing restriction secondary to scleral buckling procedure for retinal detachment repair. Horizontal incomitance looks clinically similar to sixth cranial nerve palsy.**
In horizontal gaze palsy, there is a lack of ability to generate binocular saccadic or pursuit eye movements to some eccentric orbital position.\textsuperscript{13} A compensatory head turn may resemble that seen in unilateral sixth nerve palsy. With monocular patching, the anomalous head posture should disappear in abducens palsy but will persist in gaze palsy.

Divergence palsy with a mild reduction in abduction may be confused with bilateral sixth nerve paresis. This subject is discussed in detail in Chapter 11.

**Treatment**

**NONSURGICAL MANAGEMENT**

In children up to 4 years of age, treatment of acute sixth nerve palsy is aimed at preventing amblyopia and preserving binocular fusion.\textsuperscript{5, 137, 139, 141} Although diplopia may be a prominent symptom during the acute phase, children may rapidly learn to suppress the second image, which can lead to amblyopia. Standard amblyopia therapy should be instituted as early as possible. Adopting a small face turn is beneficial in maintaining binocular vision, and parents should permit their children to assume the anomalous head posture. Loss of the head posture with persistence of an incomitant esotropic deviation is an ominous sign, indicating loss of fusion and the subsequent development of amblyopia.

Alternate occlusion to prevent secondary contracture of the MR may be tried. Patching also relieves diplopia.\textsuperscript{123} However, occlusion of the good eye may lead to disorientation and vertigo.\textsuperscript{22, 116} For small deviations, Fresnel add-on prisms may be given and reduced as the sixth nerve palsy improves. Vision may be blurred by the prisms, in proportion to the amount of prism applied.\textsuperscript{118} If a child loses binocularity and is not recovering sixth nerve function, chemodenervation of the antagonist MR may help to rapidly restore binocularity.\textsuperscript{157, 158}

In adults with acute sixth nerve palsy, intervention is directed at preventing secondary contracture of the antagonist MR muscle and creating a meaningful area of single binocular vision during the recovery period.\textsuperscript{58, 91, 105, 115, 137, 139-141} The optimal timing for botulinum toxin injection is variable. Many patients spontaneously recover, especially if sixth nerve palsy is related to diabetes. The decision to use botulinum toxin will depend on the degree of incapacitation caused by the sixth nerve palsy. If involvement is only partial and the patient has an area of binocular single vision, the ophthalmologist may consider delaying intervention. If involvement is total, some surgeons may elect to use botulinum toxin as early as 2 weeks.\textsuperscript{137, 141} Others recommend botulinum toxin if no signs of improvement are apparent within a month of onset.\textsuperscript{138} Chemodenervation reduces the length of morbidity.\textsuperscript{115, 118, 141}

In a nonrandomized study, in which the control group consisted of patients who refused chemodenervation therapy, 70\% of patients with unilateral sixth nerve palsy who did not receive botulinum toxin required surgery, compared with only 10\% of those who received the toxin. The need for surgery in bilateral sixth nerve palsies does not appear to be reduced by concomitant use of botulinum toxin during the acute phases.\textsuperscript{107}

Prospective randomized clinical trials, on the other hand, showed no evidence of any difference between the treated and untreated groups in terms of ultimate outcome. Botulinum-treated patients, however, reported subjective improvement.\textsuperscript{33, 90, 91} These studies included a large number of patients with sixth nerve palsy caused by diabetes. Such patients are known to have a high rate of spontaneous resolution. The use of botulinum toxin may reduce the number of rectus muscles requiring surgery later or may avoid surgery altogether.\textsuperscript{48, 91, 156, 174} Although not as effective as surgery, botulinum toxin is helpful in cases of chronic sixth nerve palsy with residual abduction.\textsuperscript{16, 126} Botulinum toxin is also beneficial in cases in which surgery may be contraindicated.\textsuperscript{48, 91}

One disadvantage is that patients with only a paretic LR who adopt a face turn for binocular vision may note a temporary contraction of the binocular diplopia-free field after botulinum toxin injection because of induced MR paresis.\textsuperscript{48} Chemodenervation is discussed further in Chapter 32.

**SURGICAL MANAGEMENT**

**Operative Strategies**

Six months after the onset of an acute sixth nerve palsy, the chance for spontaneous recovery is greatly reduced.\textsuperscript{16, 80}
It is at this juncture that surgery directed at correcting an incomitant esodeviation, improving abduction, and increasing the size and centration of diploplia-free binocular field may be considered.141

Forcedduction testing to identify MR contracture should be done before surgery and repeated while the patient is under anesthesia.

Surgical strategies for sixth nerve paresis (or partial sixth nerve palsy) and total palsy are different (Table 19–6). If preoperative evaluation shows any significant muscle force generation, a recession–resection of the horizontal rectus muscles is the best procedure.54, 143 Resection procedures of the LR muscle are only useful if some amount of residual LR function exists.141 Small deviations of up to 20 PD may be treated by unilateral LR resection of 5.5 to 8.0 mm. This procedure will not be sufficient if an MR contracture coexists. LR resection is usually combined with MR recession.75

An adjustable suture on the recessed MR enhances the surgeon’s ability to provide centration of the binocular diplopia-free field.140, 141 A contralateral MR recession combined with a unilateral recession–resection procedure may be needed if the primary gaze deviation is more than 40 PD.

In complete sixth nerve palsy, surgery should be aimed at achieving the following goals: (1) create an abducting force, (2) increase the binocular diplopia-free field, and (3) provide centration of the binocular diplopia-free field.

In total sixth nerve involvement, resection of the LR muscle may have a transient mechanical result but will result in poor long-term alignment.103 A mechanical effect may be achieved by performing supramaximal MR recession (10–15 mm) and large LR resections (10–12 mm) without any improvement in generation of abduction saccades.103, 106, 116

This practice has resulted in undercorrections after surgery for sixth nerve palsy. The reoperation rate is estimated to be as high as 50%. Moreover, a smaller binocular diplopia-free field is obtained after surgery when compared with transposition procedures, because of the profound limitation of both abduction and adduction.91, 118 More importantly, resection of a palsied LR muscle destroys anterior ciliary circulation from the rectus muscles. This increases the risk of anterior segment ischemia after subsequent vertical rectus transposition surgery. Surgical transposition with botulinum toxin injected into the ipsilateral MR is less effective than when done as a first procedure.91

If preoperative evaluation does not disclose significant muscle force generation, a transposition procedure should be performed as the initial operation and the paretic muscle left undisturbed to preserve the blood supply to the anterior segment.150 Vertical rectus muscle transposition of the superior rectus (SR) and inferior rectus (IR) muscles to the LR insertion improves abduction (see Fig. 19–1). It is commonly combined with a MR weakening procedure.139 MR recession is advised for esotropia of more than 25 PD.55 If the deviation is less than 75 PD, some authors advocate 4.5- to 6-mm MR recession; if the deviation exceeds 75 PD, MR recession larger than 6 mm may be required.150 Alternatively, pharmacologic weakening of the antagonist MR may be achieved by injecting botulinum toxin.30, 103, 123, 181 If MR recession is chosen, this means that three rectus muscles will be disinserted in the same eye to achieve the surgical goal. This increases the risk of anterior segment ischemia in older patients and patients with microangiopathies such as diabetes.141 Sparing the anterior ciliary vessels56 may be difficult because of the distance the muscle insertion is moved by the transposition. Other transposition procedures such as the rectus muscle union (Jensen procedure55, 74, 161) and partial transposition (Hummelsheim procedure49, 71) are preferred by some, but anterior segment ischemia has been reported in both normal and predisposed children65, 82, 155 and adults.54, 61, 150, 173 Transposition procedures, as well as ciliary vessel sparing procedures, are discussed in Chapters 36 and 39.

**Table 19–6. Surgical Strategies for Sixth Nerve Palsy**

<table>
<thead>
<tr>
<th>Partial Sixth Nerve Palsy</th>
<th>Total Sixth Nerve Palsy</th>
</tr>
</thead>
<tbody>
<tr>
<td>Medial rectus (MR) recession</td>
<td>Partial vertical rectus transposition + MR weakening</td>
</tr>
<tr>
<td>Lateral rectus (LR) resection</td>
<td>(botulinum toxin or recession)</td>
</tr>
<tr>
<td>MR recession and LR resection (R &amp; R)</td>
<td>Full tendon vertical rectus transposition + MR weakening</td>
</tr>
<tr>
<td>R &amp; R and contralateral MR recession</td>
<td>Rectus muscle union + MR weakening</td>
</tr>
</tbody>
</table>

**Botulinum Toxin**

Botulinum toxin injection of the MR instead of a recession has been proposed to enhance the results of vertical rectus muscles transposition (Fig. 19–7).48, 49, 77, 91, 100, 143 This allows operation on only two rectus muscles, decreasing the risk of anterior segment ischemia. Anterior segment ischemia has been reported after botulinum toxin injection of the MR and vertical transposition, but several associated risk factors (including systemic vascular disease and manipulation of the LR or the limbal conjunctiva) may have contributed to the ischemic process.77 Undercorrection is managed by either reinjection or recession of the contralateral MR. Botulinum toxin injection may be done at the time of surgery,100 within 15 days after surgery,138 or 4 to 11 days before the transposition procedure.49, 92

Strabismologists who prefer preoperative injection cite the following advantages45: (1) paralysis of the antagonist MR at the time of transposition surgery permits an easier maneuver because one does not need to tighten the vertical rectus against a contracted MR; (2) the toxin is injected into a normal anatomic tissue compartment and is more likely to stay within the MR; (3) the transposition surgery is reduced to a two-muscle procedure, allowing greater postoperative comfort.

We prefer administering the botulinum toxin injection several days postoperatively so that we can differentiate any induced vertical deviation from the transposition procedure. In addition, the botulinum toxin dose can be tailored to the size of residual esotropia. (Some cases will not require botulinum toxin because postoperative alignment is satisfactory after transposition surgery alone.)138 High-quality electromyographic localization with botulinum toxin injection should prevent inadvertent injection of anatomic structures other than the MR.
Figure 19–7. Same patient in Figure 19–1 after postoperative botulinum toxin injection. Top photographs were taken 1 week after injection of 5 units of botulinum toxin into the right medial rectus. Note full abduction in the right eye, exotropia in primary gaze, and pharmacologic weakening of the right medial rectus. Bottom photographs show the same patient 2 months after injection. Note orthotropia in primary position and improved abduction and adduction. Patient has 50 degrees of binocular diplopia-free field. (From Rosenbaum AL, Kushner BJ, Kirschen D: Vertical rectus muscle transposition and botulinum toxin [Oculinum] to medial rectus for abducens palsy. Arch Ophthalmol 1989;107:820–823. Copyright 1989. American Medical Association.)

Assessment

Surgical results should be evaluated using the following parameters: deviation in primary gaze, residual face turn, size of the diplopia-free binocular field, and the size of the binocular diplopia-free field in the field of the paretic LR (Table 19–7). Before the availability of botulinum toxin, Murray advocated a supermaximal horizontal recession-resection procedure (MR recession of 12–16 mm and LR resection of 8–10 mm) combined with contralateral MR recession (7–9 mm) to achieve good cosmesis and centration of binocular fields in complete sixth nerve palsy. Supermaximal surgery is recommended only for patients with no sixth nerve function. If the LR muscle has some function, this type of surgery will result in gross overcorrection. More recently, Murray acknowledged the advantages of performing a transposition procedure. Supermaximal surgery is now reserved for undercorrected patients who had previous horizontal recession-resection surgery and in whom a transposition procedure with vessel sparing is not possible.

A vertical rectus transposition procedure combined with pharmacologic weakening of the MR muscle resulted in a

Table 19–7. Results of Surgery for Sixth Nerve Palsy

<table>
<thead>
<tr>
<th>Author</th>
<th>Procedure</th>
<th>No.</th>
<th>Abduction (degrees)</th>
<th>Binocular Fields (degrees)</th>
<th>Residual Esotropia (%)</th>
<th>Residual Face Turn (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Murray et al</td>
<td>Supermaximal R&amp;R</td>
<td>12</td>
<td>11</td>
<td>25</td>
<td>25</td>
<td>25</td>
</tr>
<tr>
<td>Jensen</td>
<td>Rectus muscle union</td>
<td>16</td>
<td>&gt;25: 60%</td>
<td>69% with fusion</td>
<td>12.5</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>10–25: 35%</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>&lt;10: 6%</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Cline and Scott</td>
<td>Jensen</td>
<td>9</td>
<td>&gt;25: 26%</td>
<td>41</td>
<td>30</td>
<td>47</td>
</tr>
<tr>
<td>Frueh and Henderson</td>
<td>Jensen</td>
<td>11</td>
<td>10–25: 36%</td>
<td>50% fusion</td>
<td>20</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>&lt;10: 27%</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Selezinka et al</td>
<td>Jensen</td>
<td>16</td>
<td>18</td>
<td>50% fusion</td>
<td>38</td>
<td>31</td>
</tr>
<tr>
<td>Scott et al</td>
<td>MR recession with adjustable transposition</td>
<td>13</td>
<td>18</td>
<td>36 (range: 0–80)</td>
<td>40</td>
<td></td>
</tr>
<tr>
<td>Ciancia et al</td>
<td>Full vertical transposition with botulinum toxin to MR</td>
<td>3</td>
<td>18</td>
<td>40</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Rosenbaum et al</td>
<td>Vertical transposition + MR weakening (recession or botulinum toxin)</td>
<td>10</td>
<td>21</td>
<td>51</td>
<td>20</td>
<td>20</td>
</tr>
<tr>
<td>Sarnicola</td>
<td>Vertical rectus transposition and MR recession</td>
<td>28</td>
<td>23 (range: 15–40)</td>
<td>51.6 (range: 0–85)</td>
<td>21</td>
<td></td>
</tr>
<tr>
<td>Rosenbaum et al</td>
<td>Full vertical rectus transposition and MR recession</td>
<td>8</td>
<td>17</td>
<td>44</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Foster</td>
<td>Vertical transposition with lateral fixation suture</td>
<td>2</td>
<td>21</td>
<td>71</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Foster</td>
<td>Vertical transposition with lateral fixation and MR recession</td>
<td>2</td>
<td>26</td>
<td>76</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

MR, medial rectus; R & R, medial rectus recession and lateral rectus resection.
mean diplopia-free binocular field of 51 degrees compared with almost none preoperatively. The Jensen procedure was able to achieve improved fields of 41 degrees. From 30 to 90 PD of esotropia in primary gaze may be corrected with this technique. Adjustable MR recession as an alternative to pharmacologic MR weakening allowed centration of the binocular diplopia-free fields. Posterior fixation suture augmentation of vertical rectus transposition has been described; the preliminary results are promising in terms of improving abduction and increasing diplopia-free binocular fields.

COMMON PITFALLS

Failure to distinguish between paresis and palsy may lead to the incorrect choice of surgical strategy. Similarly, failure to identify MR contracture may lead one to forego performing an MR weakening procedure. Inappropriate selection may contribute to complications in certain patients. In Möbius syndrome, for example, in which bilateral sixth nerve palsy is associated not only with facial diplegia but also with gaze palsy and asymmetric LR fibrosis, transposition surgery may be contraindicated. A combined paretic and restrictive strabismus is observed, and recession of the tight fibrotic MR may achieve good results.

COMPLICATIONS

Induced Vertical Deviation

The major concern with vertical rectus muscle transposition is inducing a vertical deviation in patients with only a horizontal deviation. This occurs in 13% to 30% of cases after standard transposition surgeries. We have reduced the incidence by placing both the SR and IR muscles on adjustable sutures. Care is taken to cut the intermuscular septum around the SR and IR muscles, sever the attachments between the IR and the capsulopalpebral attachments, and separate attachments between the SR and the superior oblique. These maneuvers prevent narrowing of the palpebral fissure and allow for easier transposition of the vertical rectus muscles. Interestingly, vertical rotations are not compromised by this procedure. The adjustment is performed on the first postoperative day, taking care to correct for any induced vertical deviation. Residual esodeviation is treated by either MR recession or chemodenervation. Botulinum toxin, when needed, is injected a few days after the adjustment procedure.

A self-adjusting vertical transposition technique to decrease the risk of induced vertical deviation may be achieved by joining the temporal halves of each vertical rectus muscle. In some cases, the vertical deviation may be due to an associated fourth nerve palsy or to skew deviation.

Anterior Segment Ischemia

The extent of surgical manipulation in transposition procedures, as well as the need to augment the effect of the procedure with MR recession, increases the risk of anterior segment ischemia—especially in predisposed patients. This complication has been reported with both full-tendon transposition of the vertical rectus muscles and the Jensen procedure. By using pharmacologic weakening instead of surgery on the antagonist MR, the risk of anterior segment ischemia is decreased. Alternatively, the MR weakening procedure may be performed on the contralateral unininvolved eye, not only to center binocular diplopia-free fields but also to avoid surgery on a third muscle in the involved eye. In addition, ciliary vessel sparing procedures may be tried. The IR muscles have the largest ciliary vessels and may be the easiest to preserve. The same procedure may be tried with the anterior ciliary vessels on the MR (see also Chapter 39).

Overcorrection

Secondary exotropia may occur after the Jensen procedure owing to iatrogenically induced lateral restriction. The Jensen operation is difficult to take down, reverse, or relieve.

A more unique and difficult problem is the development of late secondary exotropia after transposition procedures. The approach to managing this problem is discussed in Chapter 36.

Undercorrection

Undercorrection after transposition procedures occurs less often if the procedures are combined with MR weakening. It is estimated to occur in 12.5% to 20% of cases. Performing a horizontal recession-resection when there is complete sixth nerve palsy increases the risk of undercorrection.

Conclusions

Current management of sixth nerve palsy requires the ability to differentiate partial (paresis) from total (complete) palsy. Horizontal recession-resection procedures are only effective for partial sixth nerve weakness. Misinformed surgeons may perform LR resection because of the technical ease of the procedure. This not only eliminates the blood supply from the LR but also increases the risk of anterior segment ischemia if transposition surgery is subsequently required. The advent of chemodenervation has added a new perspective in the management of patients with this affliction. In the acute phase, botulinum toxin assists in rapidly restoring binocular fields and prevents MR contracture. In the perioperative and postoperative setting, botulinum injection of the antagonist MR may preclude surgery on a third rectus muscle, prevent secondary contracture, and improve centration of an enlarged binocular diplopia-free field.

REFERENCES

118. Repka MX, Lam GC, Morrison NA: The efficacy of botulinum neuro


MONOCULAR ELEVATION
DEFICIENCY
(Double Elevator Palsy)

MONOCULAR DEPRESSOR
DEFICIENCY
(Double Depressor Palsy)

History and Terminology

Double elevator palsy is a unilateral defect of upgaze associated with ipsilateral ptosis. Bilateral upgaze paralysis was first described by Henoch in 1864. Subsequently, Parinaud in 1883, described upgaze, downgaze, and total vertical gaze palsies. White, in 1942, described a congenital deficiency of upgaze associated with hypertropia and ptosis of the affected eye (Fig. 20–1). The upgaze deficiency was attributed to paralysis of both the superior rectus (SR) and inferior oblique muscles. The term double elevator palsy was later coined by Dunlap in 1952 to describe the weakness affecting both muscles of elevation.

More recently, the SR has been shown to be the muscle mainly responsible for upward rotation of the eye. The findings of impaired upgaze can result from SR palsy alone without involvement of the inferior oblique (Fig. 20–2). Unilaterally impaired upgaze can also be caused by inferior rectus (IR) restriction and supranuclear disorders. Therefore, the term monocular elevation deficiency (MED) more accurately describes the syndrome of unilaterally impaired upgaze with associated ptosis than does double elevator palsy and will be used throughout this chapter.

Anatomy

The supranuclear pathways for upgaze are located in the midbrain pretectum. The rostral interstitial nucleus of the medial longitudinal fasciculus (riMLF) and the posterior commissure are crucial structures mediating vertical gaze. The efferent tracts for upgaze leave the riMLF, decussate across the midline in the posterior commissure, course through the pretectum, and enter the SR subnucleus of the oculomotor nucleus. After leaving the subnucleus, upgaze fibers to the SR immediately cross the midline again. Because of the double decussation of the upgaze fibers, the SR receives innervation from the ipsilateral riMLF as well as the contralateral pretectum and SR subnucleus.

Because supranuclear elevation deficiency affects the supranuclear fibers mediating upgaze, supranuclear elevation deficiency may be a type of true double elevator palsy. Kirkham and Kline suggested a supranuclear “compartmentalization” for gaze above and below the primary position. This would explain why some patients with MED have a normal vertical saccadic test below midline but an abnormal test above midline.
Figure 20-1. Congenital monocular elevation deficiency, right eye, with accompanying ptosis and hypertropia. Limited upgaze is the same in abduction, straight upgaze, and adduction.

Figure 20-2. A, Preoperative findings of monocular elevation deficiency, right eye. Suggestion of some, albeit limited, right inferior oblique rotation can be seen. B, Postoperative photographs taken after vertical recession-resection surgery on the right eye. Elevation deficiency persists in the field of the right superior rectus. Improved upgaze is seen in straight upward gaze. Overaction of the right inferior oblique is apparent. Diagnosis of congenital superior rectus palsy causing monocular elevation deficiency is confirmed.
### Table 20–1. Causes of Acquired Monocular Elevation Deficiency

<table>
<thead>
<tr>
<th>Disorder</th>
<th>Signs and Symptoms</th>
<th>Ancillary Evaluation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cerebrovascular disease</td>
<td>Acute-onset diplopia ± impaired consciousness, acute diplopia, neurologic symptoms</td>
<td>CT or MRI of brain and brain stem; neurologic evaluation</td>
</tr>
</tbody>
</table>
| Midbrain tumors\(^{1}\):  | All associated with neurologic symptoms.  
(1) pineocytomas,  
(2) acoustic neuromas,  
(3) metastatic tumors | MRI of brain and brain stem; neurologic evaluation                                      |
| Sarcoidosis\(^{40}\)      | Acute-onset diplopia, fatigue, uveitis, retinal changes ± neurologic findings | Chest radiography, ESR, ACE, CT or MRI of orbits, brain, and brain stem; neurologic evaluation |
| Tertiary syphilis\(^{38}\) | Acute-onset diplopia, pupillary findings (Argyll-Robertson), ± uveitis, neurologic and systemic findings | RPR or VDRL, FTA-ABS, CT or MRI of brain and brain stem; neurologic evaluation |

ACE, angiotensin converting enzyme; CT, computed tomography; ESR, erythrocyte sedimentation rate; FTA-ABS, fluorescent treponemal antibody-absorption test; MRI, magnetic resonance imaging; RPR or VDRL, rapid plasma reagin and Venereal Disease Research Laboratory nontreponemal antigen tests.

### Causes

Congenital cases of MED occur sporadically. The causes include supranuclear defects, primary SR paresis, primary IR restriction, and inferior restriction secondary to SR paresis. Neonatal hypoxia has been associated with some cases of congenital MED. Acquired MED is usually caused by cerebrovascular disease (hypertension, arteritis, thromboembolism). Other causes of MED include tumors, sarcoidosis, and infectious disease. The location and size of tumors causing MED determine whether associated findings are present (Table 20–1). Requests for ancillary laboratory evaluation depend on the diagnostic possibilities. Laboratory tests may include a complete blood count with differential, the rapid plasma reagin or Venereal Disease Research Laboratory test, the fluorescent treponemal antibody absorption test, an erythrocyte sedimentation rate, and the level of angiotensin converting enzyme. Potential imaging modalities include chest radiography, computed tomography, magnetic resonance imaging of the brain stem, and cerebral angiography (see section on differential diagnosis).

### Clinical Characteristics

The classic presentation of congenital MED is a unilateral limitation of upgaze above midline with accompanying ptosis. Hypotropia is usually present on the affected side. The limited upgaze is the same whether in adduction or abduction (see Fig. 20–1). The true incidence of MED is unknown. Variable head postures can be seen with MED, depending on the fixation preference and the degree of binocularity. A patient who has some degree of fusion usually tilts the head back in a chin-up position to maintain binocularity if the affected eye is hypotropic. If the affected eye is not hypotropic, or if the hypotropic eye is also amblyopic, the patient will have a normal head posture. Rarely, a patient fixes with the affected eye, causing a large secondary hypertropia in

---

Figure 20–3. Acquired monocular elevation deficiency in the right eye. Patient prefers to fix with the affected eye causing a large secondary hyperdeviation of the left eye. Note also the absence of ptosis in the affected right eye. (From Ziffer AJ, Rosenbaum AL, Demer JL, et al. Congenital double elevator palsy: Vertical saccadic velocity utilizing the scleral search coil technique. J Pediatr Ophthalmol Strabismus 1992;29:142; reprinted by permission of Slack, Inc.)
Ptosis is usually not present. Acquired MED may be associated with pupil abnormalities, convergence weakness, or downgaze paresis. Bell’s phenomenon is usually present unless secondary inferior restriction has developed. The patient often assumes a chin-up head position to maintain binocularity and decrease diplopia.

**Diagnosis**

**EXAMINATION**

It is important to ask the patient or parents when the problem started and whether there were associated symptoms. If the temporal sequence of events is unclear, the presence of amblyopia suggests a congenital etiology. With acquired MED, one should inquire about other neurologic symptoms (vertigo, ataxia, tinnitus, deafness, loss of consciousness), the past medical history (cardiovascular disorders, malignancy), and general systemic symptoms (fatigue, weight loss).

Clinical examination can distinguish between MED caused by primary SR palsy, supranuclear causes of SR weakness, and primary and secondary IR restriction (Table 20–2). Primary SR paresis or palsy is characterized by a forced duction test demonstrating no restriction to full upward rotation and slowed upward saccades both below and above midline. Bell’s phenomenon is not present. Fifty percent of patients with MED and hypotropia in primary gaze have SR palsy.

Patients with primary IR restriction or fibrosis often do not have hypotropia in primary gaze. The forced duction test demonstrates restriction to upward rotation because of a tight IR. Upward saccades are normal until stopped by the tight IR preventing further upward gaze. Secondary IR contracture can occur with SR palsy or supranuclear disorders. Hypotropia and ptosis are usually present. Forced duction testing reveals a tight IR. Upward saccadic velocity testing is normal or weak below midline. An exaggerated infraorbital lid crease can be seen on attempted upgaze in patients with IR restriction.

Supranuclear MED, which is usually congenital, is characterized by intact or mildly reduced vertical saccadic velocity below midline but abnormal or absent velocity above midline, monocular absence of vertical eye movements in superior fields of gaze, and no resistance to upward rotation by forced duction tests. Bell’s phenomenon is present, indicating an intact oculomotor nerve, fasciculus, and nucleus.

Worth four-dot and stereopsis tests should be done both in forced primary gaze and downgaze. Many patients with

---

**Table 20–2. Clinical Findings in Monocular Elevation Deficiency**

<table>
<thead>
<tr>
<th>Disorder</th>
<th>SR Weakness</th>
<th>IR Restriction</th>
<th>Upward Saccades</th>
<th>Bell’s Phenomenon</th>
<th>Forced Duction</th>
</tr>
</thead>
<tbody>
<tr>
<td>Primary SR palsy</td>
<td>+</td>
<td>−</td>
<td>Slow/floating</td>
<td>−</td>
<td>−</td>
</tr>
<tr>
<td>Supranuclear elevation</td>
<td>+</td>
<td>−</td>
<td>Slow or absent above midline</td>
<td>+</td>
<td>−</td>
</tr>
<tr>
<td>deficiency</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Primary IR restriction</td>
<td>−</td>
<td>+</td>
<td>Intact</td>
<td>−</td>
<td>+</td>
</tr>
<tr>
<td>Secondary IR restriction</td>
<td>+</td>
<td>+</td>
<td>Slow or intact</td>
<td>−</td>
<td>+</td>
</tr>
</tbody>
</table>

SR, superior rectus; IR, inferior rectus; +, present; −, absent.
MED will fuse in downgaze but suppress or develop diplopia in primary position. Cover tests are done to measure the amount of vertical deviation and to detect any horizontal deviation (Fig. 20–5). The eyes may be orthotropic in primary position, or a primary or secondary deviation may be present. A characteristic feature of MED is that the amount of attempted elevation does not change on adduction or abduction. Version and duction testing should be done in the nine diagnostic gaze positions to identify relative restrictions of gaze. The Parks' three-step test and Bielschowsky's head tilt test are used to rule out superior oblique palsy. Pupillary abnormalities may be present, as well as convergence anomalies such as convergence retraction.

The presence of Bell's phenomenon is an important feature distinguishing MED caused by supranuclear disorders (Fig. 20–6). Clinical saccadic tests can determine whether a floating saccade is present, indicating paresis, or a rapid saccade that ends abruptly, indicating restriction or supranuclear MED. To evaluate vertical saccadic velocity below midline, the examiner should have the patient look back and forth rapidly between one target held in the patient's downgaze field and the other target level with the patient's midline eye position. To check vertical saccades above midline, the patient should look rapidly back and forth between a target held in the patient's upgaze field and the other held at the level of the midline eye position.

Forced duction testing is important to rule out IR restriction. A false-negative result can occur if the eye is inadvertently retroplaced into the orbit during the test. If IR restriction is present, the affected eye will not rotate passively as high as the other eye (Fig. 20–7). Active force generation testing is useful to verify the degree of muscle paresis. If SR paresis is present, the examiner will feel a less powerful tug on the forceps than in the unaffected eye. If total SR palsy is present, the examiner will feel only a faint tug or none at all on the affected eye.

Vertical saccadic velocity studies can differentiate between inferior restriction, SR paresis, and supranuclear MED. Electro-oculography (EOG) and scleral search coil testing are objective methods of measuring saccadic velocity. Scleral search coil testing is more accurate than EOG in measuring vertical saccades. A Hess test is useful in determining the severity of hypotropia and the area of binocular diplopia-free fields.

DIFFERENTIAL DIAGNOSIS

Congenital MED is diagnosed clinically and does not require laboratory or radiologic studies. The differential di-
diagnosis of congenital MED includes Brown superior oblique syndrome, vertical Duane syndrome or co-contraction syndrome (Fig. 20–8), congenital fibrosis of the IR, congenital absence of the SR with or without absence of the inferior oblique, third nerve palsy with superior branch involvement, and anomalous insertion of the IR or SR muscles (Table 20–3). Congenital absence of the SR is often associated with craniofacial dysostosis.21, 22, 77

Acquired MED requires further testing to determine the cause. The differential diagnosis of acquired MED includes thyroid orbitopathy, orbital floor fracture with entrapment, myasthenia gravis, progressive external ophthalmoplegia, lesions of the superior branch of the oculomotor nerve, orbital inflammatory disease, orbital cellulitis, systemic amyloidosis with extraocular muscle infiltration, cerebellar tumors, and labyrinthine disorders.38 Edrophonium (Tensilon) testing is indicated to rule out myasthenia gravis if forced duction/active force generation tests and saccadic velocity measurements indicate SR paresis—especially if the degree of paresis fluctuates. Patients with a history of cataract surgery may have myotoxic injury of the SR or IR from local anesthetic.40, 62 The IR may undergo primary or secondary contracture. Radiologic tests may be necessary to evaluate disease in the orbit or central nervous system (Table 20–4).

Table 20–3. Differential Diagnosis: Congenital Monocular Elevation Deficiency

<table>
<thead>
<tr>
<th>Disorder</th>
<th>Features</th>
</tr>
</thead>
<tbody>
<tr>
<td>Brown syndrome</td>
<td>Hypotropia worse in adduction; hypotropia improved in abduction; V-pattern strabismus; positive forced duction: superior oblique restriction</td>
</tr>
<tr>
<td>Vertical Duane syndrome</td>
<td>Globe retraction on downgaze; hypotropia worse in upgaze Often familial; hypotropia worse in upgaze; other extraocular muscles can be affected; positive forced duction: inferior rectus restriction</td>
</tr>
<tr>
<td>Congenital fibrosis of inferior rectus</td>
<td>Often associated with craniofacial anomalies</td>
</tr>
<tr>
<td>Congenital absence of superior rectus ± absence of inferior oblique</td>
<td></td>
</tr>
<tr>
<td>Third nerve palsy</td>
<td>Exotropia may be present; pupillary findings may be present; limited eye movements up, down, and in adduction</td>
</tr>
<tr>
<td>Anomalous insertion of inferior rectus or superior rectus</td>
<td>Variable findings</td>
</tr>
</tbody>
</table>

myasthenia gravis, progressive external ophthalmoplegia, lesions of the superior branch of the oculomotor nerve, orbital inflammatory disease, orbital cellulitis, systemic amyloidosis with extraocular muscle infiltration, cerebellar tumors, and labyrinthine disorders.38 Edrophonium (Tensilon) testing is indicated to rule out myasthenia gravis if forced duction/active force generation tests and saccadic velocity measurements indicate SR paresis—especially if the degree of paresis fluctuates. Patients with a history of cataract surgery may have myotoxic injury of the SR or IR from local anesthetic.40, 62 The IR may undergo primary or secondary contracture. Radiologic tests may be necessary to evaluate disease in the orbit or central nervous system (Table 20–4).

**Treatment**

Not all cases of MED require intervention. Observation is recommended for patients with limited head position changes and orthotropia in primary position. Amblyopia should be treated in the conventional manner. Indications for surgery include vertical deviation in primary gaze, deviation causing suppression and amblyopia, diplopia in primary gaze, and contracted binocular fields. Pseudoptosis will resolve after successful strabismus surgery in MED. Ptosis repair should be considered only if significant ptosis persists after strabismus surgery.

**Surgical Techniques**

The goal of surgery is to improve the position of the affected eye in primary gaze, thereby increasing the field of binocular vision. It is essential that the patient and family understand that surgery will not cure the MED, particularly if it is caused by SR paresis or has a supranuclear etiology. The ophthalmologist must clearly explain preoperatively that the affected eye will not elevate as well as the other eye.
after surgery. More than one strabismus operation may be necessary. Posis surgery may also be required after strabismus surgery.

 Forced duction testing should be repeated in the operating room. If restriction to upgaze is demonstrated, IR restriction is present. An IR recession with conjunctival recession should be done in these patients. Seventy-three percent of patients with MED and restricted forced duction on upward rotation developed full upgaze after recession of the IR.54

The limbal approach is used to incise conjunctiva, and then the IR is identified and carefully hooked. A tight IR muscle is more susceptible to avulsion. As many attachments between the IR and the lower lid as possible are excised to avoid postoperative lower lid retraction. Preplacing muscle sutures before disinsertion guards against unexpected muscle escape. However, performing this maneuver on tight rectus muscles is difficult and risks perforating the globe. Modifications of the muscle hook—thinner dimensions and a central groove for protecting the globe during suture placement—were designed especially for use with tight rectus muscles (Kowal L, personal communication, 1997). Some surgeons prefer to use a spring-loaded muscle clamp (Sheppard clamp [modified Apt clamp]), which gives a more secure hold on a muscle before disinsertion and suture placement.

After the IR is disinserted, the forced duction test is repeated. The globe should now elevate freely. The IR may be recessed 5 to 8 mm, depending on the severity of hypotropia. The globe should be able to rotate 20 to 25 degrees superiorly after the IR is reattached.50 The forced duction test result is compared with that in the unoperated eye, and an attempt is made to match the two sides. The conjunctiva is recessed at least 4 mm from the limbus or up to the level of the original IR insertion. If the patient has a very large primary-position hypotropia before surgery, SR resection may be required in addition to IR recession.

In cases of secondary IR restriction, the hypotropia will persist after IR recession because of primary SR palsy. In these cases, a Knapp procedure (vertical transposition of the horizontal rectus muscles)54 should be performed in addition to IR recession. It is important to wait at least 4 months between the two procedures in adults to reduce the chance of anterior segment ischemia. If the IR recession and Knapp procedure are planned at the same session, the surgeon can spare the ciliary vessels in the rectus muscles. The vessels of the IR and SR muscles are larger and easier to dissect.

---

**Table 20-4. Differential Diagnosis of Acquired Monocular Elevation Deficiency**

<table>
<thead>
<tr>
<th>Disorder</th>
<th>Signs and Symptoms</th>
<th>Clinical Tests</th>
<th>Laboratory and Radiologic Tests</th>
</tr>
</thead>
<tbody>
<tr>
<td>Thyroid orbitopathy</td>
<td>Proptosis, diplopia, lid retraction, dilated</td>
<td>Restriction on forced duction, normal force generation, abnormal exophthalmometry</td>
<td>Abnormal TFTs; enlarged EOM bellies on orbital imaging (+) ACh-RAb; abnormal EMG</td>
</tr>
<tr>
<td>Myasthenia gravis</td>
<td>Ptosis, diplopia, orbicularis weakness, Cogan lid twitch, worsens with fatigue</td>
<td>(+) Edrophonium (Tensilon) test, no restriction on forced duction, normal force generation until fatigue</td>
<td></td>
</tr>
<tr>
<td>Orbital floor fracture:</td>
<td>(1) Diplopia, enopthalmos, anesthesia V2,</td>
<td>(1) Restriction on forced duction, normal force generation; (2) equivocal forced duction, weak to normal force generation</td>
<td>(1 and 2) Abnormal orbital imaging</td>
</tr>
<tr>
<td>(1 and 2) IR paresis</td>
<td>history of trauma</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Progressive external</td>
<td>Bilateral signs, decreased saccades in all</td>
<td>No restriction on forced duction; weakened to normal force generation</td>
<td>Ragged red fibers on muscle biopsy, ± abnormal ECG</td>
</tr>
<tr>
<td>ophthalmoplegia</td>
<td>EOM, usually sporadic, orbicularis weakness,</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>ptosis = retinal changes</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Third nerve paresis or</td>
<td>Posis, diplopia, spares pupil, muscles</td>
<td>No restriction on forced duction, weak to absent force generation</td>
<td>Check glucose, ESR, CBC + differential, RPR, FTA-ABS; CT = MRI of orbits, brain, and brain stem</td>
</tr>
<tr>
<td>palsy, superior division</td>
<td>supplied by inferior division (IO, MR, IR); sixth (LR) and fourth (SO) normal</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Orbital inflammatory</td>
<td>Globe tenderness, lid swelling, exopthalmos,</td>
<td>Possible restriction on forced duction, normal force generation</td>
<td>Check glucose, ESR, CBC + differential, RPR, FTA-ABS</td>
</tr>
<tr>
<td>disease</td>
<td>diplopia, painful EOM, chemosis</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Orbital cellulitis</td>
<td>Painful EOM, diplopia, exopthalmos, lid</td>
<td>Painful EOM; check for sinus tenderness</td>
<td>Check glucose, ESR, CBC + differential, RPR, FTA-ABS</td>
</tr>
<tr>
<td></td>
<td>swelling, possible history of sinusitis</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Previous cataract surgery</td>
<td>SR paresis from myotoxic effects local</td>
<td>No restriction on forced duction; normal to weakened force generation</td>
<td>Diagnosis of exclusion</td>
</tr>
<tr>
<td></td>
<td>anesthesia</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Labyrinthine disorders</td>
<td>Acute onset, vertigo, tinnitus, nausea,</td>
<td>Abnormal OKN</td>
<td>CT = MRI of brain and brain stem; neurologic evaluation CT or MRI of brain and brain stem; neurologic evaluation</td>
</tr>
<tr>
<td></td>
<td>nystagmus</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Cerebellar tumors</td>
<td>Progressive ataxia, ocular flutter or</td>
<td>Abnormal OKN, ataxia</td>
<td></td>
</tr>
<tr>
<td></td>
<td>dysmetria, papilledema</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

ACh-RAb, acetylcholine receptor antibodies; AFG, active force generation; CBC, complete blood cell count; ECG, electrocardiogram; EMG, electromyogram; EOM, extraocular movements; ESR, erythrocyte sedimentation rate; FTA-ABS, fluorescent treponemal antibody-absorption test; IO, inferior oblique; IR, inferior rectus; LR, lateral rectus; MR, medial rectus; OKN, optokinetic nystagmus; RPR or VDRL, rapid plasma reagin or Venereal Disease Research Laboratory nontreponemal antigen tests; SO, superior oblique; SR, superior rectus; TFTs, thyroid function tests.
than those of the medial and lateral rectus muscles (see also Chapters 36 and 39).

If the forced duction test is nonrestrictive, the patient has either SR paresis or supranuclear MED. The Knapp procedure—transposition of the medial and lateral rectus muscles to the SR—should be performed. This surgery corrects between 20 to 35 PD of hypotropia in primary position but may cause only a small improvement in upgaze. The Knapp procedure after IR recession will have a greater effect. A modification of the Knapp procedure, using partial rather than full tendon transpositions of the medial and lateral muscles, may suffice after IR recession. A partial tendon Knapp procedure is recommended in patients who have undergone IR recession and have less than 25 PD of hypotropia in primary position or those with no previous surgery who have less than 10 PD of hypotropia in primary position. For MED with more than 35 PD of hypotropia but no IR restriction (RS Foster, personal communication, 1998), reports good results using posterior fixation suture to augment superior transposition of the horizontal rectus muscles following the spiral of Tillaux (Fig. 20–9) (see also Chapter 36).

Patients with less than 25 PD of hypotropia and upgaze ability above midline may benefit from SR resection as an alternative to horizontal rectus transposition.

True ptosis will not improve after strabismus surgery and in fact will appear worse after the eye has moved from its hypotropic position to an improved midline position. If there is enough upgaze to avoid exposure keratitis, external levator resection can be done after strabismus surgery to lessen the residual ptosis.

**COMPLICATIONS**

Persistent hypotropia, overcorrection with hypertropia, and increased diplopia may occur postoperatively.

Residual hypotropia after a Knapp procedure may improve with time. For mild to large amounts of postoperative hypotropia after a Knapp procedure, IR recession may be required. Alternatively, the contralateral SR muscle may be recessed; this has the added benefit of decreasing incomitance in upgaze. For persistent hypotropia after IR recession, a modified partial Knapp procedure is recommended.

Patients who are hypertropic after a Knapp procedure usually experience worsening as time progresses. They should have the medial and lateral rectus muscles moved to a lower position. If hypertropia occurs after IR recession with or without a Knapp procedure, the IR needs to be advanced. The generally accepted rule is that 1 mm of change in the IR position causes approximately a 3 PD shift in vertical position. Results of forced duction tests before and after moving the muscle, and also between the two eyes, should be compared. The adjustable suture technique is helpful when feasible.

Increased diplopia in the reading position may occur after a large unilateral IR recession. The unoperated IR will depress the normal eye more than the eye with a weakened IR. If diplopia occurs on downgaze, the patient may require surgery to weaken the previously unoperated IR to match the limitation in downgaze and increase the binocular diplopia-free field. A fadenoperation or a small recession on the contralateral uninvolved IR can achieve these goals. Prism glasses may be needed for reading if diplopia on downgaze persists.

Lower lid retraction may result if the fascial attachments between the recessed IR and lower lid were not completely excised. Horizontal misalignment may occur after a Knapp procedure if the medial and lateral rectus muscles are not moved equally.

Anterior segment ischemia can occur if more than two rectus muscles are operated on at one time; it is more common in adults than in children. Ischemia results from disruption of the ciliary vessels that are carried in the rectus muscles. A vessel-sparing procedure can be done on the rectus muscle in an attempt to avoid this complication (see Chapter 39).
Double depressor palsy is a unilateral limitation of downgaze and will be referred to in the rest of the chapter as monocular depression deficiency (MDD). MDD is extremely rare, usually congenital, and sporadic. The etiology is unclear.

Anatomy

Monocular depression deficiency is usually caused by primary IR palsy, IR weakness from supranuclear causes, or primary or secondary SR contracture.

The supranuclear pathway for downgaze is bilaterally represented in the riMLF located in the midbrain. Only bilateral lesions of the riMLF cause downgaze paresis. Because bilateral lesions are much less common than unilateral ones, downgaze disorders are much less common than those affecting upgaze.

Vascular studies of the riMLF support the view that only bilateral lesions cause downgaze paresis. The paired posterior thalamic paramedian arteries supply blood to the riMLF and the downgaze tracts between the riMLF and the oculomotor nuclei. Percheron described three anatomic variations of the paired thalamic paramedian arteries. In the first type, each artery originates from separate basilar communicating arteries. In the second, each paired paramedian artery originates from a vascular arcade between the basilar communicating arteries. In both these conditions, bilateral vessel occlusions are required to affect downgaze. In the third variant, both paired arteries originate from a single basilar communicating artery, often having a common stem. In this variation, only a single vessel need be occluded to cause bilateral ischemic lesions of both riMLF, resulting in downgaze paresis.

The downgaze fibers, as opposed to the upgaze fibers, do not decussate through the posterior commissure. The paired posterior thalamic paramedian arteries supply blood to the riMLF and the downgaze tracts between the riMLF and the oculomotor nuclei. Percheron described three anatomic variations of the paired thalamic paramedian arteries. In the first type, each artery originates from separate basilar communicating arteries. In the second, each paired paramedian artery originates from a vascular arcade between the basilar communicating arteries. In both these conditions, bilateral vessel occlusions are required to affect downgaze. In the third variant, both paired arteries originate from a single basilar communicating artery, often having a common stem. In this variation, only a single vessel need be occluded to cause bilateral ischemic lesions of both riMLF, resulting in downgaze paresis.

The downgaze fibers, as opposed to the upgaze fibers, do not decussate through the posterior commissure. The paired posterior thalamic paramedian arteries supply blood to the riMLF and the downgaze tracts between the riMLF and the oculomotor nuclei. One case has been reported of bilateral upgaze palsy with unilateral downgaze palsy affecting the right eye. The computed tomogram showed an infarction of the right paramedian thalamus and upper mesencephalon. This lesion probably interrupted the supranuclear downgaze fibers after their decussation just before entering the oculomotor nucleus.

Other possible sites affecting downgaze include the periaqueductal gray matter (probably because of involvement of afferent downgaze fibers projecting to the riMLF), the interstitial nucleus of Cajal, and the posterior commissure.

Causes

Acquired MDD is usually caused by cerebrovascular disease affecting the pons or cerebellum. The initial presenting symptoms depend on the site and extent of the lesion, but patients usually present with impaired consciousness. Infection from acute viral hemorrhagic conjunctivitis has also been reported in association with MDD.

Clinical Characteristics

Monocular depression deficiency is characterized by unilateral downgaze deficiency of equal degree in adduction and abduction. The affected eye is hypertropic and may have upper lid retraction when the unaffected eye fixes. The lid is drawn back passively when the globe is hypertropic because of the fascial attachments between the SR and levator palpebrae. Pseudoptosis may occur on attempted downgaze when the paretic eye does not depress but the lids lower bilaterally.

Diagnosis

Examination

Clinical examination of MDD is similar to that described for MED. Vestibulo-ocular reflexes (doll’s head maneuvers) are usually intact on downgaze, indicating a supranuclear etiology with preservation of the nuclear reflexes. MDD may be associated with impaired convergence and pupillary anomalies.

Differential Diagnosis

Congenital disorders that can mimic MDD include vertical Duane syndrome, congenital fibrosis of the SR, congenital absence of the IR with or without absence of the superior oblique, and third nerve palsy. Vertical Duane syndrome can be ruled out by checking for globe retraction as a result of co-firing of the SR and IR muscles. Third cranial nerve disease involving the inferior division presents as unilateral deficiency of downgaze associated with exotropia and, possibly, pupillary abnormalities.

The differential diagnosis of acquired MDD includes a traumatic orbital blow-out fracture with associated IR paresis or entrapment and thyroid orbitopathy with SR contracture. Previous strabismus surgery with excessive recession of the IR, a large resection of the SR, or scar tissue formation may cause diminished downgaze.

Treatment and Complications

The treatment of MDD, surgical principles, and complications are similar to those described for MED. In MDD the medial and lateral rectus muscles are transposed to the IR for primary or supranuclear IR weakness. The SR is recessed for SR contractures. Postoperative complications include undercorrection or overcorrection, increased diplopia, retraction of the upper lid, and anterior segment ischemia.
REFERENCES


RESTRICTIVE STRABISMUS
Historical Perspective

Gorman, citing Rolleston, noted that an association of eye disease with goiter was recognized as early as the 12th century. The triad of hyperthyroidism, diffuse nodular goiter, and ophthalmopathy was noted by Parry in a treatise published after his death in 1825. Shortly thereafter, Graves and von Basedow also noted the same association; Graves name was bestowed on this constellation of findings. Von Basedow thought that an “intumescence of cellular tissue behind the bulbus” was responsible for the exophthalmos, rather than the global enlargement invoked by Parry and Graves. He also described physical signs such as resistance to retropulsion of the globe, staring, and skin changes characteristic of pretibial myxedema. Von Graefe expanded on the ocular signs and symptoms in 1857, highlighting their importance in the field of ophthalmology.

The biochemical and endocrinologic basis of Graves disease began in the 19th century, when iodine and its protein-binding ability were recognized. In 1932, animal studies indicated that thyroid-stimulating hormone from the anterior pituitary could produce exophthalmos. Thus, there appeared to be a linkage between pituitary overactivity, hyperthyroidism, and the eye signs of Graves disease. The excessive production of thyroid hormones in this disorder appears to result from stimulation of thyroid-stimulating hormone receptors on the thyroid cell membrane by an immunoglobulin, thyroid-stimulating antibody. This suggests a significant role for the immune system in the etiology of hyperthyroidism. In addition, several autoimmune diseases occur more commonly in association with Graves disease than in the general population; examples are myasthenia gravis, rheumatoid arthritis, and diabetes mellitus.

Because the early signs and symptoms of Graves disease may resemble those of other inflammatory orbitopathies, it sometimes has been thought to represent primary orbital inflammatory disease. The presentation may include proptosis, conjunctival injection, chemosis, and lid edema. The differential diagnosis of these conditions is outlined later in this chapter.

Epidemiology and Risk Factors

Graves ophthalmopathy occurs more frequently in women than in men and is seen most often in middle-aged patients. The overall pediatric contribution to thyrotoxicosis is less than 5%; most children are diagnosed between ages 10 and 15 years. Bartley and associates, in a retrospective study in Minnesota from 1976 to 1990, found that 86% of patients with Graves disease and oculomotor abnormalities were female. The incidence in women (16 per 100,000) peaked at 40 to 44 years and 60 to 64 years. In men, the incidence (3 per 100,000) peaked slightly later, between ages 45 and 49 years and 65 and 69 years. Ninety percent of these cases were associated with hyperthyroidism.

The ocular myopathy does not necessarily reflect the state of thyroid activity. Evaluation by an internist may reveal hyperthyroidism, euthyroidism, or even hypothyroidism. The disease is common in patients with previous undiagnosed episodes of hyperthyroidism and in those having had thyroidectomy.

The association between myasthenia gravis and thyroid disease has been recognized for many years. About 5% of patients with myasthenia gravis reportedly develop Graves disease. A smaller proportion of those with thyrotoxicosis (0.2%) subsequently develop myasthenia. The two conditions have much in common: both are mediated by autoantibodies to membrane receptors, and they share certain immunogenetic features. Patients with Graves disease often have relatives with myasthenia gravis and vice versa.

Genetics

Certain specific subtypes of human leukocytic antigen (HLA) from chromosome 6 are increased in patients with
Graves disease. An increased subtype seems to be specific for patients of white, Japanese, and Chinese origin. Genetically abnormal suppressor T lymphocytes fail to abort the proliferation of abnormal plasma cells, leading to the production of autoantibodies. These, in turn, cause target somatic cells, such as extraocular muscle fibers, to be coated with autoimmune complexes that cause damage through inflammatory changes. The plasma cell-mediated release of mucopolysaccharides and collagen formation lead to hypertrophy of the extraocular muscles. This muscle enlargement leads to congestion of the orbital contents and strabismus, with typical limitations in the range of ocular rotations.

Hollingsworth and associates described six families with Graves disease. In all these kindreds there seemed to be mother-to-daughter linkage over several generations. The likely mode of inheritance was autosomal dominant, with a predilection for females. The occurrence of Graves disease at all ages within the same families suggests a single underlying defect.

Clinical Characteristics

CLASSIC APPEARANCE

The disease may present with typical congestive, inflammatory signs of the eye and orbit. These include conjunctival injection, sometimes over the site of the rectus muscle insertions, and chemosis. In addition, edema of the eyelids and proptosis secondary to congestion in the orbit can occur. Over time (often months to 1 or 2 years), the inflammatory signs abate and the picture is one of restricted, fibrotic extraocular muscles and infiltration of mucopolysaccharide ground substance through the orbit. A few patients develop restrictive findings without passing through the inflammatory stage. This mode of presentation is less common and may confuse the diagnosis, especially if thyroid blood chemistries are normal.

The onset of diplopia is often insidious and may be closely related to the appearance of exophthalmos. Periorbital edema and limited elevation of one or both eyes may be the initial clinical findings in patients with Graves ophthalmopathy. Limited upgaze is the most common defect of ocular rotation. The next most frequent abnormality is limited abduction, followed by restricted depression and a reduced range of adduction (Fig. 21–1). In primary position, vertical and horizontal deviations may coexist, resulting in diagonal diplopia. The forced traction test reveals restricted passive movement of the globe toward the field of limited motility. The deviation is incomitant, with the greatest vertical deviation recorded in upgaze. This is a restrictive, not a paralytic type of strabismus. Although the process is bilateral, the motility abnormalities may be limited to one eye or be markedly asymmetric.

Other signs may include upper lid retraction, lid lag on downgaze (the von Graefe sign), a reduced frequency of blinking, weak convergence, an inability to hold fixation on extreme lateral gaze, a staring appearance, resistance to retrodisplacement of the eye, edema and injection of the conjunctiva, and tremor on gentle eyelid closure. Retracted upper lids lead to the appearance of staring. This is accentuated by exophthalmos and in attempted upgaze when the patient tries to elevate the globe in the presence of a tight inferior rectus (IR) muscle. According to the law of Hering, the increased innervation required by the superior rectus (SR) to counteract a tight IR spills over to the upper lid retractors.

CLASSIFICATION OF THE SPECTRUM OF CLINICAL PRESENTATIONS

In 1977, Werner, as chairman of the ad hoc committee of the American Thyroid Association (ATA), published several modifications to the original classification of the eye changes of Graves disease. The six classes may be remembered using the mnemonic NO SPECS:

Class 0: No physical signs or symptoms
Class 1: *Only* signs, no symptoms (upper lid retraction, stare, lid lag)
Class 2: Soft tissue involvement (signs and symptoms)
Class 3: Proptosis
Class 4: Extraocular muscle involvement
Class 5: Corneal involvement
Class 6: Sight loss (optic nerve involvement)

The disease need not pass sequentially through each of these classes. Some patients have limited motility and strabismus before proptosis, soft tissue signs and symptoms, and typical thyroid lid findings. It is much less common to note corneal and/or optic nerve involvement before motility abnormalities. Other methods of classification have been described but have not proved to be more useful than the Werner/ATA system.

The extraocular muscles are the most consistently involved tissue in Graves orbitopathy. Involvement ranges from minimal enlargement of a few muscles to tremendous enlargement of multiple muscles. Initially, the muscles may retain their ability to contract, but later they become fibrotic and unable to relax, resulting in restricted motility secondary to the tethering effect. During the acute inflammatory phase there may be pain or a pulling sensation on attempted ocular rotation. As the inflammation subsides, pain usually disappears. The IR is most frequently involved, followed by the medial rectus (MR), (SR), and lateral rectus (LR). Aside from hypotropia, a tight IR can cause small degrees of exocytorsion and, because it is an adductor, may cause an esodeviation with a restricted forced duction test in abduction.

Pathologically, the extraocular muscle bellies are the primary site of disease. The tendon sheaths are usually not involved at their insertions. Inflammation in the extraocular muscle stimulates fibroblasts, producing both mucopolysaccharides and collagen. As collagen accumulates, the striated extraocular muscle cells undergo degeneration. Fibrosis of the extraocular muscles accounts for restricted motility and for most of the increased soft tissue volume that produces proptosis.

**SUBTLE AND ATYPICAL CASES**

Some patients develop strabismus and diplopia without the typical signs of Graves ophthalmopathy. Exophthalmos, lid lag on downgaze, staring with upper lid retraction, lid edema, and signs of conjunctival inflammation and chemosis all may be lacking. Thyroid blood chemistries may be within the normal range. The clinician should suspect Graves disease in incontinent acquired strabismus, when limited upgaze in one or both eyes is a prominent feature, when the pattern of strabismus does not fit that expected from an extraocular muscle palsy, and when passive rotation of the globe is limited.

Paresis of the LR muscle and restriction due to infiltration and fibrosis of the muscle from Graves disease may coexist. The paresis is ascribed to pressure on the nerve supply to the muscle from enlargement in the posterior cone.

Because endocrine ophthalmopathy is frequently bilateral, a patient presenting with primarily unilateral findings may be misdiagnosed. Only one eye may appear to be restricted. Elevation may be limited on one side, whereas that of the other eye appears to be full. Careful examination of the apparently “normal” eye often reveals a subtle limitation to full ocular rotation in at least one direction. Mild resistance to retropulsion of the globe may be another helpful clue.

**Diagnosis**

**KEY CLINICAL EXAMINATION FINDINGS**

The key features of Graves disease on ophthalmologic examination include the following:

1. Soft tissue signs (lid edema, conjunctival inflammation, chemosis)
2. Exophthalmos (unilateral or bilateral)
3. Ocular motility abnormalities (often limited upgaze and limited abduction) and strabismus with diplopia (often vertical strabismus or a combined vertical and horizontal deviation)
4. Increased intraocular pressure on upgaze (a 2-mm or greater increase on upgaze, reflecting the restrictive effects of a tight IR pressing on the globe with attempted elevation)
5. Corneal staining secondary to exposure
6. Reduced visual acuity (due to both corneal exposure and optic nerve compression caused by muscle enlargement at the orbital apex)

A method of quantifying limited eye movements by measuring ductions using a modified perimeter has been described. The results are claimed to be precise and reproducible. This test might also prove useful in evaluating the effects of both medical and surgical therapy for Graves disease.

Although the disease is more common among women, it appears to be more severe in men and in patients older than 50 years—especially the ocular motility findings (Fig. 21–2). Serum antibody titers do not correlate well with the severity of the disease. A gradual, chronic onset is common in older patients, whereas a more acute onset is characteristic of younger patients. Older patients and men more commonly have asymmetric abnormalities.

Graves disease is uncommon in the young; and even among children and adolescents with this diagnosis, ophthalmoplegia is infrequent. The process is substantially more benign than in adults; increasing severity correlates with advancing age. It may be that children have less severe ophthalmopathy than adults because they have lower levels of thyroid antibodies.

**LABORATORY TESTS**

**Orbital Imaging**

Computed tomography (CT) and magnetic resonance imaging (MRI) demonstrate enlarged muscles in the orbit, contrasting with the lucent orbital fat (Fig. 21–3). In unilateral exophthalmos, when the diagnosis may be in doubt, the finding of hypertrophied extraocular muscles in both eyes strongly suggests Graves disease. There is maximal involvement of the belly of the rectus muscles with sparing of the tendons at the muscle insertion. The lacrimal gland may also appear to be enlarged. The sensitivity of CT in recognizing enlarged muscles exceeds 85%, compared with 61% for MRI.
When orbital and extraocular muscle volumes are estimated from high-resolution CT scans in patients with Graves ophthalmopathy, total and individual muscle volumes are increased compared with control values.\(^6\) The degree of clinical ophthalmopathy is related quantitatively with the amount of increased tissue bulk in the orbit.

In patients with proptosis, orbital imaging can detect causes other than Graves disease—especially orbital tumors, which are diagnosed with an accuracy of greater than 95%.\(^12\) Such orbital masses are commonly discrete, encapsulated, and unilateral. Paraorbital lesions causing proptosis also image very well. Several infiltrative orbital processes may resemble Graves disease on imaging. They include inflammatory pseudotumor, orbital myositis, and certain forms of lymphoma. Findings such as scleral thickening, anterior soft tissue edema, and bone destruction can help distinguish among these conditions.\(^11\) Most patients with Graves disease have abnormalities in both orbits, whereas other conditions are usually unilateral.

**Orbital Ultrasonography**

B-scan ultrasonography demonstrates hypertrophied extraocular muscles (Fig. 21–4) and accentuated orbital walls. Intramuscular echoes probably reflect edema separating the myofibrillar bundles. A-scan ultrasonography can show high acoustic reflectivity of the extraocular muscle bellies, caused

---

**Figure 21–2.** Severe Graves ophthalmopathy in a 58-year-old man. A, Preoperative photographs show esotropia and severe left hypertropia with limited ductions in all gazes. Strabismus surgery was done in stages. First surgery consisted of left superior rectus recession and bilateral medial rectus recession. B, Intermediate postoperative photographs showing residual esotropia and small left hypertropia with improved ocular rotations with some lid retraction of the left eye. C, Two additional strabismus surgeries were required before the final alignment of 2 PD esophoria shown in this composite was attained. Limited adduction in both eyes is apparent, as well as moderate decrease in abduction of the left eye, with bilateral lower lid retraction.

**Figure 21–3.** Computed tomography of a patient with endocrine ophthalmopathy. The muscle bellies are tremendously enlarged, although the tendinous insertions do not appear abnormal. Left photograph, axial view; right photograph, coronal view. (Courtesy of Paul Rosenberg, MD.)
by edema fluid and cellular infiltration. Generally, ultrasonograms are not as disease specific as CT scans.  

Standard echography shows increased heterogeneity and reflectivity of the thickened extracocular muscles and thereby can more specifically identify patients with Graves ophthalmopathy. Typical findings include solid thickening of the optic nerve sheaths and swelling of the lacrimal glands. Muscle thickening on ultrasound study can also represent acute myositis. In the latter condition, however, the muscles show extremely low reflectivity with thickened tendons.

Ocular Electromyography

A majority of reports on electromyography (EMG) in patients with Graves disease indicate a myopathic rather than a neurogenic problem. A reduction in amplitude of the action potential is noted, without a loss of motor units. These findings agree with those in thyrotoxic myopathy involving skeletal muscles.

Myasthenia gravis is more prevalent in patients with thyroid disease than in the general population. The EMG findings, with fatigue of motor units followed by recovery of activity on rest or after administration of edrophonium (Tensilon), may be found in some patients with endocrine ophthalmopathy.

Saccadic Velocity Measurements

Average horizontal and vertical saccadic velocity measurements, made by electro-oculography in patients with noncongestive endocrine ophthalmopathy, are normal, suggesting a restrictive rather than a paretic cause of limited motility (Fig. 21–5). Saccades generated toward the limit of ocular rotation decrease in velocity as the endpoint is reached. This may result from stretching out of the restricted antagonist muscle, slowing its movement as the end position is reached.

Active force generation studies also fail to demonstrate evidence of paresis.

One report did demonstrate slowing of downward saccades in two patients, consistent with IR weakness. This is more likely to occur in the acute congestive phase of the disease, before restrictive elements predominate and muscle elasticity declines. An older view, that toxic damage to the oculomotor nerve or the oculomotor nuclei could cause

![Figure 21-4. B-scan ultrasonography of the orbit in a patient with Graves disease. Muscle enlargement (black arrow) can be seen. (Courtesy of Gwen Sterns, MD.)](image)

![Figure 21-5. Vertical saccadic velocity of patient with thyroid ophthalmopathy. Both upward (u) and downward (d) saccades are normally rapid. Upper trace, eye position; lower trace, peak velocity. (From Metz HS, Woolf PS, Patton HG: Endocrine ophthalmopathy in adolescence. J Pediatr Ophthalmol Strabismus 1982,19:58–60. Reprinted by permission of Slack, Inc.)](image)
paralysis of the elevator muscles, does not appear to be valid. If any paretic component coexists with restrictive strabismus, saccadic velocity will be reduced, and this should be taken into account when planning surgery.

Infrared oculography has also demonstrated a decrease in peak horizontal saccadic velocity with an increasing severity of orbital disease in endocrine ophthalmopathy. After orbital decompression, the peak velocity increases.

**Forced Duction Testing**

The technique of forced duction testing is described in Chapter 3. The test may be done qualitatively in an outpatient setting using topical anesthesia or in the operating room under local or general anesthesia. In patients with Graves disease, restrictions to passive movement of the globe are usually encountered in all directions, most notably in upgaze and abduction. The findings are often found in both eyes, although they may be asymmetric. The restrictions are often described as “leashlike,” because the eye rotates fairly well up to a point, when marked restriction is encountered.

Quantitative forced ductions demonstrate increased stiffness (a steep length-tension curve) with continued upward rotation of the globe. After IR disinsertion and recession, stiffness is diminished (the length-tension curve is flattened) and the range of upward rotation is increased (Fig. 21–6).

**Blood Chemistries**

In the presence of normal thyroid chemistries, the clinician should look for the typical clinical signs of endocrine ophthalmopathy. These include acquired vertical strabismus and diplopia; limited ocular motility, most apparent in upgaze; restricted elevation on forced duction testing; exophthalmos; lid retraction; lid lag on downgaze; and enlargement of the rectus muscle bellies on CT or MRI. Regardless of the results of blood studies, if a number of the above findings are present, the diagnosis is Graves ophthalmopathy until proved otherwise.

**DIFFERENTIAL DIAGNOSIS**

The differential diagnosis of endocrine ophthalmopathy includes orbital inflammatory disease, systemic disease with orbital involvement, contiguous sinus disease, orbital cysts and neoplasms, and orbital vascular lesions.

**Orbital Inflammatory Disease**

Orbital pseudotumor and chronic orbital myositis are included in this category. Although the ocular findings are usually unilateral, they may become bilateral. Signs of vascular congestion, inflammation, and proptosis are common. When inflammation primarily involves the extraocular muscles (orbital myositis), ophthalmoplegia with pain on eye movement is a frequent finding. The response to systemically administered corticosteroids is good. The CT scan may show a thickened Tenon’s capsule and feathery orbital densities, which help make the distinction from Graves disease. B-scan ultrasonography may show sub-Tenon’s edema and a characteristic squaring off of the optic nerve as it enters the globe. In orbital myositis, the muscle tendon and not just the muscle belly itself may be involved on the CT, in contrast to thyroid ophthalmopathy (Fig. 21–7). There is generally no eyelid retraction, and limited elevation is not common.

**Systemic Diseases with Orbital Involvement**

Some of the systemic disorders that may involve the orbit and produce proptosis with limited motility are lymphoma, sarcoidosis, and amyloidosis. Lymphoma has a predilection for the trochlear area. A nodular mass felt in the superonasal
aspect of the orbit should suggest something other than endocrine exophthalmos. Sarcoidosis and amyloidosis also produce extraocular muscle enlargement. Involvement of other organ systems helps clarify the diagnosis. A biopsy may reveal the definitive cause of proptosis with restricted ocular rotations.

**Contiguous Sinus Disease**

If orbital sinus infection causes orbital cellulitis, many of the signs of orbital inflammation and congestion that are found in thyroid orbitopathy can result. In ethmoidal sinusitis, CT may demonstrate an enlarged MR muscle secondary to inflammation. Fever and an elevated white blood cell count along with other signs of an acute infectious process should alert the clinician to the possibility of orbital cellulitis. Bilateral findings on orbital imaging and enlargement of multiple extraocular muscles are more suggestive of endocrine ophthalmopathy.

**Orbital Cysts and Neoplasms**

Epidermoid and dermoid cysts occur most commonly in the orbit. Rupture of a cyst may produce inflammatory signs that may resemble endocrine ophthalmopathy. Cysts are most commonly unilateral, and their characteristic appearance on CT or MRI helps differentiate them from Graves disease.

Neoplasms arising within the orbit or metastatic to the orbit may present as proptosis and signs of inflammation. Some of the lesions to be considered include metastatic carcinoma of the breast and malignant melanoma (which can both cause extraocular muscle enlargement), meningioma, glioma of the optic nerve, lacrimal gland tumor, and rhabdomyosarcoma. Palpation of the orbit along with CT or ultrasonography usually distinguishes these lesions from thyroid orbitopathy. If confusion persists, an orbital biopsy may be required.

**Orbital Vascular Lesions**

The orbit may exhibit vascular engorgement from increased venous pressure. This can arise from a carotid-cavernous fistula or a dural arteriovenous malformation. Vascular engorgement may cause extraocular muscle enlargement as well as chemosis, conjunctival injection, proptosis, and ophthalmoplegia, mimicking the signs of Graves disease. Orbital ultrasonography and CT may show dilated orbital vessels, suggesting the proper diagnosis. Angiography is the most definitive procedure.

Demer and von Noorden reported a highly myopic patient (−28.00 D) who presented with restrictive findings suggestive of endocrine ophthalmopathy but no thyroid abnormalities. The limited ocular rotation was ascribed to contact between the elongated globes and the bones of the orbital apices.

**Treatment**

**NONSURGICAL MANAGEMENT**

**Medical Treatment**

Appropriate systemic treatment for confirmed hyperthyroidism is indicated. Generally, this does not eliminate strabismus or restrictions of ocular motility. Topical therapy with artificial tears and decongestants can be useful for patients having conjunctival injection and chemosis. Guanethidine eye drops and thymoxamine are reportedly helpful in reducing lid retraction and thus improving symptoms of corneal drying.

Corticosteroid treatment may be used in the acute, congestive phase of the disease. Inflammatory signs will be relieved; the optic nerve will be decompressed, resulting in improved visual acuity; and proptosis secondary to orbital congestion will be reduced. The ophthalmopathy usually does not improve. Long-term use of high doses of systemic corticosteroids should be avoided because of possible complications. Nonsteroidal anti-inflammatory agents have not been effective.

Immunosuppressive therapy has also been advocated for Graves disease. Cyclophosphamide and azathioprine have improved the congestive changes in most patients, improved eye muscle involvement in about half, and decreased proptosis in very few individuals. Cyclosporine is a potent immunosuppressive agent that alters the function of T-helper lymphocytes and some B lymphocytes. It interferes with cellular and humoral autoimmune processes thought to contribute to Graves ophthalmopathy. A study using both corticosteroids and cyclosporine in the medical treatment of Graves disease found corticosteroids to be more effective, although less well tolerated. Eye muscle enlargement and proptosis were reduced, and visual acuity was improved. Combination treatment helped patients who did not respond to either drug singly, while allowing a lower dose of corticosteroid. Both therapies, when effective, were begun in the acute inflammatory stage of the disease, not the fibrotic phase. Nephrotoxicity and, rarely, lymphoma are concerns with the long-term use of cyclosporine. Plasmapheresis, which attempts to eliminate autoantibodies and immune complexes, has been successful in a few patients lacking chronic exophthalmos.

**Radiation Therapy**

Radiation treatment is effective in the early stages of Graves disease and decreases the need for high-dose cortico-
steroid therapy. Significant benefit has been found when these modalities are used together. Soft tissue changes and motility disturbances improved after radiation therapy, but there was less functional improvement in restrictions and strabismus. Two thirds of treated patients eventually required eye muscle surgery. Radiation therapy begun after fibrosis is established will not confer clinical benefit. Care should be taken in planning radiation therapy because of the potential dangers of dry eye, cataract, and radiation retinopathy.

**Botulinum Toxin Therapy**

There has been some interest in the use of botulinum toxin for this problem since Scott began treating strabismus with chemodenervation in the early 1980s. In the early stages of the disease, when inflammatory signs predominate, botulinum toxin may correct the deviation and allow the patient to regain fusion. However, when both the agonist and the antagonist rectus muscles become fibrosed later in the course of Graves ophthalmopathy, this technique may not have long-lasting effects. My own attempts have yielded some good short-term results, but only until the paralytic effect of the toxin wears off. After that, the full preinjection deviation returns.

**Prisms**

Prismatic therapy can help eliminate diplopia in the primary gaze position. If the deviation is not too large (less than 12–14 PD), prisms can be ground into a patient’s glasses. Both vertical and horizontal strabismus correction can be incorporated in each lens. If the deviation is larger, as it often is, Fresnel prisms may be used.

The problem with prism treatment is that the strabismus is incomitant. Prisms provide correction in one direction but not in other fields of gaze. In addition, the size of the deviation often varies over time, requiring several changes of prism power before achieving stability. Fresnel prisms will turn yellow over time. Many patients object to the reduced visual acuity caused by the lines in the prisms, as well as to yellowing of the lens. If prism therapy fails, the patient can always occlude one eye to avoid diplopia before surgery is done.

**SURGICAL MANAGEMENT**

Before surgical treatment is planned, the patient’s motility status must be stable. The deviation should remain constant and the degree of ocular restriction should be the same for at least 6 months. Surgery performed earlier may lead to significant variations in the postoperative period, because the disease may progress and continue to alter ocular motility. As a result, reoperation will be required more often.

Surgery should be delayed in the presence of anterior inflammatory signs. The findings of conjunctival injection and chemosis, along with increasing proptosis, should warn the clinician that surgery may exacerbate inflammation and lead to postoperative complications (see section on complications) and an unsatisfactory alignment. Patients, although often eager to proceed with surgery to gain relief from diplopia and restrictions, should be cautioned that surgery done before the appropriate time can lead to a poor and sometimes disastrous outcome.

Intraoperative forced duction testing must be performed in every case, before and after surgery. Restrictions will be felt, especially during attempts to passively elevate the eye. A tight IR may also cause esotropia with decreased abduction and restricted forced duction testing. The test therefore should be repeated in abduction after the IR has been disinserted from the globe. If most of the restriction to abduction is relieved, the preoperative restriction may be ascribed to IR pathology without involvement of the MR muscle. Small degrees of excyclotorsion may also be due to a tight IR and can be corrected by IR recession alone. The goal of surgery is to align the eyes in primary gaze and the reading position with some increase in the range of rotation. It is not to restore full versions in all fields of gaze, especially in upgaze. Secondary contracture of the SR resulting in overcorrection then becomes less likely. The range of downgaze will be maximized if a full range of upward rotation is not sought.

Hypotropia with limited elevation, either monocularly or binocularly, is the most common motility abnormality. IR recession, usually of a fairly large degree, will be required to fully correct the deviation. At surgery, the tendon looks relatively normal even though the muscle belly is enlarged on the CT scan. The IR is often so tight that it is difficult to get a muscle hook beneath the insertion. Care must be taken in performing this maneuver, as well as during disinsertion of the muscle from the globe, to avoid perforating the sclera. A large IR recession in one eye can result in monocular limitation in downgaze. This may be necessary to achieve orthotropia in primary gaze. Separate reading glasses with prisms can help such a patient if there is some overcorrection in downgaze. A problem still exists when these patients walk down steps, attempt to hit a golf ball, and the like.

If both eyes demonstrate markedly limited elevation, both IR muscles can be recessed, one more than the other, to correct hypotropia. Bilateral IR recessions may lead to a postoperative A pattern. Management of this problem is discussed later in this chapter (see section on complications).

Generally, rectus muscle resections are not indicated in the treatment of endocrine ophthalmopathy. These muscles are already very tight, and even a small resection can increase muscle stiffness considerably. I have occasionally resected the SR by 1 to 2 mm in patients with very large vertical deviations to avoid recessing the ipsilateral IR further back. This approach has been successful, but it must be used judiciously. In large hypotropias, an alternative is to recess the SR of the opposite eye. The subconjunctival instillation of soluble corticosteroids minimizes postoperative scarring. Supramid sleeves have not proved helpful as an adjunctive measure in my experience.

If the eye has been hypotropic for some time, the conjunctiva is likely to be foreshortened inferiorly. Conjunctival recession can eliminate this problem.

Horizontal strabismus, usually esotropia, should be corrected at the same time. The most restricted MR muscle is usually recessed. If there is a choice, I usually prefer to do horizontal muscle surgery on the side opposite the IR procedure to avoid anterior segment ischemia.

Orbital decompression surgery, whether required for cosmetic problems, marked proptosis, corneal exposure, or optic
nerve compression, should be done before strabismus surgery.\textsuperscript{25} If performed afterward, strabismus often recurs, making another muscle operation necessary.

Because all of these patients are adults, it would seem that adjustable suture strabismus surgery would be an ideal approach. However, Ruttmann\textsuperscript{25} found that his results were no better in cases where an adjustable suture was used than with standard surgery. Fewer overcorrections resulted when adjustable sutures were avoided. The basic principle behind the use of adjustable sutures is that the surgeon has the best chance of placing the eyes in the most desirable position after surgery. Clinicians may disagree as to where the best position is; each has his or her own idea. Empirically, if ocular alignment is good immediately after surgery, it is more likely to be satisfactory in the long term. We still prefer the use of adjustable sutures in surgery for Graves ophthalmopathy but agree that the postoperative course is less predictable because of the underlying disease process.

When surgery is successful, patients are usually extremely grateful because they are no longer incapacitated and can now obtain fusion in some positions of gaze. The return of normal motility with fusion in all fields of gaze is not the goal. Patients should understand what can realistically be achieved. They should be told that reoperation may be necessary, sometimes years after the initial procedure. Dyer\textsuperscript{7} reported that 40\% of his cases required another procedure.

**SURGICAL COMPLICATIONS**

**Overcorrections and Undercorrections**

Although not true complications, overcorrections and undercorrections are clearly unsatisfactory results, especially if diplopia persists in the primary position and downgaze. Overcorrections are probably due to both excessive recession of the \textit{IR} and further tightening of the already thickened and restricted ipsilateral \textit{SR}. A positive forced traction test toward downgaze will reveal the role played by the \textit{SR}. \textit{SR} recession is recommended.\textsuperscript{7} If this will not suffice, concurrent \textit{SR} advancement may be done.

Undercorrections can be treated by recessing the contralateral \textit{SR} unless a small \textit{IR} recession was performed initially. Re-recession of an \textit{IR} that has already undergone significant weakening will likely lead to markedly limited depression with hypertropia in downgaze as well as lower lid sag. Overcorrections and undercorrections in the horizontal plane can be treated similarly, without concern over limiting depression. Again, resections are best avoided. In overcorrected exotropia, the \textit{MR} may be advanced or the \textit{LR} recessed.

**Marked Postoperative Inflammation**

Strabismus surgery should be delayed in the presence of anterior inflammatory signs, such as conjunctival injection and chemosis. In a patient with Graves ophthalmopathy who had surgery when one eye appeared quiet while the other demonstrated mild inflammation, the quiet eye was unremarkable whereas a tremendous amount of proptosis and edema was seen in the other.\textsuperscript{25} The findings included decreased vision secondary to corneal opacification, corneal thinning and vascularization, hypertropia larger than the preoperative hypertropia with downgaze limited to the midline, marked chemosis, conjunctival ecchymosis, and proptosis threatening corneal perforation and loss of the eye (Fig. 21–8). Treatment with a moisture chamber, hourly lubrication, and high-dose systemic corticosteroids saved the globe, but poor vision persisted with corneal scarring, large hypertropia, and markedly limited motility.

**Late Slippage of the Inferior Rectus**

There have been some cases of apparent late slippage of the \textit{IR} muscle after recession using the adjustable suture technique, resulting in progressive hypertropia in the months after surgery (Fig. 21–9). Scott (personal communication, 1997) believes that the \textit{SR} has usually become extraordinarily strong by isometrically exercising against the tight \textit{IR}. He has found thickened Tenon’s fascia under the \textit{IR} muscle, possibly because of its association with the Lockwood ligament. This soft tissue allows the muscle to slip back after the sutures have absorbed. This tissue can also thicken progressively and become a pseudotendon anterior to the muscle. It must be excised to allow better adhesion to the globe. It is not clear that there is any other way to prevent this complication unless the adjustable suture technique is abandoned and the cut end of the tendon is sutured directly to the globe. Alternatively, nonabsorbable adjustable sutures could prevent a strong \textit{SR} from causing late slippage.

**Lower Lid Retraction**

When a large \textit{IR} recession is required, as is common in the surgical treatment of endocrine ophthalmopathy, lower lid retraction is frequently noted postoperatively (Fig. 21–10). To prevent or minimize this change, the \textit{IR} should be cleared from any attachments to the inferior oblique muscle, the Lockwood ligament, and the lower lid retractors. Otherwise the recessed muscle will pull the lid down because the tarsal attachments are recessed posteriorly. Jampolsky (personal communication, 1996) suggested making a mark...
on the sclera to indicate the distance behind the limbus where the lid retractor attachments to the muscle were found. The retractors are reattached to sclera (not to the muscle that has been recessed) at that point so as to limit sagging of the lower lid. This maneuver supports the lower lid and prevents or minimizes retraction. If lid retraction persists after surgery, despite the surgeon’s best efforts, a plastic procedure such as lateral tarsorrhaphy or insertion of a scleral or hard palate graft spacer can be performed later.

A-Pattern Exotropia

A-pattern exotropia has been noted after bilateral IR recessions. The IR muscle has a secondary adducting action. When both are recessed, adduction in downgaze is weakened, leading to an A pattern. To prevent this, each IR insertion may be displaced nasally one-half to one full tendon width. A preoperative A pattern has been treated successfully by elevating both MR muscles, recessing the lower border 2 to 3 mm more than the upper border. Tenotomy of the posterior 75% of the superior oblique fibers has also been suggested to treat the A pattern, but I believe that nasal transposition of the IR muscles is both simpler and more predictable. With thickened muscles and restricted rotation of the globe, it may be difficult to obtain adequate exposure for operating on the superior oblique tendon. If another surgery is required once vertical strabismus has been corrected, elevation of both MR muscles or depression of both LR muscles may prove helpful.

REFERENCES


Motility disturbances associated with scleral buckling procedures are frequently transient and resolve spontaneously. The symptoms are caused by postoperative edema, decreased vision, extraocular muscle swelling, and hemorrhage. Most patients are able to adapt and eventually regain normal fusional ranges after the induced shift in oculomotor imbalance. Recovery is often rapid but the recovery period may take 3 to 6 months.

The true incidence of motility disorders after scleral buckling procedures has been difficult to determine because of the variety of retinal reattachment techniques used, as well as the difficulty of performing a complete preoperative motility evaluation. With the trend in retinal detachment surgery moving toward internal tamponade and vitrectomy, a conspicuous decline in the frequency of strabismus complications may be expected. The problem remains, however, and continues to affect a significant number of patients.

The incidence in earlier retrospective series ranged from 3% to as high as 50%. More recent studies cite a reduced incidence of less than 5%. The largest of these series reported the lowest incidence. Strabismus may have been present before the onset of retinal detachment. The possibilities include horizontal deviations such as esotropia and exotropia, as well as vertical deviations—most commonly superior oblique (SO) palsy. Graves ophthalmopathy and myasthenia gravis may also be underlying causes. Amblyopia may have been present and does not preclude the development of diplopia. Borderline fusion may be disrupted in a variety of ways: blurring of vision from the detachment; prolonged recovery

<table>
<thead>
<tr>
<th>Pathogenesis</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>PREDISPOSING FACTORS</strong></td>
</tr>
<tr>
<td>Predisposing factors in the development of motility problems after scleral buckling procedures are summarized in Table 22–1.</td>
</tr>
</tbody>
</table>

Strabismus may have been present before the onset of retinal detachment. The possibilities include horizontal deviations such as esotropia and exotropia, as well as vertical deviations—most commonly superior oblique (SO) palsy. Graves ophthalmopathy and myasthenia gravis may also be underlying causes. Amblyopia may have been present and does not preclude the development of diplopia. Borderline fusion may be disrupted in a variety of ways: blurring of vision from the detachment; prolonged recovery

<table>
<thead>
<tr>
<th>Underlying strabismus</th>
</tr>
</thead>
<tbody>
<tr>
<td>Disinsertion of extraocular muscles ± faulty repositioning</td>
</tr>
<tr>
<td>Cryoapplication</td>
</tr>
<tr>
<td>Multiple reoperations</td>
</tr>
<tr>
<td>Large buckles</td>
</tr>
<tr>
<td>Radial buckles</td>
</tr>
<tr>
<td>Encircling elements/buckles</td>
</tr>
<tr>
<td>Location under rectus muscles</td>
</tr>
<tr>
<td>Scleral dissection beneath rectus muscles</td>
</tr>
<tr>
<td>Violation of posterior Tenon's capsule causing fat adherence</td>
</tr>
</tbody>
</table>

Table 22–1. Predisposing Factors to Motility Problems After Retinal Detachment Repair

<table>
<thead>
<tr>
<th>Underlying strabismus</th>
</tr>
</thead>
<tbody>
<tr>
<td>Disinsertion of extraocular muscles ± faulty repositioning</td>
</tr>
<tr>
<td>Cryoapplication</td>
</tr>
<tr>
<td>Multiple reoperations</td>
</tr>
<tr>
<td>Large buckles</td>
</tr>
<tr>
<td>Radial buckles</td>
</tr>
<tr>
<td>Encircling elements/buckles</td>
</tr>
<tr>
<td>Location under rectus muscles</td>
</tr>
<tr>
<td>Scleral dissection beneath rectus muscles</td>
</tr>
<tr>
<td>Violation of posterior Tenon's capsule causing fat adherence</td>
</tr>
</tbody>
</table>
of vision after the repair; occlusion after surgery; nerve damage from postoperative intraocular pressure (IOP) elevation; or various factors causing haziness of the ocular media, such as corneal edema, lens injury, vitreous opacities, and anterior segment ischemia.

Disinsertion of the rectus muscles in itself predisposes to the development of oculomotor imbalance after scleral buckling procedures. Faulty repositioning of the muscle, with disruption of tissue planes, amplifies this risk.\textsuperscript{33, 61, 75, 97, 118, 120} Disinsertion avoids prolonged stretching that causes muscle damage.\textsuperscript{13, 93} Where to reinsert the rectus muscle is a matter of controversy. Some authors suggest recessing slightly to prevent overaction (to counteract the resection effect of a buckle under a rectus muscle).\textsuperscript{40, 46} whereas others favor a resection before reattachment to avoid underaction.\textsuperscript{53} We favor replacing the muscle at the original insertion site because postoperative muscle function cannot be accurately predicted.\textsuperscript{104}

Cryopexy by itself induces restrictive fibrosis, but its transconjunctival application has not been associated with strabismus.\textsuperscript{3} Several studies have shown that cryopexy causes fewer motility problems when compared with surface diathermy, especially when cryoapplication or diathermy is done beneath the rectus muscle.\textsuperscript{1, 92} This latter view, however, is not universally accepted.\textsuperscript{196}

Although motility problems may be seen after a single retinal detachment operation,\textsuperscript{10} more numerous and complex procedures increase the probability of strabismus developing.\textsuperscript{71, 74, 85, 94, 105, 118, 120} Careless or inaccurate closure of periccular tissues (i.e., Tenon’s fascia and conjunctiva) contribute to this risk.\textsuperscript{120} Repeated procedures also incite a more aggressive inflammatory response after each surgery.

It is easy to see why circumferential buckles cause strabismus.\textsuperscript{11} Encircling elements or bands, in themselves, are associated with motility disturbances.\textsuperscript{19, 61, 74} The problem is accentuated by posteriorly or obliquely placed buckles.\textsuperscript{3} Radial buckles, especially high buckles, predispose to ocoumoto-motor problems.\textsuperscript{3, 51, 120} Additionally, the more muscles that overlie the exoplant, the more fields of gaze will be affected.\textsuperscript{111} A circumferential buckle length of at least 5 mm is more likely to cause motility problems than smaller buckle segments.\textsuperscript{101} The more anteriorly placed exoplants or implants increase the pressure on the muscles, affect the blood supply, and cause stretching and even dehiscence. The eye then becomes prone to granuloma, extrusion, infection, and muscle erosion.\textsuperscript{40, 70}

In some detachment repairs, the need to isolate the tear means that Tenon’s fascia may need to be dissected farther than 10 mm posterior to the limbus. This may expose orbital fat within the muscle cone into the operative field, which incites scar tissue formation and increases the risk of progressive restrictive motility disturbances that may prove difficult to correct surgically.\textsuperscript{87, 88, 121, 122}

MECHANISMS FOR HETEROTROPIA

### Mechanical Factors (Table 22-2)

#### Adhesions

Any operative trauma can cause adhesions between extraocular muscles, sclera, scar tissue, fat, and the exoplant.\textsuperscript{3, 23, 61, 85, 91, 104, 120, 121} These fibrous adhesions may cause significant restrictions that need to be relieved if strabismus surgery is planned. Adhesions posterior to the buckle element create a posterior fixation suture effect\textsuperscript{112, 32, 85, 104} similar to the so-called fadenoperation.\textsuperscript{18} Fibrous encapsulation and hyalinization of the muscles have been demonstrated histopathologically in some cases.\textsuperscript{12}

Jampolsky\textsuperscript{12} described surgical leashes and reverse leashes in the surgical management of strabismus with restrictions. A direct leash is due to mechanical factors limiting movement of a muscle in the direction opposite to the gaze limitation. A reverse leash causes a restriction on the same side as the gaze limitation. Leashes may be due to scar tissue, muscle contracture, or adhesions. The phenomenon of leashes was reiterated by several authors who have studied the association of strabismus with retinal reattachment procedures.\textsuperscript{31, 32, 85, 91, 93, 118}

#### Mass Effect

It is not difficult to imagine how the sheer bulk of scleral elements, especially when located under the rectus muscles, mechanically restricts extraocular muscle movement (Fig. 22–1).\textsuperscript{24, 79, 77, 91, 97, 105, 107} In one study, restrictions and diplopia were related to the position of the explant in 84% to 87% of cases.\textsuperscript{111} The presence of the scleral element alters the direction of muscle pull, changing its arc of contact, the length-tension relationship, and the geometry of its action.\textsuperscript{32, 98, 118} Fusional vergence mechanisms may be adequate to overcome mild mechanical deviations caused by the presence of a buckle under a rectus muscle.\textsuperscript{1, 77}

#### Muscle Factors

##### Direct Muscle Trauma

The practice of excessive and prolonged stretching during exposure for retinal reattachment procedures accounts for most of the direct trauma to the rectus muscles.\textsuperscript{1, 23, 85, 88, 92, 111, 118} Damage to the SO tendon has also been implicated in cyclodeviation.\textsuperscript{85, 97}

##### Muscle Ischemia

Prolonged muscle compression from the buckle element alters the effective blood supply to the muscles\textsuperscript{91, 120} and may cause muscle erosion.\textsuperscript{120}

##### Muscle Slippage

Muscle slippage or “lost muscles” have been reported in association with strabismus after retinal detachment repair procedures.\textsuperscript{36, 88, 120} The pressure from the buckle, causing muscle erosion through the insertion, may lead to detachment of the muscle. In addition, erosion

<table>
<thead>
<tr>
<th>Table 22-2. Mechanisms for Heterotropia</th>
</tr>
</thead>
<tbody>
<tr>
<td>I. Mechanical Factors</td>
</tr>
<tr>
<td>A. Adhesions</td>
</tr>
<tr>
<td>B. Bulk effect</td>
</tr>
<tr>
<td>II. Muscle Factors</td>
</tr>
<tr>
<td>A. Muscle ischemia</td>
</tr>
<tr>
<td>B. Direct trauma</td>
</tr>
<tr>
<td>C. Disinsertion ± faulty repositioning</td>
</tr>
<tr>
<td>D. Muscle slippage</td>
</tr>
<tr>
<td>III. Other Anatomic Factors</td>
</tr>
<tr>
<td>A. Nerve damage</td>
</tr>
<tr>
<td>B. Oblique muscle inclusion or rotation</td>
</tr>
<tr>
<td>C. Macular ectopia or gliosis</td>
</tr>
<tr>
<td>IV. Sensory Factors</td>
</tr>
<tr>
<td>A. Breakdown in fusion</td>
</tr>
<tr>
<td>B. Sensory deprivation</td>
</tr>
<tr>
<td>C. Aniseikonia and anisometropia</td>
</tr>
<tr>
<td>D. Image distortion</td>
</tr>
</tbody>
</table>

---

MECHANISMS FOR HETEROTROPIA

### Mechanical Factors (Table 22-2)

#### Adhesions

Any operative trauma can cause adhesions between extraocular muscles, sclera, scar tissue, fat, and the exoplant.\textsuperscript{3, 23, 61, 85, 91, 104, 120, 121} These fibrous adhesions may cause significant restrictions that need to be relieved if strabismus surgery is planned. Adhesions posterior to the buckle element create a posterior fixation suture effect\textsuperscript{112, 32, 85, 104} similar to the so-called fadenoperation.\textsuperscript{18} Fibrous encapsulation and hyalinization of the muscles have been demonstrated histopathologically in some cases.\textsuperscript{12}

Jampolsky\textsuperscript{12} described surgical leashes and reverse leashes in the surgical management of strabismus with restrictions. A direct leash is due to mechanical factors limiting movement of a muscle in the direction opposite to the gaze limitation. A reverse leash causes a restriction on the same side as the gaze limitation. Leashes may be due to scar tissue, muscle contracture, or adhesions. The phenomenon of leashes was reiterated by several authors who have studied the association of strabismus with retinal reattachment procedures.\textsuperscript{31, 32, 85, 91, 93, 118}

#### Mass Effect

It is not difficult to imagine how the sheer bulk of scleral elements, especially when located under the rectus muscles, mechanically restricts extraocular muscle movement (Fig. 22–1).\textsuperscript{24, 79, 77, 91, 97, 105, 107} In one study, restrictions and diplopia were related to the position of the explant in 84% to 87% of cases.\textsuperscript{111} The presence of the scleral element alters the direction of muscle pull, changing its arc of contact, the length-tension relationship, and the geometry of its action.\textsuperscript{32, 98, 118} Fusional vergence mechanisms may be adequate to overcome mild mechanical deviations caused by the presence of a buckle under a rectus muscle.\textsuperscript{1, 77}

#### Muscle Factors

##### Direct Muscle Trauma

The practice of excessive and prolonged stretching during exposure for retinal reattachment procedures accounts for most of the direct trauma to the rectus muscles.\textsuperscript{1, 23, 85, 88, 92, 111, 118} Damage to the SO tendon has also been implicated in cyclodeviation.\textsuperscript{85, 97}

##### Muscle Ischemia

Prolonged muscle compression from the buckle element alters the effective blood supply to the muscles\textsuperscript{91, 120} and may cause muscle erosion.\textsuperscript{120}

##### Muscle Slippage

Muscle slippage or “lost muscles” have been reported in association with strabismus after retinal detachment repair procedures.\textsuperscript{36, 88, 120} The pressure from the buckle, causing muscle erosion through the insertion, may lead to detachment of the muscle. In addition, erosion
through the muscle belly can cause necrosis of the muscle tissue and possibly muscle dehiscence at this junction.

**Muscle Disinsertion With or Without Faulty Repositioning.** This mechanism has often been implicated in the incidence of extraocular muscle imbalance after retinal detachment repair. The incidence of strabismus is directly related to the number of detached muscles. Faulty repositioning of a rectus muscle may result from a failure to properly identify the muscle insertion after disinsertion.

**Other Anatomic Factors**

**Neural Factors.** Nerve damage or injury either from direct trauma or an inadequate blood supply secondary to ischemia leads to muscle paresis, which in turn can lead to secondary contracture of the antagonist. Oblique Muscle Inclusion or Rotation. Both the SO tendon and the inferior oblique (IO) muscle have been implicated in vertical and torsional deviations after scleral buckling procedures (see later discussion on cyclodeviations in this chapter).

**Macular Ectopia or Gliosis.** Patients with a reattached retina may develop an epiretinal membrane or gliosis over the macula or macular ectopia. This entity is difficult to treat because patients experience diplopia due to anatomic displacement of the involved fovea in relation to the normal fovea of the other eye. These patients may have intractable "macular" diplopia, but usually maintain good peripheral fusion.

**Sensory Factors**

**Fusion Breakdown and Sensory Deprivation.** The breakdown in fusion and sensory deprivation leads to sensory deviations, albeit usually transient. Poor vision as well as occlusion or any opacity in the media after retinal detachment surgery may transiently disrupt binocularity.

**Aniseikonia and Anisometropia.** The scleral buckle can induce both myopia and astigmatism. An increase in axial length of 0.99 mm is associated with 2.75 D of myopic shift. Cylindrical errors are usually transient, but can be as high as 6 to 7 D, and may be irregular. Increasing the height of the buckle increases the risk for induction of a refractive error. Silicone oil, used as internal tamponade in retinal detachment, has also been associated with diplopia. A large amount of astigmatism following the axis of the oil meniscus has been described. Intravitreal injection of silicone oil is associated with complex effects on refraction: in phakic eyes, hyperopia ranging from 2 to 8 D (mean, +5 D) occurred; in aphakic eyes, the refraction induced a myopic shift ranging from 4 to 9 D (mean, -6.63 D). Silicone oil extraction induced a refractive myopic shift of -5.57 D. In unilateral involvement, the difference in refractive errors induces both aniseikonia and anisometropia. Adults are less able to tolerate these problems than children. This problem is also seen in patients who become aphakic after lensectomy.

**Image Distortion.** Depending on the duration of the detachment, and the successful reattachment of the retina, image distortion can be a bothersome complication. Patients can learn to suppress this image. Eventually, a sensory esodeviation or exodeviation may develop.

**Clinical Characteristics**

Most deviations are transient and resolve within 6 months. Accordingly, definitive surgical management should not be planned before 6 months have elapsed. If the strabismus continues to improve even after 6 months, it is prudent to wait for measurements to stabilize.

**VERTICAL DEVIATIONS**

Vertical deviations are the most common form of strabismus after scleral buckling procedures. Because of smaller vertical than horizontal fusional amplitudes, the pa-
tient is more likely to be symptomatic and bothered by the vertical misalignment.\textsuperscript{85} This may also be related to the higher incidence of superior tears.\textsuperscript{104} However, no correlation between muscle scarring and the location of the implant has been demonstrated.\textsuperscript{85} Commonly, patients present with a hypotropia and a restricted forced duction test that simulates an acquired Brown syndrome (i.e., tightness felt in the SO tendon associated with limitation of elevation in adduction) (Fig. 22–2).\textsuperscript{85} Other investigators believe that the anatomy of the IO and its proximity to the orbital fat pad contribute to the hypotropia.\textsuperscript{104} Hypotropia is frequently associated with torsional diplopia.\textsuperscript{85}

**CYCLODEVIATIONS**

Patients frequently become symptomatic when cyclodeviation exceeds 5 degrees.\textsuperscript{58, 77, 85} Torsional deviation occurred in 46\% of patients with strabismus after scleral buckling procedures.\textsuperscript{16} Excycloversion was more common than incycloversion in a ratio of 9:1.\textsuperscript{16} The true incidence is probably underestimated in many series, hence the belief that cyclodeviation is less common than other heterotropias.\textsuperscript{3, 61, 77}

Knowing the possible mechanisms that may induce torsional diplopia allows one to direct surgical strategy to a suspected area of pathology. In patients with altered anatomy after repeated surgeries, minimizing unnecessary dissection may be critical. The three most common mechanisms of anomalous torsion are listed below:

1. Anterior displacement of the SO tendon during placement of the encircling element or buckle (Fig. 22–3).\textsuperscript{16, 77} Adhesions commonly form between the nasal border of the superior rectus insertion and the SO tendon. The patient presents with incomitant hypertropia, restriction of downward gaze, and excycloversion\textsuperscript{16, 85, 88} or incycloversion.\textsuperscript{11, 16}

2. Intentional\textsuperscript{107} or inadvertent\textsuperscript{10} tenotomy of the SO tendon, producing a clinical picture similar to SO palsy.

3. Presence of a large exoplant under the inferior rectus, resulting in hypotropia with restricted upward gaze and excycloversion (Fig. 22–4).\textsuperscript{16, 77}

Less commonly, adhesions may develop between the SO tendon, the superior rectus, the globe, and/or the exoplant.\textsuperscript{16, 77, 85, 88, 104} Rarely, the superior rectus may have become disinserted during a scleral buckling procedure.\textsuperscript{16} The IO muscle may be hooked, pulled, and tucked anteriorly while isolating the inferior rectus muscle.\textsuperscript{16, 58, 122} As a result, the encircling element is passed between the two sides of the "tucked" IO, causing a restrictive hypotropia and possibly excycloversion.\textsuperscript{104} Both the IO and inferior rectus muscles may be involved by scarring and adhesions leading to cyclo-deviation.\textsuperscript{16} Exoplants that are oriented radially produce astigmatism and are associated with torsional diplopia.\textsuperscript{77, 130} Larger exoplants contribute to the risk of astigmatism.\textsuperscript{85}

Patients may adopt a head tilt to achieve fusion or learn cyclofusion,\textsuperscript{76, 114, 115} especially if the torsional component is small and develops gradually over time. Their symptoms may be rather vague. Unless carefully sought by the examiner, very few patients voluntarily describe a torsional component of their diplopia.

**HORIZONTAL DEVIATIONS**

Horizontal deviations occur in 12\% to 30\% of patients presenting with strabismus after retinal detachment surgery.\textsuperscript{85, 111} They are commonly sensory deviations secondary to poor vision from retinal detachment or prolonged visual recovery after surgery and often are associated with anisome-
tropia, macular distortion, or silicone oil. In addition, this motility problem may represent a decompensated underlying strabismus preceding development of a detachment. These patients present with comitant horizontal deviations and absence of restriction on forced duction test (Fig. 22–5). Sometimes a restriction to horizontal globe rotation may be identified on forced duction testing (Figs. 22–6 and 22–7). The medial rectus is more often involved, because the intracanal and extracanal fat pads are more anteriorly located on the medial aspect of the globe. The location of the exoplant under the rectus muscle may be a clue but is not predictive.

**Diagnosis**

**FEATURES ON STRABISMUS EXAMINATION**

The diagnosis of strabismus after retinal detachment surgery is not difficult to establish. The history is usually straightforward. It is unfortunate that preoperative motility evaluations are often overlooked in patients with retinal detachments. Strabismus evaluations should include the assessment of refractive errors. The scleral buckle causes an increase in axial length, predisposing to both myopia and astigmatism.

Careful monocular duction and binocular version testing is imperative. The patient is instructed to look in the nine diagnostic gaze positions. Muscle strength may be graded on a 9-point scale, designating -4 as severe underaction, 0 as normal, and +4 as severe overaction. The details of complete duction and version testing are discussed in Chapter 1. For clinical purposes, we usually test torsion using the double Maddox rods. It is helpful to orient the cylinders vertically so that the patient sees horizontal lines through the Maddox prisms. By virtue of the dissociative nature of the red and green Maddox rods, torsion from each eye can be evaluated separately. The amount of cyclotorsion in degrees may be read directly from the trial frame (see Chapter 4). A careful biomicroscopic examination, identifying the probable sites of adhesions as well as signs of anterior segment ischemia from previous retinal detachment surgery, is included in the initial evaluation.

Fusion may be assessed using a vectograph (e.g., Titmus Fly or Randot Stereogram) and a Worth four-dot test or Bagolini striated lenses if available. Bagolini lenses are less dissociative and are preferred to the Worth four-dot test for analysis of peripheral fusion.

**LABORATORY EVALUATION**

Laboratory tests that might be helpful in this subset of patients are summarized in Table 22–3. Tests of muscle function (i.e., forced duction test, force generation test, and saccadic velocity analysis) clarify whether limited ocular

<table>
<thead>
<tr>
<th>Table 22–3. Laboratory Evaluation of Strabismus After Scleral Buckling and Glaucoma Implant Procedures</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tests of muscle function</td>
</tr>
<tr>
<td>Forced duction test</td>
</tr>
<tr>
<td>Force generation test</td>
</tr>
<tr>
<td>Saccadic velocity analysis</td>
</tr>
<tr>
<td>Fundus photography</td>
</tr>
</tbody>
</table>
rotations are restrictive or paretic. Identification of slipped or lost muscles may also be aided by applying the principles of saccadic velocity analysis and generation of muscle force. Fundus photography and blind spot mapping serve to document torsion. Binocular field studies may be done with a perimeter (e.g., Goldmann) preoperatively and postoperatively to assess the size of the patient’s diplopia-free binocular fields. The perimeter may also be used to document the extent of ductions. The presence of torsion inhibits fusion and may be misdiagnosed as central fusion disruption. Because of the high risk for torsion and macular ectopia, a synoptophore evaluation of a patient who is unable to fuse with prism neutralization in free space may help assess fusion ability. This device can neutralize torsion as well as vertical and horizontal deviation. Both the surgeon and the patient then will have a better idea of what may be expected after strabismus surgery.

Studies of retinal blood flow show a 50% reduction in average arterial flow rates after scleral buckling and encircling procedures. Removal of the buckling elements produces improvement of 44% to 73% in flow rates. When patients are at risk of developing anterior segment ischemia (findings of iris ischemia; elderly; multiple retinal or strabismus procedures), we have been conducting imaging studies of the anterior segment for documenting anterior segment circulation. Previously, only light-colored irides benefited from iris angiography with fluorescein. Using indocyanine green dye and the technology for digital videoangiographic imaging, we were successful in displaying the filling patterns of the iris circulation even in darkly pigmented eyes. Filling defects may be identified in this way. The strabismus surgeon then may opt to modify the operative plan because of compromised blood flow to the anterior aspect of the globe.

**Treatment**

### NONSURGICAL MANAGEMENT

#### Conservative Management

**Observation.** Most of the deviations that occur are transient. Patients learn to adapt to the phoria by a process known as orthophorization. Sensory adaptation to heterotropia and cyclotropia has also been described. Moreover, most patients with poor vision do not require intervention. However, diplopia may occur even in patients with poor macular vision because of viable peripheral retina.

**Suppression.** Patients may learn to suppress a blurred image or develop the ability to ignore the second image.

**Anomalous Head Postures.** Patients may adopt a head tilt to fuse a cyclovertical deviation or a face turn for horizontal deviation.

**Prisms**

If the deviation is small and comitant, prisms may be helpful, either as add-on (Fresnel) prisms or incorporated in the patient’s spectacle correction. On the other hand, incomitance and torsion may make neutralization by prisms problematic. Even so, prisms can be helpful to move the field of binocular vision closer to primary gaze or to increase the area of binocular vision without diplopia. Prisms alone may benefit approximately one third of patients.

### Blurring of the Second Image

The use of frosted glass to blur a second image has been reported to help patients who are refractory to other forms of therapy. Fogging with plus lenses has also been described. Monocular occlusion may be necessary. The ophthalmologist also may defer prescribing the appropriate spectacle correction for aphakia. None of these procedures, however, will prevent the later development of secondary sensory deviations.

#### Chemodenervation

Botulinum toxin may be used to temporarily relieve symptoms before definitive surgery is undertaken. On occasion, a complex strabismus problem is partially corrected in this way, permitting a simpler surgical procedure. In some instances, surgery may be avoided entirely. The toxin allows intermittent fusion to take place by reducing the size of the deviation. Fusional vergence amplitudes are slowly increased. The patient may eventually maintain acceptable ocular alignment.

The drawbacks of this procedure are related to the temporary effects of the toxin. Definitive surgical correction will be delayed until the pharmacologic effect of the botulinum toxin has completely worn off, which usually takes 2 to 3 months. If surgery is deferred, repeated injections may be needed. In addition, the normal anatomy and the relationship of the rectus muscle to the globe may have been altered by the scleral buckling surgery. This increases the risk of perforating the globe when administering the toxin. There has been no report of overcorrection, but vertical deviations have been induced by medial rectus injections.

In patients at high risk of developing anterior segment ischemia or retinal detachment, and those with a history of extraocular muscle transsection during prior eye surgery, the option of chemodenervation should be seriously considered. Doses used ranged from 1 to 7.5 units, correcting up to 60 PD of deviation in some cases. The effects of botulinum toxin lasted longer in patients whose binocularity and fusion were reestablished. Neither the results of the forced duction test nor the presence of a restriction were predictive of the therapeutic response. Chemodenervation is discussed further in Chapter 32.

### SURGICAL MANAGEMENT

#### Considerations in Selecting Surgical Management

**Tests of Muscle Function.** Forced duction testing, force generation, and saccadic velocity analysis suggest a logical surgical option. If the forced duction test is restricted, for example, the need to operate on the buckled eye to relieve restrictions or adhesions will be evident. If rotations are full, or the forced duction test is nonrestricted or only mildly restricted, one may consider operating on the “good” eye. Small deviations are amenable to operations on a
single muscle. Operating on the better eye makes the results more predictable and decreases the risk of retinal detachment.\textsuperscript{31, 45} If results of the forced duction test are inconclusive, one may need to explore the involved globe. A good starting point is the area of the explant. If the force generation test reveals some force and saccades are slowed, a resection procedure may be considered. Caution is needed to avoid iatrogenic restrictions caused by resections.

**Risk of Anterior Segment Ischemia.** The buckling procedure in itself decreases arterial blood flow.\textsuperscript{66} Aging, the number of reoperations, the number of rectus muscle disinsertions, postoperative infection, and intraocular pressure elevation amplify this risk.

**Type of Deviation and the Extraocular Muscle Involved.** Horizontal deviations most often are related to sensory deviations, whereas vertical deviations are usually due to mechanical restriction.\textsuperscript{85}

**Degree of Scarring.** Reoperations increase the postoperative inflammatory response. At the initial strabismus evaluation, the surgeon can gauge the extent and degree of scarring (and hence know what to expect during surgery) by careful biomicroscopic evaluation and testing of muscle function.

**Contribution of the Buckle.** The presence of the buckle and its relationship to extraocular muscle balance was discussed earlier. If the buckle is identified as a major factor contributing to strabismus, the surgeon needs to decide whether to remove the explant or encircling element used for retinal detachment repair.

**Removing an Explant or Encircling Element.** The decision to remove an explant should always be made in consultation with the patient's retina surgeon.\textsuperscript{66} Explant removal may be done safely as early as 3 months after the retinal reattachment procedure.\textsuperscript{31, 36} Some authors remove the implant alone as a logical first step to any strabismus deviation.\textsuperscript{31} This procedure alone, without strabismus surgery, is successful in only 20% of patients.\textsuperscript{31} When combined with strabismus surgery, removing the implant allows scleral reattachment of the rectus muscles and permits the muscle to move freely during an adjustment procedure.\textsuperscript{85} As a corollary, only enough buckle is excised to allow the muscle to attach to the globe. As much as possible of the encircling element is left in place to decrease the risk of exoplant migration and extrusion.\textsuperscript{85} Every effort should be made to obtain the retinal surgery report to fully understand size, location, and type of retinal element and drainage site location.

**Approach to Surgery**

**Intraoperative Maneuvers.** The intraoperative forced duction test allows one to gauge if the restriction has been adequately relieved or whether rectus muscle resection has caused a restriction. Temporary slip knots may be used for adjustments during surgery as dictated by the results of forced duction testing before the knots are permanently tied.

**Dissection and Lysis of Adhesions.** In reoperations, a limbal conjunctival opening provides the best exposure. Meticulous dissection of fibrous tissue and removal of adhesions are begun in the area of the buckle, using blunt and sharp dissection.\textsuperscript{16, 32, 45, 77, 85} The forced duction test may be repeated at several stages during the dissection to see whether the globe has been adequately freed from its restrictions.

Based on the preoperative clinical picture, the surgical strategy may be directed to a specific area of involvement.
Recession and Resection (R&R). Unilateral R&R procedures, either horizontal or vertical, may be done on either the involved or the normal eye. It may be more difficult to do the resection in an eye that already has restricted rotations. Excessive resections may aggravate the restriction and cause incomitance.

Marginal Myotomy. Marginal myotomy involves lengthening of a muscle by creating partial cuts in the muscle belly. The procedure has been largely superseded by the more predictable recession. The presence of an implant, exoplant, or encircling element used for retinal detachment repair is considered one of the indications for this procedure. If a muscle has been recessed maximally, the surgeon may consider resecting the antagonist or may choose to operate on the contralateral eye.

Fadenoperation. In incomitant strabismus, a posterior fixation suture may be used on a muscle on the better eye to match the limitation of rotation of a yoke muscle in the eye with a previous retinal detachment. This will improve the size of the diplopia-free binocular field in a specific field of gaze. Vertical Transposition of the Horizontal Rectus Muscles for Hyperdeviations. Vertical transposition of the horizontal rectus muscles may be used to correct small associated hyperdeviations of up to 8 to 10 PD. For a hypertropia, the medial and lateral rectus muscles are displaced inferiorly in the involved eye or superiorly if the procedure is done in the contralateral eye.

Surgical Procedures for Cyclotorsional Deviations

Harada-Ito Procedure. The Harada-Ito procedure, or anterior lateralization of the anterior fibers of the SO tendon, is the accepted procedure for the correction of excyclotropia—especially without associated hyperdeviation in primary gaze. Transposition surgery of the vertical rectus muscle was first described for a congenitally absent SO. Since then, excyclotropia has been corrected using this procedure. Transposition surgery of the vertical rectus muscle is an option to improve excyclotropia, especially when surgery on the SO is not feasible. For incyclotropia, nasal transposition of the superior rectus or temporal transposition of the inferior rectus may be helpful. Nasal transposition of the inferior rectus may be used to correct small associated hyperdeviations of up to 8 to 10 PD. For a hypertropia, the medial and lateral rectus muscles are displaced inferiorly in the involved eye or superiorly if the procedure is done in the contralateral eye.

Superior Oblique Tuck. Tucking of the SO tendon is generally reserved for patients with large excyclotorsion, typically more than 10 degrees, and with an incomitant hypertropia in the affected eye that is larger in downgaze. The difficulty with the procedure lies in quantifying the amount of the tendon to tuck and predicting the results. Nuances are discussed in Chapter 35.

Inferior Oblique Recession. This procedure is reserved for excyclotorsion with hypertropia in the affected eye that is greater in upgaze. One study dealt with the effect of anterior transposition of the IO on objective fundus torsion. Theoretically, any procedure that weakens the IO (disinsertion, myectomy, recession, anterior transposition) should affect torsion and correct a significant amount of excyclodeviation. Anterior seven-eighths’ disinsertion of the IO muscle was also described for excyclotorsion without hypertropia in primary gaze (similar to the principles behind the Harada-Ito procedure for the SO). Superior Oblique Weakening Procedures. For incyclotropia caused by an overacting SO with hypotropia in primary gaze, SO weakening procedures (e.g., recession, tenotomy, tenectomy) may be used. Anterior seven-eighths’ tenectomy of the SO tendon is preferred for incyclotorsion without associated hypotropia. The latter procedure corrects 7 to 10 degrees of incyclotorsion.

COMPLICATIONS

Redetachment of the retina is a legitimate concern after removal of a part or the whole scleral buckling element or the encircling band. The incidence of redetachment was 4% to 33% after removal for various reasons; e.g., infection, erosion, extrusion, pain, allergy, and excessive buckle height. A survey conducted in 1995 suggested that only 1 strabismus surgeon in 10 will encounter redetachments in an entire lifetime of practice. The risk is further decreased by waiting at least 6 months, allowing the retinal detachment repair to stabilize before considering any type of strabismus surgery.

The risk of perforating the globe during attempts at scleral passes of the muscle suture may be increased because of the altered anatomy after scleral buckling procedures. It is also common to see patients with thinned sclerae (myopes with elongated globes develop retinal detachment more frequently). The risk is also increased by the number of reoperations, as the anatomy becomes more complex and tissue planes are not easily defined. The surgeon should also exercise caution when excising scar tissue, taking care not to remove sclera and perforate the globe in the process.

An ischemic muscle is a weak muscle. Pressure from the scleral buckle may produce erosion and necrosis of segments of the rectus muscle. The muscle may rupture any time during the postoperative course or during strabismus surgery. Infection also increases the risk of muscle erosion, because it can lead to dissolution and necrosis of tissues.

Implant migration and extrusion is a real risk after removal of part of the buckle or the encircling element. This may be minimized by removing as little of the scleral buckle as necessary and by leaving the encircling element to maintain the integrity of the remaining buckle.

Persistent symptomatic diplopia may result despite the ophthalmologist’s best attempt at correcting strabismus. Some studies have found an incidence of persistent misalignment and diplopia as high as 20%. In many situations, torsion may be responsible for the inability to fuse. Macular ectopia and anisotony may aggravate fusion.

Finally, there will be a subset of patients whose central fusion will be permanently disrupted.

Prevention

The best way to control the incidence of postoperative symptomatic diplopia after retinal detachment repair is to prevent it. Close attention to meticulous surgical technique and careful handling of tissues at all times during ocular...
surgery will limit the degree of inflammation, and hence the fibrosis or adhesions that will ensue.\textsuperscript{13, 61, 62, 69, 105, 120} Mere dissection of periorcular tissue induces fibrosis.\textsuperscript{107} The surgeon should pay careful attention to preserving the relationships and function of Tenon’s fascia, muscle capsule, and sclera to prevent scar formation.\textsuperscript{2, 3} Stripping of the fascial attachments of the rectus muscles with a cotton-tipped applicator is discouraged.\textsuperscript{121} Disregard for fascia and extraocular muscle planes can lead to strabismus.\textsuperscript{23, 24} The use of viscoelastic material (e.g., Healon) has been suggested as well,\textsuperscript{26, 37, 54} but it remains in the eye for only 24 to 48 hours.\textsuperscript{121} Scarring and fibrosis may continue for several weeks to months after detachment surgery.\textsuperscript{121}

**STRABISMUS AFTER GLAUCOMA IMPLANT PROCEDURES**

**Historical Perspective**

Implantation of ocular drainage devices has become an important component in the control of intraocular pressure for recalcitrant cases of glaucoma. These devices include the Molteno implant,\textsuperscript{48, 49, 69, 108, 109} Schocket device,\textsuperscript{29} Baerveldt device,\textsuperscript{66, 67} Krupin valve,\textsuperscript{26, 56} and the Ahmed valve (Fig. 22–8).\textsuperscript{14, 15, 65} The dimensions of the different valves are summarized in Table 22–4.

**Incidence of Strabismus**

Several isolated case reports have noted the association of strabismus and diplopia with the Molteno implant,\textsuperscript{21, 83, 85, 119} the Baerveldt device,\textsuperscript{64, 95} and the Krupin disc.\textsuperscript{10, 33, 95} In larger series, the incidence of strabismus as a surgical complication ranges from 6% to 100% depending on the type of implant device used. The Molteno valve was associated with 6% to 47% incidence\textsuperscript{31, 109}; the 350-mm\textsuperscript{2} Baerveldt device with a 10% to 88% incidence\textsuperscript{66, 109}; and the 500-mm\textsuperscript{2} device with a 21% incidence.\textsuperscript{66} Strabismus was found in all patients given a Krupin disc.\textsuperscript{33, 66}

Strabismus is probably related to the technique of surgical placement, as well as to the size of these devices. The plates of the Molteno and the Ahmed valves are placed between the rectus muscles, with the interplate tube for the double-plated Molteno placed either over or under the rectus muscle. The Baerveldt device and the Krupin disc are inserted between adjacent muscles with the edges extending under the muscles (Fig. 22–9).\textsuperscript{109} The oval silastic disc of the Krupin valve had the greatest vertical height (1.75 mm)\textsuperscript{66} before the advent of the Ahmed valve (1.9 mm).

The true incidence of strabismus may be difficult to ascertain because of both a lack of preoperative strabismus evaluation and the high rate of poor visual function in patients undergoing glaucoma seton procedures. The risk is real, however, and caution should be exercised in patients at risk of developing symptomatic strabismus.\textsuperscript{33}

**Mechanisms for Development of Heterotropia**

**LEASH AND REVERSE LEASHES**

The phenomenon of the surgical leash has been postulated as the mechanism of heterotropia.\textsuperscript{4, 83, 95, 109} Limitation of

<table>
<thead>
<tr>
<th>Drainage Device</th>
<th>Surface Area (mm\textsuperscript{2})</th>
<th>Height (mm)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Single-plated Molteno</td>
<td>130–160</td>
<td>1.0</td>
</tr>
<tr>
<td>Double-plated Molteno</td>
<td>265–320</td>
<td>1.0</td>
</tr>
<tr>
<td>Baerveldt</td>
<td>200, 350, and 500</td>
<td>1.5</td>
</tr>
<tr>
<td>Krupin</td>
<td>195</td>
<td>1.75</td>
</tr>
<tr>
<td>Ahmed</td>
<td>184</td>
<td>1.9</td>
</tr>
</tbody>
</table>
movement may be confined to the field where the device is placed. Heterotropia may, however, be unrelated to the quadrant where the implant was placed.

**ADHESIONS AND SCAR TISSUE FORMATION**

Many patients with an oculardrainage device have had at least one surgical procedure in the past. The use of drainage devices is considered only after at least one failed glaucoma surgical procedure. The patients are therefore at an increased risk of inflammation and scar formation, which may eventually lead to restrictions (Fig. 22–10). If fibrosis occurs too far posteriorly, a posterior fixation suture effect may be created. Fat adherence, as described in the progressive motility disturbance after scleral buckling procedures, has also been postulated as a cause of strabismus after glaucoma implant procedures.

**MASS EFFECT**

The mass effect created by the glaucoma drainage device arises from two sources: the device itself and the cyst or bleb formation after drainage. The high rate of motility disturbance seen with larger devices has led some authors to recommend avoiding use of these implants in both binocular and monocular patients. Development of a large bleb may displace a muscle away from the sclera, altering the extraocular muscle’s length-tension relationship and affecting motility in the same way as a resection. The same mechanism has been implicated in acquired Brown syndrome in these patients because the distance between the trochlea and the SO is compromised by the presence of the bleb.

**MUSCLE FACTORS**

The location of the device or parts of it under a muscle belly can induce ischemia, necrosis, or direct trauma. The adjunctive use of antimetabolites such as mitomycin C by some surgeons may cause muscle toxicity. In contrast to this view, some authors suggest using these same agents to prevent fibrosis.

**Diagnosis**

The clinical presentations are as varied as the number of different implants and differing preimplant situations. Similar risk factors for the development of extraocular muscle imbalance apply to patients undergoing glaucoma implant procedures and those having retinal detachment repair.

The diagnosis is usually straightforward, because the relation of strabismus to the patient’s history of glaucoma surgery will be obvious. The need to identify preexisting motility disturbance stresses the importance of a preoperative motility evaluation before valve insertion. Poor vision in
these patients may disrupt fusion mechanisms. In advanced glaucoma where peripheral vision is lost, fusion mechanisms may already be compromised. A thorough strabismus evaluation, including forced duction, force generation, and saccadic velocity tests should be undertaken if indicated. Ultrasonography has been used to monitor the size of the bleb and its relation to the development of motility problems. Despite profound limitation of motility after valve placement, the forced duction test may be free.

Treatment

NONSURGICAL MANAGEMENT

Conservative management consists of observation, prisms, and chemodenervation. Prisms are of limited use because of incomitance or the large angle of the deviation common in these cases. Chemodenervation is an interesting alternative, but there has been no clinical report of its use.

SURGICAL MANAGEMENT

Strabismus surgeries are complicated by the concern for intraocular pressure control, the large bulk of the implant, and the involvement of two rectus muscles with either the Baerveldt device or the Krupin valve.

Passive traction testing will reveal mechanical limitation of rotation. If adhesions and scar tissue are contributing factors, the problem must be addressed before contemplating removal of the implant. Exploration and possibly removing the drainage device need to be discussed with the glaucoma surgeon. Replacing the implant with a smaller device may allow adequate pressure control.

A recession procedure is preferred over a resection because of the possibility of the latter procedure increasing the restriction. Transposition procedures may be difficult because the implant encroaches on the surgical field and sites of muscle attachment.

COMPLICATIONS

The foremost concern in reoperating for a motility disturbance is intraocular pressure control. Violation of the conjunctiva, the bleb, and the scar tissue may compromise control that already is tenuous by the time a patient receives a glaucoma drainage device. Further progression of glaucoma, with increasing optic nerve damage and loss of both central and peripheral vision, may be a risk not worth taking.

In certain patients with good vision, the diplopiccaused by the drainage device need to be discussed with the glaucoma surgeon. Replacing the implant with a smaller device may allow adequate pressure control.

A recession procedure is preferred over a resection because of the possibility of the latter procedure increasing the restriction. Transposition procedures may be difficult because the implant encroaches on the surgical field and sites of muscle attachment.

Prevention

The choice of an implant may reduce the risk of strabismus developing. The superior quadrant is preferred over the inferior, because the orbital roof has a greater capacity to accommodate blebs and cysts forming around the device. Patients whose motility disturbance is pronounced inferiorly have difficulty reading. If a choice can be made between the temporal and nasal quadrants, the former is preferred because there is more room in this area.

As pointed out earlier, the choice of an implant by the glaucoma surgeon should be influenced by the presence of binocularity and fusion, especially in patients who retain fairly good vision. Modifying the implant so that its height is decreased and bleb formation is more diffuse may be helpful.

REFERENCES

24. Dunlap EA: Plastic implants in muscle surgery: Plastic materials in...
84. Ogasawara H, Feke GT, Yoshida A, et al: Retinal blood flow alter-
STRABISMUS AFTER ORBITAL FRACtURES AND SINUS SURGERY

LISABETH S. HALL, MD, JOHN D. McCANN, MD, PhD, ROBERT A. GOLDBERG, MD, ALVINA PAULINE SANTIAGO, MD, and ARTHUR L. ROSENBAUM, MD

STRABISMUS AFTER ORBITAL FRACTURES

Trauma to the orbit may either be isolated or associated with other injuries to the head, neck, or body. The cause of ocular motility disturbance after orbital trauma often involves multiple mechanisms. Localized acute changes such as edema, hemorrhage, and soft tissue swelling may cause diplopia and gaze disturbances. Muscle entrapment and cranial nerve paresis cause more long-term sequelae and often require intervention by the orbital surgeon and/or the strabismologist.

Incidence

Strabismus after orbital trauma has been estimated to occur in about 58% of patients sustaining a blow-out fracture. In a study of 363 patients developing diplopia within the first week of sustaining a facial fracture from blunt trauma, 82% had resolution of diplopia within 6 months of the injury. Only 1 patient required strabismus surgery. The main risk factors for diplopia were a vehicular accident, blow-out fracture, and comminuted malar fracture. Other authors found that, although a majority of patients experience relief of diplopia, 24% require strabismus surgery. In a study of 40 patients who sustained blow-out fractures of the orbital floor, 5 showed no evidence of strabismus or diplopia and 7 had a motility disturbance consistent with an extraocular muscle or cranial nerve palsy, confirmed by forced duction testing. Diplopia had resolved in 4 of these 7 patients after 1 year.

Classification of Orbital Fractures

Orbital fractures are commonly classified on the basis of their location. The most common isolated injury affects the orbital floor. The typical patient with an orbital blow-out fracture is a young man involved in a fist fight. In one series, 90% of patients were men with an average age of 26 years. In women, orbital fractures often result from domestic violence. The youngest patient reported was an 8-month-old infant who developed inferior rectus (IR) contracture caused by orbital trauma sustained in an assisted forceps delivery.

Low-velocity blunt trauma, such as a ball striking the orbital rim, causes an acute rise in orbital pressure, resulting in displacement at the weakest point—the orbital floor (Fig. 23–1). The term blow-out fracture was coined by Smith and Regan: it should be reserved for orbital floor fractures not associated with an orbital rim fracture. Orbital floor fractures may also involve the infraorbital rim, but these are not considered to be blow-out fractures. In high-velocity injuries, displaced fractures of the malar bone, nasoethmoid complex, or maxillary buttress typically extend to involve the lateral and medial orbital walls and the orbital floor.

Medial orbital wall fractures are less common but more frequent than generally recognized. The medial orbital wall is actually the thinnest part of the orbit, but the adjacent ethmoidal sinuses buttress it and make it more resilient. Isolated medial orbital wall fractures are uncommon; they typically occur in combination with orbital floor injury. Indi-
CLINICAL STRABISMUS MANAGEMENT

Figure 23–1. Mechanism of low-velocity orbital floor fracture. Blunt trauma to the orbit causes an acute rise in orbital pressure, fracturing the orbit at the weakest point along the orbital floor, with antecedent prolapse of orbital soft tissue into the maxillary sinus. (From Nesi FA, Spoor TC. Orbital fractures. In Klein E [ed]: Ophthalmic Plastic and Reconstructive Surgery. St. Louis, CV Mosby Co. 1987:473. Reprinted with permission.)

cations for repair of medial orbital wall fractures include horizontal diplopia and enophthalmos. If both the orbital floor and medial orbital wall are fractured, these injuries will have an additive effect on enophthalmos.

Lateral orbital wall fractures are most commonly associated with a displaced fracture of the malar bone. The weakest point on the lateral orbital wall is the frontozygomatic suture, the most common point of fracture. A lateral orbital wall fracture rarely results in lateral rectus entrapment. However, it is unusual to have a displaced lateral orbital wall fracture without fracture of the orbital floor as well. Thus, a lateral orbital wall fracture may be associated with IR restriction along the orbital floor.

The orbital roof is the most resilient section and is typically fractured in high-velocity injuries. The degree of trauma required to fracture the orbital roof accounts for the high incidence of concomitant injuries of the globe, optic nerve, and nerves controlling extraocular motility. A blow-in fracture may be caused by downward displacement of the orbital roof with a decrease in orbital volume. This is a relatively uncommon condition, characterized by an inwardly displaced fracture of the orbital rim or wall that decreases the orbital volume and causes proptosis. Because of the high risk of damage to the frontal sinus, cerebrospinal fluid leakage, and cerebral contusion, these fractures are often managed in collaboration with a neurosurgical team.

Mechanisms of Strabismus After Orbital Fracture

The proposed mechanism of extraocular muscle dysfunction after orbital fracture has undergone evolution over the years. Lerman described the effect of IR muscle “entrapment” on motility and concluded that, if entrapment is located posterior to the equator of the globe, restriction of depression results. Correspondingly, entrapment anterior to the equator leads to restricted elevation. Anterior orbital floor fractures may also cause inferior displacement of the globe, which further impairs its elevation.

True entrapment of the IR muscle is rare. Instead, the fibrous orbital septa pull the IR muscle toward the fractured orbital floor, resulting in extraocular muscle restriction (Fig. 23–2). In one study, all patients with an orbital floor fracture showed evidence of IR thickening or displacement toward the fracture site. Imaging studies failed to demonstrate true entrapment of the IR muscle in any patient.

Blunt trauma may also injure an extraocular muscle or its innervation, resulting in paresis without restriction. Wojno and associates described 7 of 40 patients with blow-out fractures who had palsy of a single extraocular muscle. These patients showed no evidence of entrapment on radiologic studies or forced duction testing. All but 1 of them had spontaneous resolution of symptoms. Patients may also present with simultaneous restriction and paresis of a muscle. This is important to recognize because release of the incarcerated tissue may cause the paretic component of strabismus to become manifest, thereby transiently increasing the angle of strabismus.

Clinical Characteristics

The classic blow-out fracture caused by blunt trauma exhibits a discontinuity of the orbital floor, as shown on plain films and CT scans (Fig. 23–3). Considerable advancements in imaging techniques have made many previously undetected—and often unsuspected—fractures identifiable, particularly those of the medial orbital wall. The “typical” strabismus patient with an inferior orbital floor fracture and entrapment of the ipsilateral IR will have hypotropia on primary gaze with limited supraduction (Fig. 23–4). Depending on the location and the degree of entrapment, the size of the deviation in primary gaze may vary. The function of the IR will also be determined by the nature of the

Figure 23–2. Restriction of the inferior rectus in association with orbital floor fracture rarely is caused by true muscle entrapment. The typical cause is herniation of orbital fat and associated fascia into the maxillary sinus with tethering of the inferior rectus to the prolapsed soft tissue. (From Koornneef L: Current concepts on the management of orbital blow-out fractures. Ann Plast Surg 1982;9:185. Reprinted with permission of Lippincott-Raven.)
entrapment as well as the degree of damage to the muscle and/or nerve.

Varied patterns of strabismus are seen after orbital trauma depending on both muscle paresis and restrictive factors. The most frequently encountered types are discussed here, giving attention to the treatment of complex problems. Rare motility disturbances reported in the literature include acquired Brown syndrome (Fig. 23–5), several cases of retraction syndrome,27–57 loss of accommodation with convergence insufficiency,1 and inferior oblique palsy.96

**Diagnosis**

**CLINICAL EVALUATION**

The evaluation of strabismus in patients who have suffered orbital trauma is particularly challenging. A motility disturbance may result from cranial nerve palsy, extraocular muscle weakness or entrapment, edema, hemorrhage, or incarcerated soft tissue. The clinical findings vary considerably, depending on the timing of the evaluation in relation to the injury. Knowledge of the anatomy and appropriate diagnostic and surgical principles is necessary to properly manage these complex cases.

Taking a careful history is crucial in assessing and treating a patient with orbital trauma. The date and nature of the injury must be determined, as well as the possibility of previous orbital trauma or strabismus. Often patients presenting with an unexplained motility disorder will have a vague history of prior facial trauma that is elicited only by careful history taking. In acute cases the diagnosis will be evident. The differential diagnosis includes any disorder that could give rise to an incomitant vertical strabismus (Table 23–1). A thorough history will greatly narrow the differential diagnosis, but, again, a clear history of trauma may not always be obtained. An occult fracture must always be suspected in a patient with vertical diplopia and limited upgaze, even in the pediatric population (Fig. 23–6).

When orbital trauma is suspected, particular attention should be paid to certain aspects of the examination. Facial symmetry should be observed from both a direct frontal and a superior view (above the patient). Exophthalmometric measurements will disclose relative enophthalmos and/or exophthalmos. Palpation of the orbital rim may reveal a bony “step off” characteristic of a fracture. Crepitus on palpation around the orbit implies communication with the sinuses. Intraocular pressure that increases in upgaze suggests a tight IR muscle.

Observation of the patient when relaxed, fixing both in the distance and at near, should precede cover testing. Any anomalous head position may suggest that the patient is able to fuse. In assessing versions and ductions, special attention must be given to any restrictive or paretic muscles creating the illusion of an overactive yoke muscle. For example, a tethered IR muscle may decrease elevation in abduction.

<table>
<thead>
<tr>
<th>Differential Diagnosis of Strabismus After Orbital Trauma</th>
</tr>
</thead>
<tbody>
<tr>
<td>Myasthenia gravis</td>
</tr>
<tr>
<td>Thyroid orbitopathy</td>
</tr>
<tr>
<td>Orbital tumor</td>
</tr>
</tbody>
</table>

**Figure 23–3.** Coronal CT scan of the mid orbit demonstrating classic appearance of blow-out orbital floor fracture. Note opacification of the maxillary and ethmoidal sinus with prolapse of the orbital soft tissue into the maxillary sinus.

**Figure 23–4.** The most common presentation after orbital floor fracture is restriction of the ipsilateral inferior rectus. Note that there is right hypertropia in primary gaze that increases in upgaze from restriction of the right inferior rectus. Downgaze is preserved, indicating that innervation to the right inferior rectus is intact.

**Figure 23–5.** Acquired Brown syndrome in the right eye resulting from facial physical assault. Note limitation of elevation on adduction of the patient’s right eye (top right) and facial scars involving the trochlear region. There was no evidence of right superior oblique palsy on examination; downgaze in adduction is full (bottom right).
causing the yoke of the superior rectus (SR)—the contralateral inferior oblique—to appear overactive (Fig. 23–7). Metz\textsuperscript{39} describes an orbital floor fracture causing a typical direct leash-type restriction. Clinically, the "leash" will not limit movement until the muscle reaches a certain point, where the eye will suddenly stop. In cases of trauma with excessive scarring, a "reverse leash" effect may be seen.\textsuperscript{17} This results from a shortening of conjunctiva or Tenon's tissue, causing a restriction on the same side to which the eye is attempting to move.

When measuring the angle of deviation in cases of paresis and/or restriction, it is important to consider primary and secondary deviations. Alternate prism cover testing should be carried out in all cardinal positions of gaze. Measurement of the prism neutralization needed to achieve fusion in free space will help determine the potential for fusion after surgery. All patients with incomitant strabismus must be measured during forced head tilt to the right and left to identify a possible superimposed fourth nerve or inferior oblique palsy. Sixth nerve palsy has also been reported after orbital trauma.\textsuperscript{41}

It is important to distinguish between strabismic deviations caused by a restriction and those caused by a paresis or palsy. Some patients may exhibit features of both. Forced duction and force generation testing are required for all patients suspected of having a restrictive or paretic muscle. These may be performed under topical anesthesia in the office and are described in detail in Chapter 3. Saccadic velocity analysis assesses the degree of paresis of an extraocular muscle (Fig. 23–8). It is also helpful in determining the degree of recovery of muscle function.

Both paretic and restrictive processes may cause ocular torsion and subsequent torsional diplopia. Often the patient is not clearly aware of abnormal torsion. One clinical sign of torsional diplopia is noted when appropriate vertical and horizontal deviations are corrected with prisms in a fusing patient. Although the images are reported as "single," they are still not perceived clearly. The double Maddox rod test may serve to evaluate subjective torsion. Objective torsion can be assessed during the dilated fundus examination. Static observations are made using the indirect ophthalmoscope. A dynamic examination is also helpful and is done by having the patient look in the desired field while observing the degree of fundus torsion generated by the muscle in question. For example, an IR restriction may cause excyclotorsion that increases when the patient looks down. In the case of IR paresis, fundus incyclotorsion may be evident and may increase in downgaze.

**LABORATORY EVALUATION**

Force generation and saccadic velocities can be estimated clinically as described in Chapter 3. Quantitative analysis becomes important where paresis is suspected but uncertain, and the timing of the surgery is in question. In cases of muscle paresis the surgeon should allow a full 6 months to elapse to allow the paresis to resolve spontaneously.

Binocular diplopia-free field testing may help in planning surgery.\textsuperscript{12, 42} Often a major improvement in motility is not a realistic goal. Nevertheless, transposition surgery may expand the patient's binocular field and/or reposition it closer to the primary position, thereby correcting an anomalous head posture (Fig. 23–9). Some patients may experience instability of this diplopia-free binocular area when fatigued or when generating refixation saccades.\textsuperscript{13} The binocular diplopia-free field is usually larger when the object of regard moves from within the field of binocular single vision and smaller when the object moves from the periphery toward the binocular diplopia-free field.\textsuperscript{51}

The Lancaster red-green or Hess test may also be used when planning surgery for incomitant deviations and is valuable for postoperative evaluation of alignment. The possibility of disrupted central fusion should be entertained in a patient who has sustained significant head trauma. A synoptophore evaluation may be helpful.

![Figure 23-7](image-url)
RADIOGRAPHIC EVALUATION

Historically, Caldwell and Waters roentgenograms of the face were used to detect displacement of the orbital walls, outline prolapsed orbital contents, and demonstrate air-fluid levels in the periorbital sinuses. Computed tomography (CT) is excellent for delineating orbital detail (Fig. 23–10). Magnetic resonance imaging (MRI) is less sensitive than high-resolution CT when imaging fractures in the maxillofacial area. In rare cases, MRI may add useful information about entrapped soft tissue (Fig. 23–11). Orbital ultrasonography may also aid the diagnosis of orbital fracture, but its usefulness correlates directly with the examiner’s expertise.

A screening CT study typically includes only thick axial cuts through the brain and orbit. One may find signs of orbital fracture, such as blood in the sinuses or air in the orbit, on a screening examination, but the chance of visualizing a small orbital floor fracture is nil. In contrast, these fractures are always seen on a coronal projection, which provides excellent images of the medial and lateral extent of the fracture. If it is crucial to delineate the anteroposterior extent of a fracture, sagittal re-formations should be obtained. The coronal and oblique sagittal projections are also ideal for evaluating fractures of the orbital roof. The axial projection is most helpful for evaluating the medial and lateral orbital walls.

All patients with a suspected orbital fracture should benefit from nonenhanced, thin-slice, direct axial and direct coronal CT scans of the orbits. If more detail of the anteroposterior extent of the fracture is needed, oblique sagittal re-formations of the images may be obtained along the axis of the IR muscle (Fig. 23–12). Three-dimensional reconstruction of orbital CT scans may add useful information on complex fractures involving the malar bone.

Interpreting CT images of orbital fractures should focus...
on three factors: their location, the effect of a fracture on orbital volume, and its effect on the orbital soft tissues. The location of a fracture site is crucial in guiding treatment. A loss of orbital volume posterior to the equator of the globe will present as enophthalmos, whereas a loss of volume anterior to the equator will manifest as hypoglobus (Fig. 23–13). Posterior fractures are more likely to result in muscle entrapment, which may be difficult to release. True entrapment of the IR muscle is unlikely to occur in the anterior orbit because of the distance of the IR muscle from the orbital floor, but it may occur in the mid and posterior orbit because the IR is more intimately associated with the floor as one proceeds posteriorly into the orbit. Biesman described a 95% (19/20) prevalence of posterior floor fracture in patients having persistent diplopia after surgical intervention. In those with a posterior blow-out fracture, hypertropia, and limited depression, forcedduction testing may be nonrestrictive but the CT scan may suggest posterior entrapment of the muscle (Fig. 23–14). In such
cases, orbital exploration and placement of an orbital floor implant may eliminate or improve the diplopia.\textsuperscript{30} Flaccid paralysis of the IR muscle anterior to the fracture site may be a possible mechanism.\textsuperscript{71}

The effect of a fracture on orbital volume is also crucial. A study showed that none of 263 patients with nondisplaced fractures of the orbital floor who were observed without surgery developed enophthalmos.\textsuperscript{40} Patients presenting late with an un repaired orbital fracture involving more than one fourth of the orbital floor exhibit significant enophthalmos.\textsuperscript{34} Orbital expansion and herniation of orbital contents through the fracture site on CT correlate with late enophthalmos.\textsuperscript{26}

A conflict of terminology exists between orbital surgeons and radiologists. Most orbital surgeons agree that true entrapment of an extraocular muscle in a fracture site is unusual. It is more common to find orbital fat and associated fibrous septa entrapped in the fracture site, resulting in muscle restriction.\textsuperscript{30, 61} Given the relative rarity of actual entrapment on surgical exploration, it is surprising how often it is noted on CT scans. In one study, all patients without restriction of the IR had lost their diplopia within 2 months, whereas all those with entrapped muscles required surgical intervention.\textsuperscript{26} Only patients with strong evidence of muscle entrapment on CT require surgery for diplopia.\textsuperscript{10} Nonvisualization of the IR muscle on sequential coronal slices through the mid orbit suggests IR muscle entrapment.\textsuperscript{3}

**Treatment**

The treatment of strabismus after orbital trauma is intimately related to the timing of the incident and must be carefully coordinated with the orbital surgeon in the immediate postinjury period. Treatment is designed to meet the needs of an individual patient and must be planned only after thorough evaluation of the motility disorder. Treatment options include early intervention to repair the orbital defect and free entrapments or restrictions, prism spectacles, botulinum toxin, strabismus surgery, and observation.

**TREATMENT OF ORBITAL FRACTURE**

**Indications for Repair of Orbital Fracture**

Diplopia resolved in 76% of patients with orbital fracture who were managed without surgery.\textsuperscript{62} On the basis of these results, it was recommended that all patients with blow-out fractures be observed for 4 to 6 months before considering repair. Other studies also support the thesis that diplopia after a blow-out fracture will resolve without treatment in more than 3 of 4 patients.\textsuperscript{13, 19} It is difficult to compare results from surgically and nonsurgically treated patients, because the former usually have more severe involvement. However, review of the literature demonstrates that only 2 of 3 patients operated on early for an orbital floor fracture will have resolution of diplopia,\textsuperscript{9, 19, 23, 34, 76} and only 1 of 3 operated on 4 to 6 months after injury will be diplopia free.\textsuperscript{14, 34} These data suggest that spontaneous resolution of diplopia after a blow-out fracture is common. A patient with a large angle of strabismus and evidence of muscle entrapment or restriction is likely to improve with the release of incarcerated tissue, but many of these patients will still need strabismus surgery to alleviate diplopia.
In 1983, Hawes and associates suggested that patients with a fracture involving more than 50% of the orbital floor should be operated on within 2 weeks of the injury. This important paper points out that enophthalmos is common in unrepaired patients with fractures of this extent and that repair of enophthalmos is more successful if performed early. The greatest benefit patients receive from early surgery for orbital floor fracture is a reduced risk of cosmetically unacceptable enophthalmos.

Orbital surgery to correct enophthalmos and diplopia is more effective when done before fibrosis and scarring of tissue into the fracture site develop. The surgeon’s dilemma is to determine which patients will develop diplopia or enophthalmos if surgery is not performed. Most patients with diplopia will recover without operative intervention. In the first few days after trauma it can be difficult to evaluate enophthalmos because it is masked by orbital swelling. This may be partially overcome by treating patients with prednisone in a dose of 1 mg/kg for 10 days after injury. Corticosteroids decrease the time needed for diplopia to spontaneously improve; reduce orbital swelling, unmasking enophthalmos; and retard the formation of scar tissue.

Protocol for Treatment of Orbital Fractures

All patients who suffer orbital fracture should undergo a complete ophthalmic examination to rule out damage to the globe and optic nerve. Special note is made of infraorbital anesthesia, periorbital swelling, enophthalmos, ductions, the angle of strabismus, and step off of the orbital rim. If an orbital fracture is suspected a CT study is obtained. If fracture repair is being considered, the patient is started on an oral antibiotic and prednisone as just described. If the fracture involves more than 50% of the orbital floor, surgery is recommended 10 to 14 days after injury. If there is evidence of true muscle entrapment on a CT scan or gross, clinically evident restriction, we tend to operate earlier. For fractures involving more than just the orbital floor, surgery is recommended if the expansion of orbital volume exceeds 4 sq cm. Surgical intervention is also suggested if there is significant displacement of the malar bone or zygomatic arch.

Patients with smaller fractures are reevaluated 10 days after injury. If 2 mm or more of enophthalmos is present at the second evaluation, surgery is recommended. If diplopia is present, clinical evidence of entrapment is sought. An elevation of intraocular pressure to more than 45 mm Hg in upgaze is taken as evidence of entrapment. Forced duction testing is done in the office, and surgery is recommended if the test shows unequivocal restriction of upward rotation.

Surgical Considerations in Early Fracture Repair

We perform more than 90% of all orbital floor fracture repairs using the lower eyelid transconjunctival approach. This route offers better visualization of the orbital floor and is not plagued by a high incidence of late lower eyelid retraction. Repair begins with a forced duction test as a baseline for judging the success of surgery. Great care is taken to identify and cauterize the vessel passing from the infraorbital neurovascular bundle to the IR to prevent hemorrhage. Successful repair requires freeing all incarcerated tissue from the fracture site. Typically, releasing tissue from the posterior aspect of the fracture is most difficult. It is imperative that dissection be carried to the depth required to create posterior support for the orbital implant. The posterior orbital floor has an upward slant where the implant should be placed (differentiate it from the maxillary sinus). Sewall orbital retractors and a bright head lamp are useful in this dissection.

An orbital implant is fashioned in the size and shape needed to re-create a normal contour of the bony orbit (Fig. 23-15). In many large fractures it is difficult to find medial support for the implant because of comminution of the medial orbital wall. In these cases, dissection is carried high onto the junction of the medial orbital wall with the orbital roof to locate support for the implant. Typically, we friction-fit implants, but if the medial wall is comminuted and there is little superomedial support, the implant is fixed to prevent its migration. The implant should be contoured to re-create the oblique coronal projection of the medial orbital wall. It is important that no soft tissue be allowed to prolapse between the implant and edge of the fracture, because this may create restrictive strabismus. After inserting the implant the forced duction test is repeated to confirm that the operation has reduced muscle restriction.

Surgical Considerations in Late Fracture Repair

Late fracture repair is best avoided by appropriately prompt surgical intervention. Unfortunately, some patients who need early intervention will not be identified even with the protocol just described. In addition, some patients will have concomitant globe or head trauma that precludes early intervention.

Orbital surgery should be reserved for patients with cosmetically unacceptable enophthalmos, hypoglobus, or restrictive strabismus. If forced duction testing gives evidence of restriction, orbital surgeons usually recommend freeing the prolapsed tissue to reduce the amount of restriction but warn the patient that strabismus surgery probably will be needed to alleviate diplopia in primary gaze. If a large

Figure 23-15. A titanium implant shaped to fit the contour of the junction of the orbital floor with the median orbital wall is fixed in this position.
recession of the IR is performed, simultaneous surgery on the lower eyelid retractors may prevent inferior scleral show and lagophthalmos.

Surgery to correct a narrow palpebral fissure caused by enophthalmos after a blow-out fracture often is better directed at the narrow fissure than at the enophthalmos itself. Adequate cosmetic results may be achieved by correcting ptosis alone. Patients may also suffer from paradoxical eyelid retraction after a blow-out fracture with enophthalmos (Fig. 23–16), which can be corrected by excising Müller muscle or by recessing the levator aponeurosis.

If enophthalmos cannot be disguised adequately by eyelid surgery, the orbital volume should be supplemented. The complexity of surgical management in these late cases exceeds the scope of this chapter.

TREATMENT OF STRABISMUS

Nonsurgical Management

Observation is appropriate when there is no radiographic evidence of entrapment in the acute phase. Conservative management is recommended in this setting even if disabling diplopia is present. With extraocular muscle or cranial nerve palsy, a majority of patients improved after 1 year without intervention. Puttermann and associates reported patients with edema and hemorrhage in the orbit or within a muscle causing a motility disorder, with restriction confirmed by forcedduction testing. A majority of these cases resolved over time, although clinically they may mimic entrapment of an extraocular muscle.

Optical solutions to diplopia are difficult to achieve, because most deviations are incomitant. Prisms may be helpful in the primary position, but the range of binocularity will be small and patients are not typically satisfied with prismatic correction. Selected patients with small vertical deviations in primary gaze do respond to prisms, but they will require separate reading glasses for downgaze with a different prismatic correction.

Treatment of the antagonist with botulinum toxin to help prevent contracture may be considered if paresis is confirmed. The use of chemical denervation in cases of orbital fracture is not specifically described in the literature, but it may be useful in some cases.

Surgical Management

Because of extreme variability in clinical presentation, surgical planning must be tailored to the individual patient. The timing of surgery for strabismus in cases of orbital fracture depends on multiple factors. In specific cases, immediate repair will relieve the restriction and correct strabismus. Later, after excessive scarring has occurred, it is better to treat strabismus alone without exploring the orbit. Even in patients with radiographic and clinical evidence of entrapment, late repair rarely helps to resolve the strabismus. These patients must be approached on an individual basis by a multidisciplinary team. Several types of patient presentations are described along with suggestions for treatment.

Inferior Rectus Restriction; Hypotropia in Primary Gaze. The most common extraocular complication of an orbital floor fracture is restriction of the ipsilateral IR muscle. This is usually a result of the initial trauma but has also been reported after repair of a fracture with a Teflon implant. Commonly, there is hypotropia in the primary position owing to the restricted IR muscle. Upgaze is limited above the midline, but the patient is able to fuse in downgaze where IR function is not compromised. The downward vertical saccade is normal, and forcedduction testing reveals restricted elevation. Force generation testing reveals no evidence of IR paresis (see Fig. 23–4). In this case an IR recession is desirable, with the goal of correcting the deviation in the primary position without causing hypertropia in downgaze. An adjustable suture technique helps to prevent an overcorrection in downgaze. Some restriction to full upward rotation of the globe may persist.

Inferior Rectus Palsy. Paresis of the IR often causes diplopia in patients incurring blow-out fractures. It may be the result of direct damage to the extraocular muscle or cranial nerve. Patients typically present with hypertropia in primary position and downgaze, and they may develop secondary SR contracture (Fig. 23–17). Some authors speculate that hypertropia results from a posterior entrapment that causes a change in the effective origin and insertion of the
CLINICAL STRABISMUS MANAGEMENT

Figure 23-18. Inferior rectus paresis and restriction after orbital floor fracture. Combined inferior rectus paresis and restriction may cause orthotropia, hypotropia, or hypertropia in primary gaze. This patient adopts a small chin-down position to fuse, as evidenced by the inferior scleral show in both eyes. Photographs demonstrate right hypotropia in upgaze (restriction) and right hypertropia in downgaze (paresis). Note pseudo-overaction of the left inferior oblique due to fixation duress.

If weakness of the IR is partial and the hyperdeviation is less than 10 PD, the IR may be resected. If the hyperdeviation is large, the antagonist SR should be recessed in addition to performing a resection of the paretic muscle. Other authors recommend posterior fixation sutures on the contralateral eye, which may be combined with IR recession.

If IR palsy is complete, or for some reason a resection of the IR is not feasible, horizontal rectus transposition may be beneficial (see also Chapter 36).

Combined Paresis and Restriction. Patients with both restriction and paresis of the IR muscle after a blow-out fracture of the inferior orbital floor require more complex surgical management. The patients may be orthotropic, hypotropic, or hypertropic in primary gaze and may exhibit hypotropia in upgaze with hypertropia in the reading position (Fig. 23–18). They experience diplopia in downgaze caused by hypertropia when the paretic IR fails to fully depress the eye. Diplopia on upgaze occurs because the IR restricts upgaze. Ocular rotations will be at least moderately limited in both upgaze and downgaze. Forced duction testing will reveal restriction of upward gaze. Some patients will exhibit a chin-up posture to fuse, but the diplopia-free binocular fields are usually limited.

Recessing both the IR and the SR of the injured eye may expand the binocular diplopia-free field (see also Fig. 23–9). However, any recession performed on a paretic IR may aggravate downgaze limitation in the involved eye unless the IR paresis is mild.

For a small hypotropia (less than 8–10 PD) the contralateral SR may be recessed. If the hypotropia is large, a small (4 mm) recession of the IR to improve upgaze may be combined with contralateral SR recession for primary position correction. A posterior fixation suture on the contralateral IR will expand the diplopia-free binocular field in downgaze.

If the patient is orthotropic in primary position, a resection of the ipsilateral IR should be done to improve downgaze. This will create a hypertropia in primary position that will require a contralateral SR recession.

If the primary-position deviation is hypertropia, a small resection of the traumatized IR may improve primary-position and downgaze deviation but aggravate it in upgaze. If IR function is totally absent, one should consider horizontal rectus transposition with possible ciliary vessel sparing.

If the surgeon believes that the restriction in upgaze will be aggravated by an IR resection, an alternative is to perform horizontal rectus transposition to straddle the IR insertion (inverse Knapp) if the hypertropia in primary deviation exceeds 15 PD (Fig. 23–19).

Inferior Rectus Restriction, Orthotropia in Primary Gaze. A patient with a restricted or entrapped IR may be orthotropic in the primary position but have upgaze limitation (Fig. 23–20). Very few patients or occupations require...
expansion of the binocular diplopia-free field in upgaze. Such patients usually can be managed conservatively. If the binocular diplopia-free fields need to be improved in upgaze, a posterior fixation suture (fadenoperation) on the contralateral SR, matching the restriction in upgaze, will expand the diplopia-free fields. Introduced by Cüppers in 1976, suturing of a muscle to the globe 10 mm posterior to its insertion weakens the muscle in its field of action (see Chapter 37). An ipsilateral IR recession is best avoided in this situation because of the risk of causing a vertical deviation in the primary position.

**Medial Rectus Entrapment or Restriction.** Fractures of the medial wall are less often encountered but may occur even from minor orbital trauma. The most significant strabismic complication is entrapment of the medial rectus (MR) muscle, but MR palsy may also occur. Clinical signs of restriction or entrapment include horizontal diplopia, retraction of the globe on abduction, severe enophthalmos, and an abduction deficit. Plain films and CT scans are often not helpful in making the diagnosis, but forced duction testing may be more informative. Saccadic velocity analysis can be invaluable for detecting a superimposed sixth nerve paresis that may follow head trauma. The treatment algorithm is similar to that for isolated IR restriction, as described earlier, except that surgery is directed at the MR muscle.

### STRABISMUS AFTER SINUS SURGERY

There have been numerous reports of ocular injury caused by sinus surgery. Dutton estimated that some type of orbital injury occurs in up to 3% of patients having paranasal sinus surgery. Damage to an extraocular muscle or its innervation is only one of the ocular complications reported (Table 23–2).

#### Mechanism of Injury

Open sinus surgery that creates a window in the frontal sinus employs an incision in the superonasal quadrant of the orbit (modified Lynch incision) that extends halfway between the inner canthus and the anterior aspect of the nasal bones. The trochlea is a U-shaped cartilage that encases the tendon of the superior oblique (SO) and is attached to the frontal bone a few millimeters behind the orbital margin. This surgical approach is associated with complications affecting the SO muscle/tendon complex. SO palsy may occur in the following situations: (1) the incision is made or extended more inferiorly; (2) excessive periosteal stripping near the trochlea is carried out and not repaired; (3) direct muscle damage to the SO tendon or trochlea occurs from instruments such as a drill bur, chisel, or rongeur; (4) the blood or nerve supply to the SO tendon is disrupted or compromised. Acquired Brown syndrome causing limited elevation in adduction may occur after cicatrical adhesions form in the area of the SO. Orbital hemorrhage resulting in IR palsy has also been reported after open sinus surgery. The mechanism may be compression of the IR muscle or its blood and nerve supply against the orbital floor.

Intranasal surgery, usually performed for nasal polyps or anterior ethmoidectomy, may lead to direct injury of the MR and/or the adjacent SO. More recently, the trend toward performing endoscopic sinus surgery risks damage to extraocular muscles specifically when the procedure extends into the middle or posterior ethmoidal sinus. The bony wall between the ethmoidal sinus and the orbit is normally very thin and may even be eroded by chronic sinusitis that has continued for months or years. Sinus surgeons are trained to recognize the presence of orbital fat as a warning sign that the endoscopic surgical dissection has extended into the orbit. However, blood may obscure the orbital fat, or it may be recognized only after actual entry and orbital tissue injury have already occurred.

Because of its proximity, the MR is at risk of endoscopic trauma. The SO tendon, which is only a few millimeters above the MR muscle, may also be damaged. Injuries range

---

**Table 23–2. Ocular Complications of Sinus Surgery**

<table>
<thead>
<tr>
<th>Enophthalmos</th>
</tr>
</thead>
<tbody>
<tr>
<td>Optic nerve atrophy</td>
</tr>
<tr>
<td>Orbital cellulitis</td>
</tr>
<tr>
<td>Orbital hemorrhage</td>
</tr>
<tr>
<td>Orbital wall defect</td>
</tr>
<tr>
<td>Floor</td>
</tr>
<tr>
<td>Medial wall</td>
</tr>
<tr>
<td>Proptosis</td>
</tr>
<tr>
<td>Ptosis</td>
</tr>
<tr>
<td>Strabismus</td>
</tr>
<tr>
<td>Exotropia</td>
</tr>
<tr>
<td>Third cranial nerve palsy</td>
</tr>
<tr>
<td>Medial rectus scarring, paresis, or severance</td>
</tr>
<tr>
<td>Superior oblique palsy or restriction</td>
</tr>
<tr>
<td>Inferior rectus paresis</td>
</tr>
</tbody>
</table>

---
from mild contusion to almost total destruction of the extraocular muscle.

**Diagnosis**

Forced duction and force generation testing demonstrate the extent of muscle paresis and the presence or absence of mechanical restriction. Clinical observations of saccadic velocity also help in judging the degree of muscle function that remains.

Dynamic MRI is particularly helpful in these cases to confirm the presence and degree of actual muscle destruction or to identify both ends of a transected or tenotomized muscle (Fig. 23–21). This information helps the surgeon decide whether exploration can lead to recovery of the traumatized muscle. If extensive muscle destruction has occurred, imaging shows whether the remaining proximal muscle segment is still functioning. Serial saccadic velocity analysis indicates whether muscle function is improving.

**Treatment**

If dynamic MRI reveals an intact but paretic MR muscle, botulinum toxin injection of the ipsilateral antagonist (lateral rectus) may prevent secondary contracture (see Fig. 23–21).

Definitive surgical intervention is based on the degree of muscle recovery present 6 months or longer after the initial insult. General surgical principles concerning isolated muscle palsy are covered in Chapter 19. Which procedure is chosen depends on the type of induced muscle deficiency. Immediate repair of a muscle laceration or transection is indicated. Absorbable muscle sutures (e.g., Vicryl 6-0) are preferred for repairing the muscles. If extensive muscle damage is noted intraoperatively, the sinus surgery is completed and the patient is referred to a strabismus specialist promptly.

If MR injury is minor, patients may develop signs of MR paresis and consequent exotropia. Appropriate horizontal rectus surgery then may be performed at a later date. If a portion of the MR muscle was removed, the strabismus surgeon should determine whether the muscle can be repaired or should be resected.

If dynamic MRI reveals a large segment of muscle destruction, surgery may be done earlier because spontaneous improvement in muscle function will not occur. In this situation, full vertical rectus muscle transposition to the MR and botulinum toxin injection of the ipsilateral lateral rectus may be necessary. Medial transposition of the vertical rectus muscles creates slack by reducing the angle between the muscle plane of the vertical rectus and the optical axis (see Fig. 36–13). For optimal effect, each transposed muscle is resected to compensate for this slack (see also Chapter 36).

In some cases a segment of MR has been destroyed but a large functioning proximal segment of MR remains. This segment may be recovered via a medial orbitotomy incision and attached to the globe. If required, a bridge of temporalis fascia between the muscle and the globe may be used to provide active muscle force from the remaining muscle seg-

---

**Figure 23–21.** After endoscopic sinus surgery with septoplasty and turbinectomy, this patient complained of horizontal diplopia. He was able to fuse in far right gaze with a 35-degree left face turn. A, Ocular rotations revealed inability to adduct the right eye past midline. There was marked slowing of adducting saccades of the right eye. Findings were consistent with right medial rectus palsy. B, High-resolution magnetic resonance imaging (MRI) demonstrated an intact right medial rectus from insertion to the annulus of Zinn, but ragged contours suggest injury at several sites due to instrumentation. (The right medial rectus was contracting poorly by dynamic MRI evaluation.) C, Nine months after sinus surgery without any strabismus intervention, the primary position deviation was 5 PD of exophoria, with mild right adduction deficiency, and 20 PD of exotropia persisting on left gaze. A posterior fixation suture on the left lateral rectus was considered to expand his binocular diplopia-free field.
Strabismus after Orbital Fractures and Sinus Surgery

Figure 23–22. Damage to the medial rectus muscle after sinus surgery. A, Clinical photographs demonstrate inability to adduct the left globe past midline. Left exotropia and left hypotropia improves slightly on left gaze but is worse on right gaze. B, Dynamic magnetic resonance imaging with surface coil demonstrated a 20-mm segment of the medial rectus muscle missing with only the proximal segment functioning. C, Excellent alignment with fusion in primary position was achieved with a temporalis fascia bridge between the proximal segment of the extirpated medial rectus and the globe and supplementary botulinum toxin injection of the left lateral rectus. (A and C from Shin GS, Demer JL, Rosenbaum AL: High resolution dynamic magnetic resonance imaging in complicated strabismus. J Pediatr Ophthalmol Strabismus 1996;33:282. Reprinted with permission of Slack, Inc.)

References


SELECTED STRABISMUS SYNDROMES
The focus in this chapter is on the management aspects of Duane syndrome (DS).

This condition occurs in approximately 1 in 50 patients with strabismus. My purpose is to simplify the diagnosis and treatment of this syndrome, based on the following concepts and principles:

1. The anatomic/histologic pathology shows that during the narrow window of 4 to 8 weeks of gestation there is maldevelopment or injury to developing structures (absent or partial development) of the sixth nerve nucleus and nerve(s). Branches from the third nerve are then redirected to the lateral rectus (LR). Thus, the LR may exhibit a wide spectrum of mixed anomalous (third nerve) and (sub)normal (sixth nerve) innervation. No credible evidence exists that the sixth nerve branches are redirected to innervate any third nerve muscles, and thus there is no relevant anomalous innervation of these muscles.

2. Rarely, the medial rectus (MR) may have subnormally developed innervation, presumably owing to its usual redundant complement of developing nerve fibers being redirected to the LR. This may result in a paucity of MR innervation, with noninnervated areas of MR becoming fibrotic.

3. Other development anomalies and syndromes developing at the same time may show DS-like patterns, syndromes including actual DS with so-called cluster anomalies.

4. Electromyography (EMG) clarified the nature of the LR anomalous innervation, but misinterpretations and artifacts of EMGs have led to unnecessary confusion concerning alleged anomalous innervations of the third nerve muscles. EMG of muscles taking up the slack created have been misinterpreted as misdirected anomalous innervation of some third nerve muscles. A detailed analysis of EMGs in DS is beyond the scope of this presentation.

5. There are secondary muscle changes due to ordinary (non-DS) strabismus factors, which may be due to (a) long-standing eye position; (b) excess innervation and overactions; and (c) relative globe immobility or lack of muscle contraction. Most common are secondary superior rectus (SR) contracture as a result of severe upshoots and secondary MR contracture that may result from increasingly large-degree esotropic DS. Or, the MR may be secondarily stretched out (elongated) in large-degree exotropic DS.

6. Relative immobility of the DS eye (in unilateral DS with fusion, and in many cases of bilateral DS with fusion) may manifest a progressive, gradually diminishing rotatory capability of the shortened/stiff MR. Such secondary muscle changes should not be confused with anomalous innervation in these muscles, which does not occur.

7. All of the DS patterns are fundamentally due to anomalous innervation/mechanics of the LR (mixed with nil or some [sub]normal LR innervation), with secondary muscle changes sometimes superimposed. Subnormal MR innervation (but not anomalous innervation) may rarely occur. This enormously simplifies the diagnosis and treatment of DS (Fig. 24–1).

**Terminology**

A horizontal gaze innervation is defined here as the right eye fixing in right gaze and the left eye fixing in left gaze (cross fixation is not included in this definition). The terms same-side gaze, and opposite gaze are used instead of ipsilateral and contralateral gazes. The term duction applies to monocular movement of the fixing eye, or the eye being examined by forceps force tests. The term positive forced duction must be clinically quantified (e.g., –1 to –4 denotes 25% decrements of rotation). The term abduction is an active innervational function of abduction, to be clearly differentiated from an eye position of exodeviation.
Anomalous muscle innervation is that which is received from a cranial nerve that does not normally supply that specific muscle.

**Clinical Examination of Duane Syndrome Patterns**

The diagnosis of DS is established by the examiner being aware of, and recognizing, the various clinical DS patterns, and knowing that all DS patterns are caused by LR anomalous innervation and mechanics in different gazes, with varying degrees of severity, with secondary muscle changes. Muscles other than the LR are not anomalously innervated and are not anomalously co-contracting. Rarely, the MR may have deficient innervation. Thus, DS pattern recognition is essential to knowing where and how to look for the LR anomalies. The examiner must first determine (using ordinary strabismus examination procedures with modifications) if, where, and how the anomalous LR innervation/mechanics acts; and further, what, if any, secondary muscle changes may have occurred that add to the DS patterns, such as secondary shortening (contracture), or elongation, or loss of contractility.

Globe retraction per se is not present in all DS patterns; it is replaced by what I call retraction escapes or equivalents. Examples are (1) the knife-edge LR/globe slip with an upshoot (or downshoot); (2) deficient inward rotation (in opposite gaze); and (3) some exotropic DS with the "splits." All of these may act as retraction escapes, substitutes, or equivalents.75

**DUANE SYNDROME PATTERNS**

Most DS patterns have anomalous LR recruitment in upgaze and/or downgaze, anomalous head positions, and retraction or equivalents.

**Unilateral Patterns**

Unilateral patterns include the following:

1. **Classic eso-DS**, anomalous LR to opposite gaze. There is no normal LR function to same-side gaze.
2. Similar to **classic eso-DS** but with both anomalous LR and normal LR activity.
3. **Eso-DS** with significant upshoot (or downshoot) in opposite gaze.
4. **Relatively immobile globe** in primary zone, marked retraction, and upshoots/downshoots.
5. **Exodeviation** with or without upshoots and downshoots, with retraction in moderate exodeviations.
6. **Simultaneous abduction** of each eye (the splits), usually with no retraction or equivalents.
7. **Fixation with DS eye** with marked overshoots/undershoots of fellow eye.
8. **Y (or λ) pattern**, normal or slightly deficient abduction. There are no retraction/equivalents (anomalous LR only in upgaze or downgaze). In pseudo-Brown syndrome there is a Y pattern but leash restriction in up and in rotation.
9. **Subnormal LR**—without anomalous LR. Horizontal rotations are limited with no retraction/equivalents.
Bilateral Patterns

1. **Bilateral DS with fusion.** There are usually (not always) limited eye movements and/or relatively immobile globes, often without retraction or equivalents.

2. **Bilateral DS without fusion.** There are usually (a) marked esodeviations (including occult DS with infantile esodeviation) and (b) marked exodeviations (including splits).

3. **Bilateral Y (or λ) pattern.** The LR activity in all patterns may be as in classic eso-DS or similar to classic eso-DS or combinations as described in Figure 24–1.

A qualification is that, in DS, the anomalous LR in upgaze (and much less frequently in downgaze) is perhaps the most common of all DS patterns. Secondary strabismus-related muscle length changes may be superimposed upon the just-described patterns. Of course, the most common are the stretched-out MR in constant esodeviation and the SR contracture in long-standing severe upshoots. These are easily identifiable.

**EXAMINATION SEQUENCE**

It is assumed that all the usual strabismus diagnosis and management procedures are in use. I shall focus on modifications of the usual, or on different test procedures applicable to DS diagnosis.

**History, Refractive Error, and Amblyopia.** Hyperopia is common in DS, and optical correction of hyperopia may so reduce the deviation in an eso-DS with head turn that the turn may be satisfactorily ameliorated. Anisometropic amblyopia may or may not be more common in DS.\(^{89, 187}\)

**General Ocular Inspection.** One should be alert to the developmental cluster syndromes (Möbius syndrome, general fibrosis, gaze palsy, Goldenhar syndrome, and others) that have a similar developmental time course.\(^{117-119}\) These may merge one into the other or occur in various combinations, so that when one makes a diagnosis of DS, one should be on the lookout for other associated developmental anomalies, and vice versa. These are adequately discussed in the literature, and further discussion is beyond the scope of this presentation except to emphasize the importance of an adequate neurologic examination, including hearing tests for deafness which occurs in 10% of DS cases.\(^{89, 163b}\)

**Fusion.** Quantitative traditional cover-uncover tests to determine the presence or absence of bifoveal fusion are done in whatever anomalous head position the patient habitually prefers. Or, the patient is simply asked to assume the position that he or she believes is best to determine whether fusion is present in some unusual position.\(^{82}\)

Compensatory head positions are frequent, and intended to (1) obtain fusion or (2) attain or maintain fixation when there are imbalanced forces in the primary zone. Such fixation restrictions are governing.\(^{69}\)

Infants may mask DS by choosing not to look toward the affected side or by settling for less than a completely compensatory turn. Additionally, the infant’s near world and the examiner’s arm’s length examination, in themselves, compensate for some esodeviations that may exist for more distant points and further mask the DS. The moderate and severe anomalous LR innervation and early mechanical muscle fibrosis may not produce early manifestations of DS patterns, which only appear with the progressive development of more severe mechanical changes.\(^{22}\) Adults may mask the DS by a hard-to-notice prolonged blink accompanying head movements. This may also occur in apraxic patients. Kennedy\(^{83}\) has done an excellent study of abnormal head posture in DS.

DS patients usually have normal binocularity in a preferred fusing position despite the incomitance. In this regard they are similar to non-DS patients fusing in one field of gaze but not in another, with the anticipated sensory anomalies in the nonfusing positions.\(^{5}\)

**Deviation-Quantitative Cover Tests (with Prisms).** These tests are performed in both the habitual head position just described and in the forced straight-ahead primary position. The often-described mismatch between the fusion status and deviation in the primary position, compared with the habitual compensatory turn, frequently occurs because there is a poor abducting saccade of the DS eye during cover testing, making it less accurate. A simple auxiliary measure in the forced primary position is to force fixation with the DS eye with the prisms in front of it and with the fellow normal eye observable under partial cover. During changes of prism the examiner moves the head from right to left to maintain fixation and to easily observe the overshoots, out and in, of the fellow eye until there is better prism neutralization. This fixation duress exponentially drives the fellow covered eye in or out, until its straight-ahead position is obtained with the appropriate prism before the forced fixing DS eye. This procedure eliminates the need to attempt assessing an endpoint with a poorer abducting saccade.\(^{75}\)

My colleagues and I have shown that the deviation in eso-DS is usually less than 30 PD.\(^{197}\) However, in bilateral nonfusing DS, or in unilateral DS fixing with the DS eye, there may be very large degrees of esodeviation. Elsas\(^{86}\) has emphasized the occult DS that may be embedded in a true infantile esotrope with large esodeviations.

Deviation measurements may be markedly different for (1) distance versus near, (2) upgaze versus downgaze, and (3) right versus left gaze fields. This is especially evident in patients with bilateral DS who are fusing with an A pattern. All of these differences affect management strategies.

**Ocular Rotations.** Ocular rotations, of course, form the cornerstone of the diagnosis and differential diagnosis of DS. The examiner seeks to determine what are anomalous LR neuromechanical actions (misbehavior); whether there are any normal or subnormal LR actions in the horizontal plane; and what, if any, secondary strabismus muscle shortening/lengthening exists. The examiner must control gaze innervations through the horizontal primary positions, as well as horizontal rotations in upgaze and in downgaze (Fig. 24–2). It is of utmost importance in the eso-DS to determine if there is any abduction in the horizontal plane beyond -4, as in Figure 24–1E and F, and especially to note the amount of outward rotation in upgaze and downgaze, which is due to the LR recruiting (increasing motor unit activity) in upgaze and/or downgaze. Figure 24–2A also depicts the corresponding matching of adduction deficiencies in horizontal rotations not only across and through primary gazes but also horizontal rotation during upgaze and during downgaze. The examiner should note the deviation in extreme opposite gaze, because whether there is a slight esodeviation or esodeviation...
tion is important in the differential diagnosis of mild cases of eso-DS.\textsuperscript{76} It is very important to note whether there is any (sub)normal LR recruitment, as in Figure 24–1E and F (see later discussion of eso-DS). Figure 24–2C depicts anomalous LR recruitment only in upgaze, from which the Y pattern is derived (or the \( \lambda \) pattern in downgaze).\textsuperscript{75} Because patterns merge one into the other, it is important to realize that the almost invariably anomalous vertical gaze LR recruitment occurs as a part of the horizontal opposite gaze anomaly and that the pattern in Figure 24–2C is only a pure, more severe extension of the pattern seen in Figure 24–2A.

Figure 24–2D depicts no anomalous LR recruitment, only a (sub)normal LR (limited abduction) and a restrictive adduction limitation derived from the fibrotic/shortened LR (see pattern discussion).

Globe retraction may be absent in many DS patients, and one should search for that horizontal plane of rotations (usually in some downgaze) where retraction is maximized, because it is not always in the expected horizontal primary rotations (Fig. 24–3, column A, bottom).\textsuperscript{75}

Physiologic globe retraction frequently occurs in perfectly normal individuals in extreme down and out rotation, owing to the long wraparound arc of contact of the SR in that position.\textsuperscript{199} Acquired pseudo DS patterns may follow trauma, surgery, or a painful eye due to myositis, or metastatic cancer, but these patients do not acquire the LR anomalous innervations, nor do they exhibit the other important characteristics of DS patterns.\textsuperscript{35, 123, 123a, 133, 134, 167, 168, 186b}

Lid fissure narrowing in opposite gaze accompanies globe retraction. Duane\textsuperscript{34} pointed out the lower lid ascent as well as upper lid descent, an observation also made by Isenberg and Urist.\textsuperscript{36} Lid fissure narrowing is one of the least dependable diagnostic signs in mild or moderate DS. A convenient observation of globe retraction during left to right rotations

\textbf{Figures 24–2.} A through D. Upper row depicts various combinations of lateral rectus (LR) anomalous actions (A), combined with (sub)normal LR recruitment in B and (sub)normal LR only in D. Lower row shows the derived clinical patterns. (See text for explanation.)

\textbf{Figure 24–3.} Duane syndrome in left eye, with “limited in and out eye movements” (almost approaching the relative immobility stage). Upper picture in C shows fusion with straight head, and all of column D shows left gazes actively innervated (left eye abduction), which is characteristically more in either or both up and out, and down and out gazes than in primary horizontal outward gaze. This curved outward rotation pattern is uniquely characteristic of DS. It is owing to anomalous LR innervation in up and down gazes, not (sub)normal LR innervation of abduction as might be erroneously inferred. This is also depicted in Figures 24–1D and 24–2A. Column B shows the knife-edge upshoot and downshoot, which occur suddenly at either side of the gaze of maximum retraction (second from bottom). Bottom figure in column A also shows this position of maximum retraction in a somewhat down and out right gaze.
may be made by manually spreading the lids apart and observing the globe retraction from the side or from below. Normally there is a narrowing of the fissure in opposite gaze and a wider fissure in abduction with a peak elevation of the upper lid at the inner one-third junction.166 Every Smith-Kettlewell—trained fellow has been subjected to planned discomfort in not being fully aware of this normal variation. Similarly, if one suspects multiple sclerosis, the normal temporal disc pallor may perceptually become a bit more pale.

**Upshoots and Downshoots.** Upshoots and downshoots in opposite gaze occur with greater frequency in increasingly severe anomalous LR recruitment (see Fig. 24–3). Duane24 explained it this way: "The left eye shoots obliquely upward, particularly if the eye is elevated slightly above the horizontal plane." The most eloquent description of this unique upshoot and/or downshoot was made by Souza-Dias,172 followed by his unique discussion of the "bridle effect." At the same meeting, Scott159 also described the sliding of the shortened anomalously innervated LR over the globe in this condition.

The difference between the sudden (anomalous LR) versus gradual (non-LR) upshoot/downshoot rotations in opposite gaze has been well described by Khawam, Warrak, and Shahine.86 Romero-Apiis145,146 described them as "shoots and hooks," and Kraft171 has also addressed these differences. The authors discussed the gradual upshoot, which may be due to inferior oblique overaction or SR contracture. We have previously discussed secondary SR contracture in prolonged upshoots, with hypertropia and characteristics of the SR contracture syndrome.

**The Y Pattern (and the Less Frequent Lambda Pattern).** These patterns were so named in my 1988 Jules Stein Eye Institute lecture on DS73 to distinguish them from the long recognized V pattern. The Y pattern is depicted only from primary gaze to upgaze, owing to LR anomalous innervation in vertical gaze (see Fig. 24–2C). In unilateral cases the DS eye should be observed under cover (to dissipate fusion) during upgaze to avoid a sudden Y-pattern flick-out at the point of fusion loss in greater upgaze.

A pseudo-Brown syndrome is closely allied to the Y pattern.77 Others have reported Brown syndrome in DS, but, in my experience, the forceps force tests, which differentiate between restricting leash and the reverse leash of a true Brown syndrome, clearly make the distinction.66,135,136 The forced traction test alone, in both the pseudo-Brown and true Brown syndromes, shows restriction to up and in rotation. Scott187 has shown that, in true Brown syndrome (a superior oblique [SO] anomaly), the continued upward rotation forceps force reveals a reverse leash with more restriction to upward rotation while the globe is being pushed back into the orbit.

**Falling or Rising Eye Movement.** A falling or rising eye movement of the nonfixing abducted eye may occur consequent to the DS eye fixing with vertical fixation duress in adduction fixation.69 In non-DS, the fixation duress may be due to any vertical force imbalance during adduction, such as oblique muscle imbalance in this adduction fixation, whether it is part of a superior oblique palsy with fixation of the involved eye (described by Bielschowsky) or part of a common non-DS esodeviation oblique imbalance. DS cases with adduction fixation of a DS eye, in which a rising or falling nonfixing abducted eye may occur as a result of the DS eye’s LR restraining forces, result from nonlinear or irregular (fibrotic and innervational) forces in the anomalous co-contracting LR when this eye is made to fix in adduction. This may occur especially in bilateral nonfusing DS.

**Fixation with a Duane Syndrome Eye.** Fixation with a DS eye must be identified, because it may occur in unilateral DS with amblyopia of the fellow eye and in some nonfusing bilateral cases. This may produce not only a rising or a falling eye but sometimes wildly bizarre overshoots and undershoots of the fellow eye from fixation duress in all positions. Especially in these conditions, abnormal and normal forces must be determined to avoid unpredictable surgical results.

**Exotropic Duane Syndrome.** Exotropic DS, although more common in adults, may also occur in children. This is an example of how a pattern itself may change with progression from esodeviation to exodeviation. The LR innervations (normal and/or abnormal) remain the same, but there are increasing mechanical LR shortening changes. The patient usually has a compensatory head turn to obtain fusion.

With extremely exotropic DS, and in the splits, the now shortened inelastic LR diminishes the possibility of its wrap around the globe against the opposing MR, so that a previously existing upshoot/downshoot in opposite gaze may disappear with exotropic DS.

The examiner must carefully distinguish between an exotropic eye position and a true, actively innervated LR abduction function (see Fig. 24–1E and F) and must not assume the active abduction function from the eye position (see forceps force tests later).

**Simultaneous Abduction.** Simultaneous abduction (the splits) is easily recognized by simple observation of this dramatic display of the DS eye turning outward as the normal fixing eye fixes in controlled opposite gaze.77 When the LR has both normal and anomalous recruitments (recruits in both directions), the more severe splits demonstrate eyes locked into the external canthi (see section on management of the splits).

**Bilateral Duane Syndrome with Fusion.** Bilateral DS with fusion often displays an A (or λ) pattern and/or an acquired progressive heterophoria owing to changing imbalance in the bifixation process.84 The near deviation may be significantly different from the distance deviation, depending on the sometimes progressively diminishing MR rotatory capability.

**FORCEPS FORCE TESTING**

Modifications of the forced traction, force generation, forced augmentation, and force degeneration tests provide necessary information on what muscle functions are operating and where and when they act and identify both restrictive and innervational anomalies (normal and anomalous LR recruitments). Forceps force testing is selectively indicated in alert patients and sometimes is performed under anesthesia (including, as a first step, under surgical anesthesia) but, more importantly, whenever possible, is done under topical surgical anesthesia (described later).185 I have discussed elsewhere the use of the cotton-tipped applicator stick for pressure anesthesia and of forceps without teeth but with clamp action tips.82

*The forced traction of the LR in opposite gaze measures...*
both the anomalous innervation as well as the mechanical restricting forces. The distinction between the two cannot be made with the simple forced duction (traction) test alone (see force degeneration test later).

The **forced augmentation test** that I have described is a way to test the voluntary abduction limit to the forceps forced augmentation (possibly extension) of this limit. The ability to demonstrate forceps-improved outward globe rotation during the same-side directed gaze simultaneously reveals the presence of paralysis of the agonist muscle and the forceps-determined antagonist restriction that quantifies the true limit of outward globe rotation. This may easily be done in most alert adults and many children, and in all infants under surgical anesthesia (with muscle relaxants). The forced augmentation test eliminates the false assumption that the MR in the usual childhood DS has abnormal contracture force in the primary position (see discussion of eso-DS).

The **force generation test,** first suggested by Scott, is a sensitive method for gauging active innervation LR abduction force, regardless of eye position (esotropia, orthotropia, or exotropia). The information obtained from this test is of special importance in diagnosing the DS pattern. In some cases such as bilateral DS with fusion, extreme bilateral exotropia, or the splits, it is not always easy to have the patient place the eyes where there is sufficient room to feel generated force. However, it remains essential to perform the test when rotational observations alone do not suffice. A subnormally innervated MR may exist rarely in some exo-DS and in simultaneous abduction. An actively innervated abducting LR force (normal or subnormal) cannot be assumed from an exotropic position of the eye. These are not the same. To determine that there is an actively innervated abducting force, the eye must either be observed or felt during forceps force generation to actively abduct from its position, whether esotropic, orthotropic, or exotropic.

The **force degeneration test,** described by Romero-Apis, is an innovative way of using forceps to demonstrate anomalous LR innervation as a DS eye goes into opposite gaze. It is most useful in selected cases in which differential diagnoses need to be clarified. The details of this test are summarized in Figure 24–4.

**Surgical forceps tests** of forced duction under general anesthesia (with muscle relaxants) allow evaluation of pure muscle mechanics. They should be performed in all cases regardless of whether testing has been done in the alert state. The spring-back tests (described elsewhere) reveal the state of balance or imbalance of mechanical forces, which is of special relevance in DS.

**Surgical forceps tests under topical surgical anesthesia** (alert patient) provide valuable diagnostic and predictive information by directly observing the ocular rotations after detachment of selected muscles. Active force innervation and rotation ability can be determined by directing the patient to gaze in certain directions. In essence, these maneuvers serve as a therapeutic trial for predicting the results. It is in the most difficult DS management problems that such tests decisively suggest the appropriate surgical management (Fig. 24–5).

Under topical anesthesia, patients with a knife-edge sign exhibit a thin blue sclera under the LR muscle. The location of the blueness confirms the direction of LR pressure indentation in a particular DS pattern. Finding this sign is especially reassuring in a patient with pseudo-Brown syndrome.

**Treatment**

<table>
<thead>
<tr>
<th>Table 24–1. Diagnostic Factors Necessary to Manage Duane Syndrome Patterns</th>
</tr>
</thead>
<tbody>
<tr>
<td>I. Diagnosis</td>
</tr>
<tr>
<td>A. DS pattern recognition; severity of dominant features.</td>
</tr>
<tr>
<td>B. LR anomalous innervation/mechanics (severity), in gazes opposite, same side, up/down. Unilateral or bilateral.</td>
</tr>
<tr>
<td>C. Any normal LR active innervation, in same-side gaze.</td>
</tr>
<tr>
<td>D. Secondary muscle changes.</td>
</tr>
<tr>
<td>E. Is there fixation with the Duane eye(s)?</td>
</tr>
<tr>
<td>F. Is fusion present, and where? Compensatory head turn?</td>
</tr>
<tr>
<td>G. Are there cluster neurologic anomalies, including impaired hearing?</td>
</tr>
<tr>
<td>H. The strabismus deviation in the primary position.</td>
</tr>
<tr>
<td>II. Treatment</td>
</tr>
<tr>
<td>A. Indications and relative contraindications (surgical; nonsurgical).</td>
</tr>
<tr>
<td>B. What is the goal, result predictions (risk/benefit)? Will there be ameliorization of compensatory head position and bizarre eye movements?</td>
</tr>
<tr>
<td>C. What is the surgical strategic plan?</td>
</tr>
<tr>
<td>1. Must one perform surgery on the abnormal LR, and if so what weakening/manipulative surgical options are contemplated?</td>
</tr>
<tr>
<td>2. What other secondary muscle changes require surgery?</td>
</tr>
<tr>
<td>3. What if any surgical options exist for uninvolved muscles in the normal eye or in transposition (superior rectus/inferior rectus) of the involved eye?</td>
</tr>
<tr>
<td>4. What intraoperative diagnostic/treatment tests are anticipated?</td>
</tr>
<tr>
<td>D. Neurologic and hearing examination clearance?</td>
</tr>
</tbody>
</table>

The outline presented in Table 24–1 summarizes the special diagnostic factors that one needs to know, and the thought sequence applicable to managing DS patterns. Of course, these are in addition to the usual strabismus management factors.

**SURGICAL INDICATIONS AND CONTRAINDICATIONS**

In any strabismus disorder, surgery is indicated to (1) obtain and/or maintain balanced alignment in the primary position (i.e., the deviation); (2) obtain as comitant rotations as are possible; and (3) accomplish good cosmesis of the lids and conjunctiva, all within a reasonable framework of the risk/benefit ratio. The word “balanced” implies minimizing restrictions in the primary zone, because this governs a head turn, regardless of whether the balanced/imbalanced eye fixates alone or as part of binocular fusion. The goal of centering the fusion field, thereby eliminating the compensatory head position, is included in the traditional goal alignment statement by implication but has special relevance (and special problems) in DS. Obtaining “as comitant rotations as are possible” is almost always a significant compromise in DS, in which the problems of bizarre eye movements are significant. The risk/benefit ratio in DS is considerably different than in usual strabismus surgeries. In fusing bilateral DS cases there is ample oppor-
Figure 24-4. The force-degeneration test is an innovative new forceps force test suggested by Romero-Apis, to demonstrate lateral rectus (LR) anomalous innervation in opposite gaze in selected cases in which differential diagnosis needs clarification. Romero-Apis suggests the following, "To demonstrate co-contraction just with the forced duction test in those cases of substitution (no VIth nerve in lateral rectus muscle). (A) We ask the patient to look halfway between the primary position and adduction. Then, we try to move the eye into more adduction, and we feel positiveness because some restriction is originated by the lateral rectus co-contraction. (B) Again, we ask the patient to look halfway between the primary position and adduction, then we hold the eye with forceps, but now, we ask the patient to look outwards, and in the meanwhile, we move the eye inwards, and we feel negativity (release of force), because the lateral rectus has been inhibited by its IIIrd nerve abnormal innervation." In other words, the forceps hold the eye in an opposite gaze position, not so far as to feel restriction but leaving some room for further forceps addition. The patient is then instructed to change gaze toward the involved side (attempted abduction). The examiner, at this moment, feels a release of the anomalous LR co-contraction and can now rotate the eye still further inward. I would suggest that one might term this test the Romero-Apis force-degeneration test. Before this force-degeneration test is done, one has determined that there is no LR normal or (sub)normal active abduction innervation. The test determines the presence of anomalous LR innervation in opposite gaze.

In DS management there are many relative contraindications to what are ordinarily traditional surgical procedures. For instance, in eso-DS, a simple modest recession of the MR in the presence of a severely misbehaving LR may produce a very significant overcorrection, the risk being compounded if there is any significant normal active LR abduction. A transposition of the SR and inferior rectus IR temporalward is contraindicated if there is severely anomalous LR function. A recession of the good eye’s LR to augment an exodeviation correction in unilateral exo-DS may be contraindicated if there is significant residual LR anomalous misbehavior in the involved DS eye. A simple, modest recession of the MR muscles in bilateral DS cases with fusion may immediately produce large degrees of exodeviation unless one has determined the degree of anomalous and normal LR innervation in each eye. These and other relative contraindications are discussed later.

MANAGEMENT OF SPECIFIC DUANE SYNDROME PATTERNS

In general, classification systems serve different purposes, that is, for logical identification of causes or for treatment grouping. A management-based DS classification should include all DS patterns and thus must address not only the usual somewhat linked progressive severity of the horizontal gaze DS patterns (from eso-DS to exo-DS) but also such widely diverse and really separate management entities as bilateral DS with fusion and without fusion (vastly different problems in themselves), Y pattern, the splits, and fixing with the DS eye. No single classification system has been found to satisfactorily cover the management of the diversity of DS patterns, and no such overriding classification system is suggested here. Attempts to classify DS patterns according to any one factor, such as the horizontal gaze LR anomalous innervations, or whether the deviation is eso, ortho, or exo, do not adequately meet the challenge. Mild or severe DS patterns may exist in childhood or become progressively severe and change over time owing to the changing LR mechanics (shortening) and secondary muscle involvements, although the LR innervation anomalies remain constant. The importance of determining LR innervation causes (both anomalous and [sub]normal) and how to apply this information in managing the wide panorama of horizontal and vertical components of DS patterns have been emphasized. For practical management purposes, it is best to discuss each of the wide variety of specific DS patterns separately based on the aforementioned essential factors and questions relating to diagnosis and treatment (Table 24-1).
Unilateral Duane Syndrome

Classic Unilateral Eso-DS. Anomalous LR recruitment occurs in opposite gaze. There is no normal LR recruitment. Some of the ever-present LR anomalous recruitment occurs in upgaze and/or downgaze and produces the very characteristic DS pattern depicted in Figures 24–2A, 24–6, and especially 24–3(column D). There is mild inward-rotation limitation in opposite gaze (due to a restraining co-contracting LR, which produces a slight but important to note exodeviation in far opposite gaze). Esodeviation in the primary position does not usually exceed approximately 30 PD. with a head turn left to obtain fusion in some degree of opposite gaze. (All unilateral examples in this presentation are of left-eye DS.)

Differentiating DS from LR paralysis and esotropia is a matter of observing abducting saccades. Slow-abducting saccades are seen in most DS and in LR palsy, which indicate absent LR recruitment (which differs from esotropia). The abduction characteristic (in upgaze to downgaze) of LR palsy is a V pattern, and in esotropia, a more or less straight vertical line, compared with the curved outward rotation pattern uniquely characteristic of DS, seen in Figure 24–3(column D). The title of a Souza-Dias publication stated: "Congenital VIth nerve is Duane's syndrome until disproven" and it also reflects the rarity of congenital sixth nerve paresis.

The compensatory head turn (to the involved side) is a main feature and an indication for surgery. A large compensatory turn for fusion may denote a mild DS, and, conversely, a small head turn paradoxically usually denotes a more severe DS pattern. This is because the incomitant esodeviation gradually diminishes toward opposite gaze owing to the increasing effect of restraint by the co-contracting LR in that gaze direction, where sooner or later the restraining LR forces diminish the esodeviation so that parallelism and fusion occur. Of course, the relevant management factors in all compensatory turns in DS are the amount of the deviation, the severity of the LR anomalous co-contraction, and imbalanced restrictive forces in the primary po-

Figure 24–5. Same patient as in Figure 24–3 photographed under topical anesthesia. Upper right photo is the same as Figure 24–3, column A, lower photo, that is, the position where horizontal rotations display maximum retraction in opposite gaze. Top photographs were taken preoperatively at the time of topical anesthesia. Bottom photographs were taken during surgery after the lateral rectus muscle has been completely detached from the globe (with severance of intermuscular membranes far back). The two lower photographs (with the lateral rectus detached) replicate the gazes in the two upper preoperative photos. There is almost complete disappearance of the maximum retraction in down and right gaze (bottom right) and almost complete disappearance of the marked upshoot in up and right gaze (bottom left). Thus, there is no question that the anomalous innervation/mechanics of the LR was the cause of the retraction and upshoot, despite any EMG recordings for other explanations (see text).

Figure 24–6. Classic Duane syndrome of left eye with complete absence of any normal lateral rectus (LR) innervation in horizontal gaze left. LR anomalous innervation in upgaze (and here depicted as downgaze also) allows the eye to rotate outward more in these vertical gazes (to different degrees of –1, –2, –3, depending on the upgaze-innervated LR), which makes it appear as if there is active abduction in these vertical gazes. One must distinguish between gaze left innervations and up/downgaze innervations. This clinically produces the picture of matching adduction deficiencies in different horizontal opposite gazes, as depicted in Figure 24–2A. The anomalous LR innervation may be only in upgaze, or only in downgaze, or both.
sition. It does not take much head turn in severe LR co-contraction to neutralize considerable esodeviation. Or, looking at it another way, a compensatory head turn position seeks a better balance between the opposing MR and LR muscles of the DS eye.

**Treatment of unilateral mild/moderate classic eso-DS** (only anomalous LR), of course, starts with the consideration of indications, goals, and result predictions. The most important strategic surgical decision is whether one must perform surgery on the abnormal LR because of the severity of the anomalous innervation and mechanics, and, if so, what weakening/manipulative procedure is contemplated? It is never necessary to resect the LR in DS patterns, because this will only make the anomalous LR worse. In this mild/moderate eso-DS, there is no need to operate at all on the LR. Let us first consider why only an MR recession of the DS eye is not a first surgical preference. As mentioned earlier, the MR in children with DS does not exhibit excessive stiffness or contracture in the primary zone, that is, it is normal and is not usually restricted until −2 forces abduction. A single MR recession of the DS eye with −4 voluntary abduction does not improve abduction, and it can only decrease inward rotation in opposite gaze; the latter sometimes decreases exponentially in the more moderate/severe LR anomalous pattern. The amount of esodeviation corrected is often insufficient for complete head turn correction (although it is ameliorated), because it substantially depends on the degree of severity of the LR anomalous co-contraction, as described in Figure 24–7.

Two surgical options are recommended: (1) asymmetric MR recessions and (2) transposition of SR/IR temporalward.

**Asymmetric Medial Rectus Recessions.** Asymmetric medial rectus recessions intentionally produce a fixation duress in the good fellow right eye (10-mm MR recession) for previously stated goals, namely, to “(1) considerably enhance the amount of esocorrection in the primary position and (2) diminish the probability of a left MR recurrent contracture via its continual inhibition through fixation duress of the right eye, even in the primary position as well as in attempted left gaze, all by virtue of the large right MR recession. One need not fear any significant long-term deficient adduction of the fixing right eye.” “Abduction will not increase, nor will an exodeviation occur in horizontal left gaze, because the LR is ‘dead’ in that direction (selectively −4 voluntary abduction). Indeed, there is no reason not to do a right MR recession (of the good eye) of considerable degree in this type of DS.” The transient −1 or −2 adduction of the good eye after such a large recession diminishes in time to a barely perceptible deficiency as the muscle takes up its slack, it being in the fixing eye. Fixation duress was first demonstrated electromyographically by altering the tonus of the medial rectus muscle in the fixing eye by procaine injection. At that time (1970), we stated that “it may be generally stated that tonic alterations which primarily affect one muscle have some influence on all muscles.” Apparently there was an increased effort by the medial rectus to maintain fixation after the procaine block, which resulted in a reciprocal inhibition of the antagonist lateral rectus muscle. It is apparent that when a muscle in the fixing eye alters its tonic or relative mechanical contribution to fixation, the other muscles are affected, and if it is the fixing eye that dictates the innervational pattern of both eyes, there are far-reaching consequences in the tonus of both eyes. It is of clinical interest that surgical alteration of one muscle in the fixing or dominant eye has more far-reaching consequences relative to the basic deviation of both eyes (through alteration of the tonic in flow to both eyes) than does a similar procedure performed on the nondominant, nonfixing eye. I first advocated creating such a DS fixation duress in 1971 as follows, “A recession of the medial rectus of the fixing eye will cause that eye to try harder to fix in the primary position.” And, in 1972, “If one does the same procedure on the fixing eye (MR), then in order to fix with that eye, it will need more adduction juice.” Smith-Kettlewell–trained fellows have enjoyed the benefits of this option in mild/moderate DS for two or three decades, and although I have published this in several more publications, from 1972 to 1996, its general acceptance is just emerging as a viable option.

There has not been a full understanding of the nature of the fixation duress created by operating on the good eye’s MR. Some have advocated substituting a posterior fixation suture on the normal eye’s MR. This certainly does not have the same physiologic effect as a large MR recession. The
effect of a posterior fixation suture is not obtained until the fixing good eye is rotated inward a considerable amount, when suddenly the posterior fixation suture takes effect. However, it is not effective in the primary zone at all so far as creating any fixation duress is concerned or correcting any esodeviation. I do not recommend the posterior fixation suture as a substitute for a large (10-mm) MR recession of the good eye in these cases.

With MR surgery (either unilateral or asymmetric bilateral), the DS eye may exhibit a small amount of exodeviation (5–10 PD) in left gaze, which may be misconstrued as “apparent abduction.” But this small amount of exodeviation is due to residual LR tone (even anomalous) in the primary zone; the slight expansion of rotation and the binocular field may be a small unanticipated favorable dividend when it does appear, although its significance should not be misinterpreted.

Transposition of the SR/IR Temporalward. Transposition of the SR/IR to the LR by a variety of techniques (including adjustable sutures) has been thoroughly discussed by Rosenbaum49, 116, 122 and is a very viable option in this specific instance. Foster39 has published a transposition technique that is combined with posterior fixation sutures of the vertical muscles and LR, augmenting the transposition procedure. It is especially effective in mild/moderate LR anomalies in eso-DS.

Transposition of the SR and IR temporalward to create a passive exo-force (rubber band) is a viable option when there is not a severe LR anomaly. One need not fear that, in transposing the SR and IR in DS, one is dealing with anomalously innervated vertical muscles. This unwarranted fear is engendered by misinterpretation of EMGs. In mild/moderate eso-DS the transposition is aided by the fact that the MR is not usually in contracture in the primary zone, as is often incorrectly thought, and thus differs from transposition in many cases of LR palsy. This important characterization of the near-normal MR in mild/moderate DS was the result of MR length-tension measurements made during surgery by Collins and myself.25, 36 Innovative modifications of transposing forces have been suggested by Spielmann,173, 175 and artificial muscle forces (magnetic) have been suggested by Bicas.10

If transposition of all of the SR and IR is contraindicated because of severe LR anomalous innervation, an alternative is conversion to a sixth nerve palsy (by eliminating the LR function) combined with an adjustable partial (temporal halves) muscle transposition of the SR and IR, which Carlson and I3 proposed. We addressed the risks of anterior segment necrosis, even in transposing the temporal one half of the vertical rectus muscles (preserving the ciliary vessels in the unoperated nasal one half), if the LR also is very markedly recessed, converting it into a sixth nerve paresis. We initially described using the operating microscope to identify anterior ciliary vessels with the aid of red-free light to avoid complications in the anterior segment circulation.25 This technique is suggested if further MR surgery is required.

Comparative outcomes of the different transposed force options and of asymmetric MR recessions require long-term follow-up to contrast the extent of abduction benefit, adduction deficit, and vertical deviation residuals.30, 39, 43

Moderate to severe anomalous LR in eso-DS (no active LR normal recruitment) clinically manifests more marked deficiency of inward rotation, more retraction, and very frequently marked up/downshoots in opposite gaze (to be discussed later). In considering severe LR misbehavior, to achieve the goals one usually decides that one must weaken the LR (with or without some manipulations). Because this increases the esodeviation and at the same time converts a severe LR misbehavior into a milder one, the just described asymmetric recessions of the MR muscles become part of this three-muscle recession procedure.174, 176, 192 The amounts of recession depend on the severity of the anomalous LR and the degree of esodeviation finally to be corrected, which is best accomplished by one or more adjustable sutures.

Eso-DS with Both Anomalous LR and (Sub)normal LR Activity. Adding any (normal or [sub]normal) active abducting LR force considerably changes the treatment strategy. Referring to Figures 24–1E and F, and 24–2B, one can see that there is no longer a −4 abduction in horizontal same-side gaze, so that creating a fixation duress in the good eye by a large MR recession may be contraindicated. This fixation duress may cause a large esodeviation in the DS eye via Hering’s law.

Of course, no surgery is indicated if there is a reasonable balance of forces without significant head turn. When the LR is both anomalous and normal, in varying degrees, there exist many combinations of the misbehaving and normal LR. Here only the eso-DS problems are discussed. There are two ends of the spectrum of relative forces of the misbehaving and normal LR: (1) a mild anomalous LR and a strong normal LR force and (2) a severely anomalous LR and a weak normal LR force. In both situations there is normal MR adduction force.

A decision with regard to the severity of anomalous LR innervation must first be made. A severe anomoly requires some form of LR weakening to decrease the DS retraction and/or equivalents, and, at the same time, of course, this diminishes any existing normal LR force. Weakening the LR converts this pattern into one somewhat resembling the classic eso-DS described earlier (with no normal LR activity), with the exception that now there may be some residual normal LR activity. The surgeon now has to address the problem of the increased esodeviation created by the LR recession. Depending on the residual LR activity and the amount of esodeviation created, MR recession is performed in the DS eye. To achieve sufficient deviation correction one may have to recess both MR muscles. However, the caveat in managing this type of patient is to avoid a large recession of the MR in the good eye that would create significant fixation duress in the presence of a substantial esodeviation-producing force (true active LR abduction, residual anomalous LR innervation, or transposition). The relative amounts of this bimedial recession depend on the residual (if any) LR normal force, the anticipated residual of the LR anomalous force (depending on the severity to start with and the amount that the LR was weakened), and the deviation.

If only mild anomalous LR innervation is present, with lack of prominence of retraction and/or equivalents, and there is a strong normal LR abducting force, the case may be treated substantially as an ordinary (non-DS) esodeviation. If the amount of esodeviation is minimal, one MR recession in the DS eye may suffice. If a greater esocorrection is required, recession of both MR muscles may be performed. The LR of the DS eye need never be resected.
In other words, the more the normal versus anomalous LR activities are imbalanced toward the good normal LR, the fewer the DS considerations. The more it resembles a regular esodeviation, such treatment is planned accordingly. The more the imbalance is tilted toward the existence of a severe misbehaving LR, which one must weaken, the more one must consider what one has done to a co-existing normal LR force and the deviation created (more eso) and fix that accordingly.

One must avoid a large fixation duress, created by a recession of the good eye’s MR in the presence of any significant residual active normal LR force. It is this caveat that is a principal distinction between the management of the classic eso-DS with no LR normal force and the varieties discussed here (with both abnormal and normal LR functions) with possible residual abducting forces.

In another instance, eso-DS with minimal anomalous LR and minimal (sub)normal LR recruitments, no treatment may be necessary. Or, if indicated, only an MR recession of the involved eye might be planned. Other instances of imbalanced normal and abnormal LR would respond equally well to only a significant MR recession of the good eye, but this may be contraindicated if there is significant normal LR activity because of the danger of Hering’s law-initiated exodeviation.

When there is both anomalous and normal LR recruitment of the rare type (see Fig. 24–1F), there are some special considerations. If surgery is indicated, one must consider that LR tone in the primary zone is less than it is in either horizontal gaze. When significantly so, depending on the relative severity of the two LR recruitments, it is wise to check the deviation in right-left gazes versus the primary zone to help elucidate the problem. The force generation tests in both directions measured from the primary position with straight-ahead fixation are helpful in clarifying the character of the imbalance. The esodeviation in the primary position is greater than in either right or left gaze, for entirely different reasons. The esodeviation decreases in opposite gaze owing to the anomalous LR misbehavior, as described earlier. On the other hand, the esodeviation decreases on same-side gaze, being diminished by the increasing normal LR recruitment that exists. Thus, the nuances of forceps force testing in different positions are helpful.

Eso-DS with Significant Upshoot (or Downshoot) in Opposite Gaze. LR innervation may be either anomalous or both anomalous and normal. The presence of upshoot and/or downshoot is discussed separately to simplify the different presentations of an esotropic DS. Upshoots and/or downshoots occur not only in esotropic DS but also in almost the entire panorama of DS patterns.13, 31, 91, 109, 114, 115, 143, 146, 164. There may be exceptions to this, such as in mild eso-DS, bilateral DS with fusion, some large-degree exotropic DS (including the splits), and the Y patterns.

Severe upshoots and/or downshoots are retraction equivalents or may be combined with some retraction. All the knife-edge types of upshoots/downshoots are due to the anomalous LR innervation and mechanics. It follows that amelioration of retraction and/or equivalents involve weakening the LR muscle. SR contracture (and rarely of the inferior oblique) develops only as a secondary muscle involvement (see Fig. 24–5). I stated in 1978 that “vertical rectus or oblique muscle surgery is fruitless” in the up/downshoot with a knife-edge LR/globe flip,66 and I find no reason now to alter that statement.

There are three surgical options: (1) recessions of both LR and MR; (2) LR bifurcation (the Y-split procedure) with recessions; and (3) LR posterior fixation suture. I prefer option 2.

Recessions of Both LR and MR of the DS Eye. For modest upshoot/downshoot and retraction, a modest but necessary recession of the anomalous LR must be counterbalanced by an MR recession.138, 171, 172 A recession of the good eye’s MR is done depending on the final degree of esodeviation to be corrected. Souza-Dias171, 172 advocated a very large recession of the LR and MR to the region of the equator and, if necessary, rebalancing this with a recession of the opposing MR. If LR recession is the chosen option, a very large recession of the anomalous LR is required with severe anomalous LR-produced defects, followed by rebalancing of other muscles as described.

For more modest LR-produced retraction and upshoots/downshoots, a more modest recession of both horizontal rectus muscles has been advocated by von Noorden192, 193 and Sprunger.170 Of course, there is a loss of eye mobility with large recessions, but, in time, muscle slack may be taken up and rotations may improve.

**LR Recession with Bifurcation (or Y split)** (Figs. 24–8 and 24–9). The LR muscle is split from its insertion as far posteriorly as possible, spreading the muscle halves 10 mm apart. The middle diagram shows the position of maximum retraction in horizontal gazes slightly down, with a bridle retraction action on the crest of the globe in this position. With only slightly more downgaze, there is a sudden downshoot. (A corresponding upshoot is shown in Figure 24–9.)
The bifurcation and recession procedure appears to have gained acceptance. For very severe cases, especially in large exodeviations, one has the option of the very large LR and MR recessions discussed earlier or converting the case into a sixth nerve paralysis by exterminating the severely misbehaving anomalous LR and then managing it as a sixth nerve paralysis. Completely eliminating the function of the LR by recession alone is not an easy task, no matter how far it is recessed. Scott has recommended sewing the LR to the orbital wall to eliminate its torque purchase on the globe, and others have suggested sequentially pulling more and more of the LR forward, removing bit by bit for maximal extirpation. This second suggestion is akin to what I once, in jest, suggested—using a miniature tonsil snare to extirpate as much of the LR as possible. This conversion of an extremely severe anomalous LR by attempts to totally eliminate the defective muscle, thus creating an LR palsy, is rarely indicated. This might be more acceptable to the reader if it is compared to the generally accepted treatment of severe jaw-winking, in which the surgeon incapacitates the misbehaving lid elevator muscle and substitutes a brow suspension.

**LR Posterior Fixation Suture.** LR posterior fixation suture was first advocated by Scott. This does not sufficiently ameliorate the defect, especially the residual retraction, because the misbehaving LR muscle is still on the crest of the globe.

I have previously discussed secondary contracture of the SR that may result from severe and persistent upshoots (or downshoots). If such a hypertropia exists in most fields of gaze and exhibits the SR overaction/contracture syndrome, an SR recession is indicated for its own sake but will not by itself affect the upshoot/downshoot because it is not the cause. In the examination section, I cited authors who differentiated between sudden and gradual upshoots. I here emphasize the DS characteristic knife-edge sudden flip-up associated with an anomalous LR. Melek has presented an excellent discussion of these rotational defects. In my experience the anomalous LR-caused upshoots are so materially improved by appropriate LR surgery that I have not found it necessary to follow this with a later inferior oblique weakening procedure.

**Relatively Immobile Globe.** This is usually accompanied by marked retraction, with or without upshoots (and less frequently downshoots), and anomalous LR innervation only or both anomalous and/or (sub)normal LR function. In unilateral cases there may not be just limited eye movements but practically no eye movements in either direction, although the globes are locked in fusion at or near the primary position. The head position may be straight or nearly so, depending on the balance/imbalance of the MR and LR in the affected eye. There is very marked retraction and often a severe upshoot with a not infrequent secondary SR contracture. Both the LR and MR muscles may be contracted/shortened and stiff. However, this assumption must be con-
firmed by forceps tests. I have hypothesized that the fusion-lock (in unilateral and bilateral DS with fusion) maintains the affected eye(s) in a stable, nonrotating situation. The isometric muscle contractures and stiffening are secondary to this, leading to stiff, short muscles that may lose contractility. This hypothesis is based on my muscle length-tension measures and electromyography, which at times shows MR recruitment but a lack of rotatory capability even after the anomalous LR is completely detached from the globe and observing the consequences of directed gazes. Remember that, with a relatively immobile globe and stiff agonist/antagonist muscles, the muscles may lose their contractility (as discussed earlier). The prediction is that usually postoperative eye movements will be unchanged from those present preoperatively, except that with time the MR may regain some contractility, with improved eye movements.

In one respect, the lack of diagnostic assurance that there is no normal LR recruitment is not so important as long as one plans to do a very large recession of the LR—which diminishes both LR anomalous and normal activity. Such large recessions of the LR and MR require adequate sharp dissection of the intermuscular membranes on either side of the muscles as far back as is safely convenient to avoid, as far as possible, any globe protrusion that may otherwise result from such procedures.

**Exo-DS.** Exotropic DS occurs with or without upshoots/downshoots and retraction in moderate exodeviations. The LR may be anomalous, or both anomalous and (sub)normal. More commonly, a unique feature of exotropic DS is that it has a severely anomalous LR innervation/mechanics (contracture). The LR has what I have termed a contracture propensity. There usually is a compensatory head turn to fuse, which allows better muscle balance in the exotropic eye position—where fusion alignment can occur. The head position is governed by the more muscle-imbalanced eye if there is fusion or when there is fixation with the DS eye.

The surgical goal is to correct the deviation and head turn and ameliorate as much of the anomalous DS eye movement as possible. Again, judging whether one must operate on the anomalous LR is an easy decision. In all but very mild cases of exotropic DS, the anomalous LR innervation may be considered to be severe. The shortened anomalous LR must be recessed. This simultaneously diminishes the DS anomalies, lessens any (sub)normal LR innervation, and partially realigns the eye. It is a constant error in exotropic DS management to underestimate the contracture propensity of the LR and its frequent postoperative recurrence. For severe LR anomaly and large-degree exodeviation, it is difficult to perform enough surgical weakening of the LR, even with recession far back of the equator and suturing on the globe. In jest, to those who still like formulas, I have commented that here the formula is “about 1-mm recession for 1 prism diopter of primary position exodeviaton correction.” The surgeon must consider the possibility of a rare subnormally innervated MR.

Resection of the secondarily stretched out (but not anomalously innervated) MR is indicated (i.e., a recess/resect procedure), sufficient to align the eye with balanced MR and LR forces at the time of surgery. The anticipated result from surgery is that the eye has been shifted from an exotropic position, with no abduction, to an ortho position, also with no abduction. The eye has simply been moved to an ortho position. This may be viewed by some parents or patients as a loss of ability to rotate the eye outward (active abduction) compared with preoperative rotations. However, there was no ability to rotate the eye outward from whatever exodeviation position preexisted, or from the now orthotropic posi-
tion. This may be a difficult concept for both the surgeon and the patient, and the anticipated result must be discussed before surgery. The patient really believes that "the eye does not go out anymore." It must be stressed that the eye did not move outward in either instance. It simply was out preoperatively and now is straight after surgery.25

What about augmenting the correction of a large exodeviation by recession of the LR in the normal eye? Exotropic DS is not an ordinary exodeviation, because one is managing ever-present gaze-related anomalies. Whether one can modestly recess the normal eye's LR depends on the severity of the LR anomaly in the DS eye and the amount of residual misbehavior after the LR has been recessed. Attempting to augment the exocorrection by recession of the LR in the normal eye can potentially create the possibility of two unwanted effects. First, for left eye eso-DS, recessing the normal eye's LR (right LR) causes a right gaze duress innervation for fixation even in the primary position. This sends unwanted central right gaze innervation to the anomalous left LR, which maximizes its anomalous co-contraction in the opposite gaze (right gaze), as well as in primary-position fixation of the right eye. If the anomalous left LR has been only modestly recessed, the ability to cause DS retraction and/or equivalents in even modest opposite gaze may still be observed. On the other hand, if the anomalous LR has been recessed by a very large amount, the same increasing fixation duresses in opposite (right) gazes may allow a still-normal left MR to overpower a very large left LR recession. This may result in an overcorrection (eso-deviation) in primary and/or right gazes.

One must carefully consider the possible unwanted results from even a modest recession of the normal eye's LR. The decision is dependent on the severity of the residual LR anomaly in the DS eye and the amount of residual misbehavior capacity it exhibits after its recession. It is not the same circumstance as in a non-DS exodeviation.

In many cases an eso-DS may be viewed as a progressive stage of an earlier classic eso-DS. It should also be recognized that anomalous congenital tissue bands rarely may hold the eye in an esodeviation position. These bands may be discovered only at the time of surgery by the inability of forceps to rotate the eye adequately after the LR has been detached from the globe. With extremely exotropic DS (and in the splits), the progressive mechanical LR shortening and loss of elasticity reduce the possibility of a muscle wrap-around extended arc of globe contact and bridle effect with the opposing MR (which may be subnormally innervated or weakened by being stretched out), so that a previously existing upshoot/downshoot may disappear. The DS pattern may change with progression of mechanical LR changes, but LR innervations (both normal and abnormal) remain the same.

Simultaneous Abduction of Each Eye (The Splits). These DS cases usually have no accompanying retraction or equivalents, and the LR may be anomalous, or both anomalous and (sub)normal. The term simultaneous abduction is descriptive and avoids the unnecessary sophistication of the term synergistic divergence, which imputes some unwarranted etiologic significance to the "divergence." 71, 11a, 11, 13, 126, 169, 174, 185, 184, 194, 196, 203 Even the term simultaneous abduction does not strictly adhere to our definition of abduction as an actively innervated abduction. Descriptive terms are always preferred to etiologic terms, because etiologic terms may change with the acquisition of new information. Somewhat in jest we have used the descriptive term "the splits,"71 which, perhaps unfortunately, has found some common use. (For a more thorough discussion of the confusion generated by mixing descriptive and etiologic meanings in the use of such terms as divergence or convergence, the reader is referred to the classic terminology papers on ocular motility by Lancaster.96, 97)

In unilateral cases (left eye example), the simultaneous abduction is exhibited when the normal right eye fixes toward opposite (in this case, right) gaze and there is a simultaneous outward rotation of the nonfixing DS left eye. These cases demonstrate the absolute necessity of controlling gaze fixation. The causes of this pattern and the management questions are unchanged from the traditional classic eso-DS, in which there is anomalous LR recruitment to opposite gaze. Whether the precipitating cause is naturally occurring or postoperative MR recession, it is the weakened MR in the DS eye that allows the co-contracting LR to overpower the weak MR in the same classic way, that is, in opposite (in this case, right) gaze. Instead of there being retraction and/or equivalents and the usual reduced internal rotation of the DS eye in opposite gaze, the LR anomalous contraction overpowers the weak MR and with exactly the same physiology, but now the imbalanced forces turn the DS eye outward and simultaneous abduction is observed.

The MR is usually normally innervated but rarely may be deficient. It is not anomalously innervated. That is, it does not receive innervation from the sixth nerve. In DS, when the redundantly available third nerve fibers are being distributed to their normal recipient (the MR), and also to their unusual recipient (the LR), it is possible that the MR may be deprived of its usual amount of innervation. Noninnervated parts of the MR muscle may become fibrous as well as suffer deficient innervation.

It is possible that this weakness of the MR (not anomalously innervated) may be responsible in part for such patterns as the splits (and some eso-DS), in which the MR may be found to have a weak generated force. However, this does not alter the important principle that the MR is not anomalously innervated, nor is management significantly affected by the sometimes weak innervation of the MR, in addition to its being stretched out (elongated) due to an exotropic position. The MR must be resected in either instance. Carlos Souza-Dias96 suggested that a medial rectus paresis was an important cause of "synergistic divergence."

I became indelibly convinced of the DS "splits" mechanism when I caused simultaneous abduction to appear immediately after only a modest MR recession in the DS eye of an "ordinary" eso-DS case. It proved to be a patient with severely anomalous LR innervation (see Fig. 24–7). My report4 emphasized the failure of using a fixed formula for the MR recession in eso-DS, when there are such vast differences in the severity of the anomalous LR innervations. The almost immediate postoperative occurrence of the simultaneous abduction often leads to a rapid severe LR contracture. The inquisitive surgeon can easily demonstrate this mechanism during the course of an indicated MR recession under topical surgical anesthesia in eso-DS by instructing the patient to gaze to the right (right eye fixing) after the left MR has been completely detached from the globe.71 In
this case, the still effective co-contracting anomalous LR overpowers the now ineffective detached MR in right gaze, resulting in the splits.

A head turn may be adopted to fuse. The direction of the head turn is dictated by the fixing right eye’s position that decreases outward rotation of the left DS eye, allowing the eyes to be aligned. There is no retraction or equivalents because the bridle effect does not exist, and the simultaneous abduction becomes the retraction equivalent. This has the same meaning as the retraction that would occur in a preexisting eso-DS in opposite (right) gaze with an MR that is not (yet) overpowered by an anomalous LR.

The answer to the question of whether one must weaken the anomalous LR (the same question as in management of all DS patterns) is, in this instance, easy: it is a severe LR anomaly and must be recessed a large amount. Here the LR is not only anomalously innervated in opposite gaze but is also allowed to contract via eye positions of extreme exodeviations. There is a double dose of the propensity to LR contracture.

The importance of controlling the gaze innervation in all DS patterns cannot be overemphasized. When uncontrolled, there may be cross fixation of the DS left eye in right gaze, in which case the fixing DS eye, under adduction fixation demand, can do nothing other than adduct, masking the entire simultaneous abduction pattern. This occurs especially in some bilateral DS cases with the splits, in which each eye may have some deficient abduction, making cross fixation easier.

The question of whether there is any normal LR recruitment in addition to the severe anomalous LR recruitment is more or less academic, because one must perform a very large recession of the anomalous LR, which simultaneously diminishes both anomalous and any normal LR innervations. But the question should be asked, and simple observations in such an instance of both anomalous and normal LR reveals the following. As the fixing right eye moves from primary to right gaze (abduction), the left eye simultaneously moves outward. As the right eye moves back toward primary position, the nonfixing left eye reduces its outward rotation because co-contraction is now diminished by decreasing right gaze innervation. If the right eye has to continue to fix from primary position toward adduction (with controlled fixation), the nonfixing left eye may move outward again if there now is (via Hering’s law) stimulation of any normal LR innervation. In any instance, the head turn chosen for fusion is where the fixing right eye’s position and innervation reduce outward rotation of the left eye, allowing alignment and fusion to take place.

After the first decision and step of a large LR recession, the secondarily stretched out, normally or subnormally innervated MR must be resected, with the amount depending on the residual deviation. In extreme cases, where the globe appears to be fixed in extreme exodeviations with extremely limited rotations, the splinted MR may lose some contractility but must nevertheless be resected, preferably on an adjustable suture.

Whether to perform LR recession in the good fellow eye should be weighed using the same parameters discussed previously under exodeviations.

**Fixation with the DS Eye.** This may occur with or without retraction/equivalents. There may be marked over-
Surgery should first address the imbalance of forces in the fixing DS eye (forces should be rebalanced in the primary position). Any surgical changes in the duress pattern of the fixing eye produce a central innervational outflow change in all the muscles in both eyes and alter the fellow eye’s overdriven/underdriven muscles as well. I previously (1971) remarked that “since the fixing eye determines the motor outflow to all the muscles in both eyes, it is obvious that the motor outflow pattern to all the extraocular muscles will change when one muscle in the fixing eye is surgically altered. This is a most important notion with far reaching consequences.”

Thus, adjustable sutures in the normal fellow eye are strongly suggested for adapting to the difficult-to-predict exaggerations and/or diminishations of overdriven muscles, following the first required step of altering the fixation duress process in the fixing eye to better balance its fixation alignment. In the rare patient with mild anomalous LR innervation, in which the DS eye has become much more esotropic, a substantial recession of the MR may be necessary for alignment purposes. In this very rare circumstance, a very modest resection of the LR may be indicated only if there is indeed a very mild anomalously innervated LR.

Y (or λ) Pattern. There is normal (or slightly deficient) abduction. Anomalous LR innervation occurs only in vertical gazes (minimal or absent horizontal gaze anomalies); thus, no retraction is observed. The Y pattern is characterized by increasing outward rotation of the DS eye from primary to upgaze and may reach large exodeviations in far upgaze. The inverted Y (or λ) is less prominent. Duane,34 the consummate observer, described it: “In spite of the almost complete inaction of its externus (the left eye) diverges markedly and progressively as the eyes are elevated, and when elevated can be abducted to quite a considerable degree.” Duane wrestled with theories of other vertical muscles being causative before the notion of paradoxical innervations was introduced in the 1950s.15 In 1961, Esslen and Papst37 explained the “divergence in upgaze” as due to anomalous recruitment of the LR. Scott,157,158 in 1968 and in 1973, demonstrated that the LR muscles are responsible for the V pattern in DS. In 1985, I stated that “one should always think of this type of Duane syndrome” in any patient where the divergence in upgaze is large, that is, “70 to 100 prism diopters.” And also that one should “note that the involved left eye adduction is worse in upgaze. In other words, the entire Duane pattern is worse horizontally all across the field in upgaze than in other positions.” In the diagnostic examination section, I discussed the importance of observing horizontal gazes, not only going through the primary position but in upgaze and downgaze across the board. The exaggerated restriction of adduction in upgaze is often remarkable. “This is one of the most interesting DS patterns because it is one of the least recognized.” In my 1988 Jules Stein lecture on Duane syndrome,75 these patterns were described as a Y and λ pattern, and I suggested that whenever one sees this pattern, “with a little deficient abduction...think Duane’s. This is not an inferior oblique as it goes across the board.”75 Kushner,41,94 in 1991 and 1997, popularized the Y pattern and termed it “cross fixation inferior oblique overaction associated with Y and V patterns.”

There may be a habitual chin-up or chin-down position to better fuse in these patterns. Characteristically, there is either minimal or no DS anomaly in the usual horizontal gazes through primary position, other than an occasional minor deficiency in abduction.

If surgery is indicated, it consists of bilateral resections of the LR muscles and elevation of the tendons, with more than the usual amounts of both recession and elevation. There is another surgical procedure that uses the Y pattern rather than getting rid of it. I described this as the “scroll down” procedure; it consists of weakening both SR muscles (10 mm each) and creating a fixation duress toward upgaze while fixing in the primary zone.73 This produces more outward rotation or “apparent” abduction in horizontal gazes through primary position. A “scroll up” may be achieved by modest recessions of both LR muscles for λ patterns.121 Of course, anomalous chin-up/down positions for better alignment may be surgically approached by reducing the deviation in the primary position by other means.

The pseudo-Brown syndrome is a subtype of the Y pattern, observed when there is a leash restricting upward and inward rotation. The diagnosis of apparent Brown syndrome associated with DS has been well noted.195 It is my experience that these cases represent only a pseudo-Brown syndrome, as demonstrated by the forceps traction restriction in upward and inward rotation but with better continued upward rotation during retropulsion of the globe, that is, the absence of a tight superior oblique reverse leash. The pseudo-Brown forceps maneuvers are well discussed by Pittar35 in his textbook. The reader should not be surprised to learn that this DS variant is due to an anomalous LR innervation and mechanical shortening that limits the upward and inward rotation as part of the Y pattern.75

When surgery is indicated, the recessed anomalous LR must be transposed upward sufficiently so that it is above the crest of the globe in that position, rather than in the preoperative below-the-crest position. This is essential for successful relief of the pseudo-Brown pattern. At surgery one can see the blue sclera underlying the restrictive muscle, which corresponds to the indentation made on the globe. When the restrictive anomalous LR is detached from the globe there is now an easy upward and inward forceps rotation. These findings do not occur in true Brown syndrome. This surgical approach improves both the pseudo-Brown syndrome and the Y pattern.

Subnormal LR Innervation. There is no anomalous LR innervation and therefore an absence of retraction and/or equivalents. Limited rotation is seen in both directions of horizontal gaze. I here report and include this as a DS anomaly because it is derived from the same roots of maldeveloped sixth nucleus and nerves, with subnormal LR innervation and marked developmental muscle fibrosis and shortening, but without any apparent anomalous LR innervation. It is identical in many descriptive ways to the “limited adduction and abduction” eye but includes no anomalous innervation. The subnormal LR innervation causes deficient abduction, and the fibrotically shortened LR limits rotation in abduction (opposite gaze), but there is no retraction or other DS retraction equivalents. Forceps force testing, along with observations of clinical rotations, establishes the diagnosis. It is here that the force degeneration test (Romero-Apis) is most informative (see Fig. 24–4), because it rules out anomalous LR co-contraction. Surgical management may
not be indicated if the rotations are appropriately balanced. If they are not, then the surgeon may rebalance the normally innervated MR and weaken the subnormally innervated and fibrotic, shortened LR as appropriate for the deviation.

**Bilateral Duane Syndrome**

It is generally accepted that bilateral DS comprises 15% to 20% of DS cases. However, the published literature on bilateral DS comprises only a fraction of that, for very good reasons. Thereinin, in strabismus management are the indications for surgery so unclear, the benefits so unpredictable and problematic, and the risks so high. The diagnosis is often clouded by the presence of other bilateral so-called cluster anomalies, such as gaze palsies, fibrosis syndrome, and Möbius-like syndromes among others. Of the little published material available on bilateral DS, only very few reports are specifically directed toward the management of these patients. Rather, the literature speaks to a scattering of unexpected and often disastrous results published by bravely honest investigators. Here I shall attempt to focus on both general and specific bilateral DS patterns and give some examples of my own frustrating experiences. First, one must recognize that the patterns and management of bilateral DS with fusion are vastly different from bilateral DS without fusion, and it is unhelpful to group their diagnosis and management together.

**Bilateral DS with Fusion.** A patient with bilateral DS and fusion usually has limited eye movements and/or relatively immobile globes. Retraction and/or equivalents are generally not observed. The diagnosis is often mistaken as congenital horizontal gaze palsy. There is usually a straight head with fusion. Inverted Y (λ) patterns are more common here than are Y patterns, and a chin-up or chin-down position may be used to aid fusion. If indeed, surgery is indicated, management is conveniently divided into patients with relatively immobile globes (severely limited eye movements) and those with reasonable, though imperfect, eye movements.

**Relatively immobile globes** may produce a bilateral pattern similar to that described under unilateral relatively immobile globe. However, when bilateral, the pattern is severely compounded by fixation duress in each eye on attempted gaze to either side, with usual short, tight MR and LR muscles, as described in the discussion on unilateral relatively immobile globe. However, one cannot assume this is so unless it is demonstrated by forceps force tests.

There may be a perfectly straight head position with orthophoria, or a head turn, usually due to gradually progressive heterophoria secondary to muscle imbalance in one of the bifixing eyes, which controls the head position. It is especially in this DS pattern that one needs to know whether there is any active normal LR innervation. In the often-encountered esophoria (with λ pattern), let us analyze the results of any amount of bilateral MR recessions. I have previously stated that patients with bilateral DS and fusion “may have no abduction, or may have some abduction, and there is a tremendous difference” and explained that with no actively innervated LR abduction it may be safe to recess both MR muscles. If there is any active abduction, as well as anomalous LR innervation, a recession of one or both MR muscles may unleash large amounts of immediate exotropia. This is because of the now combined fixation duress (either dominant eye at the time) with yoke exaggerated innervation of the now free-to-move fellow eye and some normal LR innervation. In this case, MR recession “opens the gate” to exodeviation because it is a worse case of fixation duress no matter which eye fixates and no matter in which direction, with overaction of yoke muscles. Whether there is any significant normal LR active innervation is not a trivial matter. The force generation test and other forceps force tests are essential to predict the results. On the other hand, with no normal active innervation of the LR in abduction, it may be safe to recess both MR muscles, with due regard for any λ pattern.

The management scheme cannot depend on utilizing clinical observations of “limited” eye movement in one direction more than another, because an active abduction force may be held in abeyance by the bilateral opposing stiff, shortened muscles. One must determine the anomalous and normal innervations present in all DS patterns, and especially in bilateral DS with fusion and relatively immobile globe.

Patients with bilateral DS and fusion who exhibit relatively immobile globes usually (though by no means always) have all four horizontal muscles restricting movement in all directions. This is similar to the previously described unilateral DS with fusion and immobile DS globe, but bilateral cases are compounded by bilateral fixation duress problems.

In patients with bilateral DS and fusion who exhibit some reasonable eye movements, any (sub)normal active abducting forces may be identified by simple observation, making management strategies easier. Fairly normal MR adduction and convergence may progressively decrease over time as the eye movements diminish and relative immobility develops. Sevel and Kassar reported three successive generations of bilateral DS with fusion, which emphasizes the hereditary factor in this particular group of patients. Bilateral DS, even more commonly than unilateral DS, may be mixed with the other cluster anomalies, especially gaze palsies, ophthalmoplegia, fibrosis syndrome, and Möbius-like syndromes. I have seen mild eso-DS in one eye, and fibrosis syndrome in the fellow eye.

Because of the unpredictable results and the risk of surgery in bilateral DS patients with fusion, prisms should be considered for straightening the head or alleviating symptoms from heterophoria. The surgeon should recommend surgery only when compelling indications exist. The lesser the surgical indications, the greater the likelihood of postoperative complications. Both intraoperative and postoperative adjustments should be employed. Preoperative local anesthetic injection into a muscle, as a therapeutic trial, releases only the innervation and not the mechanical aspects of the problem. It is here that intraoperative maneuvers under topical surgical anesthesia (conjunctival drops only) has its greatest benefit, because one can readily assess the immediate effects of even mild muscle position changes. Alternatively, adjustable sutures play an enhanced role here because of the unpredictability of the postoperative results.

Adjustable sutures are infrequently applicable to infants and children who either have bizarre DS patterns or have undergone previous surgeries. The sutures may be adjusted 1 or 2 hours postoperatively under selected, fast-acting anesthesia. One can observe the rotations well enough to make outcome judgments during the immediate postoperative period. I have repeated the adjustments at 1- to 2-hour intervals.
as many as three separate times, in rare instances, to achieve optimum outcomes.

Recession of all four horizontal muscles, in amounts depending on their relative state of contracture, may be indicated if there are adequate indications for surgery and a likelihood of obtaining clear benefits. The resultant postoperative eye movements may change little from those existing preoperatively.

Spielmann has reported an unusual case of bilateral DS with all four horizontal muscles short and stiff, limiting upward gaze by their vectors; it was alleviated by recession of all four horizontal muscles.

**Bilateral DS Without Fusion.** Patients usually have marked esodeviations or have marked exodeviations (including the splits). Retraction and/or equivalents are not common. Either eye or both may have anomalous and/or normal LR innervation. The deviations are usually large because of the fixation duress of the fixing eye at any time, with an overdriven/underdriven fellow eye, often in alternating fixation patterns. A falling or rising of the nonfixing abducting eye may also be observed.

The surgical management of these patients is similar to the previously described management of unilateral DS in which there is fixation with a DS eye. Of course, in bilateral cases there are overdriven/underdriven rotations in the nonfixing fellow eye, but these are now further complicated by this eye having both anomalous LR innervations and either a total absence of abduction or some active abduction. However, in bilateral DS cases, the fellow eye cannot be overdriven in the direction of an absent abduction function. This must be differentiated from ordinary alternating infantile esotropia. Thus, there may be a complex mixture of eye rotations due to fixation duresses and overdriven/underdriven fellow eyes. The goal is to ameliorate any abnormal head position by rebalancing the MR and LR in the dominant fixing eye. This changes the fixation innervation of the fixing eye, and also the yoke innervations to the fellow eye. Predictions of the surgical outcome are far from certain, often requiring adjustable suture (see the discussion of unilateral DS fixing with the DS eye).

**Bilateral simultaneous abduction** (the “splits”) is a subtype of bilateral DS without fusion. It is very much an exaggeration of the unilateral splits discussed earlier. When occurring naturally (not after MR recession), it progresses rapidly to the most extreme degrees of relatively immobile exotropia. It is as if the globes were nailed to the outer canthus.

Several innervational and mechanical (eye position) factors operate together to cause the most extreme shortening and contracture of the LR muscles. Consider the following in the example of bilateral DS with simultaneous abduction: when the right eye fixes in right gaze, the left eye also rotates outward simultaneously, as explained previously. No matter where each eye fixates in patients with bilateral DS and simultaneous abduction, the LR muscles of each eye become markedly contractured/shortened by both innervational and mechanical (exotropic position) factors.

Treatment is directed toward eliminating the function of each LR, not just a simple large recession (as discussed previously), combined with marked resections of the MR muscles to align the eyes. If the eyes are not aligned under surgical anesthesia on the operating table, they will not be aligned postoperatively. Any residual innervation or mechanical exoforce of the LR muscles will destabilize an otherwise satisfactory result. This propensity for recontracture of the LR is the problem in all cases of DS with large exodeviations, but especially in bilateral DS with simultaneous abductions.

**Bilateral Y or λ Pattern.** Bilateral Y (or λ) pattern is an exaggeration of the unilateral pattern described earlier. It is included here only for the sake of completing the categories. The more common λ (rather than the Y) may be alleviated by transposing the LR muscles downward with recessions to compensate for the increased muscle tension caused by the transposition. Alternatively, depending on the deviation in the primary position, this pattern may be relieved by corresponding transposition of the horizontal muscles, as is appropriate.

**TREATMENT OVERVIEW**

Anomalous and/or subnormal LR innervations and mechanical changes account for the DS patterns. Secondary strabismus changes of other (not anomalously innervated) muscles may occur, as well as subnormal innervated MR muscle. These considerations allow for a rational management approach.

**REFERENCES**

34. Duane A: Congenital deficiency of abduction associated with impairment of adduction, retraction movements, contraction of the palpebral fissure and oblique movements of the eye. Arch Ophthalmol 1905;34:133.
68. Jampolsky A: Paralytic strabismus—Duane’s syndrome and VIth nerve
Dry.


MacLèbois NM: Paralysis of left external rectus, movement inwards of the affected eye associated with contracture of the orbicularis palpebrum on the same side and marked retraction of the left globe. Trans Ophthalmol Soc UK 1895:16:209.


123b. Mulhern M, Keotane S, O’Conner G: Bilateral abducens nerve lesions in unilateral Type 3 Duane’s retraction syndrome. Br J Ophthal-
mol 1994:78:70.


125a. Nemet P, Ron S: Ocular saddaces in Duane’s syndrome. Br J Ophthal-

126. Noel LP, Clarke WN: Adduction deficiency following medial recti re-
cession in Duane’s retraction syndrome: Case report. Arch Ophthal-

127. O’Malley ER, Helveston EM, Ellis FD: Duane’s retraction syn-


136. Prieto-Diaz J, Souza-Dias C: Duane’s syndrome electro-oculographypattern. J Pedia-


138. Prieto-Diaz J. Duane’s syndrome electro-oculography pattern. J Pedi-


143. Remy A, Bricht B: Le syndrome de Stilling Turck Duane: Etude electromyographique. Rev Oto- 


Chapter 25

BROWN SYNDROME

DAVID R. WEAKLEY, Jr., MD, DAVID R. STAGER, MD, and DAVID R. STAGER, Jr., MD

Historical Perspective

The clinical entity commonly known as Brown syndrome was initially described in 1949 by Harold Waley Brown as the superior oblique (SO) tendon sheath syndrome. The patients were described as having a short anterior tendon sheath of the SO, which restricted passive elevation of the globe in the nasal field. Brown also described a patient showing restricted elevation in adduction with forceps while under anesthesia (what is now known as a restricted forced duction test in Brown syndrome). He initially believed that the syndrome was due to congenital paralysis of the inferior oblique (IO) muscle, resulting in secondary shortening of the anterior sheath of the SO tendon.

The accepted definition of the SO tendon sheath syndrome has evolved considerably over the years. What is now known as Brown syndrome still has as its most consistent feature an inability to elevate the eye in the adducted position, both actively and passively on forced duction testing (Fig. 25–1). However, a number of other findings have been associated with the syndrome and are discussed further.

Epidemiology

Accurate estimates of the incidence and prevalence of Brown syndrome are difficult to obtain, but it is believed to occur once in every 430 to 450 cases of strabismus. Brown syndrome is found in 10% to 24% of patients with vertical muscle paresis. If one accepts that the overall incidence of strabismus in the general population is 2% to 3%, by extrapolation the estimated incidence of Brown syndrome is approximately 1 in 20,000 live births.

Review of many series of Brown syndrome patients has failed to document consistent predilections for either laterality or sex. Although Brown initially reported a slight excess of females (58%) and right eye involvement (62%), Wilson and colleagues, in a review summarizing several series, noted roughly equal distributions. The same study showed that Brown syndrome may be bilateral in approximately 10% of cases. The true incidence is probably higher because many mild cases are asymptomatic and never reach an ophthalmologist.

Heredity

The familial occurrence of Brown syndrome has been noted. Twin studies reveal that monozygotic twins may be equally affected or that in some instances only one of the pair is affected. Affected dizygotic twins have also been documented. Cases of affected siblings, often with bilateral Brown syndrome, as well as instances of an affected parent and child have been described. Nevertheless, most cases occur sporadically. The few familial cases have led authors to postulate autosomal dominant inheritance with incomplete penetrance and variable expression, although recessive transmission has been proposed in some cases.

Etiology

The question of etiology is one of the most controversial issues surrounding Brown syndrome. This is further complicated by the existence of both acquired cases with multiple causes and congenital Brown syndrome.

The acquired variety involves secondary changes in a previously normal SO tendon or tendon-trochlear complex, producing the clinical picture of Brown syndrome. On the other hand, the congenital variety is a primary abnormality in the tendon and/or the tendon-trochlear complex, which is present from birth.

For the normal eye to elevate in the adducted position, the SO muscle and tendon must relax as the anterior pole of the adducted eye moves upward. The posterior pole of the eye, where the SO tendon inserts, moves away from the trochlea to an abducted and depressed position. This requires a certain degree of relaxation or elasticity of the SO muscle and tendon.
**CONGENITAL BROWN SYNDROME**

**“Sheath” Anomalies**

Brown continued to believe in this theory for some time, although he abandoned the concept of a short anterior tendon sheath. He maintained that the cause of a “true sheath syndrome” was still unknown but continued to refer to some of his cases in this way. He also classified a number of cases as “simulated sheath syndrome,” including acquired, intermittent, and spontaneously recovered restrictions of elevation in adduction. Brown ultimately realized that more than one factor explained the clinical findings.

**Short or Inelastic Tendon**

Girard theorized that one of several possible mechanisms of Brown syndrome was a congenital or acquired anomaly of the SO tendon, restricting its passage through the trochlea. Crawford suggested a tight SO tendon rather than its sheath as the cause. This was supported by a high success rate of SO tenotomy in these cases. Parks agreed, arguing that the SO tendon did not in fact have a sheath but instead had a fine transparent avascular capsule like the other extraocular muscles. This capsule extended the length of the tendon and could not be the primary cause of Brown syndrome. In fact, current surgical techniques that have evolved to treat Brown syndrome (including tenotomy and the SO expander) are designed to minimally disrupt this structure.

Currently, the most widely accepted theory of the cause of Brown syndrome (at least the congenital stationary variety) is that of a congenitally short or inelastic tendon. This hypothesis is supported by the frequent success in relieving the inability of the eye to elevate in adduction after a tenotomy. The success of this procedure, however, does not rule out the presence of a trochlear or tendon-trochlear abnormality. A tenotomy distal to the trochlea would still free the eye to elevate in adduction in such circumstances (Fig. 25–2).

**Trochlear or Tendon-Trochlear Complex Abnormalities**

Sevel postulated a tendon-trochlear anomaly as the cause of congenital Brown syndrome. The embryonic trabeculae between the SO tendon and trochlea, which normally become fully attenuated during development, may persist and inhibit passage of the SO tendon through the trochlea. Similarly, Wilson and associates postulated that Brown syndrome parallels a condition known as congenital trigger thumb, in which the tendon sheath of the thumb (analogous to the trochlea in the orbit) prevents free movement of the tendon. A tendon-trochlear abnormality more readily explains the spontaneous resolution that occurs in some congenital cases.

Resolution of a congenitally inelastic or short tendon seems unlikely; spontaneous resolution of an abnormal tendon-trochlear complex seems more plausible. Perhaps congenital Brown syndrome occurs in patients with either of these causes, a short tendon or a tendon-trochlear anomaly, with those who spontaneously resolve being more likely to have the latter condition.

Well-documented cases of congenital intermittent Brown syndrome (sometimes referred to as the “idiopathic click” syndrome)
syndrome) suggest a trochlear problem. These patients present with typical characteristics of Brown syndrome that may be relieved by forced elevation in adduction. This may sometimes be associated with an audible or palpable click, consistent with passage of a resistant part of the SO tendon through the trochlea. These patients should be differentiated from those with acquired Brown syndrome, who may also have intermittent symptoms and clicks ("inflammatory click syndrome")—even though the mechanism may be similar.

**ACQUIRED BROWN SYNDROME**

(Table 25–1)

<table>
<thead>
<tr>
<th>Iatrogenic</th>
<th>Inflammatory or Infectious Processes</th>
</tr>
</thead>
<tbody>
<tr>
<td>Superior oblique surgery (e.g., tuck)</td>
<td>Rheumatoid arthritis, systemic lupus erythematosus, Sjögren syndrome, and Graves ophthalmopathy have been reported.</td>
</tr>
<tr>
<td>Scleral buckling surgery</td>
<td>Improvement with corticosteroid treatment supports inflammation as a cause of Brown syndrome.</td>
</tr>
<tr>
<td>Glaucoma valve surgery</td>
<td>Traumatic</td>
</tr>
<tr>
<td>Sinus surgery</td>
<td>Injury to the trochlear area is another cause of acquired Brown syndrome. These patients frequently have associated SO palsy and were described by Knapp as having &quot;canine tooth syndrome&quot; because some injuries resulted from dog bites. Various types of injury including blunt trauma have also been reported to cause the syndrome.</td>
</tr>
</tbody>
</table>

Table 25–1. Causes of Acquired Brown Syndrome

![Figure 25–3. Photograph of freshly dissected superior oblique muscle, tendon, and trochlear pulley. Note the very narrow posterior opening of the trochlea. Stenosis of this opening or dilatation of the tendon would limit free movement of the tendon through the trochlea. This mechanism seems likely in the majority of acquired Brown syndrome cases and possibly some cases of congenital Brown syndrome as well. (From Wilson ME: Vertical Strabismus. In Wright KW (ed): Pediatric Ophthalmology and Strabismus, p 223. St. Louis, Mosby-Year Book, 1995. Reprinted with permission.)](image)

**Trauma**

Clinical Characteristics

**NATURAL HISTORY**

Although classic congenital Brown syndrome is usually permanent and stationary, there is clear evidence for spontaneous improvement in selected cases. Such resolution is probably more common than previously recognized, based on an age distribution skewed toward the younger age group (i.e., a higher prevalence in children). Nine of 10 patients with idiopathic Brown syndrome arising by age 2
years experienced spontaneous resolution without surgical intervention when observed for a mean of 13 years. Three of them ultimately attained normal motility. In some cases, resolution is associated with duction exercises that attempt to elevate the involved eye in adduction. Thus, although the actual incidence of spontaneous improvement or resolution is unknown, the possibility of its occurrence must be weighed when considering surgery. It has been suggested that the constant and idiopathic click forms of Brown syndrome lie on a single continuum, the latter representing some cases of the constant variety in an intermediate stage of resolution.

The natural history of acquired Brown syndrome is variable and depends on the underlying mechanism. When associated with inflammatory conditions such as rheumatoid arthritis, the findings may wax and wane in concert with the activity of the underlying disease. Spontaneous improvement is not expected in iatrogenic cases, such as those after scleral buckling or glaucoma filtration surgery.

CLASSIFICATION

Brown initially divided his early cases into “true” and “simulated” sheath syndromes. True sheath syndromes were further classified as either typical or atypical. The findings were considered atypical if there was significant coexisting paresis of the ipsilateral superior rectus muscle. This may, however, have simply represented more severe cases with elevation deficiency in abduction. Simulated sheath syndromes included patients with spontaneous recovery, intermittent cases, or acquired cases that could not be explained by a congenital tendon sheath abnormality.

This classification seems less valuable given our current understanding of the various causes. Classifying Brown syndrome as either congenital or acquired still remains acceptable. Most cases of congenital Brown syndrome are constant. They are less likely to improve spontaneously than acquired cases and more likely to require surgery if severe. Acquired cases are clearly not evident at birth. They may be caused by a variety of factors, with the final common denominator being structural change in the SO tendon, the trochlea, or the tendon-trochlear complex that impedes normal passage of the tendon through the trochlea. Acquired cases (especially the inflammatory variety) are more likely to be intermittent and to improve spontaneously.

Some authors prefer to classify Brown syndrome according to its severity. This has the advantage of determining which patients should undergo surgical correction. We agree with this classification, describing Brown syndrome as mild if there is no hypotropia of the involved eye in the primary or adducted position but solely in the elevated and adducted position and less so when elevated (see Fig. 25–1). Moderate Brown syndrome includes patients having the additional feature of hypotropia (or downsink) in the adducted position (Fig. 25–4). Severe Brown syndrome includes cases with the above findings and, in addition, hypotropia in the primary position (Fig. 25–5).

Jampolsky regarded the absence of deviation in any horizontal position or downgaze (our mild Brown syndrome) as “true” Brown syndrome and classified patients who also had vertical deviation in the primary position or adduction, with or without a head turn and/or chin up posture (our moderate or severe Brown syndrome) as Brown syndrome plus.

Diagnosis

CLINICAL EXAMINATION

The sine qua non of Brown syndrome is an inability to elevate the adducted eye either actively or passively. However, depending on the severity and the underlying etiology, a number of other findings may also be noted. SO function is usually normal, but overaction may rarely be seen (Fig. 25–6). Wilson and associates summarized the findings comprising the spectrum of Brown syndrome, of which the first six must be present for a definitive diagnosis:

1. Deficient elevation in adduction
2. Less elevation deficiency in midline
3. Minimal or no elevation deficiency in the abducted position
4. Minimal or no SO overaction
5. V pattern with divergence in upgaze
6. Restricted forced ductions

Figure 25–4. Moderate Brown syndrome of right eye. Note the absence of deviation in primary position. A right hypotropia (downdrift) is noted in left gaze. Deficient elevation in adduction is readily apparent (upper right). V pattern exotropia and limited elevation of the right eye in midline are also present.
Other, less consistent findings include the following:
1. Downshoot or hypotropia in adduction
2. Widening of the palpebral fissure on adduction
3. Anomalous head posture
4. Hypotropia in the primary position

Other associated findings that may be noted in the acquired type, especially inflammatory cases, include trochlear tenderness and an audible or palpable click. This click, however, may also be noted in presumed congenital or intermittent cases of noninflammatory origin. Patients with bilateral Brown syndrome exhibit findings similar to those in unilateral cases, with the V-pattern exotropia in upgaze being especially noticeable (Fig. 25–7).

**STRABISMUS EVALUATION**

Careful systematic evaluation of each patient with Brown syndrome is essential to determine the etiology, the likelihood of spontaneous improvement, and the effect of strabismus on the patient’s general and visual function. As with all strabismus patients, the initial evaluation must begin with a careful history, giving attention to the presumed time of onset of strabismus (i.e., congenital versus acquired) and to any suggestion that the syndrome may be intermittent or improving. A family history of strabismus may help determine whether a familial variety exists. A history of medical problems (autoimmune diseases, inflammatory condition, trauma, or previous surgery on the eye or ocular adnexae) known to be associated with Brown syndrome is helpful, especially in acquired cases. Patients old enough to respond appropriately should be asked to what extent strabismus interferes with their daily activities and whether diplopia is present in any field of gaze.

The examiner then proceeds with a complete ophthalmologic and strabismic evaluation. Attention to visual acuity in both eyes will detect any amblyopia. One must determine if an anomalous head posture is present and if it is assumed for fusion. The strabismic deviation is measured in the nine cardinal positions of gaze as well as right and left head tilt. Versions and ductions are carefully evaluated in each eye.
Forced ductions may be performed under topical anesthesia in older children or adults, but younger children and infants are generally tested in the operating room.

Horizontal strabismus may be superimposed upon Brown syndrome, especially if it is severe enough to disrupt fusion and induce amblyopia. A secondary strabismus such as eso tropia may be present. Sensory status (fusion and/or suppression) may be evaluated using any of the standard methods: we prefer testing fusion with the Worth four-dot test (at near and distance) and measuring stereopsis with a vectograph test. Finally, careful refraction, either manifest or cycloplegic, and funduscopic examination (including evaluation for torsion) should be performed.

Laboratory examination does not play a significant role in evaluating the majority of Brown syndrome patients, especially those with the typical congenital form. Studies may be necessary in acquired cases to identify disorders such as rheumatoid arthritis and systemic lupus erythematosus. Appropriate laboratory tests may include studies for rheumatoid factor and antinuclear antibody. A patient with acquired Brown syndrome and systemic symptoms, not previously known to have systemic disease, is best referred to an appropriate medical professional. Local radiographic evaluation with plain films, computed tomography, or magnetic resonance imaging may be of some value in acquired Brown syndrome; it should be considered on an individual basis. For example, a patient with acquired Brown syndrome and a history suggesting possible sinusitis may warrant radiographic evaluation. Imaging may also be valuable when there is a history of trauma to the trochlear area.

**DIFFERENTIAL DIAGNOSIS**

Several strabismic entities may mimic Brown syndrome, but most can be distinguished by careful clinical evaluation. Occasionally, surgical exploration and forced duction testing may be required to make a definitive diagnosis.

True IO paresis may resemble Brown syndrome, and, in fact, Brown initially believed this was the cause of secondary shortening of the anterior tendon sheath of the SO. Although rare, isolated IO palsies exist and may be confused clinically with Brown syndrome. Patients with IO palsy are expected to have overaction of the SO muscle, a positive Parks’ three-step test suggesting IO palsy, a positive Bielschowsky head tilt test, and a compensatory head tilt. Restricted passive traction testing is noted in Brown syndrome, whereas forced ductions are generally free with IO palsies.

Double elevator palsy (monocular elevation deficiency) may also be confused with Brown syndrome. Unlike Brown syndrome, these patients generally have limited elevation in abduction that is equal to or greater than that in adduction. These patients may also present with ptosis or pseudoptosis, which is not characteristic of Brown syndrome.

Congenital fibrosis syndrome is a rare familial disorder in which severe fibrosis and restriction of any or all of the extraocular muscles, including the levator palpebrae superioris, may be present. Patients with unilateral or bilateral congenital fibrosis syndrome and primarily inferior rectus restriction might conceivably be thought to have unilateral or bilateral Brown syndrome. These patients, however, have restricted elevation in abduction that is equal to or exceeds the limitation in adduction. They also frequently develop esotropia on attempted upgaze, whereas the reverse is typical of Brown syndrome.

Blow-out fracture of the inferior orbital floor may present a picture similar to Brown syndrome. Normally, the elevation deficiency will be symmetric in both abduction and adduction or worse in abduction, although cases with more marked restriction adduction have been reported. Associated findings such as a history of trauma, enophthalmos, infraorbital paresthesia, and characteristic radiographic findings should easily distinguish between these entities.

Thyroid ophthalmopathy may rarely be confused with Brown syndrome when the SO is involved in the myopathic process. Thyroid myopathy frequently presents with unilateral or bilateral hypotropia and restricted elevation. However, as with most other entities in the differential diagnosis, the elevation deficiency in abduction is the same as or worse than in adduction.

The adherence syndrome results from poor surgical technique, usually after IO surgery. With violation of the posterior Tenon’s capsule, intraconal fat may invade the anterior sub-Tenon’s space and produce adhesions that restrict attempted elevation of the involved eye. The elevation deficiency is typically worse in the abducted position, although in unusual cases the clinical findings resemble those in Brown syndrome. A history of previous strabismus surgery should lead the clinician to suspect this condition.

**Treatment**

**NONSURGICAL MANAGEMENT**

Spontaneous resolution may occur even in the presumed congenital variety, and probably more often in the acquired or intermittent type. This possibility must be considered in determining the appropriateness of surgical
intervention. Elevation in adduction exercises has been reported to improve the condition in some patients having either intermittent acquired or congenital Brown syndrome.

Oral corticosteroids or corticosteroids injected in the area of the trochlea have been widely reported to improve or completely eliminate Brown syndrome in some patients, especially those with the acquired type associated with inflammatory disease. In many cases the improvement may be only temporary and incomplete. Systemic treatment of underlying arthritic, autoimmune, or inflammatory disease has also improved or eliminated some cases of associated acquired Brown syndrome.

In general, surgical intervention in patients with acquired, intermittent, or inflammatory Brown syndrome should be avoided unless the underlying disease has been controlled medically and the syndrome persists and meets the surgical criteria outlined next. Observation may be appropriate for mild or moderate cases in which the onset is unclear. Surgery can always be performed later if necessary.

SURGICAL MANAGEMENT

Indications

General guidelines for surgical intervention in Brown syndrome have evolved over the years, yet disagreement still persists among strabismus surgeons. The surgical success rate and the likelihood of postoperative complications, when compared with observation alone, helps determine the threshold for intervention. The high incidence of postoperative complications (as compared with strabismus surgery for more common problems such as esotropia) must be carefully weighed before proceeding with surgical correction.

The only absolute indication for surgical intervention in Brown syndrome is a severe and constant congenital case that threatens loss of binocularity and development of amblyopia in the involved eye if left untreated. This includes patients who have not developed a compensatory head posture to maintain binocularity (fusion).

Relative indications include primary-position hypotropia (severe Brown syndrome or “Brown syndrome plus”), with or without an abnormal head posture, and unacceptable downshoot in adduction (moderate Brown syndrome). Patients with mild (true) Brown syndrome and limited elevation only in adduction (and less so in midline upgaze), but with good binocularity and visual acuity, are generally best observed if no abnormal head posture is present. Procedures other than strabismus surgery may be indicated in some patients with acquired moderate or severe Brown syndrome secondary to other conditions, such as removal of a scleral exoplant or glaucoma filtering valve.

Surgical Options

Surgery in Brown syndrome is designed to correct hypotropia in primary gaze, reduce objectionable downshoot, increase upgaze, and expand the binocular diplopia-free fields. It is realistic to expect improved downshoot, head posture, and binocular diplopia-free fields, but improvement of elevation in adduction is only partial. This limited upgaze may not be apparent for several months or years after surgery. The surgeon, parents, and patient all should realize that no surgery can completely eliminate or “cure” Brown syndrome.

A “sheathectomy” of the anterior portion of the SO tendon was initially recommended by Brown, on the basis of the false assumption that the sheath of the tendon causes the syndrome. Not surprisingly, the results were varied and generally unsuccessful. Attempts at maintaining the eye in the elevated and adducted position also proved disappointing.

More consistent results were reported after tenectomy of the SO (Figs. 25–8 and 25–9). Improved elevation was reported in all patients, with complete resolution in 60%. Unfortunately, in addition to correcting the motility disorder caused by the syndrome, tenectomies or tenotomies are fraught with a high incidence of complications, specifically postoperative SO palsy, which may occur in 44% to 82% of patients (Figs. 25–10 and 25–11). This should not be surprising, because Brown syndrome is generally not associated with SO overaction. Nevertheless, SO tenectomy or tenotomy has been the treatment of choice for Brown syndrome until recently.

Because of concern over the high incidence of SO palsy, Parks compared tenectomy of the posterior portion of the SO, Z-tenotomy, and SO recession to the standard tenectomy procedure. The results remained unsatisfactory in most instances, and SO palsy still developed. Crawford’s results with these procedures were no better; tenotomy and tenectomy procedures were technically easier and resulted in less scarring.

Parks and Eustis recommended simultaneous IO recessions.
Wright introduced a technique in which a segment of silicone retinal band is sewn between the cut ends of a tenotomized SO tendon to control the amount of weakening. Three of four patients with Brown syndrome improved, and none developed SO palsy. Stager reported a relatively high success rate and a low rate of mild undercorrection with this technique. In this series, 17% of eyes developed iatrogenic SO palsy, whereas 57% had an excellent or good result with near-normal postoperative versions. In some patients full correction did not occur for up to 18 months postoperatively. No patient required further surgery for undercorrection.

**COMPLICATIONS**

**Superior Oblique Palsy (Overcorrection)**

The most common complication after surgery for Brown syndrome is iatrogenic SO palsy. This is minimized by selecting the appropriate procedure, but it still occurs. SO palsy may appear in the early postoperative period after an SO weakening procedure, but more typically it develops gradually. The findings in these patients are fairly typical of SO palsy: overaction of the ipsilateral IO, underaction of the weakened SO with head tilt to the noninvolved side, and a positive three-step test with excyclotropia of the involved eye. These patients will frequently have diplopia, especially in downgaze, and this can be difficult to correct. Santiago and Rosenbaum noted that iatrogenic SO palsy after SO tenotomy or tenectomy for Brown syndrome may not always be correctable or may require a series of procedures with the possibility of residual uncorrectable diplopia or torsion. This highlights the importance of selecting patients in whom surgery is truly indicated and of tailoring the surgical procedure to minimize overcorrections.

**Undercorrection**

Undercorrections may occur when Brown syndrome is treated by any of the weakening procedures, although they are less frequent and generally less worrisome. Undercorrection does not cause new postoperative complaints such as torsion and diplopia or worsen existing ones. Early undercorrection may occur with the SO tendon expander as well as...
with simultaneous SO tenotomy and IO recession procedures, but motility may improve over a number of months. One must be very wary of reoperating on undercorrected Brown syndrome until enough time has elapsed for ductions to improve.

**Other Complications**

Other problems may occasionally occur and are related to surgical technique and specific procedures. Postoperative ptosis and damage to—or transection of—the superior rectus muscle may occur with poor operative technique. These complications are more common with blind sweep techniques. Ensuring appropriate visualization of the tendon and careful technique should prevent these types of complications.

Adhesions in the superonasal quadrant can occur with any procedure in this area and may compromise the positive effects of surgery for Brown syndrome. Tendon lengthening procedures, when performed poorly, frequently cause adhesions. Careful technique that avoids disrupting the inner layer of the intermuscular septum can reduce or eliminate this complication (Fig. 25–12).

The silicone expander may extrude, but this risk can be minimized by avoiding an excessively long spacer (more than 8 mm), maintaining appropriate surgical planes, and closing Tenon’s capsule and the conjunctiva in separate layers. Otherwise, spacers may adhere to this area and lead to restricted downgaze.

**Conclusions**

One might still ask what the best procedure is for correcting Brown syndrome. Is there any procedure in which the benefits outweigh the risks? It seems evident that a free tenectomy results in an unacceptable risk of postoperative SO palsy and should be avoided. A tenotomy in which the tendon is simply cut and no segment removed may reduce the incidence of overcorrections, especially when performed appropriately. However, in Brown syndrome with no SO

---

**Figure 25-11.** Anomalous head posture after right superior oblique tenotomy for Brown syndrome. Note left face turn to allow for fusion (left) with inferior oblique overaction secondary to iatrogenic superior oblique palsy (right).

**Figure 25-12.** Top left, Preoperative moderate Brown syndrome right eye. Top right, Appearance 3 days after tenotomy of the right superior oblique tendon with care taken to avoid disruption of the intermuscular septum of the superior oblique tendon. Lower left, Versions 4 weeks postoperatively. Lower right, Versions 6 months postoperatively. The preoperative primary position hypotropia and downshoot in adduction have been eliminated. (From Wilson ME: Vertical strabismus. In Wright KW [ed]: Pediatric Ophthalmology and Strabismus, p 225, St. Louis: Mosby-Year Book, 1995. Reprinted with permission.)
overaction, even a careful tenotomy may result in unacceptable overcorrection.

An SO tendon tenotomy combined with IO recession may yield acceptable results with fewer overcorrections. Nevertheless, we believe the best operation for Brown syndrome is a controlled weakening procedure, preferably an SO expander that does not exceed 8 mm in length. This should be placed approximately 5 mm nasal to the nasal border of the superior rectus, and then Tenon’s capsule is carefully closed to prevent its extrusion. The inner layer of the intermuscular septum should be minimally disrupted to avoid adhesions.

Tenotomy using some sort of restraining suture to prevent excess separation of the tendon ("the chicken suture" as initially described by Knapp) is also an acceptable procedure when performed correctly. This is technically easier and less time consuming than using a silicone spacer. Our preference is to interdigitate knotted 4-0 Mersilene between the cut ends of the tendon to achieve the desired length, although a simple loop of nonabsorbable prolene may give equally good results. A "chicken suture" has the benefit of no "excess hardware" beneath or close to the superior rectus, which may cause limited downgaze owing to superior rectus restriction after the silicone spacer procedure.50 67

REFERENCES

MOBIUS SYNDROME

DANIEL M. LABY, MD

Although von Graefe initially described the "congenital facial diplegia" syndrome in 1880, it was Möbius, in 1888, who recognized the association between "congenital facial diplegia" and bilateral abducens palsies. Möbius syndrome, as it was later named, remains one of the most complex and difficult surgical problems facing the strabismus specialist.

Because of the ophthalmic and systemic abnormalities characterizing Möbius syndrome, case reports are scattered throughout the literature of several disciplines. In addition, what an ophthalmologist recognizes as Möbius syndrome is oftentimes termed Hanhart syndrome or Poland-Möbius syndrome by other specialists. In an extensive review of 69 cases, Herrmann and associates attempted to organize this multispecialty syndrome and suggested that cases without extensive limb abnormalities, consisting primarily of cranial nerve involvement, be categorized as Möbius syndrome, whereas those with extensive limb abnormalities would be grouped under Hanhart syndrome. As a minimal diagnostic criterion, all cases must involve sixth and seventh cranial nerve dysfunction.

Pathogenesis

Although the precise pathogenesis of Möbius syndrome is unclear, there is general agreement as to the timing of the syndrome and the factors that trigger it. The pathologic insult appears to take place during the first trimester (fourth to sixth weeks of gestation) when the cranial nerve nuclei undergo their most rapid development. Several events at this stage, including trauma, illness, and the ingestion of specific medications (benzodiazepines or thalidomide), have been associated with later development of the syndrome. Whether the syndrome represents primary hypoplasia of the affected cranial nerve nuclei or a peripheral neuromuscular abnormality resulting in secondary brain stem atrophy has yet to be determined conclusively.

A recent theory, which has gained some acceptance, implicates a vascular etiology of Möbius syndrome. The so-called subclavian artery supply disruption sequence theory (SASDS) implicates an interruption of the embryonic blood supply during the sixth week of gestation. This theory is supported by the histologic observation of symmetric calcification (in a vascular distribution) of the dorsal tectum at the junction of the midbrain and pons. The vascular insult presumably causes hypoxia and ischemia of the affected neural tissues, including the cranial nerve nuclei.

In fact, Miller and Strömland propose renaming the Möbius syndrome the Möbius sequence owing to the likely presence of a single embryonic insult leading to the well-known cascade of secondary events.

Genetics

Recent studies point to a deletion on the long arm of chromosome 13 in Möbius syndrome. Whether this is an inherited defect or occurs early in gestation as a result of a trigger event has not been determined.

Viewed in the light of classic genetics, there appears to be a recurrence rate in siblings of approximately 2% in typical isolated cases. The syndrome affects both sexes equally and with the same frequency. Although most cases appear to be sporadic, pedigrees exhibiting autosomal dominant, autosomal recessive, and X-linked inheritance have been reported.

Clinical Characteristics

The diagnosis of Möbius syndrome is categorically made in the presence of sixth (abducens) and seventh (facial) cranial nerve palsies. Although rarely unilateral, these palsies are often incomplete bilaterally. Involvement of the sixth cranial nerve leads to loss of ocular abduction and esotropia. Seventh cranial nerve palsy causes facial weakness. The facial paresis, if partial, usually involves the upper division of the nerve. Of note is a lack of facial tissue sagging, which is often observed in acquired cases of seventh cranial nerve paralysis. It is the lack of facial nerve function that causes the distinctive "masklike" facies of Möbius syndrome.

The diagnosis is often suspected early in life from diffi-
### Table 26–1. Systemic Findings in Möbius Syndrome

<table>
<thead>
<tr>
<th>Abnormalities of extremities</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Syndactyly</td>
<td></td>
</tr>
<tr>
<td>Polydactyly</td>
<td></td>
</tr>
<tr>
<td>Brachydactyly</td>
<td></td>
</tr>
<tr>
<td>Agenesis of digits</td>
<td></td>
</tr>
<tr>
<td>Clubfoot</td>
<td></td>
</tr>
<tr>
<td>Talipes equinus varus</td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>swallow and speech difficulties</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Paralysis of V, IX, and X cranial nerves</td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Craniofacial abnormalities</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Micrognathia</td>
<td></td>
</tr>
<tr>
<td>Microstomia</td>
<td></td>
</tr>
<tr>
<td>Structural abnormalities of ears</td>
<td></td>
</tr>
<tr>
<td>Bifid uvula</td>
<td></td>
</tr>
<tr>
<td>Cleft palate</td>
<td></td>
</tr>
<tr>
<td>VII cranial nerve palsy</td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Defects</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Dextrocardia</td>
<td></td>
</tr>
<tr>
<td>Defective branchial musculature</td>
<td></td>
</tr>
<tr>
<td>Missing pectoral and trapezius muscle groups</td>
<td></td>
</tr>
<tr>
<td>Absence of sternal head of pectoralis major</td>
<td></td>
</tr>
<tr>
<td>Rib defects</td>
<td></td>
</tr>
<tr>
<td>Tongue hypoplasia (25% of cases)</td>
<td></td>
</tr>
<tr>
<td>Mild mental retardation (at least 10% of cases)</td>
<td></td>
</tr>
</tbody>
</table>

Facility in sucking, drooling of saliva, and incomplete closure of the eyelids during sleep (lagophthalmos). The characteristic facies and a lack of smiling in response to familiar stimuli occur later and confirm the diagnosis.

Many systemic abnormalities have been noted (Table 26–1). Notable among these are additional cranial nerve abnormalities. Involvement of the V, IX, X, and XII cranial nerves may lead to significant problems in nutrition and speech development. Multiple craniofacial abnormalities may adversely affect oral function as well as facial structure.

The ocular characteristics of Möbius syndrome are both diverse and significant (Table 26–2). Certainly, the most common presentation is esotropia, with decreased function of one or both lateral rectus (LR) muscles. Because of poor to absent abduction ability, a face turn may sometimes be noted in primary gaze and when the patient attempts to gaze laterally. With a nonfunctioning LR muscle, there may be a large-angle esotropia; angles up to 100 PD have been observed. Vertical eye movements and convergence ability are usually preserved. When the third and fourth cranial nerves are involved, a vertical or torsional deviation may be present.

Exposure keratitis due to lagophthalmos secondary to a dysfunctional orbicularis oculi muscle (supplied by the seventh cranial nerve) is one of the most serious complications of Möbius syndrome. Although Bell’s phenomenon is preserved, serious corneal exposure is often encountered. Varied craniofacial abnormalities have been noted, including hypertelorism, epicanthal folds, and small palpebral fissures. Ptosis and entropion have also been described. All these abnormalities affect the quality of blinking and contribute to disorders of corneal lubrication.

### Diagnosis

**OPHTHALMOLOGIC EXAMINATION**

The physical examination begins by observing the patient’s head position. Occasionally, face turns are adopted to maintain single binocular vision. Most patients, however, will not exhibit an anomalous head posture, owing to the bilaterality of the abducens palsy and a lack of binocular vision.

Visual acuity must be carefully measured. Clearly, in the face of a constant deviation, patients with Möbius syndrome are at great risk of suppression and amblyopia. The amblyopia may be either strabismic or ametropic and should be treated early and aggressively. The likelihood of developing normal binocular vision may be greatly reduced.

Evaluation of the external ocular structures, including the lids and lacrimal system, is also important. The quality of Bell’s phenomenon should be evaluated. Lagophthalmos, a poor Bell’s phenomenon, and inadequate tear distribution may impair conjunctival healing and promote corneal damage after strabismus surgery. The pupillary reflexes are usually intact. The anterior segment must be evaluated for corneal surface disease.

After administration of a cycloplegic agent, a mild to moderate refractive error is often uncovered. Several abnormalities of the posterior segment may be evident on fundus examination. Patients with Möbius syndrome may have tortuous retinal vessels and situus inversus of the retinal vasculature. Additionally, should a fourth cranial nerve palsy be present, torsion may be apparent when observing the optic nerve head in relation to the foveal region.

**STRABISMUS EVALUATION**

The angle of deviation must be measured diligently. When formulating a surgical treatment plan, attention to LR function as well as medial rectus (MR) contracture is crucial to ensure maximal success.

Several specialized tests may aid in evaluating patients with Möbius syndrome. Partial function of one or both LR muscles may be present and may be estimated by force generation testing. Forced duction testing will reveal MR contracture, which is often present in patients with large-angle or long-standing esotropia. Saccadic velocity analysis also helps to detect residual LR function (see Chapter 3). Unfortunately, because these patients often are young and have underlying medical conditions, not all these tests can
be performed in the office setting. Forced duction testing may be done immediately before planned surgery under anesthesia.

**DIFFERENTIAL DIAGNOSIS**

In addition to Hanhart syndrome, which shares the cranial nerve features of Möbius syndrome with the associated limb abnormalities, certain ophthalmic and neuromuscular conditions must be considered.

Ophthalmic conditions that simulate Möbius syndrome share its ocular cranial nerve characteristics but not the associated facial and systemic signs. For example, congenital sixth cranial nerve palsy, congenital (infantile) esotropia, and (rarely) Duane syndrome all may include large-angle esotropia with limited LR function, but none of these is associated with seventh cranial nerve palsy. Congenital horizontal gaze palsy is associated with decreased abduction and may also include esotropia. Ocular motor apraxia may resemble Möbius syndrome because of decreased voluntary horizontal gaze.

Neuromuscular conditions such as facioscapulohumeral muscular dystrophy, infantile myotonic dystrophy, Charcot-Marie-Tooth disease, and congenital myotonic dystrophy are part of the differential diagnosis. In many cases, collaboration between a pediatrician and strabismus specialist will be needed to definitively distinguish these conditions from Möbius syndrome.

---

**Figure 26-1.** Composite photograph of patient with Möbius syndrome. Upper series demonstrate preoperative primary position, left and right gazes. Note decreased abduction bilaterally and moderate sized esotropia (25–30 PD). Lower series display the same positions after three stages of surgical correction. Bilateral medial rectus recession of 6.5 mm was followed by a left vertical rectus transposition procedure. An identical procedure was performed on the right side 8 months later.

**Figure 26-2.** Patient with Möbius syndrome. Top row, Photographs taken at age 2 showing 60 PD of esotropia. There was no effective abduction of either eye. Middle row, Photographs taken after transposition procedure on both vertical rectus muscles with posterior fixation suture augmentation in both eyes. The patient still showed 30 PD of esotropia with no significant improvement of abduction in either eye. Subsequently, at age 3.5 years, she underwent a second surgery. Both medial rectus muscles were found to be tight and were recessed. Bottom left, Photograph shows excellent alignment after this procedure. Bottom center, Late overcorrection seen at age 10 with 20 PD of right esotropia and a small hypotropia. Her right medial rectus was advanced to its insertion. Although she had only 4 PD of exophoria in primary gaze, there was an X-pattern deviation of 30 PD exotropia in upgaze and 20 PD of exotropia in downgaze (not shown). Bottom right, Recurrence of large exodeviation by age 12 even in primary gaze. Currently, we favor pharmacologic weakening of the medial rectus muscle over surgery. (Courtesy of R. S. Foster, MD.)
Treatment

NONSURGICAL MANAGEMENT

The nonsurgical treatment of these patients entails the detection and treatment of amblyopia, corneal exposure, and refractive errors. Certainly a majority of patients with Möbius syndrome are initially evaluated by a strabismus specialist during the early years of life. In this setting, it is crucial to identify and treat suppression and strabismic amblyopia. By minimizing ocular suppression and surgically correcting strabismus, the patient will have the best chance of developing low-grade binocular vision. Refractive errors and a lack of corneal clarity may exacerbate strabismic amblyopia. Besides lagophthalmos and entropion, corneal surface disease may be aggravated by an anesthetic cornea secondary to fifth cranial nerve dysfunction.

STRABISMUS MANAGEMENT

The management of strabismus in patients with Möbius syndrome is both complex and multifaceted. No single surgical approach is successful in all patients. Instead, a set of surgical guidelines has been proposed that must be applied individually to optimize the surgical outcome. The surgeon and family must have realistic expectations from the surgery to achieve a high degree of satisfaction. In many cases surgery will achieve optimal alignment only in primary gaze, with manifest esotropia persisting on lateral gaze.

Before formulating a specific plan, the surgeon must perform forcedduction testing on each eye to identify a tight MR muscle. If such is the case, transposing the vertical rectus muscles to the insertion of the LR may not suffice. On the other hand, if the globe can be freely rotated into abduction, indicating no or only minimal restriction, a bilateral transposition procedure alone may be adequate. A staged surgical approach has been developed in which the vertical rectus muscles are initially transposed to the LR insertions, followed by graded MR weakening (either by recessing the muscle or injecting botulinum toxin for any remaining esodeviation). Using this approach, the surgeon maintains maximal control over the effect of each “abducting procedure” and is able to vary the second procedure depending on the effects of the initial one (Fig. 26–1).

Experience has shown that standard MR recession alone is rarely effective in this condition. This is most likely explained by a lack of abduction ability and by the frequently observed secondary MR muscle contracture. Augmented recessions of up to 8 to 9 mm have been attempted. Although they do not cause overcorrection, they often fail to align the eyes in primary position. In cases of large-angle esotropia (more than 50 PD) with restricted abduction on forcedduction testing, a large MR muscle recession will often be required after initial transposition surgery (Fig. 26–2). When the MR muscles are not contracted, the vertical rectus transposition procedure will have more of an abducting effect.

Botulinum toxin injection of the MR has been proposed as an effective adjuvant to surgical therapy. In young children, who have not yet developed contracture of the MR, botulinum toxin in combination with vertical rectus transposition surgery can be an effective alternative to classic strabismus surgery (Fig. 26–3). In cooperative older patients, staged surgery may be avoided by taking advantage of an adjustable surgical technique. As described in a recent report, adjustable vertical rectus transpositions in adults allow gradation of the abducting effect of the transposition and correct any inadvertent vertical strabismus caused by asymmetric transpositions. A similar adjustable technique may be used on the MR muscle to postoperatively adjust the weakening effect of the recession. Unfortunately, because of the young age of most of these patients, an adjustable technique is usually not possible.

COMPLICATIONS

Möbius syndrome can influence all aspects of surgical strabismus correction. Preoperative evaluation of the precise angle of strabismic deviation may be complicated by factors ranging from epicanthal folds to difficulty in speech and lack of cooperation.

During the surgical procedure itself, the anesthesiologist may be challenged by increased oral secretions that interfere with intubation and extubation and may contaminate the surgical field. Cleft palate and other craniofacial abnormalities may complicate an otherwise routine anesthetic procedure.

The surgeon may not recognize hypoplasia, aplasia, fibrous bands, or abnormal muscle insertions until the extraocular muscles are isolated. Although these findings do not alter the surgical plan, they can make an already complicated case even more challenging.

Figure 26-3. Preoperative and postoperative photographs in the primary position of a patient with Möbius syndrome. A two-staged approach was used: medial rectus muscles were weakened with botulinum toxin, followed by a bilateral vertical rectus muscle transposition procedure. (Courtesy of Brian W. Fleck, MD.)
Figure 26–4. Preoperative and postoperative photographs in the primary position of a patient with Möbius syndrome. This patient presented with large-angle esotropia and underwent a combined bilateral medial rectus recession of 7.0 mm as well as bilateral vertical rectus muscle transposition. Although alignment seems acceptable at first glance, examination revealed 30 PD of exotropia.

Postoperative wound healing may be affected by ocular surface disease resulting from lagophthalmos and an inadequate blink mechanism. Ocular alignment is often adequate after the staged approach, although late overcorrections have been observed (Figs. 26–2 and 26–4). Fortunately, most of these are relatively small and do not require additional surgery. When maximal MR weakening is performed at the same time as vertical rectus transposition, a large overcorrection often develops that requires reoperation (see Fig. 26–2).^8

Conclusions

Although Möbius syndrome is a multisystemic, whole-body disorder, the majority of signs and symptoms involve the face and ocular system. By carefully applying a set of treatment principles, both the strabismic and nonstrabismic manifestations of this syndrome can be managed. Patients who undergo surgical correction often are able to lead a nearly normal visual life with only minor cosmetic abnormalities.

REFERENCES

One of the earliest descriptions of the syndrome of ptosis and restricted ocular movements was given by Baumgarten in 1840. The first account of the familial occurrence of this condition is attributed to Heuck, who also provided postmortem findings in one patient. The term "general fibrosis syndrome" was coined by Brown to describe patients having fibrosis of three or more extraocular muscles.

Other names given to this disease include congenital fibrosis of the extraocular muscles (CFEOM), congenital ocular fibrosis, generalized fibrosis of the extraocular muscles, congenital fibrosis syndrome, congenital ocular fibrosis syndrome, syndrome of generalized fibrosis, and congenital ophthalmpoplegia. Still other terms include hereditary congenital ophthalmpoplegia, ophthalmpoplegia imperfecta, abiotrophic ophthalmpoplegia externa, congenital external ophthalmpoplegia with co-contraction, and autosomal dominant congenital external ophthalmpoplegia.

**Epidemiology and Classification**

The prevalence of congenital fibrosis syndrome is unknown. Families of various ethnic and racial origins have been reported around the globe. The generalized form of CFEOM is probably the type most commonly encountered in clinical practice. Harley and colleagues divided patients with CFEOM into those having (1) generalized fibrosis, (2) fibrosis of the inferior rectus with blepharoptosis, (3) strabismus fixus, (4) vertical retraction syndrome, and (5) congenital unilateral fibrosis. The clinical significance of this classification is not evident; although most patients may be allocated to individual categories, overlap exists between the different types.

**Genetics**

In most instances, CFEOM is inherited in an autosomal dominant manner. There is significant variability in expression of the gene, resulting in a phenotype that is more severe and symmetric in some family members than in others. We have observed three Saudi families with consanguineous marriages in which siblings or first cousins were affected, whereas the parents themselves showed no evidence of the disease. CFEOM in these families is inherited as an autosomal recessive trait. The gene for autosomal dominant CFEOM was mapped to chromosome 12 by Engle and associates in a total of seven families; but the gene itself has not yet been isolated, and its function remains unclear. There is evidence that it is involved in the development of neurons of the third, and possibly other, cranial nerve nuclei. Some patients who also have Prader-Willi syndrome are suspected of having a form of CFEOM that resides in the long arm of chromosome 15. No chromosomal abnormalities have been described in patients with CFEOM.

**Mechanisms**

Both myopathic and neurogenic mechanisms of CFEOM have been postulated. Recent evidence, however, supports the view that neurologic defects are primary. Chromosome 12-linked CFEOM is believed to result from an abnormality of the motor neurons, axons, and muscles supplied by the superior division of the trigeminal nerve. Absence of the superior division of the oculomotor nerve and its corresponding alpha motoneurons was demonstrated in one patient. Three had generalized muscle abnormalities, indicating possible primary muscle pathology. The association of neural misdirection with CFEOM led some authors to suggest that, in congenital fibrosis syndromes, a primary developmental defect precludes the establishment of normal neuronal connections.

**Clinical Characteristics**

The four components of the congenital fibrosis syndrome as described by Laughlin are (1) blepharoptosis and chin...
elevation; (2) histopathologic confirmation of fibrosis of the extraocular muscles; (3) absence of elevation or depression of the globes, with the eyes fixed 20 to 30 degrees below the horizontal meridian; and (4) little or no horizontal movement. We would add the absence of Bell’s phenomenon as a fifth feature of this disease. This results from mechanical restriction of elevation of the globe and is a major consideration when planning ptosis surgery. There is wide variation in the number of muscles involved and in the extent to which extraocular movements are impaired.

Bilateral disease may be very symmetric or so asymmetric as to be labeled unilateral. A number of such patients with unilateral disease have been described, in some of whom fibrosis was attributed to prenatal orbital inflammation or prenatal penetrating orbital injury by fetal body parts.

Patients with fibrosis of the extraocular muscles are generally healthy except for rare ocular or systemic problems in sporadic cases or single family members of otherwise typical pedigrees. The associated ocular findings may include bilateral optic nerve hypoplasia, a chorioretinal coloboma, microphthalmia, ocularcutaneous albinism, neural misdirection with synergistic divergence, globe retraction, the Marcus Gunn jaw-winking phenomenon, and enophthalmos. Optic nerve head dysplasia/coloboma was present in all affected members of one family reported by Khawam and colleagues. This family, however, may have a syndrome distinct from classic CFEOM.

Reported systemic associations include bilateral inguinal hernias and unilateral cryptorchidism, a benign mesenchymoma of the ethmoid sinus that invaded the orbit, and Joubert syndrome with or without histidinemia. Patients with the Prader-Willi syndrome and fibrosis of the horizontal muscles have been described. Patients with unilateral fibrosis of the extraocular muscles may have a smaller face on the same side and deformities of the orbital walls. One of the patients reported by Traboulsi and co-workers had arthrogryposis at birth that improved over time.

Diagnosis

CLINICAL DIAGNOSIS

Patients with generalized CFEOM have blepharoptosis and a chin-up posture. The eyes are fixed in downgaze (Fig. 27–1A and B). Ductions are severely limited (Figs. 27–1C and 27–2B). Some patients may adopt a face turn when the best eye alignment is in horizontal gaze (see Fig. 27–2A). Amblyopia is common but not universal; half the patients reported by Traboulsi and co-workers were not amblyopic. Patients occasionally have visual acuity of 20/20, but most do not. Hyperopic and astigmatic refractive errors are present in about 60% of patients. A change in refractive error that occurs in some patients after strabismus surgery may result from a redistribution of forces exerted on the globe by the stiff rectus muscles.

Progressive development of an A pattern was noted in eight patients with CFEOM. The increased esotropia on attempted upgaze was postulated to result from abnormal supranuclear innervation to the extraocular muscles. In one of our patients who had an A pattern, we ascribed it to excessive convergence on attempted upgaze, possibly resulting from the secondary adductive effect of the superior rectus muscles.

Although ptosis is common in patients with CFEOM, it rarely may be absent. Some patients may exhibit pseudoptosis because of a hypotropia (Fig. 27–3). Drooping of the lid may occasionally be very severe. Patients with marked ptosis may use one finger or the dorsum of the hand to elevate an eyelid so as to clear the visual axis (Fig. 27–4). The fellow eye is usually amblyopic. We propose to call this maneuver the digitoralpebral sign.

Upper lid retraction also may be observed. Transient periorbital swelling and ecchymosis have been described, but their etiology or significance could not be determined. Enophthalmos may develop with advancing age secondary to growth of the bony orbit and/or fibrosis of the extraocular muscles and orbital tissues, resulting in retraction of the globe.

Fusion varies greatly, but 40 seconds of arc near stereocuity may be demonstrated.

LABORATORY EVALUATION

Orbital computed tomography is recommended for investigating children with unilateral fibrosis but may not be helpful in generalized CFEOM. Imaging studies may demonstrate atrophy, absence, abnormal insertion, or “thickening” of the extraocular muscles (Fig. 27–5). Histopathologic studies in CFEOM do not reveal any characteristic abnormalities. There is fibrous infiltration of the extraocular muscles and orbit. In one autopsy case, a posterior membranous insertion of all the rectus muscles and an anterior and medial insertion of the superior and inferior oblique muscles were observed. Also documented, besides generalized extraocular muscle abnormalities, have been a qualitative decrease in the number of alpha motoneurons in all oculomotor nuclei and in the abducens nucleus, a decreased diameter of the inferior division of the third nerve, and an increase in the number of internal nuclei within myofibers of all the extraocular muscles. Because of the occasional presence of mild facial weakness, hypotonia, gross motor delay, and nonspecific abnormalities in quadriceps biopsy specimens from affected family members, it has been proposed that CFEOM plays at least a transient role in normal skeletal muscle development.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis of CFEOM includes double elevator palsy, partial or complete third cranial nerve palsy, Brown syndrome, progressive external ophthalmoplegia, Möbius syndrome, and Kearns-Sayre syndrome. It also includes myasthenia gravis, orbital periostitis, entrapment in an orbital wall fracture, olivopontocerebellar degeneration with retinal degeneration, and atypical Duane syndrome. Forced duction testing may be necessary to differentiate double elevator palsy from unilateral inferior rectus fibrosis. Many of these conditions are described elsewhere in this text.
Figure 27-1. A, Eleven-year-old boy with esotropia, hypotropia, and a chin-up posture to clear visual axis. B, Lateral view of his similarly affected 9-year-old brother shows chin-up posture. C, Composite of ocular rotations in cardinal and four directions of gaze demonstrate minimal abduction and depression of both eyes. Note better vertical ductions in the right eye.
Figure 27–2. A, Six-year-old with bilateral, but asymmetric, fibrosis of extraocular muscles, simulating a right sixth cranial nerve paresis and a left third cranial nerve palsy. The patient assumes a right head turn in an attempt to align the right fixing eye in primary position. The left eye was amblyopic with an acuity of 20/200. B, Primary position shows exotropia with left hypotropia and ptosis. Right eye shows slight weakness of abduction more evident in upgaze and downgaze. Left eye rotations are extremely limited in all gaze fields except abduction.
Figure 27-3. Twelve-year-old girl with right hypotropia and pseudoptosis in primary position of gaze, secondary to right inferior rectus muscle fibrosis. Stereopsis: 100 seconds of arc. Upgaze deficiency was more pronounced in abduction than adduction in the right eye. Recession of right inferior rectus muscle resulted in orthotropia in primary position and elimination of pseudoptosis (not shown).

Figure 27-4. The digitopalpebral sign. Three first cousins age 3 years (right), 2 years (middle), and 1½ years (left) with congenital fibrosis of extraocular muscles, using finger, thumb, or dorsum of hand to elevate lid and clear visual axis. These patients were the products of consanguineous marriages and demonstrate the autosomal recessive inheritance of congenital fibrosis of the extraocular muscles in some families.
Treatment

NONSURGICAL MANAGEMENT

The nonsurgical management of patients with CFEOM consists of optically correcting refractive errors and treating amblyopia. Amblyopia is more prevalent in asymmetric cases and in patients with anisometropia. Patching and equalization of vision in the two eyes should be instituted before surgical correction. Symmetrically good visual acuities can be achieved. Repeated refractions are necessary because of the potential for a significant change in refractive error to contribute to amblyopia, especially after strabismus surgery.

Because of the absence of a Bell’s phenomenon, some patients with CFEOM and incomplete lid closure may require ocular surface lubricants at bedtime.

A minority of patients with CFEOM may not need surgery if there is no significant abnormal head posture and if the visual axis is not occluded (Fig. 27–6). Older patients with strabismus fixus may not be interested in surgery or may be at high risk of complications (Fig. 27–7).

SURGICAL MANAGEMENT

The surgical management of patients with generalized CFEOM is difficult, but relatively good results may be expected in most patients. The goals of surgery are to remove obstacles to the visual axis in primary gaze, alleviate the chin-up posture, and align the eyes in primary position.
Ideally, strabismus is corrected first and then ptosis. Rarely the chin-up head posture is relieved completely after resection of the inferior rectus muscles, without the need for ptosis surgery. In most cases, however, extraocular muscle and frontal suspension procedures must be combined to achieve an acceptable functional result (Fig. 27–8).

Individual consideration should be given to the muscles to be operated on and to the required amounts of recession or resection. These choices are based on ocular alignment in primary gaze, ocular rotations, and forced duction testing. Because of the involvement of orbital tissues and conjunctiva, conjunctival recession may be beneficial.

Several authors have emphasized the need for a large recession or even free tenotomy of the involved muscles. Recession of the conjunctiva and the use of traction sutures are also helpful. Despite the reluctance of some strabismologists to use resection in patients with CFEOM, from fear of worsening enophthalmos, many surgeons have found it necessary to add resection of the superior rectus or lateral rectus muscle to recession of the inferior and medial rectus muscles to achieve proper alignment. We have not found the adjustable suture technique to be particularly helpful in management of the ocular fibrosis syndrome.

We rely heavily on the forced duction test, both preoperatively and throughout the surgical procedure. Large recessions are usually required. Careful, extensive posterior dissection of the rectus muscles from intermuscular septal attachments is essential. The use of two muscle hooks (instead of one) to preplace absorbable sutures and disinsert the muscle may be of great help when operating on tight muscles. Some surgeons may prefer a hang-back suture technique for large resections. However, it is feasible to resuture the muscle to the posterior sclera. We achieved good results in a majority of our patients by using large resections of the fibroed rectus muscles, regardless of whether the conjunctiva was recessed and/or the antagonist muscle resected. Exploration of the superior rectus and oblique muscles may disclose insertional abnormalities that, if treated, will improve the outcome. Stereopsis can be achieved or maintained in some patients.

Aberrant circulation has been described. Since as many as three rectus muscles in one eye may need to be operated on in a single setting, care should be taken to closely observe all patients, even the younger ones, for signs of anterior segment ischemia.

A conservative fascial lata (autologous or banked) frontalis sling procedure is still the mainstay of ptosis repair in CFEOM. Extreme care should be taken to avoid overcorrecting the ptosis. This could result in corneal exposure, especially in patients who lack a Bell’s phenomenon and rapid eye movements. The generous and prolonged use of lubrication should be prescribed postoperatively.

Conclusions

The patient’s, family’s, and surgeon’s expectations of postoperative alignment should be realistic and thoroughly discussed. A clear statement about the possibility of limited ocular motility persisting is also in order. And lastly, the approach to managing each patient, even in the same family, should be individualized. Because the phenotypic expressivity of CFEOM is so variable, so must be our therapeutic approach.

REFERENCES

strabismus with presumed fibrosis of the extraocular muscles. In Rein- 
ecke RD (ed): Strabismus II. Proceedings of the Fourth Meeting of the 
12. Harley RD, Rodrigues MM, Crawford JS: Congenital fibrosis of the 
sis, blepharoptosis, and enophthalmos syndrome. Ophthalmology 
diagnosis of the congenital fibrosis syndrome. J Clin Neuroophthalmol 
17. Jacobson DM, Johnson R, Frens DB: Joubert’s syndrome, ocular fibro-
113:714.
syndrome associated with the Prader-Willi syndrome. J Pediatr Ophthal-
mol Strabismus 1986;23:170.
ophthalmoplegia with optic nerve coloboma: Report of a family. Binoc-
20. Laughlin RC: Congenital fibrosis of the extraocular muscles. Am J 
Ophthalmol 1956;41:432.
21. Leone CR, Weinstein GW: Orbital fibrosis with enophthalmos. Ophthal-
22. Letson RD: Surgical management of the ocular congenital fibrosis 
23. Prakash P, Menon V, Ghosh G: Congenital fibrosis of superior rectus 
24. Rumpf M: Fibrose du muscle droit inférieur, anomalies d’insertions 
et aplasies musculaires: Une cause rare des troubles heréditaires et 
25. Traboulsi EI, Jaafar MS, Kattan HM, et al: Congenital fibrosis of the 
extraocular muscles: Report of 24 cases illustrating the clinical spect-
26. Vossius A: Zwei Falle von angeborenen fast vollständiger Unbeweglich-
keit beider Augen und der oberen Augenlider. Beitr Augenheilkd 
1982;5:1.
27. Wilder WM, Williams JP, Hupp SL: Computerized tomographic find-
ings in two cases of congenital fibrosis syndrome. Comput Med Imag 
Strabismus after uncomplicated cataract surgery is much better understood now than it was a decade ago. Changes in surgical techniques and heightened patient expectations have recently brought this group of disorders into prominence. There also has been a qualitative shift in emphasis on the potential underlying causes. In the 1950s, 1960s, and 1970s, the ophthalmic literature focused on the effects on fusion of the aniseikonia and anisophoria that accompany the correction of monocular aphakia with spectacles or contact lenses. It was hoped that the advent and widespread use of intraocular lenses would restore binocular function in patients undergoing cataract surgery and put this problem to rest. Instead, fusion difficulties, strabismus, and diplopia have received even more recognition and more extensive coverage in the literature in the 1980s and beyond. Problems other than anisophoria/aniseikonia have prevailed.

Surgical trauma and anesthetic toxicity to the extraocular muscles have received particular attention and currently are the most important causes of strabismus that occurs after adult cataract surgery.

The focus in this chapter is on persistent binocular diplopia after cataract surgery. Transient binocular diplopia is more common but is less important clinically because it generally resolves within a few days to weeks. During this time, patients are usually preoccupied with their improved vision and other postoperative concerns and generally do not report binocular difficulties. Possible causes of transient diplopia include small, preexisting, fusible sensory strabismus; a prolonged local anesthetic effect or transient toxicity; a fusible prismatic effect of spectacle overcorrection; and operative trauma to the orbital soft tissues. Monocular diplopia may arise from corneal epithelial irregularities, cataract in the other eye, uncorrected ametropia, intraocular lens decentration, and polycoria (e.g., exposed peripheral iridectomy and iridodialysis). Some patients exhibit both monocular and binocular diplopia. This combination may cause confusion and go undetected by the clinician for some time.

The various causes of strabismus after cataract surgery are discussed as though each is solely responsible for the patient’s binocular difficulties. Cataract surgery, however, results in numerous effects that may contribute to abnormal fusion and can “tip the scale” against normal fusion in borderline cases. The general principle is that symmetric sensory input from both retinas is required for normal fusion and that cataract surgery may, in many ways, compromise this symmetry. The possibilities include anisophoria, aniseikonia, alterations in refractive power, brightness and color disparity between the phakic and operated eyes, optically induced glare and halos, changes in depth-of-focus caused by surgical alterations in pupillary size and shape, and a disordered pattern of ocular dominance that may cause fixation-switch diplopia.

**Etiology**

The causes of persistent strabismus after cataract surgery are numerous. The clinical features depend largely on the underlying etiology. Most patients can be readily assigned to one of four broad categories (see Table 28–1), although some will be difficult to pigeonhole. A number of factors deleterious to fusion can coexist in a single patient; not all patients have a single cause for their strabismus.

**PREEXISTING HETEROTROPIA OR HETEROPHORIA MASKED BY THE CATARACT**

Thyroid eye disease is the prototype for this category. Patients usually have the clinical signs and symptoms of thyroid eye disease and exhibit diffusely enlarged extraocular muscles on orbital neuroimaging (Fig. 28–1). However, the diagnosis can sometimes be difficult to establish in the absence of classic signs or when thyroid ophthalmopathy is minimal. The diagnosis is generally one of exclusion. Subtle cases of thyroid eye disease should not be confused with cases caused by surgical trauma to soft tissues of the orbit or by local anesthetic myotoxicity. Myotoxicity of a local...
Table 28-1. Persistent Binocular Diplopia after Cataract Surgery: Four Broad Etiologic Categories

Preexisting or Concurrent Disorders Masked by the Cataract
- Thyroid eye disease
- Myasthenia gravis
- Childhood strabismus, amblyopia
- Foveal dystopia
  - Cellophane maculopathy
  - Subretinal neovascular membrane
- History of macular laser photocoagulation
- Ocular motor nerve palsy
- Previous damage to orbital soft tissue (e.g., retinal reattachment surgery)
- Intraoperative neurovascular events
- Other causes of strabismus in the elderly

Disorders Caused by Prolonged Occlusion by the Cataract
- Sensory strabismus
- Diminished fusional reserves with decompensation of heterophoria
- Central disruption of binocular vision
- Disorders unique to amblyopia
  - Elimination of suppression in amblyopic eye with intractable diplopia
  - Change in strabismus angle with drift outside the suppression scotoma
  - Fixation switch diplopia

Anesthetic Myotoxicity, Surgical Trauma
- Extraocular muscle contracture
- Extraocular muscle overaction
- Extraocular muscle paresis

Optical Aberrations Associated with Aphakia and Pseudophakia
- Anisophoria
  - Anisometropic spectacle correction
  - Intraocular lens decentration
  - Uneven segment height
- Aniseikonia
  - Induced anisometropia in an isometrope
  - Induced isometropia in a previous anisometrope

- Brightness and color disparity between phakic and operated eye
- Optically induced glare and halos
- Changes in depth of focus by surgical pupillary alterations
- Disturbance of pattern of ocular dominance
- Fixation switch diplopia

such a patient is typically exotropia, hypotropia, and ex-cyclodeviated (Fig. 28–2). It is important to test fusion in these patients on the major amblyoscope to correct for any torsional deviation before stipulating that central fusion is disrupted. The authors caution that such patients fail to improve with strabismus surgery and, in fact, their symptoms may worsen because of the proximity of the two images.

The clinician is admonished to discuss the possibility of diplopia with patients who have had their fusion interrupted by a traumatic cataract or by uncorrected aphakia for more than 2 years. A corollary to this principle is the need for timely removal of traumatic cataract to prevent loss of fusion. Some of these patients may reacquire fusion over time. A trial of glasses with prisms for at least several months is therefore recommended before surrendering to monocular occlusion.

The clinician should inquire about a history of childhood strabismus or amblyopia. These conditions may dispose to the emergence of diplopia after cataract surgery through one of three possible mechanisms: (1) Prolonged occlusion by a

anesthetic may cause segmental enlargement of one extraocular muscle (often the inferior rectus). To confound the picture, some cases may overlap: thyroid eye disease that was subclinical preoperatively may become overt after surgery, not only because visual improvement unmasks diplopia but also because the enlarged muscles are susceptible to direct injury by retrobulbar anesthesia.

DISORDERS CAUSED BY PROLONGED OCCLUSION BY THE CATARACT

Sensory strabismus from cataracts is becoming increasingly rare as lenses are removed well before vision is severely reduced. It is still encountered in patients with long-standing unilateral traumatic cataract. Pratt-Johnson and Tillson reported 24 patients with intractable diplopia after removal of unilateral traumatic cataracts. The cataracts had been present for at least several years, presumably causing central disruption of binocular vision. The affected eye in

Figure 28-1. Patient with thyroid eye disease presenting after cataract surgery on the right eye. On orbital imaging, all extraocular muscles were diffusely enlarged.

Figure 28-2. Patient with intractable diplopia after removal of long-standing left traumatic cataract. The eye is typically exotropic, with some hypotropia and ex-cyclodeviation.
cataract in an amblyopic eye may eliminate suppression and lead to subsequent intractable diplopia.\(^5\) (2) The angle of strabismus may change while the cataract is present, displacing the visual axis to retinal points outside the suppression zone and resulting in diplopia after cataract removal. (Piriform compensation to align the visual axis into the scotomatos area eliminates diplopia in these patients.) (3) Switching fixation to the ambyopic eye after cataract surgery may lead to fixation-switch diplopia. The cataract surgeon should counsel patients having a history of childhood strabismus and/or amblyopia about the risk of fusion difficulties postoperatively.

Patients with a previously compensated heterophoria such as esophoria, exophoria, or long-standing superior oblique palsy may experience diplopia after cataract surgery.\(^6\) \(^7\) \(^8\) The "supranormal" fusional reserves possessed by patients with congenital superior oblique palsy can diminish from the cataract occlusion. Their ability to fuse may be compromised further by asymmetric sensory input from the phakic and aphakic eyes. Before concluding that the patient has a long-standing superior oblique palsy, the clinician should seek corroborative evidence such as head tilting in old photographs. Intraoperative vascular events and injury to the superior oblique/trochlea complex from the retrobulbar injection are alternative explanations.

### Surgical Trauma and Anesthetic Toxicity

Some experts suspect that this category of patients is growing. This is debatable, but what is certain is an increase in reports in the literature. Historically, strabismus of this type has been attributed to bridle-suture trauma to the superior or inferior rectus muscle, trauma to muscles or related nerves from the retrobulbar needle, ischemic injury of the muscle from swelling and hematoma formation, and an inflammatory reaction to subconjunctival gentamicin.\(^9\) The "supranormal" fusional reserves possessed by patients with congenital superior oblique palsy can diminish from the cataract occlusion. Their ability to fuse may be compromised further by asymmetric sensory input from the phakic and aphakic eyes.

Most authors now believe that strabismus is mediated by an adverse effect to retrobulbar administration of local anesthetic.\(^5\) \(^9\) Both local anesthetic myotoxicity and local trauma to the muscles, orbital nerves, and other soft tissues undoubtedly are factors; the respective contributions of each factor vary. The bridle-suture—the previously favored culprit—is now generally exonerated, and scarring and inflammation due to subconjunctival gentamicin\(^10\) appear to be rare causes. However, injection of gentamicin into or near an extraocular muscle can cause toxic myopathy.\(^11\)

Clustering of cases from the same cataract practice (personal observations) tends to implicate a faulty technique of retrobulbar administration. Administration of retrobulbar anesthesia by nonophthalmologists is mentioned as a factor in some reports.\(^12\) A preponderance of left eye involvement in some series also suggests faulty technique: a right-handed surgeon is more awkward when injecting the left eye. The clinician sometimes learns that the anesthetic was particularly painful, a large amount of anesthetic was injected, or the injection had to be repeated. Most injections, however, are generally described as routine and unremarkable. No cases have been reported after topical anesthesia. The capacity of commonly used local anesthetics to permanently damage extraocular muscles is now well established. Carlson and co-workers\(^13\) injected saline or various local anesthetic agents directly into monkey extraocular muscles and showed that, in contrast to the minimal damage seen in saline-injected muscles, anesthetic-injected muscles showed widespread lesions. Damaged muscle fibers were replaced by myoblasts that formed well-organized myofibrillar bundles within the first week. In the same study using humans, saline injection again was innocuous whereas anesthetic injection of extraocular muscles in two elderly subjects (ages 79 and 80 years) caused extensive damage.\(^9\)

In contrast to monkey muscles, the human muscles were filled with dense mats of fibroblasts and no significant muscle regeneration was apparent. The authors concluded that advanced age may contribute to a poor regenerative response. Direct injection into the muscle itself may be required for significant myotoxicity to occur.\(^11\)

In summary, the susceptibility to muscular injury by local anesthetics and its degree depend on several factors, which include the volume and concentration of anesthetic injected, the site of injection (perimuscular, intramuscular, near the tendon, or deep within the muscle belly), age, the extent of muscle fiber regeneration, the degree of fibrotic reaction, unspecified individual susceptibility to anesthetic myotoxicity, additional injury from hematoma, and damage to neural structures.

### Clinical Characteristics

#### Clinical Syndromes

Extraocular muscle injury after injection of local anesthetics was initially thought to affect mainly the inferior rectus muscle due to its vulnerable location. It has since been demonstrated that any extraocular muscle may be involved. The levator muscle and even the orbiculbar muscle can also be affected, causing ptosis and lagophthalmos, respectively.\(^5\) Muscle injury can result in contracture, overaction, or paresis—accounting for the varying clinical pictures reported in the literature.

#### Extraocular Muscle Contracture

Contracture of an extraocular muscle after retrobulbar anesthesia was first described in (and thought to be limited to) the inferior rectus muscle (Fig. 28–3).\(^9\) \(^16\) \(^49\) By virtue of its location, this is the most commonly affected muscle. We now know, however, that any of the extraocular muscles can develop contracture. Specific instances of superior rectus, lateral rectus,\(^1\) and inferior oblique\(^24\) involvement have been described. I have seen patients with medial rectus contracture (Fig. 28–4). Even though retrobulbar anesthesia has been used for many decades, extraocular muscle contracture was described only recently, suggesting at first glance a new deleterious effect from retrobulbar injections. A review of the literature shows the problem to be an old one that formerly was misclassified. For example, Maurer\(^1\) found hypotropia ipsilateral to the aphakic eye in 91% of 261 patients wearing aphakic contact lenses. Maurer attributed it to downward decentration of heavy aphakic lenses due to gravity. Given the significant size of the deviation, it seems more likely that most of these patients actually had an inferior rectus contracture.
The precise pathogenesis of this complication remains uncertain. Some authors suggest that a Volkmann-like contracture arises from either increased tissue pressure due to intramuscular hematoma or direct injection of anesthetic within the soft tissue scaffolding of the muscle. Most authors now believe that the problem is one of local anesthetic myotoxicity. It is unclear at present if a contracture can develop without passing through a paretic phase. Generally, the first examination by a strabismologist has not been done until several months postoperatively, so that a preceding but unnoticed paretic phase cannot be excluded. There are well-documented cases in which the affected muscle initially is paretic and within days to a few weeks exhibits contracture or overaction. An initial paretic phase is in keeping with what we know about the sequence of events in extraocular muscles after local anesthetic injection: initial severe damage, followed by regeneration and, perhaps especially in older individuals, fibrotic proliferation and associated muscle contracture.

Cases of acquired Brown syndrome after peribulbar or retrobulbar anesthesia may reflect mechanical damage to the superior oblique/trochlea complex rather than myotoxicity and contracture. Theoretically, however, injection into the muscle belly of the superior oblique may result in sufficient contracture to cause the picture of acquired Brown syndrome.

**Extraocular Muscle Overaction**

Some patients present with overaction of an extraocular muscle (Fig. 28–5). The current literature offers three possible explanations: (1) local anesthetic myotoxicity and associated paresis of the antagonist, (2) a botulinum toxin–like effect on the antagonist, and (3) initial paresis and subsequent overaction in the same muscle. It is unclear whether all these are actual possibilities. A case of hypertropia after ipsilateral retrobulbar injection was attributed to inferior rectus paresis and secondary superior rectus overaction. Subnormal saccades were generated by the inferior rectus. Four similar cases were reported, but the inferior rectus saccades were normal. Hypertropia was attributed to a botulinum-like effect of the local anesthetic, producing temporary inferior rectus palsy and allowing the emergence of superior rectus overaction in the intervening period. In another series, patients who presented initially with muscle paresis followed by contracture of the same muscle were believed to have contracture or overaction caused by myotoxic effects on that muscle rather than its antagonist. The pattern of strabismus in patients with overacting muscles can be explained by a shift in its length-tension curve, as after a small resection.
action” with free forced duction testing, whereas more significant contracture may be associated with restriction. Stated differently, whether a muscle develops contracture or overaction depends on its response to the anesthetic agent (e.g., the degree of muscle fiber regeneration and eventual fibrotic proliferation) rather than any qualitative pathogenetic difference.

**Extraocular Muscle Paresis**

This is a pattern, described in a few cases, in which the inferior rectus is typically affected; the diagnosis is supported by subnormal infraducting saccades. Esswein and von Noorden described nine patients with apparent permanent paresis of a vertical rectus muscle—seven inferior and two superior rectus muscles. Proposed mechanisms include mechanical trauma from the injection needle to the muscle’s nerve supply and direct anesthetic toxicity to the muscle or nerve. A case of inferior oblique paresis was ascribed to a damaged nerve, which is vulnerable to trauma from the retrobulbar needle. Experience with inferior oblique denervation surgery shows a remarkable tendency for reinnervation in older children. Elderly patients may have poor regenerative capacity, making the effects of local anesthetic myotoxicity qualitatively different. Hence, the procedure may not be analogous to surgical nerve transection.

In summary, the possible responses of extraocular muscles and nerves to direct local anesthetic injection include (1) mechanical or toxic injury to the nerve supplying the muscle, causing paresis; (2) initial myotoxicity (paresis), followed by muscle fiber regeneration and recovery, especially in younger patients; (3) antagonist contracture producing ipsilateral hypertropia; (4) minimal regeneration, causing paresis and ipsilateral hypertropia; (5) some fibrotic proliferation with unrestricted forced ductions; and (6) robust fibrotic proliferation with restricted forced ductions.

**POSTOPERATIVE OPTICAL ABERRATIONS**

Aniseikonia tends to be the first optical factor that comes to mind, but its role is overestimated. Even monocular aphakes fuse with aphakic spectacles when looking through the center of the lens. Patients with long-standing, compensated aniseikonia (e.g., long-standing unilateral high myopia treated with glasses) may become symptomatic after emmetropia is achieved following cataract surgery. Because of the plasticity of the visual system in childhood, central adaptation for significant anisometropia and aniseikonia is possible. Making such patients isometropic in their later years with cataract surgery may precipitate aniseikonia and fusion difficulty.

Anisophoria is a much more important cause of diplopia than aniseikonia. Generally, anisometropia must exceed 2.5 D before anisophoria becomes a problem. Yet even with a large amount of anisometropia, people differ markedly in their capacity to adapt to it. Normal subjects can maintain
Figure 28-5. Patient with overaction of the left superior rectus muscle after retrobulbar anesthesia. Notice the left hypertropia that increases on upgaze and decreases on downgaze. The patient was successfully treated with recession of the left superior rectus muscle.

single binocular vision despite significant anisometropia by differential adaptation of saccadic amplitudes in the two eyes. Interference with this adaptive mechanism by aphakia or pseudophakia may reflect concurrent neurologic disease, diminished fusional reserves, differences in the optical quality of images between the phakic and aphakic eyes, or a switch in ocular preference after cataract surgery.

Switching ocular preference (dominance) after cataract surgery may bother some patients for other reasons. Pseudophakic patients with superior vision in the nondominant eye often complain of visual discomfort despite significantly improved Snellen acuity. Patients may have less tolerance for optical aberrations affecting the dominant eye. I have seen a few patients with switched ocular dominance who seemed unable to deal with horizontal deviation of only a few prism diopters. It may help to explain these difficulties as being roughly analogous to switching from right- to left-handedness. Kushner reported several adults who developed diplopia when they were induced to switch fixation from their previously preferred eye to the other eye by optical manipulations. His patients had a history of strabismus and amblyopia. Because the suppression that accompanies strabismus is facultative, it may not be present in the usually dominant eye when the nondominant eye is fixing. Even nonamblyopes can experience similar difficulties. Attempts to restore ocular dominance, such as cataract surgery on the dominant eye or optically blurring the nondominant eye, may be helpful.

Surgical removal of the human crystalline lens eliminates light scattering, ultraviolet absorption, and ultraviolet fluorescence. As a result, some aphakic patients notice that white appears more intensely white and that red targets appear more red. This is an impediment to fusion, as is any disparity in sensory input.

Diagnosis

The differential diagnosis of strabismus that presents after cataract surgery should include the various causes of strabismus and diplopia in the elderly. Some patients may have causes entirely unrelated to the cataract, the surgery, and the related optical changes. They include thyroid eye disease, myasthenia gravis, tumors, and preceding or intraoperative neurovascular events. A careful history and physical examination should identify the underlying cause in a vast majority of patients; only very few will require ancillary diagnostic testing. The most commonly performed studies in these patients include the edrophonium (Tensilon) test, thyroid function studies, and neuroimaging of the orbits and brain.

Elderly cataract patients may have a retinal cause of binocular diplopia, namely, foveal dystopia, that often causes a diagnostic dilemma. The clinical features, once recognized, are highly characteristic. Because of mechanical distortion of the macula, a rivalry is established between the central and peripheral fusional mechanisms. Patients usually harbor subretinal neovascular membranes (especially after laser treatment), cellophane maculopathy, or diabetic macu-
lar edema. The deviation is usually small, sometimes only a prism diopter or so, but fusion does not occur either spontaneously or with prisms. Prisms provide relief for less than an hour. They do align the displaced foveas but in so doing misalign the peripheral retina. Foveal misalignment recurs once peripheral fusion dominates its central counterpart. Patching or optical penalization is usually needed for relief.

The clinician should determine whether the patient had strabismus, amblyopia, or diplopia before cataract surgery. Is there a history of patching, prism wear, or strabismus surgery? Examination of old photographs (e.g., a picture on the driver’s license) may help determine the time of onset of strabismus and its temporal relationship to cataract surgery.

It is not always easy to determine whether a patient has primary contracture of a muscle (e.g., superior rectus contracture) or contracture resulting from (and associated with) a partially resolved paresis of its antagonist (e.g., inferior rectus paresis). Forced duction testing and saccadic velocity analysis can be helpful. However, clinical observations of saccadic velocity cannot distinguish subtle paresis from normal muscle strength; this requires quantitative measurements. The Parks three-step test is helpful in identifying an isolated, acutely paretic cycovertical muscle. It is less helpful in chronic paresis and is misleading in cases with restriction or contracture. The presence of significant fundus torsion or torsional diplopia in the setting of vertical strabismus favors oblique muscle dysfunction (contracture or paresis); the latter can be verified by an exaggerated traction test performed under general anesthesia.

Intermittent diplopia in patients with tenuous fusional reserves becomes worse with fatigue, hypoxia, or alcohol consumption. This fatigability should not be misconstrued as an indication of myasthenia gravis, although distinguishing between these disorders on clinical grounds alone may sometimes be difficult. Ancillary studies such as edrophonium testing, an acetylcholine receptor antibody assay, and electromyography may be necessary.

### Treatment

The principles of nonsurgical treatment of diplopia in adults are well known. Only major highlights are covered here. It is useful for the clinician to list factors that may be impeding fusion in each patient and try to deal with those that are correctable. The two major factors that contribute to binocular diplopia are motor misalignment and asymmetric sensory input in the two eyes (e.g., disparity in color, image size, brightness, pupil size, and depth of focus). Single binocular vision can be restored to the extent that these two major abnormalities are corrected.

Prisms are the mainstay of nonsurgical management of diplopia. They are often useful either as a permanent solution or as a temporizing measure while awaiting surgery. Fresnel prisms are particularly useful in patients with large deviations. Those with smaller deviations (less than 10 PD) can use either Fresnel or ground-in prisms.

Patients who fail to respond well to optical or surgical realignment of the eyes fall into two categories: (1) those with central disruption of binocular vision after removal of a long-standing unilateral traumatic cataract and (2) those with profound asymmetry in sensory input between the two eyes. In addition, many patients with central fusional disruption have factors disposing to asymmetric sensory input such as abnormalities in the anterior segment, healed corneoscleral lacerations, or abnormally dilated pupils with traumatic sphincter rupture or damage. Mitigating these asymmetries may, along with prismatic compensation, restore some fusional ability over time. For example, in patients with very large damaged pupils, the clinician can try to reduce pupil size with miotics or colored contact lenses (e.g., Wesley-Jessen opaque tinted contact lenses) to improve optical symmetry. In my experience some patients may relearn fusion over time if painstakingly managed with the best optical correction, prisms, and encouragement, rather than immediately surrendering to monocular occlusion.

Before embarking on strabismus surgery, the clinician must (1) make sure that the patient fuses with free prisms or with the synoptophore, because surgery is contraindicated in patients unable to fuse, and (2) ascertain stability of the strabismic angle. In most patients with muscular contracture or paresis resulting from surgical and anesthetic manipulations, strabismus can be successfully treated by weakening the contractured muscle or weakening the antagonist to a paretic muscle. In muscles with contracture or overaction, large recessions are usually needed; adjustable sutures may be helpful. Overall, these patients do very well postoperatively, and they usually are among the most grateful patients encountered by strabismus surgeons.

Experience with botulinum toxin is limited. In general, it can be useful in patients with small deviations. Strabismus surgery is preferred in patients with larger deviations.

### Prevention

Preventive measures can be taken to reduce the risk of strabismus developing after cataract surgery. To avoid unnecessary surprises, preoperative evaluation of cataract patients should include an assessment of ocular motility, taking into account the disorders discussed in this chapter. Preoperative assessment of fusion potential is difficult, if not impossible, in patients with dense cataracts, but an attempt should be made to estimate the potential for postoperative binocularity.

All cataract patients should be informed of the possibility of postoperative diplopia and the potential need for corrective strabismus surgery. This is particularly true for those with preexisting disorders, such as strabismus, amblyopia, or thyroid eye disease; sensory deviations; or long-standing unilateral traumatic cataract. Because of the apparent propensity of patients with long-standing cataracts to develop postoperative diplopia, the surgeon may suggest optical correction with contact lenses instead of intraocular lenses. If the patient develops central fusional disruption, intractable diplopia can be relieved by discontinuing the use of contact lenses. All things being equal, the cataract surgeon should operate on the dominant eye first to avoid the types of problems associated with switching ocular preference. In patients with both significant sensory strabismic deviation and cataract, strabismus and cataract surgery may be performed in one setting. The strabismus surgery is done first to avoid excessive ocular manipulation after cataract surgery.
Alternatively, cataract surgery can be performed initially, followed by strabismus surgery at a later date. In this scenario, improved visual acuity will enhance the ability to fuse.

A significant proportion of patients with diplopia after cataract surgery have local anesthetic myotoxicity or mechanical trauma to orbital soft tissues by the injection process itself. Retrobulbar anesthesia therefore should be administered only by those thoroughly familiar with the technique, the orbital anatomy, and the potential complications. These complications include ocular motility disturbances, globe perforation, optic nerve damage, central retinal artery occlusion, and subarachnoid injection with resultant brain stem anesthesia and respiratory arrest. Alternatives to retrobulbar anesthesia should be explored. Topical, sub-Tenon’s with a blunt cannula, or general anesthesia should be considered for patients with thyroid eye disease, because the chance of muscular injury from anesthetic injections appears to be higher than in patients with normal-sized muscles. The sub-Tenon’s infusion technique is an excellent substitute for retrobulbar anesthesia, offering equivalent anesthetic effects with few, if any, of the drawbacks associated with the retrobulbar route.7, 32, 44, 58

Conclusions

The body of knowledge about this group of disorders has indeed matured over the past decade, yet there remain many gaps in our understanding. Some of the persistent questions are: what is the incidence or frequency of central fusional disruption in patients with long-standing traumatic cataracts? Can we predict preoperatively which patients with long-standing traumatic cataract will develop intractable diplopia after cataract removal? Do the concurrent findings of some patients with thyroid eye disease, because the chance of muscular injury from anesthetic injections appears to be higher than in patients with normal-sized muscles. The sub-Tenon’s infusion technique is an excellent substitute for retrobulbar anesthesia, offering equivalent anesthetic effects with few, if any, of the drawbacks associated with the retrobulbar route.7, 32, 44, 58

REFERENCES

Several rare forms of neurogenic strabismus are seen infrequently by the practicing clinician. They may cause incomitant strabismus and diplopia, in addition to features of the underlying systemic disease. Diagnosis may be difficult, and, more importantly, there may be significant morbidity, which may be minimized by a rational choice of therapy. The main differentiating features are shown in Table 29–1. The interested reader should consult a text on neurology or neuro-ophtalmology for a full account of the diagnosis and pathophysiology of these disorders. In this chapter, the discussion centers on managing the ocular motility complications of these conditions.

**MYASTHENIA GRAVIS**

Myasthenia gravis is a chronic disorder characterized by weakness and fatigability of striated muscles caused by impaired transmission across the neuromuscular junction. The pathology is an autoimmune process producing antibodies directed against the motor end plate that reduce the number of acetylcholine receptors. Myasthenia may occur in a purely ocular form, affecting only the extraocular muscles along with the levator and orbicularis oculi. Alternatively, ocular involvement may precede or accompany generalized myasthenia.

### Historical Perspective

The first description is credited to Willis in 1672. Jolly coined the term *myasthenia gravis pseudoparalytica* in 1895, and Walker demonstrated the therapeutic value of physostigmine, an anticholinesterase, in 1934. Blalock popularized the use of thymectomy in the late 1930s and early 1940s. Although an association between myasthenia and other autoimmune disorders had long been recognized, anti-acetylcholine receptor antibodies were not demonstrated until the 1970s.

### Epidemiology

Myasthenia may affect patients of any age, but the mean age at onset in women is 28 years, and in men it is 42 years. Although generalized myasthenia is more common in women (ratio 3:2), ocular myasthenia is slightly more common in men. Between 50% and 80% of all myasthenics present with ocular signs and symptoms. Half to two thirds of patients presenting with strictly ocular signs will develop generalized disease. If myasthenia remains ocular for 3 years or longer, generalized disease is rare. From 10% to 20% of ocular myasthenics will remit spontaneously, but some may later relapse.

Myasthenia occurs in all racial groups, although ocular myasthenia is said to be three times more common in Chinese than in whites. Estimates of prevalence range from 1 in 50,000 to 1 in 10,000. There is a strong association with several tissue histocompatibility factors, including HLA-A1, B8, C7, DR3, and DQw2.

### Subtypes of Myasthenia

Myasthenia may present in childhood. It may be neonatal, occurring in one in seven children born to myasthenic moth-
Table 29-1. Differentiating Features of Selected Forms of Neurogenic Strabismus

<table>
<thead>
<tr>
<th>Disorder</th>
<th>Site of Lesion</th>
<th>Age at Onset</th>
<th>Causation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Myasthenia gravis</td>
<td>Myoneural junction</td>
<td>Any age but peak in young adults</td>
<td>Autoimmune</td>
</tr>
<tr>
<td>Chronic progressive external</td>
<td>Extraocular muscles and mitochondrial DNA</td>
<td>Gradually progressive from adolescent to middle age</td>
<td>Genetically determined mitochondrial DNA defect</td>
</tr>
<tr>
<td>ophthalmoplegia</td>
<td>Medial longitudinal fasciculus</td>
<td>Adult</td>
<td>Demyelination or ischemia</td>
</tr>
<tr>
<td>Internuclear ophthalmoplegia</td>
<td>Brain stem</td>
<td>Adult</td>
<td>Ischemia, trauma, tumor, neurosurgery</td>
</tr>
<tr>
<td>Skew deviation</td>
<td></td>
<td>Adult</td>
<td>Compression and radiation therapy</td>
</tr>
<tr>
<td>Ocular neuromyotonia</td>
<td>? Cavernous sinus</td>
<td>Adult</td>
<td></td>
</tr>
</tbody>
</table>

Clinical Characteristics

Congenital myasthenia may be caused by several rare syndromes that affect neuromuscular transmission. Most are familial with autosomal recessive inheritance.

Juvenile myasthenia is identical to adult-onset myasthenia and is distinctly more common in girls, with a ratio of 4.5:1 in the first decade (Fig. 29–1). The prognosis for juvenile myasthenia is good. In one study,20 a spontaneous remission rate of 22.4 per 1000 person-years was found. (The article evaluated 149 patients over a median period of 17 years yielding 2533 person-years.)

Adult-onset myasthenia has been classified by Osserman17, 18 into four groups:

- **Group I. Ocular myasthenia.** The disease is localized to the extraocular muscles, levator, and orbicularis oculi. Prognosis is good, especially if no progression to systemic disease occurs within 2 years.
- **Group IIA. Mild generalized myasthenia gravis.** Onset is slow, sparing the respiratory muscles. Response to therapy is good, with a very low mortality rate.
- **Group IIB. Moderate generalized myasthenia gravis.** There is a gradual onset with greater involvement of skeletal and bulbar musculature. Respiratory muscles are not involved. There is a less satisfactory response to drug therapy than in groups I and IIA. The mortality rate is low.
- **Group III. Acute fulminant myasthenia gravis.** Onset is rapid, and progression is complete within 6 months. There is severe bulbar and skeletal muscle involvement with early involvement of respiratory muscles. This group has the highest percentage of thymomas. Response to therapy is poor, and the mortality rate is high.
- **Group IV. Late severe myasthenia gravis.** Late exacerbation of disease occurs at least 2 years after presentation, with group I or II symptoms and signs. This group has the second highest percentage of thymomas. Response to drug therapy is poor, as is the prognosis.

Clinical Characteristics

Ptosis occurs in a majority of patients with ocular myasthenia, either alone or in combination with diplopia due to extraocular muscle involvement. It is commonly variable, may be unilateral or bilateral, and usually worsens on prolonged upgaze. The lid may quiver spontaneously, and the Cogan lid twitch sign may be produced by eliciting a rapid eye movement from downgaze to primary position. An overshoot of the lid is noted; the lid returns to its previous position after a series of twitches.

Upper lid retraction may be seen in the nonptotic lid and may disappear on covering the other eye and on downgaze. This may also be a sign of concurrent Graves ophthalmopathy, which may be present in 4% to 10% of myasthenics.

Underaction of extraocular muscles is the second most common presenting symptom. A history of variability through the day, with symptoms worsening when the patient is tired, is typical. Any muscle or combination of muscles may be affected, and myasthenia may mimic any disorder of eye movement. However, in my experience, isolated underaction (without restriction) of one or both inferior rectus muscles should be assumed to be due to myasthenia until proved otherwise. Myasthenia should also be considered in the differential diagnosis of acquired superior oblique underaction.

The more florid types of eye movement disorder, such as gaze palsies and generalized external ophthalmoplegia, may be misdiagnosed as due to ischemic or infiltrative brain...
stem disease and investigated by costly neuroimaging. Such patients are normally very well apart from their ocular disorder, whereas they typically would exhibit other signs or symptoms if the problem was of central origin. A particularly spectacular presentation is that of “pseudo-internuclear ophthalmoplegia,” with unilateral or bilateral adduction weakness and dissociated nystagmus in the abducting eye.

Myasthenics typically have saccadic eye movements that are faster than normal. The pupil and ciliary body are not clinically affected by myasthenia, although subtle abnormalities have been reported. The orbicularis oculi is usually weak in patients with ocular myasthenia. The examiner should be unable to open the eye of a normal person attempting forced lid closure, but the myasthenic orbicularis is quite easy to overcome by gentle digital force.

**Diagnosis**

**STRABISMUS DIAGNOSIS**

**History**

The history may be vague and consist of no more than a complaint of intermittent diplopia. Care should be taken to establish the time course of the diplopia, in particular, whether it is worse at the end of the working day, when tired, and so on. Myasthenia may be precipitated by a systemic illness or worsened by certain drugs (see later). A family history of autoimmune disorders, such as Graves disease or rheumatoid arthritis, may be elicited. Family members should be asked if one lid closes spontaneously, because the patient is not always aware of this happening.

**Examination**

The patient may have an obvious ptosis or manifest deviation. There may be an abnormal head posture related either to ptosis or incomitant strabismus. The cover test will reveal a tropia or phoria in diagnostic positions of gaze, but the three-step test is of little value in most cases, because one is usually not dealing with a single muscle paresis. The use of graphic methods such as the Hess-Lancaster screen is particularly valuable. Patients with myasthenia typically have normal retinal correspondence, which may easily be documented. Because studies are performed in an identical manner on repeat clinic visits, variation is easy to detect even when subtle. A field of binocular single vision on a bowl perimeter will document the patient’s functional deficit and will also allow the physician to advise on the safety of driving a motor vehicle.

Ocular movements should be tested carefully, asking the patient to report diplopia if it is experienced. Special care should be taken to assess inferior rectus and superior oblique function, because these muscles, along with the medial rectus, are commonly weak in ocular myasthenia. Cover testing or questions about diplopia in extremes of gaze will identify the responsible muscle(s).

Variability and fatigability, the key clinical features, may be demonstrated by serial assessments of lid position and ocular motility at different times of day and after prolonged eccentric gaze. Using graphic methods of documentation such as the Hess-Lancaster screen, and improvement with edrophonium (Tensilon), may be more valuable than prism measurements (Fig. 29–2).

Nystagmus may be related to prolonged eccentric gaze but may occasionally be present in primary position. In the special subtype of pseudo-internuclear ophthalmoplegia there is gross underaction of one or both medial rectus muscles on side gaze, with abducting nystagmus of the fellow eye. These signs disappear dramatically after intravenous administration of edrophonium.

Saccadic velocities are typically faster than normal in myasthenia, especially during the initial phases of saccade generation. This and other features of saccades in myasthenia are discussed in greater detail in Chapter 3.

**Pharmacologic Testing**

Pharmacologic testing with anticholinesterases is the standard diagnostic test for myasthenia. A positive response is valuable. Unfortunately, a negative response has no diagnostic value because false-negative results are common, especially in ocular myasthenics. Edrophonium, a short-acting anticholinesterase, is injected intravenously in an adult dose up to 10 mg and 0.15 mg/kg in children. It is usual to give 1 to 2 mg as a test dose, wait 1 minute, and then inject the

---

*Figure 29–2. A, Ocular myasthenic at beginning of day. B, Same patient when fatigued, at end of same day. Note bilateral ptosis, worse on right.*
rest in small increments, stopping when a positive response is detected. A placebo injection of normal saline may be given first to detect functional problems.

If there is marked ptosis, diagnosis of a positive response is easy. Video recording of ocular movements, eye movement recording, Hess-Lancaster screening, and intraocular pressure monitoring may provide evidence of an effect. Edrophonium may cause systemic cholinergic effects, and intravenous atropine should always be available. Patients known to have cardiac disease should be monitored during the injection and should not be tested in the office. Both false-positive and false-negative responses occur; the latter are relatively common in purely ocular myasthenia, especially when of long standing. If there is strong clinical suspicion but the edrophonium test is negative, a trial of oral pyridostigmine will occasionally show a response, particularly in chronic cases.

LABORATORY DIAGNOSIS

Acetylcholine receptor antibody assay, if positive, is diagnostic of myasthenia. Unfortunately, the test is negative in one half to two thirds of ocular myasthenics. Antithyroid antibodies should be assayed at the same time, in view of the known association between myasthenia gravis and thyroid disease.

Single-fiber electromyographic and repetitive supramaximal motor nerve stimulation are usually performed in a neurophysiologic laboratory and may be particularly useful in dubious cases with negative antibody tests. Mediastinal imaging may detect a thymoma or thymic hyperplasia. Skeletal muscle biopsy with receptor assay is highly specific for myasthenia.

Nevertheless, there is a diagnostically puzzling group of patients who have signs and symptoms typical of ocular myasthenia but negative findings on all diagnostic tests.

DIFFERENTIAL DIAGNOSIS

Moorthy and associates have reported eight patients with "pseudomyasthenia" or "myasthenia plus," who were originally confidently diagnosed as having myasthenia gravis but later were found to have other pathologic processes instead of, or in addition to, the myasthenia. In four cases the problem was believed to be entirely due to the presence of a mass lesion (two parasellar meningiomas, one internal carotid aneurysm, and one chondrosarcoma). In the other four, the diagnosis of myasthenia was not in doubt but further investigation led to the discovery of other pathologic processes (two parasellar meningiomas and two sphenoid meningiomas, one of them recurrent).

Chronic ocular myasthenia may be confused with chronic progressive external ophthalmoplegia.

Although myasthenia and thyroid-related ophthalmopathy may coexist, it is my experience that diagnostic confusion is rare. Proptosis, local inflammation, and muscle enlargement on orbital imaging never occur in myasthenia. Forced duction testing will reveal the underlying restrictive pathologic process in thyroid-related ophthalmopathy.

A variety of drugs, of which the best known is d-penicillamine, may cause myasthenic syndromes. d-Penicillamine is unique in causing a syndrome completely indistinguishable from idiopathic myasthenia. Antireceptor antibodies can be detected, and there may be a diagnostic response to edrophonium. The myasthenia remits on stopping the drug in 70% to 78% of cases but may recur after remission. d-Penicillamine may also cause systemic lupus erythematosus, Guillain-Barré syndrome, thrombocytopenia, and glomerulonephritis.

Other medications may interfere with neuromuscular transmission and worsen existing myasthenia. They include aminoglycoside antibiotics such as streptomycin, gentamicin, kanamycin, and the like, which reduce the amount of acetylcholine released in response to nerve stimulation. A more heterogeneous group of drugs, which includes ampicillin, erythromycin, chlorpromazine, morphine, quinine, procaïnine, and β-adrenergic blockers, may worsen the defect in neuromuscular transmission.

Treatment

NONSURGICAL MANAGEMENT

Treatment of myasthenia is usually offered by the neurologist and most often involves the use of oral anticholinesterases and corticosteroids, although cyclosporine and azathioprine are also sometimes used. Although thymectomy is normally reserved for systemic myasthenia, it may also be of value in ocular myasthenia. Ocular symptoms frequently respond poorly to systemic therapies, at which time the ophthalmologist may be called on to provide treatment for ptosis and diplopia.

Ptosis may be managed by ptosis props mounted on the patient's spectacle frames (Fig. 29–3) or by taping the affected upper lid to the brow, taking care to avoid corneal exposure and drying. Once ptosis becomes chronic, cautious lid surgery has been reported by several investigators to be of value. Both tarsal resection and brow suspension procedures have been successful.

Diplopia may require initial occlusion, if very incontinent or variable. Alternatively, the patient may choose to open one lid only with a ptosis prop. Concomitant deviations may be managed with Fresnel membrane prisms while they are stabilizing and by built-in prisms when stable.

Botulinum Toxin Treatment. Our group at Moorfields Eye Hospital, London, England, has tried botulinum neurotoxin A in a group of five myasthenics, all females aged 37 to 52 years. Four had exotropia measuring between 6 and 60 PD, and one had esotropia of 18 PD. One patient went on to have extraocular muscle surgery, and two others declined further treatment. The remaining two patients have continued with injections for symptomatic relief of diplopia. Both had had one previous surgical procedure. One patient had five injections to the left medial rectus; the other had seven injections to both lateral rectus muscles and two to the right inferior rectus. The interval between injections averages approximately 6 months in both cases.

SURGICAL MANAGEMENT

Patients should be observed over an extended period, with serial examinations every 3 months, until it is certain that strabismus and ocular motility defect are chronic and non-progressive. I believe that a minimum of 12 months of fully
documented stability is required before active strabismus management is contemplated. Although other authorities favor a 6-month interval, the principle is the same.

Miller has stated that “extraocular muscle surgery is never warranted in patients with myasthenia gravis and diplopia.” However, groups from London and Los Angeles have reported the results of surgery in a total of 11 patients having chronic myasthenia and stable clinical signs. All but one were exotropic, and many had inferior rectus underaction and an A pattern. The functional field of single vision improved in all the patients, and the results were stable during a follow-up period averaging 38 months, with a minimum of 7 months (Fig. 29–4). All patients of Acheson and associates were adults, aged 16 to 52 years. In the series of Davidson and colleagues, 2 patients were children aged 4 and 7 years. All patients had horizontal muscle surgery, sometimes with adjustable sutures. Superior oblique weakening was required in 4 of the 11 cases, reflecting the frequency of A-pattern exotropia with inferior rectus underaction. No problems arose from administering general anesthesia.

It is therefore clear that strabismus surgery may be of value in selected cases of myasthenia. These patients should have a stable strabismus angle documented for at least 6 months, and the systemic disease should be stable with or without medications. Patients may require recessions and resections, depending on the preferred fixing eye. Many benefit from a selective weakening of superior oblique action to balance weak inferior rectus muscles. An adjustable suture technique helps achieve the desired alignment in these patients when the response of a weak muscle may be unpredictable.

Figure 29–3. A and B, Two types of ptosis prop.

Figure 29–4. A, A 33-year-old myasthenic woman with distance deviation of 25 PD of exotropia and 20 PD of right hypertropia. Rotations show marked limitation of adduction and depression of the right eye and mild limitation of adduction and elevation of the left eye. B, At 38 weeks after recession of both lateral rectus muscles, resection of the right inferior rectus, and recession of the left inferior rectus (adjustable), stable angle of 8 PD of exophoria with improved adduction and depression of the right eye and adduction and elevation of the left eye was achieved. (From Davidson JL, Rosenbaum AL, McCall LC: Strabismus surgery in patients with myasthenia. J Pediatr Ophthalmol Strabismus 1993;30:292. Reprinted with permission of Slack, Inc.)
COMPLICATIONS

The greatest risk of surgery in myasthenia gravis is lack of symptomatic improvement. This is perhaps not surprising, because the condition is not truly curable by surgical means, owing to inherent weakness of the extraocular muscles. Nevertheless, it seems reasonable to consider active management in patients with stable disease and troublesome symptoms, because systemic therapy often has little effect on the ocular symptoms or causes unacceptable side effects. Prisms have limitations when the deviation is markedly incomitant. The higher powers are expensive when built-in, and blur vision in membrane form. The choice may therefore lie between chronic occlusion and surgery, with botulinum toxin as a possible adjunct. Patients should be aware that several procedures may be needed to achieve the final result, but that there is little risk of ending up significantly worse than at the outset. My colleagues and I, in our surgical series, reported on two patients who had one operation and three who required a further procedure to ensure the final result.

The other consideration sometimes raised regarding surgery is the risk that either the systemic or ocular disease will progress. It is known to be extremely rare for ocular myasthenia to progress to systemic myasthenia after 2 to 3 years. I and others have found that, if guidelines for establishing stability (see earlier) are followed, further evolution of the ocular signs does not occur and active management of the motility defect tends to be successful.

No anesthetic complications have been noted either by our group or by Rosenbaum’s group in Los Angeles. In my view, patients having chronic myasthenia with troublesome symptoms of diplopia or ptosis should be considered as candidates for surgery unless some contraindication exists.

CHRONIC PROGRESSIVE EXTERNAL OPHTHALMOPLEGIA

Characterized by progressive loss of all extraocular movements and bilateral ptosis, chronic progressive external ophthalmoplegia has long been known to have a familial tendency. In addition, there is a recognized association with other neurologic abnormalities such as pharyngeal weakness, cerebellar ataxia, deafness, peripheral neuropathy, optic atrophy, and dementia. Some patients also have involvement of other organs, such as the retina, heart, skin, endocrine glands, and skeleton. The term ophthalmoplegia plus was used to describe such cases. Familial cases with pigmentary retinopathy and cardiac conduction defects are usually designated as Kearns-Sayre syndrome.

These disorders are caused by abnormal mitochondria inherited by cytoplasmic transmission through the maternal line. Muscle biopsy may reveal characteristic ragged red fiber abnormalities on light and electron microscopic study (Fig. 29–5).

Clinical Characteristics

Patients may present at any age, but the majority are 35 to 55 years old. The history at presentation is often several years in duration. Ptosis and external ophthalmoplegia are the key clinical signs (Fig. 29–6). The patient may adopt a compensatory chin-up head posture to permit vision past the drooping upper lids. Frontalis muscle overaction is common. Ocular movements are nonexistent, or there may be small excursions laterally or downward. Bell’s phenomenon is usually absent. The ocular posture is usually exotropic. If the deviation is large, there may be a face turn away from the preferred eye to bring it into a satisfactory position for fixation. It is often stated that such patients do not experience diplopia, owing to the gradual onset of the condition. I, however, have certainly seen a number of patients who were aware of diplopia, although the images tend to be widely separated and cause little difficulty in separating the real from the false.

The facial muscles are frequently affected, leading to a typical “myopathic” appearance. In the Kearns-Sayre variant there is a characteristic diffuse pigmentary retinopathy, as well as cardiac conduction defects, leading to complete heart block.

Diagnosis

For the ophthalmologist the diagnosis is based on the history and clinical signs, although referral for neurologic and genetic studies will allow a more detailed characterization of the mitochondrial defect. Cardiologic investigation should also be performed, although most patients have no conduction defect.

Figure 29–5. Ragged red fibers in orbicularis oculi muscle of a patient with mitochondrial myopathy (progressive external ophthalmoplegia). (Masson’s trichrom stain at 300X magnification.) (Courtesy of Narsing Rao, MD. University of Southern California, Doheny Eye Institute.) (See Color Plate 6.)
if treatment is contemplated. Patients may have learned to ignore the image from the nonfixing eye but may be made aware of it with a red lens or similar dissociative maneuver. The potential for fusion or suppression should be assessed either by prism correction or on the synoptophore. Patients who cannot suppress or fuse diplopia should be warned that, if they elect to have surgery, they may need to wear an occlusive contact lens to eliminate a bothersome second image and perform tasks such as driving.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis includes chronic myasthenia gravis and the heterogeneous group of sporadic and familial disorders variously known as congenital ocular muscle fibrosis, congenital external ophthalmoplegia, and others (Fig. 29–7). These conditions, when familial, exhibit an autosomal dominant pattern of inheritance, and present as congenital bilateral ptosis and grossly limited ocular movements—especially vertically. Forced ductions are abnormal.

Treatment

PTOSIS MANAGEMENT

In most cases, symptomatic treatment of ptosis with lid props on the patient’s spectacles is all that can be offered. Cautious levator surgery may be of value in extreme cases. Despite a poor Bells’ phenomenon, exposure problems are rare.

Lane and Collin11 reported results in 7 patients (13 lids) who underwent anterior levator advancement, and 8 patients (14 lids) who had brow suspension procedures. Satisfactory results were achieved in 14 patients (25 lids).

STRABISMUS MANAGEMENT

It is impossible to provide either ocular movement or to make the eyes work together, but some patients wish to
have either a less abnormal head posture or an improved appearance. Extraocular muscle surgery has a limited but useful role.

The usual procedure is a lateral rectus recession (8–9 mm) coupled with a large medial rectus resection (7–8 mm) on one or both eyes, using adjustable sutures. If diplopia is a problem, an occlusive contact lens may be worn, but this is usually unnecessary. Even with bilateral “supramaximal” surgery, undercorrection is common. When general anesthesia is contraindicated because of cardiac problems, local anesthesia may be used.

One report described the experience with surgical management in four patients: one esotropic, one exotropic, and two hypertropic.23 In three cases diplopia was the presenting problem, and the fourth (exotropic) patient was concerned about appearance. One surgical procedure was performed in three cases, with a tendency to undercorrection of the deviation. The remaining patient, with hypertropia and exotropia, had two procedures on vertical rectus muscles before a diagnosis of oculopharyngeal dystrophy was made. Four further procedures and one injection of botulinum toxin achieved a final position in which single vision in primary position could be maintained using glasses with built-in prisms. The article reviews five other cases of surgically managed chronic progressive external ophthalmoplegia, all exotropic, which also showed a tendency to early or late undercorrection.

I have tried using botulinum toxin treatment in three patients with chronic progressive external ophthalmoplegia, generally with little success. It is of interest, however, that in most cases the ocular position may be altered, presumably by changing resting tone in the injected muscle. Two patients were male and one female, and the age range was 38 to 70 years. Two patients were exotropic, with deviations of 16 to 30 PD, and one was esotropic with an angle of 25 PD. Electromyographic signals were notably lower than normal, measuring 100 to 200 mV. (Typical amplitudes of signals for ocular electromyography in my unit are 300 to 800 mV for medial rectus and 200 to 600 mV for lateral rectus.) All injections produced a reproducible change in ocular position but no true long-term change in alignment.

The underlying mitochondrial cytopathy may be treated with corticosteroids and ubiquinone (coenzyme Q), but this does not seem to affect ocular muscle function in established cases.

**INTERNUCLEAR OPHTHALMOPLEGIA**

Internuclear ophthalmoplegia is caused by lesions of one or both medial longitudinal fasciculi (MLF), distinct paired fiber tracts that run from the interstitial nucleus of Cajal rostrally to the spinal cord caudally. In the brain stem these tracts lie medially and near the floor of the aqueduct and fourth ventricle, close to the nuclei of the third, fourth, and sixth cranial nerves and the vestibular nuclei. The MLF carry interneurons from the abducens nucleus/paramedian pontine reticular formation complex to the medial rectus subnucleus of the contralateral third nerve nucleus, thereby coordinating ocular rotation on lateral gaze. The MLF also carry fibers responsible for holding vertical eye position, for vertical smooth pursuit, and for the vertical vestibulo-ocular reflex.

**Clinical Characteristics**

Lesions of the MLF interfere with adduction of the ipsilateral eye on attempted conjugate gaze (Fig. 29–8). The weakness may be gross, with complete inability to adduct past the midline, or subtle, with only a reduction in velocity of the adducting saccades. Convergence is frequently spared, and these cases are often designated as “posterior” internuclear ophthalmoplegia. The other eye exhibits horizontal abducting nystagmus, with a nasalward drift and fast outward recovery movement. Nystagmus is either absent or reduced in the adducting eye and therefore is termed dissociated.

**Figure 29–8.** Asymmetric bilateral internuclear ophthalmoplegia: A, Right gaze—gross left adduction deficit. B, Left gaze—mild right adduction deficit. (Courtesy of Mr. John Elston.)
Bilateral cases of internuclear ophthalmoplegia exhibit bilateral adduction weakness and abducting nystagmus, as well as abnormalities of sustained vertical gaze. When an extensive lesion disrupts the paramedian pontine reticular formation on one side, internuclear fibers of the ipsilateral MLF may also be affected after they have crossed the midline from their origin in the contralateral abducens nucleus. The effect is a complete horizontal gaze palsy when looking toward the affected side and paresis of adduction on gaze to the opposite side. This is termed a one-and-a-half syndrome, and it is often associated with an exotropia, as is bilateral internuclear ophthalmoplegia, hence the acronym WEBINO, for wall-eyed bilateral internuclear ophthalmoplegia (Fig. 29–9). When there is exotropia with no horizontal movements, the term paralytic pontine exotropia is used.

**Diagnosis**

Unilateral internuclear ophthalmoplegia is typically caused by ischemia but may occur as a result of demyelination from multiple sclerosis or a variety of other pathologic processes. Bilateral internuclear ophthalmoplegia is most commonly due to multiple sclerosis, but it may also be caused by ischemia and other conditions such as syphilis, the Chiari malformation, and trauma. One-and-a-half syndrome is usually due to ischemia or multiple sclerosis.

**STRABISMUS DIAGNOSIS**

**History**

Many patients with unilateral internuclear ophthalmoplegia have no symptoms at all, and because many patients recover, most ophthalmologists will tend to see only acute cases. These patients may present with episodic gaze-related diplopia, which typically is noted on rapid glancing to one side as when crossing the road and disappears when the eyes are back in primary position.

Chronic bilateral cases and patients with one-and-a-half syndrome may be referred by neurologic colleagues when exotropia and complaints of diplopia and/or an abnormal head posture are present. Patients with chronic demyelination due to multiple sclerosis may have, in addition, consecutive optic atrophy and pendular nystagmus with oscillopsia.

**Examination**

The usual deviation is an exotropia, which may be enormous, measuring 90 to 100 PD, or quite subtle. Unless there is significant optic atrophy, patients are able to fixate well; and cover testing in diagnostic gaze positions will show limited adduction on conjugate gaze to the opposite side. The defect may also be documented by Hess-Lancaster screen testing. Convergence is typically spared; it should be documented, and the near point measured.

A sensory evaluation should be performed in patients with large angles, especially if surgery is contemplated. In my experience, cases due to ischemia have a good potential to regain binocularity, whereas patients with advanced multiple sclerosis may have significant barriers to fusion owing to nystagmus or optic atrophy.

**Treatment**

Most cases are either asymptomatic or only transiently troubled. A majority of patients referred for symptomatic treatment have large-angle exotropia and limited adduction of both eyes. Prisms are of no value, but large bilateral lateral rectus recessions and medial rectus resections may improve ocular position and provide a small central field of single vision. When there is associated vertical incomitance, transposition of the muscle insertions up or down may be helpful. Figure 29–10 shows a case of WEBINO before and after exotropia surgery.

I have treated five patients having strabismus and internuclear ophthalmoplegia with botulinum toxin. Two were male and three were female, with an age range of 23 to 50 years. Two cases were due to intracranial hemorrhage, and three were due to multiple sclerosis. Four patients were exotropic with deviations of 35 to 75 PD and were treated with injection to one or both lateral rectus muscles. One had a symptomatic hypotropia and received a single injection to an inferior rectus without improving. One patient has had
ocular realignment surgery without restoration of fusion, but with an improved appearance. Another is due for surgery in the near future. Both are young men who had sustained intracranial bleeding.

**SKEW DEVIATION**

Skew deviation is a vertical misalignment of the visual axes due to imbalance of prenuclear inputs. It may show a constant angle in all gaze positions or vary with gaze position (Figs. 29–11 and 29–12). Most cases occur in association with other signs and symptoms of brain stem and/or cerebellar disease such as internuclear ophthalmoplegia, when the hypertropic eye is usually on the side of the lesion. Many cases are transient, occurring in comatose or obtunded patients, but some patients—usually those with an intracranial vascular event or trauma—may have persisting problems and seek ophthalmologic assistance.

**Differential Diagnosis**

The main differential diagnosis is from acquired fourth nerve palsy. Most cases of acquired fourth nerve palsy exhibit some degree of excyclotorsion, whereas patients with skew deviation do not have subjective cyclotorsional symptoms (although they may show objective torsion on fundus examination).

**Treatment**

Most cases are transient and require no treatment. Prisms are of minor value, but chronically symptomatic patients may benefit from surgery. I have treated a case of incomitant...
Figure 29–13. A, Preoperative Hess-Lancaster screen of 52-year-old man with skew deviation. Marked deficit of left depression is evident. B, Hess-Lancaster screen after inverse Knapp procedure of the left eye shows some improvement of depression.

skew deviation with uniocular depression deficit, which followed a cerebrovascular accident, in a 52-year-old man who had a 20-month history of vertical diplopia on depression and whose Hess-Lancaster screen is shown in Figure 29–13A. He had a marked deficit of left depression, which was managed initially by occluding the lower part of his left spectacle lens. When the signs seemed stable he underwent downward transposition of the horizontal rectus muscles of the left eye (inverse Knapp procedure) with some improvement, as shown in Figure 29–13B. Diagnostic injection of botulinum toxin into the right inferior rectus led to further subjective improvement, and he finally underwent an adjustable recession of the right inferior rectus with a good long-term result (see Fig. 29–13C).
This rare ocular motor phenomenon is characterized by persisting spasm in a muscle supplied by a particular cranial nerve. It usually occurs when the eye moves into the direction of gaze of the affected muscle and may be unilateral or bilateral (Fig. 29–14).14

The condition was first described in 1970 by Ricker and Mertens,15 who confirmed the diagnosis by electromyography. About 18 patients have since been reported. A report by Ezra and associates adds three cases, two affecting the third, and one, the fourth, cranial nerve. The former two cases were triggered by voluntary gaze, whereas in the patient with fourth nerve involvement spasm was induced only by alcohol consumption. Other reports besides Ezra and associates’ series are those of Newman and co-workers16 and Fu.17

Of 19 reported cases, 11 have involved the third cranial

**Figure 29–14.** Ocular neuromyotonia. A through C, A partial chronic right third nerve paresis is present during quiescent periods with a right hypotropia (A), right upper lid retraction on downgaze (B), and weakness of right elevation (C). C through E, Right levator neuromyotonia. On sustained upgaze (C), levator spasm is induced, resulting in increased upper lid retraction, particularly on downgaze (D and E). F through I, Right medial rectus neuromyotonia. During quiescent periods, right lateral gaze is full (F). Sustained left gaze (G) induces right medial rectus neuromyotonia with restriction of right lateral rectus action (H). With resolution of the episode, full abduction of the right eye is restored (I). Simultaneous neuromyotonia of the superior rectus–levator complex and the medial rectus could not be elicited. (From Ezra E, Spalton D, Sanders MD, et al: Ocular neuromyotonia. Br J Ophthalmol 1996;80:350. Reprinted with permission of the BMJ Publishing Group.)
nerve, of which 6 had a chronic third nerve palsy and 5 were normal apart from the neuromyotonia. Two patients, including Ezra and associates' alcohol-induced case, have fourth nerve involvement. The remaining 6 cases involved the sixth nerve. The interval from the initial problem and the onset of symptoms, when known, varied from 2 months to 4 years.

Morrow and colleagues have reported a case of bilateral oculomotor neuromyotonia affecting muscles innervated by both oculomotor nerves in a 45-year-old woman. A pituitary tumor had been treated surgically 18 years previously, and a recurrence 2 years later was treated by external-beam irradiation. There was no concurrent cranial nerve palsy. The neuromyotonic symptoms began 18 months before presentation (i.e., more than 14 years after radiation therapy).

The majority of cases have followed irradiation of a pituitary tumor or other neoplasm of the skull base, although one case reported by Ezra and associates was due to third nerve compression by an internal carotid artery aneurysm. In five reported cases, no preceding factor could be identified.

The pathologic process is unclear. Electromyography has shown sustained neural discharge in the affected muscle. It has been speculated that this may be due to interneural transmission in the form of a self-perpetuating circuit. Ephaptic transmission has been suggested and may be facilitated by an increased extracellular potassium level. The effect of alcohol is not understood.

**Treatment**

Treatment is medical and is based on the severity of symptoms. Carbamazepine, a membrane-stabilizing drug, has been helpful in most reported cases and has produced at least one remission. There is no place for surgery.

**REFERENCES**

The craniosynostosis syndromes have been well described and studied in the past century and a half. However, before Tessier’s pioneering craniosurgical approach, little substantive or structural alteration of the face could be achieved. The rarity and complexity of these syndromes dictate that treatment is best undertaken by a coordinated team approach including a pediatric ophthalmologist and strabismologist, neurosurgeon, plastic surgeon, otolaryngologist, anesthetist, oral surgeon, psychiatrist, geneticist, speech therapist, and orthoptist.

Craniosynostosis syndromes are recognized very soon after birth (Fig. 30–1); the visual system usually is more involved than other organ systems. Cranial surgery performed in the first 3 to 4 months of life allows optimal brain growth and the attainment of normal developmental milestones (Fig. 30–2). Otherwise, premature cranial suture closure arrests bony growth 90 degrees away from the axis of the suture involved. Half of untreated patients suffer headache, and 13% develop mental retardation. Strabismus is seen in a majority of individuals.

**Strabismus Mechanisms**

Anatomic and sensorimotor factors contribute to the development of strabismus associated with craniosynostosis. The anatomic features include abnormally shallow and exorotated orbits, which provide poor support for the globe; short orbital walls that alter the arc of contact of the globe with the extraocular muscles; and disordered structure of the cranial base and trochlea, influencing extraocular muscle function. Magnetic resonance imaging demonstrates excyclorotation of the entire extraocular muscle cone, resulting in altered vectors of muscular action and leading to clinical pseudo-overaction and pseudo-underaction of the extraocular muscles.

Specific extraocular muscle abnormalities may include complete absence, anomalous insertion, and anomalous microanatomy. In a series of 11 patients with Apert syndrome and clinical bilateral superior oblique (SO) paresis, 5 were found to have no SO tendon on either side and 2 others had only a fibrous remnant. Anomalous insertions such as bifid medial rectus (MR) muscles have also been observed.

**V-PATTERN STRABISMUS**

V-pattern strabismus is more common in both esodeviations and exodeviations; reported rates range from 59% to 100% (Fig. 30–3). Several explanations have been proposed:

1. Inadequate support of the globe and orbital contents by the short orbital floor forces the globe to rest on the inferior oblique (IO) and inferior rectus muscles. This increases the arc of contact with the globe and results in increased activity, particularly of the IO. After orbital floor reconstruction, excessive reliance on the inferior muscles for support of the globe is relieved and less overaction is noted.

2. The shallow medial orbital wall increases the normal angle (54 degrees) between the SO tendon and visual axis and eliminates its mechanical advantage as a depressor. If the craniosynostotic SO tendon makes an angle of 90 degrees with the visual axis, the SO will function mainly as an incyclorotator, allowing unopposed action of the ipsilateral IO (Fig. 30–4).

3. Some amount of IO overaction leading to a V pattern may be related to anomalous muscle vectors because of exorotation of the globe. If the muscle cone is excyclorotated, the surgeon should consider the possibility that the displaced muscles gain additional vectors, and thus an added effect on muscle action. For
Figure 30–2. A patient with Crouzon syndrome having a frontal advancement procedure. The frontal flap is reflected inferiorly and covers the nose. It is being held down with a tissue rake. Note that both trochleas have been effectively disinserted. After these procedures, it is best to wait for tissue to heal and edema to subside before proceeding with corrective strabismus surgery. (Courtesy of Albert Biglan, MD.)

Figure 30–1. A, Congenital severe craniostenosis (clover-leaf skull) demonstrating shallow exorotated orbits, exotropia, and proptosis. B, Profile shows occipital flattening, oxybrachycephaly, and proptosis.

Figure 30–2. A patient with Crouzon syndrome having a frontal advancement procedure. The frontal flap is reflected inferiorly and covers the nose. It is being held down with a tissue rake. Note that both trochleas have been effectively disinserted. After these procedures, it is best to wait for tissue to heal and edema to subside before proceeding with corrective strabismus surgery. (Courtesy of Albert Biglan, MD.)
Figure 30-3. Craniofacial synostosis associated with V-pattern exotropia. Note V-pattern in midline gaze position. Both inferior oblique muscles are overacting. Right superior oblique shows underaction.

**Hypothesis**

Primary S.O. Underaction
Mechanical? Angulation S.O. not 54°

Figure 30-4. Line drawing of possible cause of mechanical disadvantage of superior oblique function with shortening of the anteroposterior dimension of the medial orbital wall. Left drawing reflects normal angle (54 degrees) that the superior oblique tendon makes with the visual axis. Right drawing shows shortened medial orbital wall, with proptosis of the globe. The superior oblique tendon in this illustration makes an angle of 90 degrees with the visual axis, allowing it to function only as an incyclorotator, eliminating the optimum position to perform its other functions of depression and abduction.

example, on left gaze, with the left eye fixing, both the left superior rectus (SR) and lateral rectus (LR) may be innervated. By Hering’s law, anomalous contraction of the contralateral synergists—the right IO and MR muscles—takes place and may explain some of the V pattern observed. The frequent overelevation in adduction may be due to the combined vectors of action and the anomalous new muscle pairing. Surgery on either muscle alone may not resolve overelevation in adduction.

4. Relative IO overaction that results from underaction or absence of the SO may lead to a V pattern.

**Clinical Characteristics**

In 1906, Apert described patients with oxycephaly (a flattened occiput and steep forehead), horizontal groove above the supraorbital ridge, hypoplastic midfacial region, parrot-beak nose, hypertelorism, proptosis, and strabismus (Figs. 30–5 and 30–6). The incidence of Apert syndrome has subsequently been reported as 1 in 160,000 live births.

Crouzon, in 1912, described a group of patients with hypertelorism and exophthalmos who had poor midfacial bone growth, an increased angle of orbital divergence (greater than 90 degrees), reduced orbital volume, and an increased medial and lateral interorbital distance (Fig. 30–7). Esotropia, exotropia, and vertical strabismus are common correlates.

In 1964, Pfeiffer reported another clinical variant of craniosynostosis that was associated with broadened thumbs and great toes. The patients also had variable and partial soft tissue syndactyly in the hands and feet, oxybrachycephaly with maxillary hypoplasia, hypertelorism, and proptosis (Fig. 30–8). These patients tend to have more airway problems and exorbitism than other groups.

In all the syndromes just described, central nervous system
involvement may include hydrocephalus, developmental delay, and nystagmus. Cranial nerves are rarely involved but compression of the optic (second cranial) nerve may be observed with delayed cranial surgery, as well as sixth (abducens) nerve paresis secondary to hydrocephalus and increased intracranial pressure.

**Diagnosis**

**CLINICAL DIAGNOSIS**

As a crucial member of the craniofacial team, the ophthalmologist must be prepared to perform a thorough baseline ophthalmologic evaluation before any planned craniofacial reconstruction. The ophthalmologist should be consulted for problems such as exorbitism, subluxation of the globe (Fig. 30–9), and strabismus, both before and after cranial surgery. The diagnostically helpful craniofacial features include head shape, orbital divergence, parrot-beak nose with a flat midfacial region, antimongoloid slanting of the lid fissures, and tearing problems. Peripheral features include short stature, broad thumbs and toes, and syndactyly of the hands or feet. Anomalous head positions are common—alternation of head turn with alternating fixation may occur, as well as a compensatory head position for nystagmus or to promote fusion in alphabet-pattern strabismus.

These children require a particularly caring and “light” atmosphere when examined. Virtually all of them have had multiple surgical procedures since their earliest months, and some may be very frightened of medical personnel. A gradual, deliberate approach will help to gain their trust.

**STRABISMUS DIAGNOSIS**

**Amblyopia**

To have the best chance of regaining normal vision and developing binocular (bifoveal) fusion, strabismic children with craniosynostosis should be treated like other strabismus patients and receive appropriate amblyopia therapy before and after craniofacial and orbital surgery.
Figure 30-7. A, Crouzon syndrome at age 10 months. Patient at age 3 years in primary position (B) and in upgaze (C) demonstrating increased exodeviation (V pattern).

Figure 30-6. A, Patient with Pfeiffer syndrome requiring temporal tarsorrhaphy at age 10 months because of exorbitism. B, At age 18 months, tarsorrhaphy performed earlier remains adequate. C, Broad thumbs. D, Closer view showing proptosis and exotropia.
is caused by strabismus, ametropia, or ptosis much more often than structural abnormalities of the globe (e.g., cataracts, optic atrophy, corneal opacities). It responds well to traditional therapy with glasses, occlusion, and atropine penalization.

Severe lagophthalmos may lead to corneal decompensation, infected corneal ulcers, and descemetocoele (Fig. 30-10). After craniofacial reconstruction (Fig. 30-11), edema may close one eye more than the other, leading to stimulus-deprivation amblyopia (Fig. 30-12). Orbital repositioning for hypertelorism frequently causes esotropia, with subsequent suppression and amblyopia (Fig. 30-13). In addition, esodeviation may result from iatrogenic LR palsy, caused by stretching of the LR innervation when the periorbita is incised as far posteriorly as 25 mm. The orbit becomes esodrected after reconstruction, exerting increased tension on the peripheral part of the sixth nerve. Trochlear damage, or failure of the trochlea to reattach to the periorbita after subperiosteal orbital dissection, may lead to SO paresis.

The ophthalmologist should remain involved after cranial and facial repair to manage amblyopia and other ocular problems.

**Measuring the Deviation**

Pattern strabismus is the rule rather than the exception in craniosynostotic strabismus. Fixation targets must be colored, detailed, and fun for children to follow and analyze, because motility measurements often are time consuming for the physician and onerous to the child. The most repeatable measurements are obtained when children are fresh and rested; every effort should be made to schedule appointments early in the day.

Suitable distance accommodative targets include remote (foot switch)-controlled clapping bears, rabbits, or other non-threatening animals. A laser disc system that switches over to a video system will help patients remain relaxed and entertained between portions of the examination.
Figure 30-12. A, Complete bilateral mechanical ptosis after forehead advancement. B, Unilateral complete ptosis after forehead repair. The patient is at risk for stimulus deprivation amblyopia.

Figure 30-13. A, Hypertelorism with full abduction of right eye seen through a congenital upper lid coloboma. B, Photograph at age 2 years in primary gaze after repair of coloboma and hypertelorism. C, Diminished abduction of left eye after hypertelorism repair. Best corrected vision remained at 20/400 (6/60) despite adequate patching therapy for amblyopia. D, Left abduction improved with time.
The prism cover test must be recorded in all standard gaze positions at distance (6 m or 20 ft) and at ½ m (13 in), including primary gaze, direct upgaze and downgaze, gaze right and left, gaze up and down right, and gaze up and down left (the oblique corners). Head tilts are best performed with the parent gently directing the tilt, leaving the physician with both hands free to measure. Confirmation that distance fixation is maintained may be gained by asking patients about the distant fixation target.

Versions are highly informative. Most individuals will demonstrate at least some apparent oblique dysfunction. The most common is IO overaction, followed by SO underaction and LR underaction. Ductions should be examined when abnormal ocular rotations are detected on version testing.

Active force generation and passive forced duction testing in this group of patients are generally not possible in the office. Clinical evaluation of saccadic velocities when comparing one side to the other is helpful. If a “user-friendly” electronic eye movement recording system is available, and can be administered by an experienced operator, valuable added information may be provided. However, if the child is intimidated by the instrumentation, it may take some time before trust is regained.

Fusion

The ability of the cortex to function normally in response to disparate normal input and to show the expected adaptations to altered visual input is surprising when one considers how unlikely it is for the sensory system to escape unscathed. Despite aggravating conditions, fusion has been demonstrated in 33% of patients with Crouzon, 25% with Apert, and 28% with Pfeiffer syndrome.

Binocular status may be tested in younger children using the Lang test or the 4-PD base-out prism introduction test. In older children, TNO stereo test, Titmus stereo test, Lancaster-Hess screen, and synoptophore evaluation may be performed, and the field of single binocular vision mapped.

If the child spontaneously holds the head in an eccentric gaze position, it is probable that fusion is present in that position. Before a cover test is performed, fusion should be tested in this preferred gaze. Sensory testing is repeated in the “forced primary” position, with the parent supporting the child’s head. The absence or presence of fusion will clarify the reason for the anomalous head posture. Some children may adopt a compensatory head position for a nystagmus null region. The examiner should be alert to this possibility and examine fixation movements as well.

Torsion

Ocular torsion is difficult to measure objectively in young patients, but fundus torsion is easily observed by indirect ophthalmoscopic examination of the dilated fundus. It has been suggested that failure of fusion induces torsion, which in turn causes an alphabet-pattern strabismus (“sensory torsion theory”). Anatomic excyclorotation of the muscle cone may cause torsion of the globe, with anomalous force vectors and innervational abnormalities of the muscles.

LABORATORY DIAGNOSIS

Orbital images are as varied as the clinical features of craniosynostosis syndrome. Whenever possible coronal computed tomography should be performed in reoperated patients, particularly in the presence of a poor Bell’s phenomenon. Imaging studies will yield valuable information when documentation of previous surgery is unavailable, when orbital repositioning has been performed, or when there is marked apparent oblique dysfunction (such as IO overaction, or SR or SO underaction). Computed tomography or magnetic resonance imaging of the orbits also demonstrates anomalous or aberrant insertions and muscle absence. In Figure 30–14, medial and upward rotation of inferior (IR) and MR muscles in both eyes is apparent, whereas the LR muscles remain in the appropriate anatomic position. The findings of imaging studies may influence the planning of strabismus surgery.

Treatment

NONSURGICAL MANAGEMENT

Botulinum toxin injection of the ipsilateral antagonist MR may be indicated for both diagnosing LR recovery (if masked by MR contracture) and to avoid secondary contracture. In most instances the patient’s age precludes adequate placement of the needle under electromyographic guidance. Sedation with ketamine or propofol is often required. The former, however, may result in visual hallucinations and nystagmus, whereas the latter depresses the electromyographic signal. In addition, children with these syndromes commonly have V-pattern strabismus and may also have associated cyclovertical deviations.

Botulinum toxin may be useful for creating ptosis to protect the globe, especially if the child is too sick to undergo tarsorrhaphy under anesthesia before lower orbital...
advancement surgery. It could be used even in patients with nystagmus in this setting, but the limitations associated with using general anesthesia should be considered (see also Chapter 32).

**SURGICAL MANAGEMENT**

**Timing of Surgery**

Until the mid 1980s it was routine to postpone strabismus surgery until major craniofacial reconstructive efforts had been exhausted. Most modern strabismologists now prefer to operate earlier in the developmental period rather than later, to provide the best opportunity for fusion. Experience has shown that orbital repositioning rarely results in postoperative shift of the strabismus deviation. Of 140 children examined ophthalmologically before and after reconstruction, only 10 had a postoperative shift in deviation and only 4 experienced a shift of more than 10 PD. The change in strabismus deviation was more common in Apert syndrome than in Crouzon syndrome, and even less frequent in isolated craniofacial syndromes (without systemic findings). Early repair also enhances the child's psychosocial development.

The timing of craniofacial reconstruction is best left to the surgeons involved. In general, early surgery is preferred because full-thickness bone graft, which has the ability to regenerate bone, may be used rather than partial-thickness grafts. Urgent surgery is indicated when the globe or vision is threatened, as in severe corneal exposure due to exorbitism or severe lid retraction or the development of optic atrophy.

After initial craniofacial repair, usually at 3 to 4 months of age, strabismus surgery may be contemplated when the ophthalmologist believes that the usual clinical indications are met: reversal of amblyopia after patching or penalization, appropriate optical correction, and reliable cover tests or light reflex measurements. Of course, other members of the craniofacial team must be consulted with regard to the child's general health, ability to undergo general anesthesia, and possible need for other, more urgent intervention. It is imperative, however, that the ophthalmologist inform other members of the craniofacial team about the relative urgency of ocular realignment.

**Surgical Approach**

Deviations are often incomitant. The goal of strabismus repair is to achieve orthotropia in primary position and as wide a field of single binocular vision as possible.

**Conjunctival Incision.** A limbal incision is preferred so that anomalous extraocular muscle insertions or structures may be safely recognized. Some muscles may be inserted closer to the limbus than normal. Radial relaxing incisions are recommended, taking the opportunity afforded by a general anesthetic to evaluate the position and integrity of all the rectus and oblique muscles and their insertions. Direct observation of the muscles provides more accurate information than imaging studies. This knowledge provides valuable information when reoperation may be required.

**Managing V-Pattern Strabismus.** V-pattern strabismus in patients with craniosynostosis is usually not due to simple IO overaction. The causes may include excyclorotation of the globe; anomalous vectors of muscle action; a shorter orbital floor, causing increased arc of contact; and loss of fusion, as described earlier. When imaging studies show torsional anomalies of the horizontal rectus muscles, the best outcome is with vertical transposition in the appropriate direction by at least one-half tendon width to collapse the V. For example, if a scan shows vertically displaced LR muscles in a patient with V-pattern exotropia despite "clinical" IO overaction, it is prudent to elevate the LR tendons half a tendon width at the time a recession procedure is performed on the muscles.

V-pattern exotropia is the more common finding and often is associated with torsional anomalies in rectus muscle positions, especially the LR. The preferred surgical approach is suprplacement of both LR muscles, combined with recession if the exodeviation in primary gaze is more than 15 PD. If the LR muscles have anomalous insertions, the postoperative response may not be as predictable as in other patients.Parents must be informed preoperatively of the unpredictability of the surgical response and the possible need for more than one procedure. If esodeviation rather than exotropia is present, the MR muscles may be recessed and transposed inferiorly or the LR muscles resected and transposed superiorly. A-pattern strabismus is rare. (See also Chapter 14.)

On the other hand, if the LR muscles are at the approximate normal horizontal meridian, this may represent true IO overaction that may or may not be associated with SO underaction or paresis. These patients require some form of IO weakening procedure. Because of altered muscle length-tension properties, and globe and arc contact, they may be prone to upgaze restriction simulating an acquired Brown syndrome. If the surgeon favors IO surgery, Bell's phenomenon should be documented preoperatively to avoid inadvertent nocturnal lagophthalmos (Fig. 30-15). If Bell's phenomenon is strong and IO overaction is severe, anterior transposition of the IO may be appropriate. However, if the overaction is moderate and there is some doubt about the integrity of Bell's phenomenon, particularly in children who have yet to undergo an inferior LeFort III orbital advancement, a less fortified IO weakening procedure such as simple recession, myectomy, or Z-tenotomy might be safer. The results are similar to those of IO weakening procedures performed in other patients. If there is no IO overaction, and no displacement of the LR muscles on imaging, the V pattern may be associated with SO underaction. In such a case, bilateral SO tucks may be considered.

**Managing Absent or Fibrotic Muscles.** The presence of a poorly functioning extraocular muscle is easily noted on clinical examination. For example, the classic "falling eye" seen as the globe moves from adduction to abduction may herald a fibrous or absent SR. Confirmation that the muscle is merely a slip or strand of fibrous tissue, however, requires operative observation. Unlike the congenital fibrosis syndrome in which muscles are tight, the fibrous strands in these cases usually have very little tension and the muscle behaves as paretic rather than restrictive.

The surgeon must have planned several options preoperatively and discussed them with the parents at the time informed consent is obtained. In the individual with an already excyclorotated muscle cone, predicting the outcome from
surgery is difficult. If there was a complete LR palsy and no evidence of muscle displacement on the computed tomo-
gram, one might consider transposing the vertical rectus muscles. In over a decade of experience I have not used such a procedure, because I believed that there was a greater chance of the patient’s condition becoming worse rather than improving postoperatively. This problem deserves further study, possibly in a multicenter setting, because of the rarity of cases (see also Chapters 27 and 36).

**COMPLICATIONS**

Corneal exposure is an almost unique risk to these pa-
tients, much like adults with dysthyroid strabismus. The powerful combination of an insufficient Bell’s phenomenon in a shallow, rotated orbit and inadequate support for the lower lid from the floor can lead to a corneal ulcer that requires emergency care. The presence of dacryostenosis may also be worrisome because of the risk of endophthal-
mitis. Preoperative antibiotics and probing (and lacrimal apparatus reconstruction in some cases) are indicated.

Heroic surgery on muscles that insert together, such as the LR and IO, may lead to unpredictable postoperative results with worsening of the deviation vertically and/or hori-
izontally. Even with a huge V pattern, I will often delay surgery in patients whose eyes are straight in the primary position and fusing. Only when the primary-position deviation changes will surgery be considered.
Reoperation, most often for undercorrection, is frequently necessary in these patients. Surgeons should use meticulous surgical technique and handle tissues gently to minimize scarring. Deep sweeps of the muscle that may bring orbital fat forward should be avoided.

Regular postoperative evaluation is an important aspect of ongoing care. Patients who undergo repeated cranial surgery may temporarily defer patching or penalization for amblyopia. Treatment needs to be intensified once amblyopia progresses or vision deteriorates before strabismus repair is possible.

**Conclusions**

Strabismus in craniosynostosis syndromes is a most challenging problem. The ophthalmologist has to assess unique and often bizarre motility patterns in the context of multiply involved ocular and other organ systems. The treatment, as much as possible, should be the same as for any child with nonsyndromic strabismus. However, repeated cranial and plastic reconstructive procedures, concern about developmental milestones, and the need to coordinate ophthalmic assessment and treatment efforts with those organized by other members of the craniofacial team make the overall situation more complicated and involved. Despite these challenges, the medical center approach seems to be the ideal way in which to deliver the highest standard of coordinated care to these otherwise severely affected patients.

**REFERENCES**

NYSTAGMUS: CLINICAL EVALUATION AND SURGICAL MANAGEMENT

MICHAEL X. REPKA, MD

The management of nystagmus has interested ophthalmic surgeons for many years. Although the etiology and the timing of onset differ for the various types of nystagmus, the associated symptoms can be quite similar. Both medical and surgical approaches have been used in an attempt to relieve the ophthalmologic symptoms. The most important and troubling symptoms include decreased vision, oscillopsia, and anomalous head posture or torticollis.

Historical Perspective

Attempts to minimize the symptoms of nystagmus have continued for much of the 20th century. An optical approach to correcting torticollis was suggested by Metzger in 1950. Prisms were used with the apex oriented toward the null point of the nystagmus in an effort to improve both head posture and binocular vision.

Surgical methods were proposed even earlier. Colburn, in 1906, attached the rectus muscles to the periosteum of the orbital wall in an attempt to dampen the muscle movement and thus reduce the amplitude of the nystagmus. This method has no contemporary support. More recently, a variety of surgical approaches have been tried to stabilize the eye. Kestenbaum, in 1953, suggested an operation to pull the eyes away from the null point to reduce the anomalous head posture. Each eye undergoes recession and resection, with both eyes being moved in the same direction. In the same year, Anderson described recessing the yoke rectus muscles that pulled the eye in the direction of tonically directed gaze. Both head posture and visual acuity improved after each of these operations.

In 1954, Goto performed resections on the medial rectus (MR) of one eye and the lateral rectus (LR) of the fellow eye. The surgical design was to pull the eyes away from the direction of the anomalous head posture. Blatt and Kruzun suggested an operation in which the rectus muscles in the field of gaze of the head posture are transposed, minimizing their effect in the field of action. Arruga reported that the posterior fixation operation seemed to reduce the amplitude of nystagmus in patients who have had retinal detachment surgery.

Clippers suggested creating an exotropia surgically. This induced exotropia led to convergence effort and dampened the nystagmus in patients with binocular fusion. The rationale was based on the widespread observation that convergence improves vision in patients with congenital nystagmus.

Operations weakening all the rectus muscles in the plane of the nystagmus (horizontal or vertical) have also been performed. A procedure involving four retroequatorial horizontal rectus muscle recessions was reported by Bietti. Keeney and Roseman suggested that free tenotomies of the rectus muscles might serve to dampen the ocular excursions. Neither of these operations has passed the test of time.

The current surgical management of nystagmus has evolved from these early reports of optical measures and surgical procedures. It is safe to say that there is—as yet—no single best method. Treatment must still be individualized. Modern management involves (1) correcting or relieving anomalous head postures and (2) dampening the nystagmus to decrease oscillopsia or improve visual acuity. Surgical, pharmacologic, and optical methods are employed.

Genetics

The genetics of congenital nystagmus usually correlate with the sensory cause of the nystagmus. For example, autosomal recessive inheritance characterizes most forms of oculocutaneous albinism. Patients with no sensory deficit and generally good visual acuity are considered to have the “motor” form of nystagmus (also known as congenital
Idiopathic nystagmus). Both the sensory and motor forms of nystagmus may be inherited in an autosomal dominant, autosomal recessive, or sex-linked manner. Congenital idiopathic nystagmus predominates in males, many of whom likely have the X-linked mutation. The X-linked phenotype of congenital idiopathic nystagmus exhibits variable expressivity and incomplete penetrance.

**Clinical Characteristics**

Nystagmus has typically been divided into congenital, acquired, and latent types. The most common type evaluated and managed by strabismologists is congenital nystagmus. Most of the discussion in this chapter is therefore directed at the management of this condition.

**CONGENITAL NYSTAGMUS**

Earlier in this century, congenital nystagmus was divided into motor and sensory forms with jerk and pendular waveforms. This division now is recognized as arbitrary and should be discarded from contemporary thought about congenital nystagmus.

Congenital nystagmus is an involuntary, bilateral ocular oscillatory movement. It usually begins during the first 6 months of life. However, waveforms compatible with congenital nystagmus can be seen on eye movement recordings to develop much later in life. The magnitude, frequency, and tracing of the nystagmus waveform seems to vary from day to day. Many workers in this area as well as parents comment that the magnitude of nystagmus decreases as the patient grows older. Congenital nystagmus has even been reported to disappear over time.

**Clinical Findings**

The eye movements in patients with congenital nystagmus include a predominantly horizontal pendular movement disturbance in primary position, which becomes a jerk waveform in lateral gaze. The position of minimal movement may be straight ahead or may be in an eccentric direction of gaze, either horizontally or vertically. This position is known as the null zone or minimal intensity zone.

**Waveform.** Eye movement recordings demonstrate a slow drift of the fovea away from the target (slow phase), followed by a saccadic refixation (fast phase). Both eyes move conjugately and at the same rate. Ocular occlusion does not seem to affect the amplitude or direction of the movements. The slow phase of congenital nystagmus exhibits an increasing velocity. This form of nystagmus disappears during sleep. In some patients with congenital nystagmus, the fast phase follows the rotation of the drum or stripes.

Horizontal jerk-type nystagmus is by far the most common waveform manifest of congenital nystagmus. Vertical, elliptical, and torsional waveforms do occur, but less often. Additional unusual waveforms are also seen. They include periodic alternating nystagmus, which accounts for as many as 10% of cases. Congenital see-saw nystagmus without any imaging abnormality has also been reported. This form of nystagmus exhibits alternating incyclotorsion and elevation of one eye with excyclotorsion and depression of the fellow eye. See-saw nystagmus is often associated with abnormalities of the diencephalon. If the patient is young or the history of onset is vague, neuroimaging is necessary to exclude a tumor or hydrocephalus.

**Visual Acuity.** Visual acuity is reduced in nearly all patients with congenital nystagmus. The degree of reduction is related to the type of sensory defect. For patients with a neurologic cause of nystagmus, vision is compromised by both the eye movement disorder and any abnormality of the afferent visual system. For patients with no evident sensory defect, there is a mild diminution of distance visual acuity, roughly correlating inversely with the intensity of the nystagmus.

Children with most forms of congenital nystagmus generally have better visual acuity at near fixation than at distance fixation. The relative improvement at near is believed to be secondary to convergence, which damps the eye movements. This change would presumably improve the foveation time or foveation fraction (the fraction of time the fovea is positioned on target divided by the total time). However, electro-oculographic measurements of eye movements have shown that visual acuity improves at near fixation even when there is no apparent reduction in the amplitude or frequency of nystagmus. Thus, the reason for the improvement remains uncertain.

**Accommodation.** Accommodative ability is deficient in patients with most types of congenital nystagmus. These patients have a much greater depth of focus, suggesting imprecision of the accommodative system. This is related in part to abnormal visual experience very early in life and presumably a coexisting amblyopia. There was no clear correlation in this study between accommodative ability, the magnitude of the nystagmus, and its etiology. It would seem prudent to take this deficiency into account when considering spectacle correction.

**Head Position.** Most patients tend to hold their head nearly straight ahead, with the null zone very nearly coincident with the primary position of gaze. However, when the null zone is not in primary position, most patients adopt a head position that moves the eyes to a position in space where the null zone is directed toward the target. The nystagmus will reverse direction on either side of this position.

Head posturing is usually not constant. It tends to be noted mainly when recognition of fine detail is required. This is especially true for distance fixation (for example, while viewing television or the blackboard in a classroom). At other times patients do not use the eccentric gaze position. Head postures seem to be used far less often for near vision. This distance-near disparity in torticollis is noted in about 70% of patients with anomalous head postures. In about 30% of patients, however, there is a mismatch between the null zone and the anomalous head posture; that is, the null zone and the head posture were not coincident.

Other as yet unidentified factors thus play a role in head posture in many patients with congenital nystagmus. Some of these potential factors include the most recent gaze direction and the length of time spent viewing in that direction. The important finding with respect to management is that repositioning the eyes to the point of minimal nystagmus intensity may not always provide the optimal position. It is correction of the torticollis itself rather than a recording of
eye movements that must be used to determine appropriate therapy and to judge the outcome.

Congenital nystagmus may also be associated with head oscillations, which are seen most often during visual tasks. Most of the time this adaptation does not appear to be compensatory, except in a few patients with abnormally deficient vestibulo-ocular reflexes. It has been postulated that there may be benefits—as yet unmeasured—of the head shaking to visual performance.

Modulating Factors. The amplitude of nystagmus in many patients seems to diminish with near fixation, attempted convergence, and increasing age. Clinically measured visual acuity often is found to be better at near than at distance. This is true even when eye movement recordings show no change in the waveform. More recently, studies have failed to show consistent improvement in nystagmus intensity or visual acuity with near fixation. Thus, this widely accepted clinical finding is not yet well explained.

It has long been taught that convergence suppresses the waveform in patients with congenital nystagmus. Convergence produces a secondary esotropia, a clinical entity called nystagmus blockage syndrome (described later). Electromyography has demonstrated co-contraction of the LR and MR muscles during convergence. Theoretically, this could suppress the nystagmus. Nystagmus intensity is reduced in 10% of patients on near fixation. This is believed to be due to convergence, not accommodation. Convergence has rarely been associated with an increase in horizontal nystagmus.

In cases of latent nystagmus, it has been confirmed that damping of the nystagmus is associated with the development of esotropia. Convergence also plays a role in altering the waveforms of acquired nystagmus. In patients whose nystagmus is associated with multiple sclerosis, convergence may either reduce or increase its intensity.

Convergence also takes place as an effort to modulate the waveform in vestibular nystagmus. Even in the absence of vision, convergence has suppressed the nystagmus amplitude by 47%. The waveform of congenital nystagmus diminishes in amplitude with advancing age. Alternatively, some patients may reduce their nystagmus by adopting a peripheral gaze position of adduction. The tonic innervation needed to maintain this alignment will reduce or mask the underlying nystagmus. This can lead to improved visual function at the expense of torticollis. The amplitude of nystagmus may also be reduced by active eyelid closure.

Anxiety will often make the nystagmus waveform more intense and degrade visual function just when the patient may be especially depending on it. This situation may make visual tasks such as driving more difficult.

Vestibulo-ocular Reflex Abnormalities. Many patients with congenital nystagmus have an abnormal vestibulo-ocular reflex (VOR). The normal VOR allows the eyes to remain fixed on a target by moving opposite and equal to any head motion. This is readily demonstrated by viewing the optic nerve with a direct ophthalmoscope and gently moving a patient’s head back and forth. If the VOR is normal the optic nerve will stay fixed. With an abnormal VOR, however, the image of the optic nerve will not remain in the center of the viewing field, allowing the VOR to be used to reduce the nystagmus intensity.

Oscillopsia. Oscillopsia is an illusion that the visual environment is moving. Dell’Osso and Leigh have suggested that foveation periods promote visual stability and suppress oscillopsia. A loss of visual stability and consequent retinal slip will reduce visual acuity and produce oscillopsia. A change in lighting of the visual environment also can lead to oscillopsia in patients with congenital nystagmus.

Complaints of oscillopsia seem related to high-intensity oscillations. The need to minimize oscillopsia may cause head posturing. Although common in acquired nystagmus, this is uncommon in congenital cases.

The magnitude of oscillopsia is usually less than that of the nystagmus. Thus, the brain must provide some compensation for the retinal slip. Motion detection may somehow be deficient in patients with congenital nystagmus. However, this cannot be the sole reason for a lack of oscillopsia in patients studied under normal conditions, because oscillopsia may occur if the eyes are artificially stabilized.

Well-developed foveation periods of sufficient duration must occur in the same plane as the nystagmus. These periods may be as brief as 15 milliseconds.

Etiology

Sensory and neurologic causes of congenital nystagmus and idiopathic cases are recognized. Similar waveforms are seen with each of these types. The neurologic substrate of congenital nystagmus remains unknown. There has been much interest in the abnormal afferent visual pathways seen in albinism and in how this miswiring may relate to the nystagmus. This anomalous afferent input, however, cannot be the sole cause of the nystagmus.

The particular mix of congenital nystagmus cases that a physician might see is strongly related to the type of practice. Several large retrospective studies suggest that sensory deprivation from a multitude of causes is the underlying etiologic factor in most cases. Careful examination can demonstrate a sensory cause in 91% of patients. The most common sensory defects are associated with the many phenotypes of oculocutaneous albinism or with optic nerve anomalies. Each collectively represents 30% to 50% of reported cases in retrospective studies. The optic nerve anomalies include both hypoplasia and optic atrophy. The third most frequent group of causes relate to retinal abnormalities such as the heterogeneous group of disorders termed Leber congenital amaurosis.

Other causes include congenital cataracts, congenital glaucoma, achromatopsia, congenital stationary night blindness, and aniridia. For these patients an electroretinogram (ERG) can be invaluable in arriving at the correct diagnosis. Down syndrome also seems to be frequently associated with congenital nystagmus; reported rates vary from a low of 9% to nearly 20%.

Of 200 patients from my practice with congenital nystagmus (unpublished observation), clinical examination, ERG, and neuroimaging established a sensory deficiency in 90%. Sensory diagnoses were equally divided among optic nerve hypoplasia/atrophy, albinism, and a variety of anterior and posterior segment ocular anomalies. The remaining 10% of patients, almost all with excellent vision, were believed to have nystagmus secondary to abnormalities in the efferent mechanism. They included patients with neurologic disease and others without any structural cause in the central nervous system. Patients without either a sensory defect or neurologic
The cause of this asymmetry based on the fixing eye is eyes are open. The waveform is identical to that of true creased or not apparent at all when both eyes are uncovered. Consistent with Alexander law as applied to latent nystagmus recording of the waveform readily distinguishes latent and congenital nystagmus. Many patients with esotropia and manifest nystagmus (both latent and congenital) have been mistakenly thought to have nystagmus compensation syndrome.

The syndrome is seen in a patient with early-onset variable esotropia who is orthotropic with manifest nystagmus while inten tiv e and esotropic with minimal nystagmus while attentive. These patients likely become esotropic because of sustained convergence. Specific attention to pupillary constriction during esotropia will aid the diagnosis. Inferior oblique muscle overaction and dissociated vertical deviations occur much less frequently than with infantile esotropia.

**LATENT NYSTAGMUS**

Latent nystagmus is a type of congenital nystagmus that may not always be recognized in the first 6 months of life. It is generally observed after one eye is covered. The nystagmus is conjugate and binocular. The direction of eye movements as related to the covered eye is diagnostic. The viewing eye drifts slowly toward the nose (slow phase) with the refixation fast phase directed toward the temple. When the cover is moved to the fellow eye, the slow phase shifts direction, with the viewing eye again moving toward the nose. The alternating direction of the nystagmus is pathognomonic of this form. Its accurate recognition may obviate the need for excessive neurologic evaluation.

The amplitude of nystagmus tends to increase on abduc tion and decrease on adduction of the fixing eye. This is consistent with Alexander law as applied to latent nystagmus. The amplitude of latent nystagmus is markedly decreased or not apparent at all when both eyes are uncovered. A type of latent nystagmus may also be observed when both eyes are open. The waveform is identical to that of true latent nystagmus, and it has been termed manifest latent nystagmus. In this situation, one eye fixes on the target. The cause of this asymmetry based on the fixing eye is unknown. Patients who develop amblyopia or lose vision in an eye may develop the manifest form of latent nystagmus.

Clinical examination of nystagmus waveform cannot reliably differentiate between types of nystagmus. Eye movement recordings of the waveforms readily distinguish latent nystagmus from the congenital and vestibular types. The most important feature is the change in direction of the fast phases with a change in the fixing eye. The clinical observer will note that the amplitude of the nystagmus increases with occlusion of one eye. A coexisting esotropia will remain approximately the same size under cover. Latent nystagmus exhibits fast phases of decreasing velocity. In contrast, manifest congenital nystagmus will not change its direction with placement of a cover but may change with a concomitant alteration in size of the esotropia.

Many patients with infantile-onset nystagmus may have both the latent and congenital forms. In Dell'Osso’s series of patients with early-onset manifest congenital nystagmus, about 20% had a latent component.

Latent nystagmus seems to occur almost exclusively in patients with infantile-onset strabismus. It may be found in up to half of patients with infantile esotropia. It is also frequent in the setting of dissociated vertical deviation and inferior oblique overaction. Latent nystagmus does not indicate a structural or progressive abnormality of the central nervous system. Neuroimaging is not indicated if clinical examination shows the typical change in direction of the waveform with occlusion.

Although the cause of latent or manifest latent nystagmus remains unknown, a number of theories have been proposed. They include abnormal proprioception, abnormal cortical binocularity, asymmetries of monocular optokinetic nystagmus, and defective egocentric localization. The monocular optokinetic reflex is more brisk for objects moving nasally than temporally in patients with latent nystagmus and in children younger than 4 months of age. With the establishment of binocular vision, cortical control takes over the optokinetic reflex and the asymmetry disappears.

The management of this form of nystagmus is limited to correcting strabismus with spectacles and/or surgery and treating any coexisting amblyopia. Successful vertical strabismus surgery in a 13-year-old patient eliminated the manifested portion of the disease. Similarly, amblyopia therapy has been proposed for converting nystagmus to the latent form when visual function improves.

**Infantile Unioocular Blindness with Bilateral Nystagmus**

A peculiar form of bilateral horizontal nystagmus may occur in patients with uniocular blindness. The nystagmus is of latent type, in which the waveform shows slow phases of decreasing velocity. It may be associated with media opacities or retinal anomalies in the macular region. The blind eye acts as the occluder, making manifest what might otherwise have been a latent nystagmus. The nystagmus disappears in adduction, leading to the preferential gaze position. Helveston and his colleagues called this entity "unilateral esotropia."

**ACQUIRED NYSTAGMUS**

Patients with acquired nystagmus have prominent symptoms that are often disabling. Many of them are unable to pursue their usual work. The most common symptoms include oscillopsia and decreased visual acuity, both related to image motion of stationary objects on the retina. Torticollis is less frequent than in congenital nystagmus, but it may occur. Vertigo is a common feature of nystagmus associated with lesions of the vestibular pathways.

**Periodic Alternating Nystagmus**

Periodic alternating nystagmus (PAN) is a conjugate horizontal jerk nystagmus characterized by periodic changes in
direction. The changing direction of the waveforms is caused by an actively shifting null zone. This form of nystagmus occurs most frequently as an acquired abnormality in patients with multiple sclerosis, posterior fossa malformations, or lesions of the vestibulocerebellum and vestibular nuclei. It may also develop after acute visual loss.

Less commonly, PAN occurs in infancy with no associated pathology. Until recently, PAN was considered to be an uncommon form (5%–9%) of congenital nystagmus.2,41 PAN was frequently associated with albinism. Anomalous head postures are seen in up to 95% of cases but vary over time. The cause of all forms of PAN remains unknown.

Oculopalatal Myoclonus

Oculopalatal myoclonus is a central nystagmus usually caused by brain stem or cerebellar infarction. Postmortem examination of the brain demonstrates hypertrophy of the inferior olivary nucleus.43 The ocular movements are generally pendular and horizontal, but they may also be vertical and are at times disconjugate. This form of nystagmus may correspond to intrinsic scopolamine hydrobromide. Unfortunately, such therapy is only transiently beneficial and is difficult to use in ambulatory patients. Retrobulbar injection of botulinum neurotoxin A may help some patients for 6 months or longer.78

Vestibular Nystagmus

Vestibular nystagmus is caused by vestibular end-organ dysfunction or damage to the brain stem. Peripheral vestibular nystagmus is associated with the acute onset of severe vertigo and exhibits a mix of horizontal, vertical, and torsional movements. Visual fixation dampens the nystagmus and suppresses vertigo. There may be significant nausea and vomiting. Vestibular nystagmus of central or brain stem origin is usually purely horizontal, vertical, or torsional. Visual fixation does not seem to affect the waveform intensity. There is mild vertigo, and other brain stem signs are prominent.

The slow phase obeys Alexander law and varies with orbital position. It is greatest when the eyes are turned in the direction of the fast phase. This probably reflects the superimposition of gaze-evoked nystagmus on the vestibular nystagmus.

Upbeat and Downbeat Nystagmus

Vertical nystagmus in either an upbeat or downbeating direction is generally associated with the central disruption of vestibular connections.

Lesions of the cranio-cervical junction are associated with downbeat nystagmus. Many other associated conditions have been reported, including multiple sclerosis, brain stem infarction, head trauma, use of anticonvulsants, lithium therapy, vitamin B12 deficiency, Wernicke encephalopathy, and alcohol abuse.64 The nystagmus increases when the eyes are directed inferiorly (Alexander law). The fast phase also seems to increase on lateral gaze. An unusual form of downbeat nystagmus may be seen transiently in normal infants,48 disappearing during the second half of the first year of life.

Upbeat nystagmus is mild in the primary position and increases in upgaze. There is no change on lateral gaze. Causes include cerebellar disease, multiple sclerosis, tumors of the brain stem, and congenital abnormalities of the afferent visual system. It also may occur transiently in normal infants.65

Opsoclonus

Opsoclonus is a conjugate eye movement disorder featuring chaotic multidirectional saccades without an intersaccadic interval. The movements persist during sleep. It occurs most commonly after nonspecific encephalitis. Its occurrence in children should prompt a search for occult neuroblastoma. The workup should include estimating urinary catecholamines and imaging of the head, chest, and abdomen. The eye movement disorder of opsoclonus is believed to represent a paraneoplastic effect of the tumor, resulting in antibody-mediated neuronal injury. Opsoclonus may also result from a paraneoplastic effect of other carcinomas in adults, such as oat cell carcinoma of the lung and adenocarcinomas of the breast and endometrium.

Treatment with corticosteroids, adrenocorticotropic hormone, and clonazepam provides some symptomatic relief. Some moderation of the opsoclonus follows successful resection of an associated tumor, although this response is neither immediate nor always complete. Children with opsoclonus secondary to neuroblastoma eventually recover after treatment with adrenocorticotropic hormone.54

Diagnosis

CLINICAL EXAMINATION

Measurement of Visual Acuity

The accurate measurement of visual acuity is complicated by the presence of nystagmus. Retinal slip may degrade visual acuity in many patients. A latent component may further reduce the measured acuity if special precautions are not taken. These issues commonly arise with patients having infantile strabismus with latent nystagmus. Binocular acuity should be recorded before occlusion.

Methods of measuring monocular vision while avoiding total occlusion include fogging the other eye with plus spheres, polarizing lenses, central field occlusion, and the red-green dichromatic slide test. The particular method used should be recorded. Consistency between examinations will avoid artificial changes in visual acuity arising from different testing techniques. Because many patients will have better visual acuity at near, consistent test distances are needed to ensure confidence in the results (see also Chapter 1).

Patients with nystagmus have reduced visual acuity; superimposed amblyopia is also frequent. It is often difficult to determine the relative contributions of amblyopia and nystagmus to reduced acuity.

Measurement of Head Turn

Many earlier reports on the surgical and medical management of nystagmus have estimated head postures. Over the past few years it has become standard practice to attempt to quantify the torticollis. The head turn is measured in degrees of rotation of the anteroposterior axis of the head away
Figure 31–1. A goniometer, a device commonly used to measure the range of joint mobility, can be used to measure the degree of head turn. To measure head turn, the patient is asked to fixate a detailed target at distance, adopting any needed head posture to see the target clearly. A videotaped program is a good target for children and adolescents. The center of rotation is placed over the center of the skull. One arm of the goniometer is directed along the visual axis toward the target. The other arm is oriented along the anteroposterior axis of the head. The angle between these two is read from the scale of the built-in protractor, quantifying the head turn in degrees.

from the visual angle. An articulated protractor called a goniometer (Fig. 31–1), found in orthopedic examination rooms, is very helpful. One arm of the device is oriented along the visual axis and the other along the anteroposterior axis of the skull (see also Chapter 1).

Anterior Segment Evaluation

Examination of the pupils is an important part of the workup for every nystagmus patient. Bilaterally sluggish pupils or an afferent pupillary defect suggests an optic nerve or retinal etiology. In patients with nystagmus and poor vision, careful attention must be paid to the direction of pupillary movement when illuminated. Dilation in response to increased light, or so-called “paradoxical pupil,” is common in patients with congenital photoreceptor abnormalities such as achromatopsia, Leber congenital amaurosis, cone dystrophy, and congenital stationary night blindness.

The iris is carefully examined for transillumination defects. This finding is often indicative of ocularcutaneous albinism. Such defects are found in nearly every patient with ocularcutaneous albinism but in only half of those with ocular albinism. Punctate defects occur in a small proportion of normal persons. The diagnosis of oculocutaneous albinism should be made in the presence of reduced pigmentation, macular hypoplasia, and, occasionally, optic nerve pathology.

Posterior Segment Evaluation

The posterior segment should be examined giving very close attention to the optic nerve and the macula. Because of the magnified view, optic disc examination with a direct ophthalmoscope through dilated pupils is more sensitive than indirect ophthalmoscopy. Both optic nerve atrophy and hypoplasia are more difficult to diagnose accurately using the indirect ophthalmoscope. Whenever possible, one should consider using a 90-D or 78-D lens (both contact and non-contact) to provide a three-dimensional evaluation of the optic nerve head and macula. The macula should be assessed for the absence of a normal pigment and vascular pattern surrounding a well-delineated fovea (Fig. 31–2). Hypoplasia of the macula is most easily identified from lack of a normal foveal reflex, suggesting ocularcutaneous albinism. This is unusual in normal patients or in other disease states.

Figure 31–2. Clinical photographs of the retina of a 1-year-old girl with congenital sensory nystagmus diagnosed with incontinentia pigmenti on the basis of a neonatal rash and typical skin scarring seen subsequently. Careful examination of the retina of these children is crucial to determining the correct diagnosis. A. Photograph of the posterior pole of the left eye demonstrates no clear foveal reflex and large retinal vessels passing within the fovea, suggesting abnormal macular differentiation. B. The early arterial phase study demonstrates an absent foveal avascular zone in the setting of a poor foveal reflex. The nystagmus in this infant likely is a result of the inability to develop an adequate fixation reflex from the abnormal fovea.
In older patients, fluorescein angiography may be used to document the normal vascular anatomy of the fovea, including the presence and size of the foveal avascular zone (see Fig. 31–2B).

**LABORATORY EVALUATION**

**Electroretinography**

Electroretinography is an important part of evaluating patients with congenital nystagmus. This test is not uniformly available, nor is there widespread familiarity with performing it in children. Many laboratories have not established accurate normal values for infants and toddlers. Nevertheless, if the test is available it should be considered in all children having congenital-onset nystagmus, normal irides, normal optic nerves, and reduced vision. By eliminating those children with other obvious causes of sensory loss, the number of tests needed is dramatically reduced.

The test may be done using topical anesthesia, conscious sedation, or general anesthesia. A 30-minute period of dark adaptation with pupillary mydriasis is advisable to maximize the response. Monopolar contact lens electrodes are preferred for most patients because of the stronger signal-to-noise ratio. A foil electrode on the eyelid may also be used. When the study is done using topical anesthesia, a pacifier or bottle should be available to help calm the infant or toddler. Minimal electrodiagnostic evaluation includes testing with a white light stimulus and single flash (scotopic ERG) and flickering flash with a rate of more than 10 Hz (photopic ERG).

ERG serves to detect retinal disease causing sensory congenital nystagmus. The possibilities include Leber congenital amaurosis, achromatopsia, and congenital stationary night blindness. The test may also be indispensable in diagnosing some neurodegenerative disorders such as neuronal ceroid lipofuscinosis, particularly the late infantile and juvenile subtypes.

**Neuroimaging**

Infants and children selected for diagnostic neuroimaging usually will have reduced vision, a normal ERG, and abnormal optic nerves. Patients with atypical congenital nystagmus waveforms, such as see-saw nystagmus and periodic alternating nystagmus, should also undergo imaging studies. Additionally, patients in whom a congenital onset is uncertain may require imaging. On the other hand, if an older patient is seen in consultation many years after the onset of nystagmus, imaging may be deferred even in the absence of a confirmed history of early onset.

Neuroimaging with enhancement is an important part of assessing patients with acquired nystagmus. Special attention is given to the suprasellar cistern and posterior fossa. However, neuroimaging is unnecessary in evaluating one form of acquired nystagmus—uncomplicated latent nystagmus. This diagnosis is made by the clinician based on a history of strabismus and reversal of the jerk phase with alternating occlusion. Latent nystagmus has not been associated with structural neurologic abnormalities.

**Eye Movement Recordings**

Eye movement recordings performed over the past several decades have led to remarkable advances in our understanding of the physiology of normal and abnormal eye movements. They have allowed neuroscientists to postulate mechanisms for both normal and abnormal eye movements.

Clinical examination of a patient with nystagmus can usually determine that the nystagmus is horizontal, vertical, torsional, or elliptical but does not reveal the specific waveforms. From eye movement recordings, the waveforms of congenital nystagmus have been divided into at least 12 different pendular, jerk, and mixed-pattern waveforms. These seem to occur in both sensory and idiopathic forms of congenital nystagmus, without predilection for any particular type. Furthermore, the characteristics of the waveform in congenital nystagmus correlate only loosely with the clinical findings.

Eye movement recordings elegantly illustrate the particular waveform present at the time of the examination (Fig. 31–3). This may change over time, often beginning as a pendular wave but adopting jerk characteristics as the infant matures. Waveform analysis does not accurately differentiate the sensory and idiopathic forms of congenital nystagmus.

Noninvasive techniques of eye movement recording such as Purkinje image tracking have extended these studies from the laboratory to the clinical setting. For the most part, such tracings will confirm that a patient has congenital nystagmus, latent nystagmus, or PAN. Only when PAN is identified would eye movement recordings alter the treatment plan.

**Treatment**

**NONSURGICAL MANAGEMENT**

The cardinal intervention for nystagmus patients is ensuring that an accurate refraction has been performed and appropriate spectacles prescribed. Children should be allowed to sit in the front of their classrooms, hold reading materials close, and adopt any anomalous head posture that enhances their visual performance. Many such behaviors are initially rejected by educators who are unfamiliar with what is required for optimal visual function.

**Treatment of Amblyopia**

Patients with nystagmus are at risk of developing amblyopia. It is often superimposed and requires close attention to correcting risk factors of strabismus such as anisometropia and astigmatism. The approach to a patient having both nystagmus and amblyopia is similar to that used in amblyopic patients lacking nystagmus. All anisometropia and astigmatism are corrected initially, and acuity is monitored for several months. If vision does not improve, amblyopia must be treated. Both penalization and occlusion methods can succeed. It may not be necessary to distinguish between subtypes of congenital nystagmus when deciding on the best treatment of amblyopia for a child with nystagmus.

The treatment of amblyopia in patients having both congenital and latent nystagmus is difficult. Acuity in the poor eye is degraded by amblyopia, nystagmus, and any sensory abnormalities that are present. For patients with latent nys-
NYSTAGMUS: CLINICAL EVALUATION AND SURGICAL MANAGEMENT • 411

Figure 31-3. Eye movement recordings of a patient with square-wave jerks before (top tracing) and 1 month after (bottom tracing) the injection of 25 units of retrobulbar botulinum toxin. The amplitude of the eye movements was significantly reduced after the injection. There was no clinically significant ophthalmoplegia in the orbit that was injected. Visual acuity during the 2 months after the injection improved from 20/100 to 20/30. Upward deflections in the tracing represent movements to the right, downward deflections represent movements to the left.

tagmus, occlusion of the sound eye further degrades acuity in the amblyopic eye. Therefore, methods that do not occlude the eye and induce latent nystagmus seem preferable for these patients. The examiner should consider using penalization methods such as fogging with plus lenses and atropinization. Atropine drops, either 0.5% or 1.0%, are placed in the sound eye each morning. Compliance is monitored by examining the pupil. For any penalization method to be effective, fixation must be shifted to the nonfixing eye. This may induce latent nystagmus, increase retinal slip, and potentially degrade visual acuity. The patient should be periodically reexamined to ensure that fixation is maintained by the amblyopic eye at the appropriate fixation distance. For instance, the atropinized amblyopic eye should be preferred for near. This approach is not a panacea. Some patients with latent nystagmus will develop manifest nystagmus even with appropriate pharmacologic penalization. Nonetheless, the clinician may continue the treatment for 6 to 10 weeks to detect a response.

Theoretically, penalization methods that keep both eyes open might be better for patients with latent nystagmus or congenital nystagmus with a latent component. However, occlusion should not be ruled out in the management of amblyopia and nystagmus. For patients with congenital nystagmus, treatment is identical to that used for any other patient having refractive or strabismic amblyopia. Monitoring focuses on sequential visual acuity to disclose both improvement and occlusion amblyopia.

Perhaps unexpectedly, occlusion is also an effective treatment for amblyopia in the setting of latent nystagmus and may be preferred by other specialists. Many practitioners reduce the number of treatment hours to make occlusion more acceptable. This approach has, however, proved ineffective; patching for longer periods—days instead of hours—is advantageous. Longer patching lessens the difference between the intensity of nystagmus with either eye fixing. There is less nystagmus and no oscillopsia when the less preferred eye is fixing. The improvement in nystagmus is accompanied by as much as one line of improvement in visual acuity.

Like all amblyopia therapy, compliance and the initial level of visual acuity are the best predictors of a successful outcome. Consistent with amblyopia treatment in general, patients with anisometropic amblyopia have better outcomes. Sensory defects are common in patients with congenital nystagmus, and these patients will not gain the same level of improvement as those with normal visual sensory systems. The clinician must titrate the intensity of therapy against the likelihood of success and discontinue treatment when it clearly is not succeeding.

Spectacle Correction

Treatment of Refractive Error Including Astigmatism.

Many patients with congenital nystagmus will have significant ametropia. This refractive error should be corrected as part of the initial treatment. Careful cycloplegic refraction should be carried out. Repeated refractions are necessary as the patient grows older. A change in refractive error may not be noticed because of poor visual acuity.

Accommodation may be deficient in patients with congenital nystagmus. Dynamic retinoscopy should be performed before dilation. In this test, the examiner asks the patient to look at a distant target and then at a near accommodative target held just below the light source of the retinoscope. The examiner will see the reflex change toward neutrality. This confirms the presence of accurate accommodative ability. If accommodation is deficient, a bifocal correction should be provided.

Minus Lenses. Overminus treatment with spectacles or contact lenses may reduce the amplitude of nystagmus and improve visual function. These effects are related to stimulation of accommodative convergence. Patients chosen to have this treatment must show evidence of good accommodative facility.

Partial Field Occlusion. An unusual optical method of decreasing the intensity of congenital nystagmus, based on partial field occlusion, was suggested by Sasso. "Porthole" glasses that had a clear 10-degree central field but an occluded periphery were prescribed. Thirty-eight children treated for an average duration of 5 years showed mild...
improvement in visual acuity, but advancing age was not taken into account.

For some patients having torticollis and nystagmus, occlusion of parts of the peripheral visual field reportedly alleviates anomalous head posture. However, the data are inadequate for thoroughly evaluating this concept. This approach lacks corroboration in the literature. It probably would not be well accepted by many patients today. The method should be reserved for patients who wish to try any measure, regardless of efficacy or cosmetic problems.

Prisms

Prisms may be used in two different ways to improve the function of a patient with nystagmus: (1) they may be placed before both eyes to correct an anomalous horizontal or vertical head turn; or (2) they may be so oriented as to improve visual acuity, usually by stimulating vergence eye movements.

Head Posture. Prisms minimize a head turn by reorienting the visual axis toward primary gaze. The eyes remain in the preferred gaze position (zone of minimal intensity) relative to the anteroposterior axis of the head. The head, however, can resume a more natural relationship with the environment because the prisms have redirected the visual axis. Often the preferred eye is held in adduction. Base-out prism before the fixing eye will allow a more normal head posture.

Equal-power prisms are placed before each eye and oriented in the same direction so that the apex is in the direction of preferred gaze, and the base in the direction of the head turn. For a patient with eyes held in right gaze with a left head turn, the apices of both prisms are oriented to the right. Theoretically, prisms may be oriented in any direction to correct horizontal, vertical, as well as diagonal head posture. Prisms of similar power are used in fusing patients; the power can be varied in patients with any heterotropia.

Large-power prisms are needed to correct even modest head turns. Typically, treatment will begin arbitrarily with 20-PD prisms. Press-On prisms (Optical Sciences Group, Petaluma, CA) made with Fresnel optics are most suitable for this application. These prisms are fairly inexpensive, but they are difficult to clean and often blur images, further degrading the quality of vision. This method is best for older patients who cannot undergo surgery but is also acceptable for long-term use in younger patients. A successful short-term trial of prism correction (a type of prism adaptation) can help some patients appreciate the potential benefits of surgical repair. These patients may gain a more realistic expectation of the planned surgery.

Prisms also may be used to correct residual head posture after surgery for torticollis. Although Fresnel prisms may be effective, large ground-in prism optics can be used at this stage.

Visual Acuity. The reason to use base-out prisms to improve visual acuity is to stimulate convergence and so reduce the intensity of nystagmus. In many patients this results in improved distance and near visual acuity. Responsive patients have binocular vision. The prism stimulates fusional vergence and thereby improves visual acuity. The prism power is usually split between the two eyes. Both ground-in and membrane-style prisms may be used, but the latter can be adjusted to achieve the best correction with the least asthenopia or diplopia. This therapeutic option seems best reserved for highly motivated patients having relatively good vision, normal fusion, and only modest nystagmus.

Contact Lenses

Corneal contact lenses, both soft and hard, have been used to improve visual acuity in patients with congenital nystagmus. Some patients experience dampening of nystagmus and consequently improved visual acuity. The mass of the contact lens is believed by some to cause inertial damping of the eye, but this was refuted by studies showing that a search coil contact lens has a negligible effect on eye movement. Another explanation for this phenomenon is sensory or tactile feedback from movement of the lens edge against the palpebral conjunctival surface. This theory is supported by findings that topical anesthesia abolishes the phenomenon.

Contact lenses have not worked for many of my patients. However, there are enough clinical reports in the literature to warrant a clinical trial. Transient oscillopsia has been reported after removal of contact lenses. A rebound phenomenon was suggested, in which the waveform intensity increased or the foveation fraction changed when the lenses were removed. Patients having a trial of contact lenses should know that short-lived oscillopsia is a possibility.

Red contact lenses may help improve visual function when nystagmus is associated with achromatopsia. The reason for this improvement is not clear. Use of the lens has been suggested for preserving long-term retinal function.

Other Optical Devices

This treatment category endeavors to improve the symptoms of nystagmus by stabilizing the retinal image. Such a strategy reduces the magnitude of image movement by limiting image slip along the retina. When this occurs, visual acuity may improve through an increase in the foveation fraction. In addition, some patients experience a decrease in oscillopsia because of reduced image motion.

An optical device to accomplish this feat was proposed by Rushton and Cox. It consists of a high-minus contact lens combined with a high-plus spectacle lens. Rigid contact lenses of – 58.00 or – 28.00 D are paired with spectacle lenses of +32.00 or +20.00 D. Stabilization of up to 90% is possible using this system. The prismatic effect of the large plus lens is the key feature. This lens images all objects at the center of rotation of the eye, irrespective of the exact direction of the visual axis. Thus, if the eyeball rotates, light rays from the object will remain focused at the same virtual point within the eye. The minus contact lens then refocuses the image on the retina. Because the contact lens moves with the eye, there should be no prismatic effect from the high power lens. This has the added benefit of magnifying the central 30 degrees of the visual field. The peripheral field is not stabilized, leading to a ring scotoma. The depth of focus is dramatically reduced, making it difficult for patients to position reading material.

This device is useful for specific tasks while at rest, such as reading, writing, or watching television. It has not proved useful for mobility because of the altered perceptions of the environment. For the same reason it is contraindicated for
Pharmacologic Management

There is little enthusiasm for treating congenital or acquired nystagmus with systemic medications. Most drugs hypothetically augment inhibitory neurotransmitters or inhibit excitatory neurotransmitters within the central nervous system. The most common drug used is baclofen for congenital nystagmus, and periodic alternating nystagmus. Minimal efficacy also is claimed for 5-hydroxytryptophan in treating congenital nystagmus.

Currie and Matsuo treated 10 patients having downbeat nystagmus and oscillopsia with clonazepam. The patients had primary-position nystagmus that intensified in downward gaze. A dose of 1 to 2 mg reduced nystagmus intensity, usually within 1 hour, and the effect lasted 2 to 6 hours. Visual acuity often improved, but sedation was a common side effect. The authors suggested taking a smaller dose (0.25–0.5 mg) before reading or other important visual activity. Enthusiasm for this drug has waned in recent years.

Perhaps the most useful drug for treating eye movement disorders has been carbamazepine, which is widely used for superior oblique myokymia. Careful hematologic monitoring is required when this drug is given (see Chapter 15).

Pharmacologic Denervation

Botulinum toxin type A blocks neuromuscular transmission by binding to receptor sites on the cholinergic nerve terminals to restrict acetylcholine release. Investigators have used this drug in two distinct ways to dampen nystagmus. The first method involves injecting each of the four horizontal rectus muscles. Approximately 3 units of toxin reconstituted in a volume of 0.05 mL are injected into each muscle under electromyographic guidance. The second method places a larger single dose of drug into the retrobulbar space. This is a simpler and less expensive method, and is easier to use as an ongoing therapeutic option. Twenty to 25 units of botulinum toxin are given, using a standard retrobulbar injection technique through the lower lid or inferior conjunctival fornix.

All reports note that the amplitude and intensity of the nystagmus are reduced. Many have included eye movement recordings, which objectively document improvement in eye movements (see Fig. 31–3). Common side effects include diplopia and ptosis. Ophthalmoplegia is greater with direct muscular injections than when using the retrobulbar technique. The effectiveness of these methods has been extremely variable; the effects typically last only a few months.

Acquired Nystagmus. Varying results have been reported when treating patients with oscillopsia secondary to acquired nystagmus after brain stem infarction. Better success was achieved using the retrobulbar route. The treatment effect lasted 5 to 13 weeks. Ptosis was less common than with direct muscle injection, especially of the superior rectus (SR), possibly because an intracranal injection is directed away from the levator muscle. Filamentary keratitis is a possible complication.

There may be subjective and objective improvement in distance visual acuity. Eye movement recordings have confirmed a reduction in nystagmus intensity using this technique. Visual improvement lasted about 8 weeks for most patients. For those with oculopalatal myoclonus, improvement after each injection lasted up to 6 months. Troublesome diplopia may occur in some patients with binocular vision, limiting the usefulness of the procedure in the patient’s view. In addition, movements of the fellow, noninjected eye may worsen. Repeated injections may be required.

Botulinum toxin treatment may be of some value in selected patients. Ptosis is relatively infrequent. Diplopia is probably the greatest concern, because it is potentially more debilitating than the nystagmus and oscillopsia. The method is best reserved for patients who already have an oculomotor neuropathy and diplopia or unilateral optic neuropathy. These patients are more likely to improve and to be happy with the outcome. All patients must be prepared to occlude one eye should diplopia become bothersome.

The retrobulbar technique has a therapeutic advantage over direct rectus muscle injections. Logistically, two or more perimuscular injections are avoided. Also, the method of administration is familiar to every ophthalmologist. Finally, the retrobulbar injection may be less expensive because special monopolar electrode needles and an electromyographic device are not needed.

Congenital Nystagmus. There is less information on the efficacy of botulinum treatment for congenital forms of nystagmus. Using direct muscle injection, four adult patients with vision ranging from 20/40 to 20/70 in their best eye all experienced improved vision to at least 20/40 and resumed driving. Long-term improvement stabilized at about 1 line of acuity.

I have been disappointed with the results of retrobulbar injection of 20 units of botulinum toxin in patients with congenital idiopathic nystagmus and congenital sensory nystagmus associated with oculocutaneous albinism. No more than 1 Snellen line of improvement was found 4 weeks after the injection. One patient unexpectedly capitalized on the induced partial ophthalmoplegia and concomitant reduced eye movement intensity to insert her contact lenses by herself and keep them centered on the cornea; she continues to have quarterly injections. In none of these patients have ocular excursions been significantly limited.

Biofeedback and Acupuncture

Both biofeedback and acupuncture have been touted as effective ways of improving visual acuity and lessening nystagmus. Unfortunately, no peer-reviewed reports substantiate these anecdotal reports.

Surgical Management

Surgery may be performed to improve anomalous head posture or visual acuity (Fig. 31–4). Although the exact mechanics of the procedures have changed, these indications are the same as they were 40 years ago. In the first indication, the goal of surgery is to pull the zone of minimum intensity (null zone) toward the primary position. In the
second, dampening eye movement increases the foveation fraction and consequently improves visual acuity.

The best time at which to perform surgery has never been rigorously studied. It is reasonable to delay surgery in orthotropic patients having congenital nystagmus and an anomalous head posture until school is imminent. By then the child will be 4 years of age and will have had several repeatable and more reliable examinations demonstrating head posture and ocular alignment. For children with heterotropias, earlier intervention may provide the best opportunity for bifoveal fixation to develop. With an acquired nystagmus and torticollis, it is prudent to wait at least a year to be certain that the abnormal eye movements are consistent and the head turn is stable.

**Horizontal Anomalous Head Posture**

The most popular procedure for horizontal head postures is a modification of the operations described by Anderson and Kestenbaum. Most surgeons today operate simultaneously on all four rectus muscles when there is no ocular misalignment. A recession of the LR in the abducted eye is combined with a resection of the ipsilateral MR muscle, and a recession of the MR muscle of the adducted eye is coupled with a resection of the ipsilateral LR muscle.

**Kestenbaum-Anderson Procedure (Classic Maximum).** The most widely accepted modification is that of Parks, consisting of variously graded resections and resections. A surgical dosage guideline using a “5-6-7-8” rule was described in which each eye receives the same total of 13 mm of surgery. Measurements are based on a 5-mm MR muscle recession coupled with an 8-mm resection of the ipsilateral LR muscle; the fellow eye undergoes a 7-mm recession of the LR muscle coupled with a 6-mm resection of the MR. These guidelines were established at a time when resections of more than 5 mm on the MR muscle were believed to be unwise. This surgery is known today as the “classic maximum.”

Eye movement recordings performed on a few patients before and after this operation have shown that it decreases nystagmus intensity in practically all directions of gaze. The zone of minimal intensity is broadened and shifted toward the primary position.

Undercorrections are common for head turns of more than 30 degrees and may exceed 10 degrees in some cases. Visual acuity did not improve in one series, but many patients had associated strabismus and amblyopia, precluding a meaningful conclusion with regard to the effects of this surgery on visual acuity.

**Augmented Modification of the Kestenbaum-Anderson Procedure.** Realizing that “classic” surgery was not adequately correcting patients with large head turns, a 40% increase in surgical dosage has been suggested (“classic plus 40%”). Head postures are improved, but patients may be left with aduction deficit. Rarely, a new heterotropia is induced.

**Modified Anderson Procedure.** Von Noorden has revived interest in the Anderson operation, in which only two recessions are performed, but suggested a 12-mm recession on the LR muscle of the abducted eye and a 10-mm recession of the MR of the fellow (adducted) eye. The outcome of this enhanced Anderson operation is unknown. The procedure leaves two horizontal rectus muscles unoperated, making it particularly advantageous for patients having torticollis present in both the horizontal and vertical directions. Surgical procedures may be performed in both directions with a minimal risk of anterior segment ischemia.

**Recommendations.** There is little standardization of head posture measurements preoperatively or after surgery, nor are masked outcome examinations available for any of the reported studies. Similarly, the decision to perform additional surgery for recurrence is largely left up to the individual practitioner. It would seem that undercorrection is common and that the classic procedure should be avoided in all but...
Table 31–1. Surgical Dosage (in mm) for Correction of Horizontal Anomalous Head Posture*

<table>
<thead>
<tr>
<th>Magnitude of Horizontal Head Turn</th>
<th>&lt;20°</th>
<th>21°–44°</th>
<th>≥45°</th>
</tr>
</thead>
<tbody>
<tr>
<td>Right head turn (move eyes to the right)</td>
<td>Right medial rectus recession</td>
<td>6.0</td>
<td>6.5</td>
</tr>
<tr>
<td>Right lateral rectus resection</td>
<td>9.5</td>
<td>10.5</td>
<td>11.25</td>
</tr>
<tr>
<td>Left medial rectus recession</td>
<td>7.25</td>
<td>7.75</td>
<td>8.5</td>
</tr>
<tr>
<td>Left lateral rectus recession</td>
<td>8.5</td>
<td>9.0</td>
<td>9.75</td>
</tr>
<tr>
<td>Left head turn (move eyes to the left)</td>
<td>Right medial rectus recession</td>
<td>7.5</td>
<td>7.75</td>
</tr>
<tr>
<td>Right lateral rectus resection</td>
<td>8.5</td>
<td>9.0</td>
<td>9.75</td>
</tr>
<tr>
<td>Left medial rectus recession</td>
<td>6.0</td>
<td>6.5</td>
<td>7.0</td>
</tr>
<tr>
<td>Left lateral rectus resection</td>
<td>9.5</td>
<td>10.5</td>
<td>11.25</td>
</tr>
</tbody>
</table>

*Surgical dosages rounded to the nearest 0.25 mm.

the smaller head turns, using the 40% augmented surgeries for most patients with a larger head turn. New heterotropias are distinctly uncommon, a reassuring finding.

These patients should be carefully examined and counseled to ensure realistic expectations of the outcome. These include improvement in the head turn and limitation of gaze. Improvement in visual acuity is not a certainty. Prior to performing surgery, it is crucial to be certain that the head posture is consistent over time. Examination on several different days and over a period of at least 30 minutes is essential. The quality of the nystagmus can change, leading to an altered head posture. The best known example of this phenomenon is periodic alternating nystagmus, but there are other examples of torticollis changing over longer periods. The angle of torticollis is measured while the patient views a distant accommodative target. This is easiest when performed with a goniometer (see Fig. 31–1). The exact amounts of surgery may be read from a table (Table 31–1).

Outcome. The stability of surgical correction of anomalous head postures is incompletely reported in the literature (Fig. 31–5). Scott and Kraft found that 4 of 7 patients observed for 4 years or longer were stable, whereas 3 drifted to a larger undercorrection. Biglan and co-workers, in a study of 34 patients, found that head turn in 13 was sufficiently undercorrected to warrant a second procedure.

For patients with strabismus, particularly those who have had previous strabismus surgery, generalizations about surgical dosages are irrelevant. If there is a heterotropia, the surgical approach and dosages need to be modified. For these patients, the required amounts of surgery are difficult to predict. There are three basic approaches: (1) perform the procedure on both eyes with “best guess” dosages; (2) perform staged surgery, correcting the fixing eye first for torticollis and correcting the resultant strabismus at a subsequent surgery; or (3) perform surgery on both eyes simultaneously, but use one or more adjustable sutures to provide flexibility in correcting coexisting strabismus.

For small heterotropias, it is usually sufficient to adjust surgical dosages to account for the position of the nonfixing eye. I prefer to avoid the first option of estimating the effects of adjusting the surgical dosages. It is simply too unpredictable. I would rather use the second option of two-stage surgery for young patients and some adults. In this sequence, surgical dosages for strabismus are determined only after the results of the first surgery are clear. I stage the procedures at least 6 weeks apart. For some adults, the use of adjustable sutures on the nonfixing eye may allow surgery to be performed in one stage.

Figure 31–5. A, Preoperative photograph of a 7-year-old girl with congenital idiopathic nystagmus and 40-degree left face turn. B, Two months postoperatively, the head turn is gone. At near fixation, there is 50 seconds of arc stereoacuity and no heterophoria, but there is a new intermittent left exotropia at distance fixation.
Vertical Anomalous Head Posture

Vertical-plane torticollis induced by nystagmus, manifest in either the chin-up or chin-down position, is less common than horizontal face turns. The head position orients the null zone of nystagmus toward the target. Because of its rarity, various procedures have been tried by several authors (Table 31–2), and outcomes can only be inferred from several cases series.

Both patients in whom Pierse performed either of his recommended procedures had improved head posture and visual acuity. Parks originally recommended 4-mm resections of the appropriate vertical rectus muscles, and 4-mm resection of the antagonist when the anomalous posture exceeds 25 degrees. This procedure was often associated with undercorrection, leading to current recommendations of an 8-mm recession and 8-mm resection for large torticollis. Roberts and colleagues combined recession of the inferior rectus (IR) muscle, often by 5 mm, with a 7-mm resection of the SR muscle for chin-up position. Undercorrection necessitated additional surgery in two of seven patients. They then agreed that an 8-mm recession and 8-mm resection should be performed.

For chin-down posture a large SR muscle recession (8–9 mm) combined with inferior oblique recession and anteriorization to the IR yielded not only improvement in head position but also better visual acuity in all seven patients who underwent the procedure. Visual acuity generally improved by only 1 line but was not studied in a consistent and masked manner.

Recommendations. In light of the limited data available, the best surgery for vertical-plane torticollis remains uncertain. Nonetheless, for chin-up postures, large recessions of the IR muscle and resections of the SR are required to obtain a satisfactory correction. I recommend 6 mm of recession surgery for torticollis of less than 25 degrees and 8 mm of surgery for angles greater than 25 degrees. Bilateral IR recessions may cause an A-pattern deviation because of weakened adduction in downgaze. The IR may be transposed nasally to avoid creating an A pattern (see Chapter 14).

For combined torticollis, there are insufficient data to suggest an appropriate approach. Scott and Kraft proposed that combined torticollis might be treated by vertically transposing the horizontal muscles subjected to an Anderson-Kestenbaum type operation. A second approach uses the exaggerated Anderson procedure to correct horizontal face turn first. The patient is reevaluated for vertical torticollis and could then undergo vertical recession surgery, possibly coupled with a resection.

Surgery to Improve Visual Acuity

Four Horizontal Muscle Recession Surgery. This procedure was described by Bietti to place the rectus muscles behind the equator to decrease their rotational effect on a given eye movement. The nystagmus is dampened and the foveation fraction increased, improving visual acuity. Nine of 14 treated patients experienced improved visual acuity.

Later results supported similar optimism. The frequency and amplitude of nystagmus were reduced in most patients, as demonstrated by electronystagmography. The amount of objective improvement in visual acuity is often modest, but the subjective improvement noted by patients tends to be greater. Aesthetic improvement is observed with a reduction in apparent ocular motion.

Von Noorden and Sprunger performed 10- to 12-mm recession on all four horizontal rectus muscles. Helveston and his colleagues placed the MR muscle 11.5 mm from the limbus and the LR muscle 13.0 mm from the temporal limbus. No patient had a reduction in best corrected binocular acuity or induced heterotropia. A preexisting head posture may improve. Surprisingly, ductions were only slightly limited.

Head posture surgery that shifts the null point will aggravate the unoperated direction of gaze in patients with PAN. These patients, however, may benefit from a retroequatorial recession procedure. A patient in von Noorden and Sprunger's series had an improved head posture and subjectively improved reading ability with attenuation of the nystagmus.

Recommendations. This procedure and its relatively limited outcome should be discussed with the patient. It seems best reserved at present (in the absence of good long-term outcome data or substantial improvements in acuity) for highly motivated nystagmus patients whose expectations are realistic.

The standard approach to performing surgery in a nystagmus patient must be followed. The surgeon should determine if there is a deviation or fusion. The LR muscles should be recessed 10 mm from the insertion and the MR recessed about 7.5 mm from the insertion. Greater recessions of the MR muscles have been associated with the development of consecutive exotropia.

Outcome. This surgical approach has been discussed for
almost 40 years, but the functional outcome remains unclear. There is a low risk of adverse complications, such as induced heterotropias and ductional limitations. Heterotropias have not yet been reported in the immediate postoperative period. More studies are needed to properly assess the value of this surgery for improving visual function in patients with congenital nystagmus. Long-term outcome assessments will document the development of new heterotropias (most likely exotropia) and the change in visual acuity.

**Artificial Divergence.** Patients who experience successful damping of nystagmus with large base-out prisms may benefit from surgery simulating these conditions. Good candidates must have fusion and must not develop a manifest exodeviation. A unilateral recession of the MR may be combined with resection of the LR. Alternatively, bilateral MR recession surgery may be chosen. Either of these surgeries will induce an exodeviation, which may then be reduced by fusional convergence to minimize nystagmus. In many cases, visual acuity will improve.

Spielmann and Laulan recommend MR recession surgery of 5 to 12 mm, based on the response to prisms. The prism power is increased until visual acuity improves without inducing diplopia. However, only 26% of 75 patients treated by the same authors had fairly mild improvement in visual acuity. More disturbing are the 9 patients who developed nonfusible exotropia.

### Torsional Torticollis/Head Tilt

There is a small body of literature dealing with the management of head tilt produced as an adaptation to torsional nystagmus. It is believed that fundus torsion (relative to the environment) may be associated with a compensatory moderation in nystagmus intensity. Surgery in these cases is designed to re-create that relative torsion with the head held upright. The surgery creates a tilt of the patient's subjective visual environment. A great deal of planning is necessary because surgery on cyclovertical muscles—if asymmetric and unbalanced—may create significant heterotropia.

Conrad and de Decker rotated the eyes by advancing or recessing the anterior portions of the appropriate oblique muscles. Resection in combination with advancement was required to obtain a satisfactory result. Presumably, this augmented the restriction of rotation back to the original position.

In a more recent publication, de Decker suggested a simpler procedure in which torsional movement of the eyes is created by using offsets of the horizontal rectus muscles. For a right head tilt, suprarelacement of the right LR muscle, infrarelacement of the right MR and left LR muscles, and suprarelacement of the left MR are carried out. The tendons are generally shifted one full tendon width. The muscles are reattached to the globe at the same distance from the limbus as they were before disinsertion.

Spielmann suggested slanting the insertions of each of the four rectus muscles. To produce exyclorotation, the temporal border of the SR muscle is recessed, as are the inferior border of the LR muscle, the nasal border of the IR muscle, and the superior border of the MR muscle. The biggest drawback to this procedure is the need to sacrifice all the rectus muscles that provide circulation to the anterior segment.

Two patients with a 30-degree tilt toward the shoulder were presumably adopting the posture that caused relative excyclotorsion of the eye on the side of the tilt. Pratt-Johnson designed a procedure to re-create excyclotorsion on the ipsilateral side, with an associated contralateral excyclotorsion. Tenectomy of the anterior two thirds of the ipsilateral superior oblique muscle was combined with contralateral inferior oblique myectomy and anterior lateralization (Harada-Ito) of the superior oblique. In the second patient, with severe amblyopia in one eye, surgery was performed only on the fixing eye with normal vision.

Von Noorden and colleagues suggested an alternative approach using the vertical rectus muscles rather than the oblique muscles. For a head tilt to the right, excycloduction of the right eye and incycloduction of the left eye are needed (Fig. 31–6). To accomplish this, the right SR muscle is transposed nasally and the right IR is transposed temporally, causing excyclorotation of the right eye. In the left eye, temporal transposition of the left SR muscle and nasal transposition of the left IR muscle provide the necessary incycloduction. Head tilt was eliminated in four of five patients operated on by this technique. One patient was lost to follow-up. Visual acuity improved slightly. Electro-oculographic eye movement recordings showed improved nystagmus amplitude when the head was held upright. There were no complications such as diplopia, new heterotropia, or a sensation of vertical tilting of the environment.

The best choice among these methods is not clear. The decision will be influenced by a history of prior strabismus surgery and the need to repair a coexisting heterotropia. Using rectus muscle offsets is often technically easier and does not seem to induce strabismus. Oblique muscle surgery may be advantageous when compromising the vascular supply to the anterior segment is a concern.

### Nystagmus Blockage Syndrome

Bilateral MR recessions, with or without posterior fixation sutures, and unilateral or bilateral recession-resection operations may be performed for nystagmus blockage syndrome. There is no clear best procedure. The rate of undercorrection and overcorrection is high irrespective of the method chosen. This argues against the use of posterior fixation sutures because of the possibility of reoperation. Von Noorden and Wong suggest large bilateral MR muscle recessions of 6 mm or more. The effects of this surgery have not been studied in a large population.

Prism adaptation (as described for accommodative esotropia) is a clinical method of determining the largest target angle for surgery that would not be associated with significant exotropia at distance. Patients with nystagmus blockage syndrome may benefit from preoperative prism adaptation if some fusional convergence is present. After determining the largest angle, and observing the clinical effect at distance fixation, prisms may be reduced as long as the patient remains orthotropic or slightly esotropic at distance. The amount of recession of the MR muscles is based on the prism-determined amount.
COMPLICATIONS

Induced Heterotropia

Any of the procedures described in this chapter may induce a new deviation. The risk seems remarkably low with the Kestenbaum-Anderson type of balanced surgery. A much greater risk exists with the four-muscle recession operations, after which many patients develop consecutive exotropia. Such new deviations are also possible after induced divergence operations. All patients who are contemplating such surgery to correct torticollis or improve visual acuity should be informed that subsequent surgery may be needed.

Long-Term Undercorrection and Overcorrection

Correction of torticollis is often associated with undercorrection. Even patients considered to be successes in the literature may have residual torticollis of as much as 15 degrees. The need for additional surgery may not be apparent until several years later and reoperation difficult. Repeated resections or—if less than maximal resections were performed—repeated recession surgery may be required. Such surgery is more likely to induce a new heterotropia.

Development of “New” Head Postures

Some patients will undergo apparently successful surgery in one dimension only to exhibit a significant posture in another direction. This possibility should be kept in mind when planning initial surgery. This is perhaps the strongest reason to consider von Noorden’s modification of the Anderson procedure.

REFERENCES

CHEMODENERVATION
CHEMODENERVATION THERAPY: TECHNIQUE AND INDICATIONS

KEITH W. McNEER, MD, ELBERT H. MAGOON, MD, and ALAN B. SCOTT, MD

In this chapter we review the background of botulinum toxin injection for strabismus, the technique of its use, and the application of this treatment to various strabismic disorders. Once one becomes adept with the technique and comfortable with its applications, it will seem indispensable for many cases in which one wants to change ocular alignment but does not wish to operate. It should not, however, be regarded as a general replacement for incisional surgery.

Historical Perspective

The first thorough investigations of botulinum toxin were undertaken by Kerner (see Gruesser for review of this topic) and published in 1817–1822. He followed the clinical course of 230 patients with botulism, making many original observations about causation, diagnosis, prognosis, and treatment. More importantly to our topic, he extracted the toxin from sausages, demonstrated its effects in animals, correctly concluded that it paralyzed skeletal muscles and parasympathetic function, and proposed its use as a therapeutic agent in neurologic diseases such as chorea that are characterized by excessive motor movement. Just when his experiments were defining the potential medical effects of botulinum toxin, his request to the Bavarian king for support was apparently turned down, because he abandoned his researches and turned toward holistic medical practice and romantic poetry, a scenario not foreign to us nowadays.

Over the next 75 years little was added until van Ermingen in 1896, after an epidemic of intoxication caused by improperly cured ham, described in great detail the organism, its toxins, and their effects in several mammalian species. Clinical botulism is mostly a result of improper food preparation and/or preservation. Public health and commercial interests at that time then supported the research, and a large literature developed on the organisms, their toxins, and the prevention and treatment of botulism. But it was 160 years after Kerner before the idea of therapy with botulinum toxin was implemented.

We began in 1970/1971 with injection of various drugs into extraocular muscles (EOMs) as an alternative to surgical treatment for strabismus. Among these was botulinum toxin, considered also by others (Crone, Bach-y-Rita, Jampolsky, Maumenee—personal communications) and actually used by Drachman to experimentally paralyze animal hind limbs without inducing systemic toxicity. Using crystallization techniques developed by the U.S. Army Chemical Warfare Department, Schantz prepared and generously supplied us and many other investigators with botulinum toxin. We chose type A botulinum toxin because, of the several types involved in epidemics, it had a strong muscle-paralyzing effect. This approach is still used to prepare Allergan’s Botox. The type A botulinum toxin from Porton Laboratories, Dysport (dystonia—Porton), is purified by slightly different techniques. Mouse LD/50 potency tests on these products done in our laboratory gave results in units that were equivalent in side-by-side trials in human blepharospasm (Botox, right eye; Dysport, left eye of the same patient). However, as labeled by the manufacturers, the clinical potency equivalence is variable; the average of several reports is about 3 units Dysport to 1 unit Botox.

Chemistry

Botulinum toxin molecules are long protein sequences (150,000 daltons) with three domains (active portions of the molecule) (Fig. 32–1). The first domain binds to one of six receptors found exclusively on nerve terminals; these are specific receptors for clostridial toxin. At this time, nobody knows why they are there. Endocytosis then brings the toxin into the nerve terminal within vesicles. The second domain allows the toxin to enter the nerve cytoplasm from the vesicles. The third domain acts as an enzyme, each toxin...
type cutting a different and specific site on one of the three proteins required to dock vesicles to the nerve membrane for exocytosis of acetylcholine.

In the past few years, the seven toxin types, the six nerve membrane receptors, and the three substrate proteins involved in exocytosis have been identified and sequenced and the specific amino acid sequences of the botulinum toxin molecule that are involved in receptor affinity and enzymatic activity have been identified, all in great detail.

**Mechanism of Action in Strabismus**

Initially, we thought that botulinum toxin treatment of comitant strabismus worked because the EOMs were permanently weakened by the induced denervation paralysis. Spencer and McNeer have shown changes in the outer (orbital) muscular layer of botulinum toxin–treated primate EOMs, lasting for many months. It is possible that this may reduce primary-position force (normally 8–10 g for a human horizontal rectus) without showing up in full active contraction (normally 60–100 g). But active contraction force returns fully to these muscles. We now believe that the muscles alter alignment of the eyes mostly by having changed their length when they were stretched or relaxed. Botulinum toxin is just a way of getting the eye into a new position so that the muscles can adapt their length. The propensity for this internal sarcomere reorganization varies from one individual to another. It also is dependent on the dose response, on creating a large angular change of alignment from the botulinum toxin injection paralysis, and on its persistence for at least 1 month. Therefore, it is not surprising that clinical responses to initial botulinum toxin injection are variable.

**Indications and Clinical Results**

The indications for botulinum toxin injection in strabismus are similar to indications for surgery, but there are important differences. Botulinum toxin injection in EOMs is most efficient in nonrestrictive strabismus. Severe restrictions caused by scarred, inelastic muscles favor surgery, because botulinum toxin depends on reciprocal agonist-antagonist readjustments of length-tension elasticity. Scarring and paralysis restrict the inherent ability of the muscle to alter its length-tension characteristics. Smaller deviations (horizontal or vertical) with minimal sensory changes have a superior prognosis and may routinely be managed successfully using botulinum toxin injection. Large-angle deviations, more frequently found in long-standing adult strabismus, can be expected to respond favorably, although repeated injections often are needed, resulting in a longer interval from the initiation of therapy to optimal correction. Despite this apparent disadvantage, a significant number of adults choose chronic botulinum toxin injections to achieve the desired result in place of surgery.

Tables 32–1 and 32–2 show that larger deviations are less fully corrected by one injection, that esotropia and exotropia generally respond similarly, and that children and adults respond alike. In general, there is a 30% to 40% chance that one injection will correct the deviation to 10 PD or less. Biglan and colleagues obtained a 38% correction rate with 1.3 injections, and Carruthers and Kennedy achieved a 29% rate in large-angle exotropia with one injection. Botulinum toxin has only a transient alignment effect in strabismus resulting from permanent nerve paralysis but an important effect on contracture of the antagonist to the paralyzed muscle. Botulinum toxin fills a void as a nonsurgical therapeutic method.

**PARALYTIC STRABISMUS**

**Sixth Nerve Paralysis**

*Acute cases* of any age or origin are followed without treatment for 3 to 4 weeks. If healing begins within a month, it typically will be progressive and complete. Adults will be rehabilitated and children seldom lose binocularity if alignment is restored in a month. After 1 month, if disabling diplopia persists and recovery is not progressing, or if a child remains esotropic in all gaze positions so that binocularity is threatened, then injecting the medial rectus (MR) on the affected side(s) is appropriate, even though many of these cases would resolve over time. In a randomized trial in ambulatory adults with paresis of diabetic or vascular origin, there was little long-term difference in recovery between a botulinum toxin–treated (86%) and an observed control group (80%). Thus, for these patients who have a generally good prognosis, the value of botulinum toxin treatment lies in earlier rehabilitation.

For more severe paresis of traumatic origin or those caused by intracranial pathology, recovery is often delayed several months or takes place very slowly. During this time, MR contracture progresses and esotropia increases. Botulinum toxin injection of the MR in these nonacute cases allows the lateral rectus (LR) to recover against an equally weak MR of normal length, rather than against a strong and shortened MR. There is virtually no risk of permanent overcorrection and the patient’s alignment is improved, serving both functional and cosmetic goals while awaiting healing of the LR. The data from Metz and Mazow indicate that there is a higher percentage of correction in such cases (Table 32–3).

Persistent esotropia 6 to 12 months after onset may be due to limited LR recovery or to MR contracture in varying
Table 32–1. Effect of Botulinum Toxin Injection on Strabismus in Children*

<table>
<thead>
<tr>
<th></th>
<th>No. of Patients</th>
<th>Average Deviation (PD)</th>
<th>Final Deviation of 10 PD or Less</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Preinjection</td>
<td>Postinjection</td>
<td>% Change</td>
</tr>
<tr>
<td>Esotropia</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>By Number of Injections</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>1 injection</td>
<td>146</td>
<td>28</td>
<td>9</td>
</tr>
<tr>
<td>&gt;1 injection</td>
<td>115</td>
<td>35</td>
<td>11</td>
</tr>
<tr>
<td>All patients</td>
<td>261</td>
<td>31</td>
<td>10</td>
</tr>
<tr>
<td>By Initial Deviation</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>8–24 PD</td>
<td>81</td>
<td>16</td>
<td>7</td>
</tr>
<tr>
<td>25–39 PD</td>
<td>108</td>
<td>30</td>
<td>11</td>
</tr>
<tr>
<td>40+ PD</td>
<td>72</td>
<td>50</td>
<td>11</td>
</tr>
<tr>
<td>All patients</td>
<td>261</td>
<td>31</td>
<td>10</td>
</tr>
<tr>
<td>Exotropia</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>By Number of Injections</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>1 injection</td>
<td>66</td>
<td>26</td>
<td>12</td>
</tr>
<tr>
<td>&gt;1 injection</td>
<td>29</td>
<td>27</td>
<td>15</td>
</tr>
<tr>
<td>All patients</td>
<td>95</td>
<td>26</td>
<td>13</td>
</tr>
<tr>
<td>By Initial Deviation</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>10–24 PD</td>
<td>43</td>
<td>17</td>
<td>7</td>
</tr>
<tr>
<td>25–39 PD</td>
<td>32</td>
<td>29</td>
<td>16</td>
</tr>
<tr>
<td>40+ PD</td>
<td>20</td>
<td>42</td>
<td>19</td>
</tr>
<tr>
<td>All patients</td>
<td>95</td>
<td>26</td>
<td>13</td>
</tr>
</tbody>
</table>

*Results in 356 patients (ages 2 months–12 years) with follow-up of 6–65 months (average, 27 months). Reduction rate of strabismus is broken down by number of injections and initial deviation as measured in prism diopters.

Table 32–2. Effect of Botulinum Toxin Injection on Strabismus in Adults*

<table>
<thead>
<tr>
<th></th>
<th>No. of Patients</th>
<th>Average Deviation</th>
<th>Final Deviation of 10 PD or Less</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Preinjection</td>
<td>Postinjection</td>
<td>% Change</td>
</tr>
<tr>
<td>Esotropia</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>By Number of Injections</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>1 injection</td>
<td>216</td>
<td>28</td>
<td>9</td>
</tr>
<tr>
<td>&gt;1 injection</td>
<td>159</td>
<td>32</td>
<td>14</td>
</tr>
<tr>
<td>All patients</td>
<td>375</td>
<td>30</td>
<td>11</td>
</tr>
<tr>
<td>By Initial Deviation</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>10–24 PD</td>
<td>132</td>
<td>17</td>
<td>6</td>
</tr>
<tr>
<td>25–39 PD</td>
<td>142</td>
<td>29</td>
<td>10</td>
</tr>
<tr>
<td>40+ PD</td>
<td>101</td>
<td>49</td>
<td>18</td>
</tr>
<tr>
<td>All patients</td>
<td>375</td>
<td>30</td>
<td>11</td>
</tr>
<tr>
<td>Exotropia</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>By Number of Injections</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>1 injection</td>
<td>133</td>
<td>28</td>
<td>11</td>
</tr>
<tr>
<td>&gt;1 injection</td>
<td>154</td>
<td>34</td>
<td>13</td>
</tr>
<tr>
<td>All patients</td>
<td>287</td>
<td>31</td>
<td>12</td>
</tr>
<tr>
<td>By Initial Deviation</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>10–24 PD</td>
<td>93</td>
<td>17</td>
<td>8</td>
</tr>
<tr>
<td>25–39 PD</td>
<td>112</td>
<td>30</td>
<td>11</td>
</tr>
<tr>
<td>40+ PD</td>
<td>82</td>
<td>50</td>
<td>18</td>
</tr>
<tr>
<td>All patients</td>
<td>287</td>
<td>32</td>
<td>12</td>
</tr>
</tbody>
</table>

*Results in 662 patients (ages 12–90) with follow-up of 6–83 months (average, 17 months). Reduction rate of strabismus is broken down by number of injections and by initial deviation as measured in prism diopters.
proportions. If there is abduction beyond the midline, good abduction saccades, or good LR force (30 g or more), then release of the MR contracture by botulinum toxin injection alone will often be fully curative. Of course, these patients respond well to simple recess-resect surgery, but the almost magical effect of botulinum toxin injection of the MR makes this simple office procedure attractive (Fig. 32–2).

In cases of permanent paralysis there is no place for botulinum toxin injection alone, because recurrence is inevitable. Surgery by lateral transposition of the superior rectus (SR) and the inferior rectus (IR) gives the largest amplitude of single binocular vision. The value of botulinum toxin here is twofold. First and most important, botulinum toxin injection lengthens and preserves the short MR so that this muscle, now the only active horizontal mover, has a large range of contraction-relaxation. In contrast, surgical recession of an already short and contracted MR restores alignment but leads to further shortening and a further reduction in range of motion. Also important is that botulinum toxin leaves the anterior ciliary artery supply of the MR intact, obviating the threat of anterior segment ischemia. The ability of botulinum toxin to release MR contracture makes it appropriate to wait a full 6 months after the onset before undertaking transposition surgery. Botulinum toxin injection may be done a few weeks before surgery or deferred until the time of surgery to allow accurate traction testing of the MR. It is then easily done under direct visualization. Injection is best deferred until postoperative alignment can be determined in cases in which MR contracture is mild—it may be unnecessary, and overcorrection could result. Injection several months after surgery has corrected several undercorrected transposition cases and may need to be repeated.

Third Nerve Paresis

Saad and Lee corrected three of four cases that had some residual MR function, and Metz and Mazow similarly corrected a majority of their cases using botulinum toxin. Botulinum toxin is very useful in late aberrant third nerve regeneration, where small deviations interfere with primary-position alignment. The muscles seem to be especially sensitive, and small doses should be used.

Fourth Nerve Paresis

Botulinum toxin injection is beneficial in unilateral fourth nerve paresis. Lozano Pratt corrected 9 of 9 acute cases with 17 to 30 PD of vertical deviation before injection at 12 months’ follow-up. Buosanti corrected 9 of 15 (3 others partially improved). One of us corrected 4 of 6 patients observed over 12 months. It is interesting that a series of inferior oblique (IO) injections done 15 years ago did less well, so that this treatment approach was (temporarily) abandoned. Higher doses and better technique explain the current results. All cases just mentioned were injections of the overacting IO muscle. Garnham and colleagues found botulinum toxin injection of the yoke IR more useful in both acute and chronic fourth nerve paresis. We agree that this is valuable in cases in which the vertical deviation is more pronounced in downgaze.

![Figure 32-2. Seven-year-old boy with comitant 30 PD of esotropia 3 months after spontaneous onset of left lateral rectus palsy. Top, Before injection. Bottom, One week after 1.2 units of botulinum toxin was injected into left medial rectus. Left abduction is full, indicating that the esotropia was due to medial rectus contracture.](image)

Table 32–3. Botulinum Toxin Correction of Nonacute Sixth Nerve Paresis

<table>
<thead>
<tr>
<th>No. Patients</th>
<th>% Recovered</th>
<th>Botulinum Toxin Treated</th>
<th>Control</th>
</tr>
</thead>
<tbody>
<tr>
<td>Unilateral</td>
<td>34</td>
<td>70</td>
<td>31</td>
</tr>
<tr>
<td>Bilateral</td>
<td>11</td>
<td>90</td>
<td>42</td>
</tr>
</tbody>
</table>


![Figure 32-3. Infantile esotropia. Top, Age 6 months, before injection. Middle, Age 9 months, 3 months after first bilateral medial rectus injection at 6 months of age. Bottom, Age 14 months, 5 months after second bilateral medial rectus injection.](image)
CHILDHOOD STRABISMUS

Infantile Esotropia

Treatment of infantile esotropia by simultaneous bimedial botulinum toxin injection is quite successful (Figs. 32–3 and 32–4). Table 32–4 shows the results of several independent series.

Table 32–4. Infantile Esotropia Treated by Botulinum Toxin (No Prior Surgery)

<table>
<thead>
<tr>
<th>Authors</th>
<th>No. Patients</th>
<th>No. Injections</th>
<th>% Corrected to 10 PD or Less</th>
</tr>
</thead>
<tbody>
<tr>
<td>McNeer et al</td>
<td>76</td>
<td>89</td>
<td></td>
</tr>
<tr>
<td>Scott et al</td>
<td>61</td>
<td>66</td>
<td></td>
</tr>
<tr>
<td>Gomez de Liano et al</td>
<td>107</td>
<td>73</td>
<td></td>
</tr>
<tr>
<td>Campos et al</td>
<td>50</td>
<td>76</td>
<td></td>
</tr>
</tbody>
</table>

The following comments are pertinent:

1. Perform simultaneous bimedial injection.
2. Inject as early as age 3 months. The good results in the series of Campos and associates were all in patients injected before age 8 months.
3. Repeat simultaneous bimedial injection with recurrence of esotropia exceeding 15 PD, increasing the dose unless ptosis is a limiting side effect.
4. Examine patient frequently, correcting any hyperopic refractive error.

Figure 32–4. Photographs of infantile esotropic patient treated with bimedial botulinum toxin injection. A, Patient at preinjection age of 12 months displays characteristic deviation of infantile esotropia. B, Patient receives bimedial 2.5 units of botulinum toxin in intramuscular injection under nitrous oxide anesthesia. C, Patient is orthotropic at both distance and near fixation 53 months after injection.

Acquired Esotropia

Acquired esotropia here refers to strabismus developing after infancy that is neither paretic nor accommodative. Bimedial injection of 2.5 units of botulinum toxin can be expected to provide motor alignment, but it is less stable and effective with intractable amblyopia. The best responses

Figure 32–5. Loss of fusion from removal of hyperopic correction. Top, Esotropia while wearing full contact lens correction. Middle, Two months after injection of right medial rectus with botulinum toxin. Bottom, Eighteen months after injection of right medial rectus.
are achieved if amblyopia can be improved or reversed before injection. Unilateral MR injection will often enhance or create hypertropia. Therefore, the hypertropic eye should be injected even if it is the fixing eye to help correct rather than worsen such vertical deviations.

**Intermittent Exotropia**

In children, the recommended schedule is simultaneous 2.5-unit doses in both LR muscles. This avoids the secondary vertical deviations and ptosis common after higher doses or unilateral injection. As with surgery, an exotropic deviation recurs in the majority of patients. An acceptable result thus means a lesser exotropic angle that can be comfortably controlled. In an investigation limited to botulinum toxin treatment of childhood intermittent exotropia, 31 32 patients were treated by one or more simultaneous bilateral Botox LR injections and followed 3 years from the final injection. Sixty-eight percent developed a stable deviation of 10 PD or less. Seven of the patients required surgery during the study period when injection failed to prevent recurrent exotropic drift. The response between ages 3 and 5 years was much better than in younger or older patients. With evidence of exotropia exceeding 10 PD, injection should be repeated.

**Cerebral Palsy**

Strabismus surgery for neurologically impaired infants and children is less predictable than in normal subjects; it has a high rate of overcorrection, and side effects of general anesthesia are more frequent. Botulinum toxin injection offers an alternative with a very low overcorrection rate (about 3%). Low-dose bimedial injections of 1.25 to 2.0 units are preferred with esotropia because the dose-response ratio is less predictable. Exotropia responds less strongly. The usual beginning dose is 2.5 units. Reinjections are often necessary, increasing the dose as indicated (Table 32–5). Although overcorrection is possible in esotropia, it has not been seen in exotropia. Strabismus in older children with cerebral palsy is treated by unilateral injection to avoid past pointing and balance problems from the induced paralysis.

### TREATMENT OF ADULT STRABISMUS

**Multiply Operated Strabismus**

Multiply operated strabismus patients unhappy with the surgical outcome are frequent candidates for botulinum toxin injection. The strabismus is often complicated by cicatricial restrictions, excessive muscle recession, and poor fusion. Evaluation of comitance, versions, and forced ductions will be useful in making a prognosis. Botulinum toxin injection will be less predictable, often yielding small corrections requiring multiple injections. But the ease of injection and the frequently excellent outcome warrant a trial in most such cases. Because suppression and amblyopia are frequent, the effects of botulinum toxin-induced paralysis on the sensory status is often of no concern. The series of Carruthers and Kennedy, 32 with 29% correction after one injection, is representative of this group.

There is a significant incidence of restrictions from injury or prior retinal or strabismus surgery, of secondary strabis-

### Table 32–5. Botulinum Toxin Dosage for Initial Injections

<table>
<thead>
<tr>
<th>Strabismus Type</th>
<th>Units</th>
</tr>
</thead>
<tbody>
<tr>
<td>Horizontal strabismus*</td>
<td></td>
</tr>
<tr>
<td>Under 25 PD</td>
<td>2.5</td>
</tr>
<tr>
<td>Over 25 PD</td>
<td>2.5–5.0</td>
</tr>
<tr>
<td>MR injection for LR palsy</td>
<td></td>
</tr>
<tr>
<td>Early (1–3 months)</td>
<td>1.0–2.0</td>
</tr>
<tr>
<td>Later or in conjunction with transposition surgery</td>
<td>2.5</td>
</tr>
<tr>
<td>Later for partially or fully healed palsy but with MR contracture</td>
<td>2.5</td>
</tr>
<tr>
<td>Vertical muscles</td>
<td></td>
</tr>
<tr>
<td>IR for comitant deviation</td>
<td>2.5</td>
</tr>
<tr>
<td>IR for thyroid</td>
<td>5.0</td>
</tr>
<tr>
<td>IO</td>
<td>2.5</td>
</tr>
<tr>
<td>SR (rare)</td>
<td>2.0</td>
</tr>
<tr>
<td>Children with infantile esotropia or exotropia (bilateral injections)</td>
<td>2.5</td>
</tr>
<tr>
<td>Weak muscles—myasthenia, external ophthalmoplegia, aberrant regeneration, cerebral palsy</td>
<td>1.0–2.0</td>
</tr>
<tr>
<td>Retrobulbar injection for nystagmus</td>
<td>25.0</td>
</tr>
</tbody>
</table>

*MR, medial rectus; LR, lateral rectus; IR, inferior rectus; IO, inferior oblique; SR, superior rectus.

*Further refinements: Smaller doses for MR, larger for LR. Smaller doses for smaller squints, larger for larger squints. Smaller doses for small women (occasionally very sensitive).

**Post-Retinal Detachment Strabismus**

The prognosis for curing strabismus resulting from retinal detachment surgery is guarded if there is severe scarring (Fig. 32–6). These patients have normal binocular vision in their favor. In most cases, the strabismic angle is modest, and it is always worth trying botulinum toxin injection. Scott 36 corrected 60% of cases with one injection, and Pettito and Buckley 31 corrected 80%. Scarred EOMs often require larger doses or a repeat injection. Injection of two muscles (e.g., the LR and IR simultaneously) is common.

**Thyroid Ophthalmopathy**

Botulinum toxin is useful in acute or active cases to relieve diplopia (Fig. 32–7). Maintaining alignment to prevent or treat chronic cases is much less secure; surgery
is avoided in 25% to 30% of cases. Most late EOM restrictions are fibrotic scars, precluding benefit from botulinum toxin injection. However, an occasional remarkable loosening of IR contracture shows that some restrictions are caused only partly by fibrosis and that internal muscle shortening plays a role. There is no way to predict this clinically except to perform a therapeutic trial.

Thyroid-induced esotropia offers the best prognosis for botulinum toxin injection. The medial rectus muscles are frequently affected in thyroid disease and the horizontal deviation is usually modest. Because multiple muscle surgery in thyroidal eye disease carries a higher risk of ischemia, use of botulinum toxin for the esotropia while operating on the vertical deviation is very useful.

**Post-Cataract Strabismus**

Immediate diplopia after cataract surgery in previously fusing adults usually presents as hypotropia of the operated eye secondary to IR contracture after retrobulbar anesthesia. When prisms are inadequate and the patient wishes to avoid IR recession, botulinum toxin injection of the IR will correct over 60% of cases with a single injection. A second useful application is in long-standing unilateral cataract with exotropia and diplopia after cataract removal. Injection of the LR will restore alignment and subsequently maintain fusion in over half of these patients.

**Postoperative Adjustment**

Avoiding multiple surgical procedures is an integral part of strabismus treatment. Botulinum toxin injections are an alternative to additional surgery for strabismus, particularly if the intended goal has not been achieved by recent surgery. Botulinum toxin may be used either early or late in the postoperative period or in conjunction with surgery, especially if the planned surgical procedure (e.g., transpositions, additional surgery on rectus muscles) may compromise the vascularity of the anterior segment.

**Intrinsic Muscle Disorders**

Unexercised EOMs and other tissues stiffen markedly in some persons. Strabismus in chronic myasthenia and progressive external ophthalmoplegia is surprisingly responsive to botulinum toxin when the eye is stiff on traction testing. The response is much less when the eye is readily moved.

**Nystagmus**

Carruthers' improved vision by 3 lines after direct intramuscular injection of botulinum toxin in 3 of 4 cases with purely horizontal congenital nystagmus. This resulted in vision of 20/40 (6/12) or better for several months. Acquired nystagmus with oscillopsia and reduced vision usually has a vertical or rotary component. A single retrobulbar injection creates ophthalmoplegia for 3 to 4 months to dampen such movements. This has dramatically improved vision in several cases from 20/400 to 20/40 (6/120 to 6/12). Because the induced ophthalmoplegia creates diplopia and marked spatial/balance problems, this works best for wheelchair-bound patients. Only one eye of an ambulatory patient should be injected in this manner.
Technique

INFORMED CONSENT

Informed consent should include reference to the intended overcorrection and its cosmetic implications, as well as possible diplopia and spatial disorientation lasting 1 to 2 months. This will require patching in about a third of adult cases, mostly for the first week or two, followed by adaptation that allows its discontinuance. A trial of patching should be considered for 1 or 2 days before injecting adults without suppression. Some elderly persons are physically unstable, and many busy adults are unable to drive and work with only monocular vision. Children adapt in a few days, even if fixing with the injected eye. About half of patients need retreatment within 2 years. Eye perforation has occurred in 1 in 1000 cases, but no patient has lost vision. Partial ptosis and vertical deviation occur mainly with MR injection, at a rate of about 25%. Fewer than 1% of patients still have ptosis 6 months after the injection. It helps patients to know that the injection itself is more frightening than painful, that the eye is made numb so that pain is minimal, and that the technique takes less time than the preoperative discussion and paper work. Advantages include the use of local anesthesia and doing a less-invasive procedure.

PREPARATION

The amplifier should be checked to ensure that the batteries are working. The needle should have an intact tip and a good coating of Teflon. A spare should be available in case the needle gets damaged or contaminated. The minute amount of white material in the bottom of the botulinum toxin bottle is the correct content. It is mostly salt, some protein, and 100 units of the toxin itself. Reconstitution with 2.0 mL of nonpreserved saline yields a solution of 5 units/0.1 mL; 4.0 mL gives 2.5 units/0.1 mL. Tuberculinsyringes are remarkably accurate; fractions of the dose may be accurately given by varying the volume injected. First, one should aspirate the volume of the proposed dose plus an additional 0.1 to 0.2 mL into the syringe. Then the injection electrode is firmly attached, and the excess is injected through the needle electrode to check for patency and spot a leak at the syringe/needle junction. Because an awkward position is often required to read the syringe marks, one should fill with the exact volume desired and empty it completely into the EOM. If there are two muscles to inject (e.g., bimedials in children), a second syringe can be filled, or the needle transferred to the second syringe, again with the exact dose.

DOSAGE

Nearly all initial doses are 2.5 units (see Table 32–5). Subsequent doses may be increased as much as 100% depending on the response to the earlier injection. Notice that dosage is not decreased for children and may even be increased for those with large deviations.

ANESTHESIA

Proparacaine (or tetracaine, cocaine, or 4% lidocaine) in a minimum of a drop a minute for three doses is used. A vasoconstrictor drop (e.g., epinephrine), given before instilling anesthetic drops, makes it easier to avoid the anterior ciliary vessels and enhances anesthesia. After the drops, a subconjunctival injection of 0.1 to 0.4 mL of depolarizing muscle over the muscle does not diminish electromyographic (EMG) activity and reduces discomfort, especially in a previously operated patient with scar tissue.

SEDATION

Diazepam or a similar oral drug, given an hour before the procedure, is helpful for the very apprehensive patient.

INJECTION TECHNIQUE

Placement in the muscle is crucial. The neuromuscular junctions, centered halfway back in the EOM, should be the target. Injection outside the muscle reduces the treatment effect and increases the likelihood of affecting adjacent muscles. Because the drug diffuses along needle tracks, a single accurate placement is most reliable. Steps that help in injecting horizontal muscles in an alert patient using an electromyogram amplifier are as follows:

1. Prepare the syringe with the appropriate dose of botulinum and attach the monopolar needle electrode as described earlier. Note the position of the needle bevel so the orientation in the orbit can be determined.
2. Attach a ground lead to the patient—at the forehead for the MR and laterally for the LR. Wipe this area beforehand with alcohol to ensure adhesion of the usual electrocardiographic leads.
3. Turn on the amplifier to half the volume and test the connections by touching the needle tip to the conjunctiva; it should make a loud “tick.”
4. Have the patient gaze at a target away from the field of action of the muscle. (For example, the patient should look temporally during injection of the MR.)
5. Insert the needle electrode, its bevel facing the muscle, through the conjunctiva 8 to 10 mm from the limbus and, avoiding large vessels, push it 5–6 mm posteriorly, keeping to the orbital wall side of the muscle, away from the globe. Slowly move the gaze target to the primary position to activate the target muscle.
6. While listening to the EMG signals, advance the needle tip toward the area giving off the loudest sound. When a crackling sharp EMG signal is heard, inject the fluid slowly. It is a good sign if the EMG sound diminishes with the injection, indicating that the solution pushed the nearby muscle fibers away from the tip. Leave the tip there for 15 to 30 seconds until the pressure of the solution diminishes. Withdraw the needle slowly while the patient maintains primary gaze. Make a note of how reliable the EMG response and injection were, for future reference.

Special Considerations in Children

A child should know what is happening and why, couched in positive terms. After age 6 years in an intelligent child, the technique can often be performed as in adults. Because it is hard to predict which child will cooperate, we often attempt it in the office, knowing that a substantial number...
of procedures will not be completed. For the successful ones we have saved time, anesthesia, and cost.

1. A quiet setting is essential. The parents should be present and holding the child’s hands. Proceed slowly. Telling the patient that to fear something approaching the eye is a normal protective mechanism helps to gain cooperation. The child is shown that the eyes can be anesthetized with drops and that the second set of drops (a vasoconstrictor such as epinephrine 0.1%) will be cold but will not sting. It takes more to anesthetize children: plan on five instillations. Demonstrate that a moist cotton-tipped applicator will be able to touch the eye and not be uncomfortable. The child should be told that, toward the end of the procedure, there may be some aching or sense of pressure, but that this signals the end of the procedure. (These techniques are similar to those one uses for other procedures with children such as foreign body removal.) A somewhat abbreviated version may be useful for anxious adults.

2. For children aged 1 to 6 years and for those unable to cooperate, ketamine or nitrous oxide sedation can be added (see Fig. 32–4B). Intravenous ketamine, 0.5 to 1.0 mg/kg, will preserve EMG activity and keep the patient relatively quiet for 2 to 5 minutes. This is less than the usual anesthetic dose. The anesthesiologist should be informed that general anesthesia is not desired but rather some degree of akinesia, amnesia, and light sedation. Remarkably little hallucination and postoperative effects are noted with intravenous ketamine. Topical anesthetic drops given before ketamine are essential. Intramuscular ketamine, 2 to 3 mg/kg, has a much longer duration of effect. The recovery period is also longer and hallucinations do occur.

Inhalational anesthesia is an acceptable alternative that does not require intravenous access. However, the EMG signal is much diminished, and many injections will be given without the benefit of complete EMG guidance. This is probably acceptable for the medial rectus muscle, which is the usual target in children and the least variable in position. A small posterior conjunctival incision allows injection with the needle tip 10 to 15 mm posterior to the insertion under direct visualization. This is an excellent alternative when access to an efficient facility and anesthesia coverage are available.

3. From age 3 to 12 months, depending on the strength of the child, it is usually possible to anesthetize the eyes with drops and simply hold the infant down for the injection. A papoose board and additional hands to hold the head of a strong infant cannot be held still, terminate the procedure and plan to do the injections under anesthesia.

Injecting Cyclovertical Muscles

Inferior Rectus. The IR is injected through the lower lid in thyroid or other cases with restricted supraduction. The needle must be angled nasally 23 degrees (straight back puts the needle into the LR!). If the IO is encountered, continue right through it—the IR should be behind it. One should be sure to wait 30 seconds after injecting and remove the needle slowly to avoid back-flow of botulinum toxin into the IO.

Superior Rectus. SR injection is just as effective as injection of any other EOM; but because of the induced ptosis, the SR should be treated only when a hypertropia cannot otherwise be treated. The ipsilateral IO is usually injected simultaneously. Full ptosis can be expected for 2 months. Sometimes residual ptosis of 0.5 mm may persist.

Inferior Oblique. The IO is very close to the conjunctival surface inferotemporally when the eye is looking upward. Injection is done rather anteriorly to avoid the IR.

Superior Oblique. Injecting this muscle is not recommended. SO overaction and SO myokymia have been the indications, but ptosis occurs uniformly; there is a resulting SO palsy, and the basic condition recurs after a few months.

TECHNICAL COMPLICATIONS AND DIFFICULTIES

Sometime the target muscle cannot be found. An unusually positioned muscle may be found by carefully tilting and translating the whole needle and syringe. Multiple needle thrusts should be avoided because they can cause hemorrhage, especially if the thrusts are made deep into the orbit.

The LR takes a course going backward and downward below the horizontal in some patients. Recognizing this normal anatomic variant and repositioning the needle and syringe as just described are usually effective.

The EMG signal will be decreased for many weeks after the first injection and may remain so even after clinical muscle activity has returned to normal.

Partial ptosis occurs in about 16% of adults and 25% of children. Complete ptosis is truly rare, and no case of amblyopia due to ptosis has been documented.

Vertical strabismus after treatment of horizontal rectus muscles of one eye occurred in 17% of adult patients. It persists for longer than 6 months in fewer than 1% of patients and is much more common in MR injection than in LR injection. The induced vertical deviation is usually a hypertropia after MR injection and a hypotropia after LR injection. One can take advantage of this phenomenon by injecting the MR of the hypertropic eye for esotropia and the LR of the hypertropic eye for exotropia whenever possible. The induction of vertical strabismus is much less common in children treated bilaterally; any vertical effect seems to balance out.

The rate of overcorrection at 6 months is 1.7%. Cases with muscles that respond strongly to botulinum toxin, resulting in overcorrection, are also easily reversed by in-
jecting the antagonist. Smaller doses (1.0–2.0 units) should be used.

Scleral perforation may be prevented by keeping the needle tangential to the globe until it is posterior to the equator and using the EMG signal to guide it. Highly myopic eyes, eyes where the injection is near a previous surgical site, and eyes with scleral buckles are particularly at risk.

Diplopia is the most common and annoying side effect. The patient should appreciate that an overcorrection is necessary for a long-term beneficial effect. Adults may require patching but may also be able to use a head turn to align the eyes. Patching is avoided in visually immature children except to prevent or treat amblyopia.

Because we usually treat the nondonominant eye, few patients develop spatial disorientation or past pointing because of the induced paralysis. Those who do are usually alternators or have part-time fusion. Patching the eye treats the problem, and adaptation usually occurs after a week.

Retrobulbar hemorrhage has occurred in about 0.2% of cases, usually after prior surgery. There is report of one eye decompressed by conjunctival incision, but this case and all others resolved without visual complications. The expected paralytic effect is not reduced.

Pupillary dilatation (Adie’s pupil) has occurred rarely, probably from needle injury to the ciliary ganglion.

Systemic effects have not been encountered with the doses used to treat strabismus.

Estimating the surgical dosage for residual strabismus after unsuccessful injection requires waiting for stabilization to occur, typically 5 to 6 months.

**Future Developments and Other Locally Acting Muscle Drugs**

Toxin types B, C, F, and G will be helpful in large-muscle disorders in which development of antibodies to large doses of type A has become a problem. There is no advantage of an EOM and as a therapeutic trial in difficult cases before antibodies to the small doses do not develop and systemic effects do not occur.

Large doses of local anesthetics are unpredictably myotoxic and are not therapeutically effective.

Lidocaine 2%, 0.2 to 0.5 mL injected as for botulinum toxin, is useful diagnostically to reduce suspected overaction of an EOM and as a therapeutic trial in difficult cases before botulinum toxin injection or EOM weakening surgery. No adverse myotoxicity has occurred in this clinical application.

Collagenase has been injected into fibrotic EOM (e.g., thyroid, orbit injury) in eight clinical cases. Only transient effects have resulted so far, but this does indicate the range of unexplored approaches to strabismus management.

**REFERENCES**


SECTION 8

SELECTED SURGICAL COMPLICATIONS AND PROCEDURES
ADJUSTABLE AND NONADJUSTABLE RECESSION AND RESECTION TECHNIQUES

LANCE M. SIEGEL, MD, MAURICIO J. LOZANO, MD, ALVINA PAULINE SANTIAGO, MD, and ARTHUR L. ROSENBAUM, MD

Purpose

The goal of the strabismus surgeon is to achieve both motor alignment and sensory improvement with a minimum number of procedures. To obtain the best long-term results, the amount of surgery performed is based on measurements of stable preoperative deviations. Tables and algorithms for the amount of muscle surgery have been formulated on the basis of averages from a large patient population for a given deviation in primary position. These numbers are modified by the surgical technique used, axial length of globe, variation in ocular rotations, incomitance, and a history of reoperation or restrictive phenomena. Neurologic and sensory factors such as fixation preference, fusion, and muscle paresis and other unforeseen factors contribute to the final surgical outcome and may compromise the postoperative results. Better long-term success rates depend on controlling overcorrections and undercorrections in the immediate postoperative period. The ability to modify the site of a muscle insertion and control the deviation in the immediate postoperative period to enhance the surgical outcome forms the basis for adjustable suture surgery.

Although the adjustable suture technique postpones the decision of the target surgical angle it is not a substitute for a careful and complete preoperative strabismus evaluation.

Nonadjustable surgery is preferred in children and uncooperative adults. Many of the surgical steps are identical to adjustable suture surgery and will be described in this chapter.

Historical Perspective

The ability to adjust a muscle’s insertional position on the globe to a more desirable one was first described in 1941.

Interest in adjustable strabismus surgery was renewed by Jampolsky in 1975. The technique improved the accuracy of alignment in many types of strabismus, especially those occurring in adults and older children.

Several surgical approaches have been described. A technique based on the O’Connor cinch consisted of a small adjustable resection of less than 4 mm; it has been replaced by other procedures. Scleral fixation (stay) sutures that exit through the skin have been used for positioning and traction. Both cul-de-sac and limbal conjunctival approaches have been used.

As the feasibility of suture adjustment became more apparent, techniques for increasing the effective outcome and simplifying the adjustment were reported. Bedrossian described an adjustable procedure based on the light reflex before and during surgery. Adjustments performed 3 to 4 days after surgery were found to decrease patient discomfort and vasovagal responses. Suture adjustment has been reported as long as 2 weeks after surgery.

Intraoperative adjustment using topical anesthesia with prism cover testing (one-stage adjustable) has also been described.

Problems with intraoperative adjustment include blurry vision caused by corneal drying and hemorrhage, pupillary dilatation, and bleaching of macular visual pigments by the operating lights.

Advantages and Disadvantages

ADVANTAGES

Perhaps the greatest advantage of the adjustable suture technique is the ability to modify the position of the muscles
and consequently the deviation of the eye in the immediate postoperative period, when results are less than optimal. More than one muscle may be placed on an adjustable suture, making it possible to adjust combined vertical, horizontal, or torsional components. This improves the chance of placing the eye close enough for sensory fusion to maintain alignment, varying primary-gaze motor alignment to position the eye in an area of scotoma for patients who develop visual confusion and diplopia, or moving the eyes away from orthotropia if central fusion is disrupted. Adjustment (or muscle movement) may not, however, always be required. The surgeon can place the globe into a desired position immediately, instead of waiting for postoperative drift or fusional mechanisms to achieve realignment. This may be important in exotropes, for whom the desired postoperative alignment is slight esotropia, by taking them out of their hemiretinal suppression scotoma; or in esotropes who will benefit from alignment within the monofixational range.5,7

For suture adjustment to be advantageous, three criteria must be met: (1) stability in the immediate period of adjustment, (2) long-term predictability, and (3) reduction of the reoperation rate. Stability in the immediate postoperative period is ensured with secure scleral bites and knots. Others suggest making a Z-track by means of a second pass under the insertion.16 Significant muscle "creep" does not seem to occur with this technique.44 Although mild force may be required to move the muscle,20,25 sutures can still be adjusted up to 24 hours postoperatively.

Theoretically, long-term predictability may be better using the adjustable technique than with conventional surgery because of the surgeon’s ability to set the eyes in a position of optimal postoperative alignment. Studies have shown excellent long-term results from adjustable strabismus surgery in patients followed for 6 to 13 months.5,31

The reoperation rate after conventional surgery is estimated to be 19% to 35% for conventional procedures, compared with 4% to 10% using the adjustable technique.34,41,67 The success rate for adjustable vertical rectus strabismus surgery is 78%.50 The adjustable technique succeeded in 64% of strabismic patients with thyroid ophthalmopathy, compared with 38% for those having nonadjustable surgery.54 The adjustment procedure can alter up to 23 PD of deviation in the postoperative period.58

### DISADVANTAGES

Skeptics reason that the muscle attachment site is not known for certain when the adjustable technique is used.12 Although it can be argued that this may be of minimal importance if the muscle maintains function and motility, the possibility of muscle movement or "creep" cannot be disregarded.44 However, no such movement was found in two cases reoperated by Scott.61

In studies using dogs, both conventional and hang-back techniques produced a small amount of advancement or forward creep from the intended recession; the final position of the medial rectus muscle was more variable than that of the lateral rectus. The adjustable technique produced more creep than conventional surgery (Table 33–1).10 In addition, studies of the hang-back technique have shown consistent results, comparable to those of conventional surgery.31,55

Perhaps the biggest disadvantage is the fact that the technique cannot be used in most children or in some patients with neurologic abnormalities. It is unlikely that a patient who does not cooperate optimally during preoperative evaluation will do so for postoperative adjustment.

### Indications

The indications for adjustable sutures are vast and still increasing as postoperative results are better quantified and outcome analyses become available. The use of adjustable sutures often is limited only by the patient’s cooperativeness.

It can be argued that the adjustable technique may be used in all adult patients and even cooperative children undergoing strabismus surgery. More importantly, the technique is optimal for cases in which the response to surgery is unpredictable. Some suggested indications are summarized in Table 33–2. The adjustable suture technique has also been used for transposition,27 Harada-Ito,43 and vessel-sparing procedures.18 The reader is referred to the appropriate chapters covering these topics.

### Table 33–1. Comparison of Muscle Creep in Conventional Versus Adjustable Suture Technique in an Animal Model

<table>
<thead>
<tr>
<th>Muscle</th>
<th>Surgery Type</th>
<th>Advancement/Creep (mm)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lateral rectus</td>
<td>Conventional</td>
<td>0.35 ± 0.58</td>
</tr>
<tr>
<td>Lateral rectus</td>
<td>Adjustable</td>
<td>0.60 ± 0.60</td>
</tr>
<tr>
<td>Medial rectus</td>
<td>Conventional</td>
<td>1.55 ± 0.68</td>
</tr>
<tr>
<td>Medial rectus</td>
<td>Adjustable</td>
<td>2.00 ± 2.44</td>
</tr>
</tbody>
</table>

### Table 33–2. Optimal Candidates for Adjustable Suture Technique

- Sensory anomalies
- Large sensory deviations with poor vision
- Central fusion disruption
- Adult strabismus with scotoma
- "Weak" muscles
- Paralytic strabismus
- Myasthenia gravis
- Restrictive phenomena
- Graves ophthalmopathy
- Post scleral buckling and glaucoma valve surgery
- After pterygium surgery
- After cataract and postanesthetic injury
- Scarring from trauma
- Slipped (recovered), lost, and disinserted muscle
- Reoperations
- Combined vertical, horizontal, or torsional deviations
- Large eyes (axial myopia) and small (microphthalmic) eyes
There are few contraindications to adjustable strabismus surgery; probably the most significant is an uncooperative patient. Although age is important, it does not preclude adjustment except in very young children (probably younger than 8 years old). Very high myopes with thin sclerae may not be good candidates because of the risk of perforation and detachment of scleral fixation sutures. Occasionally friable, threadlike muscles may be encountered; they should be sutured securely to sclera to avoid muscle slippage. Some believe that the inferior rectus muscle should not undergo adjustment because of the increased risk of muscle slippage owing to the short arc of contact and underlying Tenon’s connective tissue, which impedes strong reattachment of the insertion.

Adjustable and Nonadjustable Techniques

PREOPERATIVE CONSIDERATIONS

The ability of a patient to cooperate should be determined at the preoperative evaluation. Touching the conjunctiva on its medial and lateral aspects with a cotton swab can help screen out fearful or unwilling patients. Alternatively, the ease with which the examiner can perform applanation testing of intraocular pressure may be a sign of the patient’s ability to cooperate.

The patient’s age affects the decision to make a limbal or fornix conjunctival incision. Fornix adjustments should be avoided in patients older than age 50 years, owing to their thin and friable conjunctiva, which may tear easily.

Any sedative, antinausea, or pain medication that diminishes the patient’s alertness may interfere with accurate adjustment. A short-acting anesthetic agent such as propofol should be used if same-day adjustment will be performed.

Careful thought should be given to the choice of the muscle for adjustment. It is preferable to place the adjustable suture on a recessed muscle, especially if that muscle is stiffer, than on a resected one. It is easier to tighten or pull forward a recessed muscle than to recess it further. Resected muscles tend to be under more tension, increasing the risk of a slipped muscle or tearing out of the suture.

THE SLIP KNOT (HALF BOW TIE) TECHNIQUE

A preferred method for performing adjustable sutures is the half bow tie technique. An approach modified from its original description is shown, with photographic correlation, for both recessions (Fig. 33–1) and resections (Fig. 33–2). Our techniques for nonadjustable conventional recession and resection are the same, except for the final steps of fixation of the muscle to the sclera (Fig. 33–1A–L and 33–2A–H).

Adjustable Recession

Partial thickness scleral traction sutures (e.g., 6-0 Vicryl) are passed 90 degrees away from the muscle 1 to 2 mm from the limbus (see Fig. 33–1A and B). They are used to rotate the eye and provide maximum exposure of the limbal area overlying the muscle. With the eye in this position, the sutures are clamped to the drape with a hemostat (see Fig. 33–1C). A limbal conjunctival incision is fashioned in the area of the rectus muscle (see Fig. 33–1D). A marking suture (e.g., 6-0 Vicryl) may be used to identify the edge of the conjunctiva (see Fig. 33–1E). Two radial relaxing incisions are then made in the oblique quadrants at 90-degree angles from the peritomy (see Fig. 33–1F). The conjunctival flap is elevated with minimal dissection of perilimbal adhesions, being careful to stay clear of the muscle insertion and overlying vessels. Access to the rectus muscles is gained by puncturing Tenon’s capsule in the oblique quadrant.

The muscle is isolated by passing a Jameson hook against the globe through the opening created in Tenon’s capsule, keeping its tip against the sclera to avoid splitting the muscle (see Fig. 33–1G). A second pass may be needed just posterior to the first if the entire muscle is not obtained. Tenon’s fascia on the end of the hook is cleaned off with Westcott scissors. The muscle is cleaned of its intermuscular septum using both sharp and blunt dissection. Lateral rectus attachments to the inferior oblique should be severed, and the superior rectus freed from superior oblique attachments. The inferior lid retractors should be separated from the inferior rectus. These maneuvers prevent changes in the position of the eyelids, conjunctiva, and caruncle. The location of the muscle insertion in relation to the limbus is measured at this point before disinsertion (see Fig. 33–1H).

A spring-loaded clamp (e.g., a modified Apt or Sheppard clamp) is placed posterior to the muscle insertion. Wet-field or low temperature bipolar cautery may be used for hemostasis before disinserting the muscle stump with scissors (Westcott or Manson-Aebli) (see Figs. 33–1I and 33–1J).

A 12-inch double-armed muscle suture (6-0 Vicryl) on a spatulated needle (e.g., S-29) is passed in the middle of the muscle 0.5 to 1 mm from its end and tied in a single overlap knot with equal lengths remaining. One end of the suture is woven from the midpoint to the lateral edge of the entire muscle stump (0.5 to 1 mm from the edge of the disinsertion site). An additional bite is taken at the edge and secured with a locking bite. The other end is passed in a similar fashion to the opposite edge (see Fig. 33–1K). The stability of the muscle sutures can be verified by holding the two ends of the suture (see Fig. 33–1L). We prefer this technique because full thickness of the muscle fibers can be identified with certainty when passing the sutures. Sutures placed before disinsertion may inadvertently pass through the muscle sheath only, without incorporating muscle fibers.

If conventional nonadjustable recession is the goal, the muscle is sutured to the sclera with three scleral attachments at the desired recession distance (Fig. 33–3).

If adjustable recession is the goal, two scleral bites are taken at the insertional stump, angulated 35 to 45 degrees toward the midline (see Fig. 33–1M). The scleral passes should avoid incorporating Tenon’s fascia or fibrous muscle sheath, which will impede postoperative adjustment.

A bolster suture (e.g., black silk 6-0) is placed under a double overhand tie (see Fig. 33–1N), and the muscle is allowed to fall back to the desired recession distance. Measurements are confirmed with a caliper or curved ruler (e.g., Scott ruler); the latter is preferred for recessions greater than...
Figure 33-1. Adjustable recession, limbal approach. A. Traction sutures used to control the globe are passed 1 to 2 mm from the limbus, 90 degrees away from the muscle to be operated on. The surgeon should make sure that conjunctiva and at least episcleral tissue are incorporated. B. Traction Vicryl sutures are completed in two quadrants 180 degrees apart. C. Traction sutures are used to expose the area of globe where muscle surgery will be performed. D. Conjunctival opening is created at the limbus. E. Marking sutures are used to identify the edges of the conjunctiva. F. Radial relaxing incisions (dotted lines) in the oblique quadrant are made. G. The rectus muscle is secured with a Jameson hook. Note the thick connective tissue overlying the muscle. H. After the muscle has been cleaned of its intermuscular septum and Tenon's fascia, the insertion is measured from the limbus using a caliper (denoted by ruler). I. Cautery is applied before disinsertion.
9 mm, beyond which there are significant differences between arc and cord length (see Fig. 33–1O). The amount of desired recession is added to the location of the muscle insertion determined earlier and measured from the limbus.

A slip knot (half bow tie) is completed over the bolster suture. The muscle sutures are cut to a length convenient for the adjustment, usually 2 to 3 cm. The overlying silk bolster then is tied into a 2-mm loop overlying the adjustable suture (see Fig. 33–1P). This bolster may be used to loosen the adjustable suture from adherent conjunctiva or connective tissue.

When marking sutures are used, the ends of the conjunctiva can be identified easily (see Fig. 33–1Q). The conjunctiva is recessed just behind the insertional stump and closed with interrupted sutures (e.g., 8-0 Dexon) (see Fig. 33–1R). If the conjunctiva is redundant, part of it may be carefully excised to expose the knots of the adjustable suture.40 An interrupted suture (e.g., 6-0 Vicryl) is used to create a “bucket handle” loop midway between the muscle insertion and the limbus (see Fig. 33–15 and T). The bucket handle will be used for postoperative adjustment.

For conventional nonadjustable recession and resection, the conjunctiva is resutured to the limbal edge of the conjunctival incisions using the marking sutures as a guide to ensure edge to edge approximation using 8-0 Dexon suture (Fig. 33–4). A 0.5- to 1-ml subconjunctival injection of dexamethasone (24 mg/mL) is given to lessen postoperative pain and inflammation. This minimizes the need for postoperative medications until the time of adjustment. The sutures may be tucked into the fornix, placed under the conjunctiva, or taped to skin.1 Ocular ointments may blur vision and should be avoided, especially when same-day adjustment will be performed; antibiotic-corticosteroid eye drops are preferred.

**Figure 33-1** Continued J. After hemostasis, the muscle may be disinserted using scissors. K. Correct technique of muscle suture placement. Locking bites should be taken at both ends. Using a spring-loaded clamp and disinserting before suture placement ensures that sutures are placed in the substance of muscle (and not just the sheath). L. Spread out the two ends of the suture to ascertain that it is tied securely. M. Technique of securing the muscle to sclera near the insertional stump is shown. N. Silk bolster suture is shown under a double hand throw. O. The amount of recession (curved ruler) is measured from the limbus using a Scott ruler.

Illustration continued on following page
Adjustable Resection

The adjustable resection begins in a manner similar to the recession procedure, from scleral traction sutures to muscle isolation. At this point a second muscle hook is passed posterior to the first (e.g., Green hook), and the muscle is placed under tension (see Fig. 33–2A). The amount of resection is measured with calipers from the insertion to where the muscle sutures will pass (see Fig. 33–2B). A 12-inch double-armed muscle suture (e.g., Vicryl 6-0) on a spatulated needle (e.g., S-29) is passed in the middle of the muscle and tied in a single overhand knot with equal lengths remaining (see Fig. 33–2C). Each of the suture ends is woven from the midpoint to the lateral edge (see Fig. 33–2D).

A spring-loaded clamp (e.g., a modified Apt or Sheppard clamp) is placed posterior to the suture and firmly locked into place (see Fig. 33–2E). A Hartmann clamp is placed just anterior to the suture and clamped for 10 to 15 seconds to provide hemostasis, and cautery is applied in the same area (see Fig. 33–2F). Sharp, fine scissors are used to cut along the cauterized site, taking care to avoid cutting the muscle sutures (see Fig. 33–2G). The muscle stump is disinserted at the insertion using scissors (Westcott or Mansson-Aebli) (see Fig. 33–2H). If conventional nonadjustable resection is the goal, the muscle is sutured to the insertional stump with 4 running scleral fixation attachments (Figs. 33–5 and 33–6).

If adjustable resection is the goal, two scleral bites are taken at the edges of the insertion site at a 35- to 45-degree angle toward the midline of the muscle stump (see Fig. 33–2I). A 6-0 silk bolster suture is placed under a double overhand tie (see Fig. 33–2J). The resected muscle may be allowed to fall back slightly (it is easier to pull it up than let it slip back), and the suture is secured with a slip knot.
(half bow tie). The spring-loaded clamp then is released carefully. The underlying bolster is tied into a 2-mm loop overlying the adjustable suture (see Fig. 33–2K). The conjunctiva is recessed to the insertional stump and closed with interrupted 8-0 Dexon (see Fig. 33–2L). Redundant conjunctiva is common in resections and may be trimmed to expose the knots of the adjustable muscle suture. A “bucket handle” suture is placed. The final suture arrangement is shown in Figure 33–2M. Medications are similar to those used in recession.

NOOSE TECHNIQUE

A noose-type slip knot for use in adjustable strabismus surgery has been described, using either a limbal or fornix approach. After securing the rectus muscle with muscle sutures as previously described, the adjustable sutures come out almost next to one another, often using a “crossed swords” technique (Fig. 33–7A). The two side sutures are called the pole sutures. The muscle sutures are then pulled up toward the insertion. They are clamped together 7 to 8 cm from the insertion, and a square knot is tied abutting the clamp. Using the clamp for traction, a second suture (the noose suture) is tied around the pole sutures using several square knots (Fig. 33–7B and C). The muscle is pulled snugly up to the insertion. A caliper is used to measure the desired recession from the insertion along the pole suture. Significant tension must be maintained on the pole suture as the noose suture is moved to this point. This serves as a stop when the muscle is allowed to fall back (see Fig. 33–7C). A scleral “bucket handle” may be placed for traction during postoperative adjustment. The sutures are carefully tucked into the inferior fornix.

POSTOPERATIVE ADJUSTMENT

Principles

The patient should be alert and cooperative during the adjustment and able to fixate on an accommodative target. Refractive error must be corrected with spectacles or contact lenses; contact lenses are preferred for patients with high ametropia. Measurements are obtained using alternate prism cover testing whenever possible. If the deviation is less than optimal, the adjustable sutures can be used to place the eye at the exact desired position.

Adjustments may be performed on more than one muscle, but the sutures must not be intertwined. This requires careful identification of the sutures at the time of adjustment. Differently colored sutures may be used to identify the various sutures; undyed sutures are not satisfactory.

Cover test measurements must be obtained not only in primary gaze but also in right and left gaze for horizontal adjustments and in upgaze and downgaze for vertical adjustments. These measurements will detect excessive recession and show the extent of ocular rotation and overcorrection or undercorrection in a specific gaze field (comitance of the alignment). Careful attention should be paid to muscle rotations. Patients with a nonparietic deviation should not be left with more than a mild duction deficiency. If resection of an antagonist muscle has restricted rotation in the opposite gaze field, advancing an adjustable recession will have little effect in overcoming the gaze limitation. Similarly, overcorrection in primary gaze caused by a resected antagonist will not be relieved by adjusting a recessed muscle. Especially with resected muscles, horizontal incomitance in left or right gaze may be bothersome when driving. Taking near measurements are also important, especially in older adults who might be presbyopic. Excessive medial rectus recession may decrease convergence and create bothersome symptoms at near.

Regression analysis shows that 1 mm of adjustment causes a 2.54-PD change in ocular alignment. There is no difference in this relationship whether adjustment is performed on the medial or lateral rectus or between a recession or resection.

Recommendations for the desired postoperative alignment may vary among different strabismologists. Some suggest leaving esotropic patients orthotropic; esotropic patients, 5 to 7 PD esotropic; and hypertropic patients, 0 to 2 PD hypotropic. Others advocate leaving nonsensory, intermittent, or decompensated exotropes 4 to 16 PD or 10 to 20 PD esotropic. These recommendations may change if patients have coexisting disease such as thyroid ophthalmopathy or myasthenia gravis.

Technique of Adjustment

The Adjustable Suture Kit. All needed equipment should be readily available at the time of adjustment. We recommend a sterile suture adjustment kit consisting of a lid speculum, two tying forceps, scissors, toothed (0.3) forceps, needle holder, suture, cotton swabs, and gauze pads (Fig. 33–8). Tetracaine in individual sterile dispensing units is used for anesthesia. The lid speculum is used only if necessary. We have found that patients tolerate manual opening of the lids by the assistant better than the lid speculum. The toothed forceps are used to lift adherent conjunctiva or apply fixation to the globe at the site of the traction suture or on the sclera/episclera. Another needle holder (not shown) may be advisable for additional tension or traction or to hold the overhand knot to prevent slippage when tying is difficult. A blunt-tipped Westcott scissors is used to trim the sutures.

The Adjustment Procedure. We wait a minimum of 6 hours, and preferably 24 hours, to ensure that patients are free from the effects of anesthesia and sufficiently awake to cooperate with testing and adjustment. Rarely, in complex cases, the adjustable suture may be left in place after initial adjustment and the alignment allowed to stabilize. We have readjusted on the second postoperative day without encountering adhesions or technical difficulty.

The eye patch is removed, and the eyes carefully cleansed with gauze moistened with sterile saline. Care is taken to avoid pulling on the sutures. The patient should be reminded not to rub the eye. Two to three drops of tetracaine are placed in the cul-de-sac. Corrective glasses are worn. If contact lenses are preferred for high ametropic errors, the surgeon should place the lenses in the patient’s eyes and remove them at the end of adjustment. Then the deviation is measured, ductions and versions are observed, and incomitance is checked. The surgeon should take a moment to plan the adjustment.

The patient is slowly placed in a reclined position. An assistant applies gentle traction on the patient’s lids to open the eyes. The patient is instructed to fix on an object in the
Figure 33-2. Adjustable resection, limbal approach. A, Two muscle hooks beneath the muscle are used to apply tension on the rectus muscle. B, top, The amount of muscle resection is measured with calipers from the insertion to where the muscle suture will pass. B, bottom, The measurement is verified after the needle is placed in the middle of the muscle. C, top, With the needle in place, the amount of resection is represented by the caliper. C, bottom, After the suture is tied in the middle of the muscle, the measurement is again verified. The same steps (Fig. 33–2B to 33–2C) should be performed as well after the sutures are passed to the ends of the muscle. D, The appearance of the rectus muscle after proper suture placement. White arrow points to a locked bite at the edge of rectus muscle. E, A spring-loaded clamp is placed posterior to the suture and firmly locked in place. F, top, Hartmann clamp just anterior to the suture for hemostasis is applied. F, bottom, Cautery applied to area clamped. White arrows indicate Vicryl muscle suture retracted away from clamp and cautery.
Figure 33-2 Continued

G, top, Blue arrows show site of cautery.

G, bottom, Sharp scissors used to cut muscle along the cautery site.

H, top, Cut rectus muscle shown. Rectus muscle with solid outline represents the amount of resected muscle.

H, bottom, The stump of muscle that remains attached to the insertion is removed.

I, Technique of securing bite using a spatulated needle (white arrows) at insertion (red arrow) is shown.

J, Silk bolster suture is shown beneath the double hand throw. The surgeon slowly brings the spring-loaded clamp toward the insertion.

K, The overlying bolster suture is tied into a 2-mm loop over the adjustable half-bow suture (white arrow).

L, Conjunctiva is closed just behind the knots of the adjustable sutures to keep them accessible. If necessary, redundant conjunctiva covering these sutures may be carefully excised.

M, Scleral "bucket handle" (black arrow) is placed to assist in applying traction during the adjustment procedure.

(Dotted lines in A–D, F–M indicate conjunctival opening; solid lines in A–H, J, and K indicate outline of rectus muscle (R); white arrows in D, F, J, L, and M indicate Vicryl adjustable muscle suture; red arrows in J–M indicate silk bolster suture.)
direction that provides the best exposure and access to the suture. All sutures are carefully identified.

**The Bow Tie Technique.** With the use of the silk bolster, the adjustable knot is exposed if necessary or the suture is loosened from its attachments to the sclera or conjunctiva. Only the silk suture is carefully cut after the adjustable sutures have been identified with certainty. If the patient does not need adjustment, the bow should not be untied. The loop is opened and pulled through to create a square knot. The suture is tied once or twice more with square knots.

If the patient requires adjustment, the loop is untied as if it were a shoelace. This should leave the double overhand throw visible. To advance the muscle and increase its action, the surgeon should pull forward on the suture following its scleral tracts, cinch it down, and retie the bow. Pulling straight up on the sutures may disinsert them from the scleral tunnel. To recess the muscle and decrease its action, the suture overhand tie is loosened by 1 to 2 mm and the patient asked to look toward the side of adjustment (e.g., for the medial rectus the patient looks nasally). This will cause the muscle to contract and pull the suture tight. Countertraction may be exerted by holding the bucket handle and/or applying force in the opposite direction while the patient looks in the direction of the muscle’s action. The suture is retied in a slip knot (half bow tie) fashion. The surgeon should watch the suture as it unravels. Occasionally, the increased tension pulls down on the overhand knot and causes it to slip before the bow tie can be secured. An assistant may hold the area of the overhand tie either with a needle holder or tying forceps until the surgeon can secure the half bow tie.

The patient is raised up slowly and cover test measurements are rechecked. The adjustment is continued until alignment is satisfactory or maximal recession or advancement has been achieved. Again, when the patient is at the desired position, the adjustable suture knot is secured by pulling through the loop of the half bow tie and additional square knots (three to four total). The suture is trimmed with a 2-mm tail and tucked beneath the conjunctiva. The traction/bucket-handle scleral suture is removed, being careful not to cut the suture to the muscle.

The advantage of the slip knot (half bow tie) approach is that the adjustable suture is easily exposed and manipulated. Tension on the suture is needed only during adjusting, not tying. It is important that both halves of the suture be pulled forward, otherwise only one end of the muscle may move. This method also requires untying and retying until the desired alignment is reached. A relative disadvantage is the inability to know how much the muscle has been pulled forward or recessed. With experience, the surgeon will be able to estimate the amount of suture and muscle movement.

**The Noose Technique.** Adjustment is conducted as follows: traction is applied to the pole sutures using a needle holder (Fig. 33–7D). To advance the muscle, traction is maintained until the knot has been raised by the needed amount above the scleral insertion, preferably in 1-mm increments. The knot then is slipped down flush to the insertion. If the muscle needs to be recessed, the knot can be moved up the pole sutures by the necessary amount and the muscle allowed to fall back. The patient may be asked to look in the direction of the operated muscle’s action to place tension on the muscle and help the knot slide forward while the muscle moves posteriorly. Countertraction may be provided using the scleral bucket handle.

When the eye is adjusted to the desired position, the distal knots on the pole sutures are cut off. A square knot and overhand knot are tied firmly against the noose to secure it in place, taking care not to pull forward on the pole sutures when tying the knot. The excess pole suture, noose bucket handle, and scleral bucket handle are trimmed with Westcott scissors. A preplaced conjunctival suture may be used to close the conjunctiva.

**Figure 33-4.** A, Conjunctiva is placed at limbus using pre-placed marking sutures as a guide (arrows). B, Conjunctiva closure completed using 8-0 absorbable sutures.
Figure 33-5. A. Resected muscle secured to insertional stump with 4 running scleral attachment points (arrows). Muscle is secured in spring loaded clamp which is used to advance muscle to insertional stump and reduce tension on suture as it is advanced. B. Resected muscle reattachment to insertional stump. At this stage, muscle clamp (arrow) is removed.

Advantages of the noose procedure include the ability to quantify recession or advancement with equal movement of both muscle edges. There is no need to continually tie and untie the knot if multiple adjustments are necessary. This technique is also helpful when loosening the suture on a tight muscle may cause it to fall back. However, the slip knot may become loose, a large central sag may be created when trying to put the crossed swords close together, and moving the knot at the time of adjustment may be problematic. In the fornix approach, it may be difficult to retract the conjunctiva to gain adequate exposure. The conjunctiva may tear when traction is applied. In both the fornix and limbal approaches using the noose technique, a larger suture mass remains after adjustment and may irritate the patient.

**Common Pitfalls**

There are three major pitfalls that need to be avoided in adjustable surgery: (1) the muscle is not adequately secured; (2) scleral bites are insufficient; and (3) the adjustable suture knots are not exposed. These all may be avoided by using proper surgical techniques as described previously.

In addition, some authors recommend placing the muscle in a recessed position 1 mm more than desired rather than at the exact predetermined site, to avoid overcorrection. It is easier to advance a muscle than to let it retract. Tight muscles tend to retract during adjustment and may induce recession. Unintended slippage should be avoided. During adjustment, an extraocular muscle should not be recessed so far as to profoundly limit rotation.

Anesthesia must be chosen carefully; the amount of sedation and the use of antiemetics may greatly affect the patient's alertness. Droperidol should be avoided because it may result in a lethargic patient. The anesthesiologist should be informed of the surgeon's desire for an alert patient during adjustment.

**Complications**

Rarely, a muscle will not move during adjustment. It is important to first be sure that the muscle has not been advanced to the insertion or is not recessed maximally, in which case the maximal effect from the surgery has been obtained. The scleral bucket handle is useful in cases in which the muscle does not adjust easily. Sometimes applying more tetracaine and gently cleaning around the suture site, as well as using 0.3-mm toothed forceps to free any scarred or adherent conjunctiva, will help loosen the muscle. The patient should be asked to look into the extreme positions of gaze to help muscle movement during the adjustment.

A broken or cut suture at adjustment is a feared complication. Unraveling of the knot is also a possibility. If the knot unravels, the ends often can be found and retied and the adjustment can proceed. If a suture remains, it should be visualized and secured. If the scleral bite is still present and enough suture exists to adjust or tie the muscle, this should be done. Otherwise the patient will have to be brought to the operating suite to resecure the muscle under more controlled conditions. Permanent curved surgical needles may be used to attach the loose end of the suture to sclera (Fig. 33–9). The loose suture is threaded through the curved surgical needle and secure scleral bites taken before tying the suture to the other end.

In the event that both sutures are cut or broken, the patient should be instructed not to rotate the eye. One may gently lift up the overlying conjunctiva and identify the muscle. The muscle is grasped securely with 0.3-mm toothed forceps. Any suture available is used to pass a bite anywhere in the mid-muscle insertion, if possible a few millimeters from the insertion edge. A single knot is tied and the suture allowed to hang out of the eye. The needles are removed. This suture will identify the muscle should it retract on the way to the...
operating room. If the muscle slips completely or is lost, appropriate management is required (see Chapter 40).30, 52, 68

If the suture accidentally pulls out of the scleral tract, the same surgical curved needle may be used to anchor the loose suture back to the sclera, tying it securely.

During muscle manipulation, the risk of an oculocardiac reflex is an important potential complication.68 Stimulating the extraocular muscles reflexly increases vagal tone, releasing acetylcholine into the atrioventricular node and generating bradycardia or dysrhythmia. Bradycardia and syncope caused by the oculocardiac reflex occur in 4.5% to 65% of patients.3, 35, 68 This may occur intraoperatively and also during adjustment.2, 3, 27 Studies have attempted to relate intraoperative oculocardiac reflex to its occurrence during postoperative adjustment.24 The presence of a vasovagal response has been closely associated with an oculocardiac reflex and is accompanied by nausea, light-headedness, hypotension, diaphoresis, a perceived temperature change, and vomiting.24

The risk of the oculocardiac response is thought to be decreased by using topical anesthesia, avoiding a lid speculum, and placing the patient supine during adjustment.35

Careful observation of the patient is most important. Most patients who experience a vasovagal reflex can proceed with adjustment after a short observation period and reassurance. Because of the transient nature of this phenomenon, cardiac monitoring is usually not indicated except in patients with a significant history of heart block and cardiac disease.

Another problem is that of changing measurements following completion of the adjustment. Unstable adjustments are found in patients with conditions such as myasthenia gravis, Guillain-Barré syndrome, and orbital fracture.52 Progressive overcorrection after inferior rectus recession with adjustable sutures is well recognized, especially in patients with restrictive thyroid ophthalmopathy.53

The suture ends may cause mild, transient irritation.34, 56 Absorbable sutures usually soften quickly; continued irritation or allergic reaction is rare.68 Persisting suture irritation can be resolved by removing the suture after sufficient time—at least 2 weeks—has elapsed for muscle adhesion and healing to take place.15, 52, 68 Dellen have been described secondary to abnormal perilimbal edema.52 Hydration and conservative management are recommended.
Rare complications include hyphema developing at the time of adjustment\textsuperscript{19} and adhesions forming between the lids and superior rectus in a patient having simultaneous lid surgery and adjustable superior rectus surgery. The adhesion was easily lysed.\textsuperscript{90} Temporary muscle paralysis occurring after topical anesthesia also has been reported.\textsuperscript{90}

REFERENCES

The inferior oblique (IO) muscle is peculiar in its anatomy, function, appearance, and response to surgery. There may be more diversity of opinion concerning the proper operation for this muscle and specific indications for surgery than for any other extraocular muscle.

**Surgical Anatomy**

Before surgery is performed on the IO muscle, a number of anatomic peculiarities must be considered. The IO is the only extraocular muscle that does not originate at the orbital apex. It arises instead from the orbital plate of the maxilla near the opening of the nasolacrimal duct. It then progresses posteriorly in a lateral direction beneath the inferior rectus (IR) muscle to insert beneath the lateral rectus (LR) muscle. The insertion ranges from 9 to 14 mm in width and stretches from 10 mm posterior to the inferior edge of the LR muscle insertion posteriorly to approximately 4.5 mm from the optic nerve. The insertion thus is within only 2 mm of the macula. The insertion itself is unusual in that the tendon is the shortest of any extraocular muscle, being less than 2 mm long. In an anatomic study of 200 autopsy eyes using high magnification with a stereomicroscope, single classic insertions were found in 100 eyes and two to six distinct and separate insertions in the other half.12

The muscle as a whole runs the shortest course of any extraocular muscle—36 to 37 mm. It is the only muscle to come in contact with two other eye muscles—the IR and LR muscles. It is initially in contact with the periosteum of the orbital floor but farther on runs adjacent to the orbital fat near the IR and LR muscles.

The nerve to the IO muscle is a branch of the inferior division of the oculomotor nerve that travels within a neurovascular bundle to enter the muscle on its upper surface just lateral to the IR muscle. The parasympathetic innervation to the pupil is nearby. The muscle is vascularized by the medial muscular branch of the ophthalmic artery and a branch of the infraorbital artery and is drained by the inferior orbital vein. The IO muscle also passes a number of vortex veins in its course around the eye.

**Historical Perspective**

The first surgical procedure recommended for the IO muscle was proposed in 1906, when Duane1 described a transcortaneous tenotomy at its origin. Since then, the most beneficial procedure for weakening an overacting IO muscle has remained controversial.

In 1929, Dunnington2 suggested disinsertion at the scleral insertion as the preferred method. A recession at the insertion site was described by White3 in 1943. He believed that myotomy at the insertion was unpredictable, whereas a recession could be titrated based on the degree of muscle overaction.3,4 Over the next two decades, the prevalent weakening operation on the IO muscle was recession of an arbitrary, graded amount. In 1962, Dyer5 advocated the disinsertion operation after reviewing his results in 45 cases. Thereafter, this operation gained popularity as experienced strabismologists turned to it. Ophthalmologists who previously had avoided IO muscle surgery began to perform disinsertion.

In 1972, Parks6,24 in a controlled study of four different IO muscle weakening operations (disinsertion, recession, and myectomy performed at two different sites), concluded that the recession operation is most effective and long lasting. Others later assessed denervation and extirpation for markedly overacting IO muscles and found it to be an effective procedure.5,14,15

In 1978, Apt and Call1 carefully studied the anatomic position of the IO muscle in 200 consecutive autopsy eyes.
The investigation provided information for locating the exact point on the globe for a desired amount of recession. They found that a slight anterior displacement of the muscle occurred with some commonly performed recessions. For example, recession of the IO muscle to what initially had been considered by Parks to be an 8-mm recession point \(^{23,24}\) was in fact not only a 10.4-mm recession but also a 1.0-mm anterior displacement of the muscle.\(^1\)

In recent years it has been realized that anterior displacement of a recessed IO muscle enhances its weakening effect. The idea of anteriorly transposing the IO muscle by attaching it anterior to the equator so as to weaken its action was first advocated by Gobin.\(^13\) Reattaching the IO muscle anterior to the equator shifts the new insertion site closer to the origin of the muscle, thereby enhancing the recession.\(^11\) The anatomic landmarks relating recession of the muscle to the LR and IR muscles indicate that recommended 8- and 10-mm recessions result in some degree of anterior transposition (Fig. 34–1).\(^1\)

Collapsing the posterior and anterior borders of the recessed IO muscle when suturing it to the sclera enhances the weakening effect of the surgery and decreases the excyclotorsion force of the muscle.\(^1\) Excellent results were obtained when an overacting IO muscle was weakened by moving the muscle insertion just anterior to the temporal insertion of the IR.\(^11\) In patients with unilateral fourth nerve palsy, anterior transposition, when performed 2 mm or more anterior to the IR insertion, caused postoperative hypotropia in the primary position and limited upgaze.\(^2\) Other authors, however, found no hypotropia after unilateral anterior transposition surgery in patients with superior oblique (SO) palsy.\(^16,21\)

### Indications and Contraindications

**INDICATIONS**

Surgery to weaken the IO muscle is indicated when the muscle is overacting. Whereas the first two indications listed in Table 34–1 are straightforward, the others require clarification.

For many years, IO muscle surgery was generally thought not to affect dissociated vertical deviation (DVD). However, it is now recognized that anterior transposition of the IO

### Table 34–1. Indications for Inferior Oblique Weakening Procedures

<table>
<thead>
<tr>
<th>Condition</th>
</tr>
</thead>
<tbody>
<tr>
<td>V-pattern strabismus with inferior oblique overaction</td>
</tr>
<tr>
<td>Primary inferior oblique overaction</td>
</tr>
<tr>
<td>Dissociated vertical deviation</td>
</tr>
<tr>
<td>Superior oblique palsy</td>
</tr>
<tr>
<td>Excyclotorsion</td>
</tr>
</tbody>
</table>
muscle can generally control DVD, especially if associated with IO muscle overaction.\textsuperscript{3, 19, 22} If a patient has simultaneous overaction of the IO muscle and a small to medium-sized DVD, an anterior transposition should be considered as a single treatment for both disorders. These two conditions must be carefully differentiated because DVD alone can to some degree simulate overaction of the IO muscle (see Chapter 17). A 10-mm recession of the IO muscle, placed 3 mm posterior and 2 mm temporal to the lateral border of the IR muscle insertion, includes a small anteriorization and therefore may have a minimal effect on DVD. Other IO muscle surgical procedures have no documented effect on DVD.

When ophthalmologists observe overaction of only one IO muscle, usually demonstrated by unilateral overelevation on adduction, SO muscle paresis is suspected. Considering this possibility entails performing a workup tailored to SO palsy. Diagnosing isolated unilateral IO muscle overaction must be done with caution. An initially inapparent overaction of the IO muscle in the fellow eye may become evident after the obviously overacting IO muscle is weakened.\textsuperscript{34}

In most cases of SO muscle paresis there is secondary overaction of the ipsilateral IO muscle. Thus, if surgery is to be done, a weakening procedure of the IO muscle is usually indicated, sometimes in association with surgery on other vertically acting muscles. The operation of choice depends on the clinical presentation of the paresis. The IO muscle may be recessed, and this may be combined with the appropriate procedure on another vertically acting muscle (see Chapter 15).

There is more disagreement among strabismologists about when to perform unilateral anterior transposition. Anteriorization of the IO muscle has been found to significantly affect elevation.\textsuperscript{41} Compared with recession, anteriorization further decreased ocular elevation by approximately 15 PD.\textsuperscript{41} If this is done unilaterally, an evident hypotropia and diplopia may appear in upgaze and even in primary position.\textsuperscript{2, 41} These complications seldom occur if the procedure is performed bilaterally. The problem results from the fact that the IO muscle can act as a depressor or anti-elevator after anteriorization, limiting the action of the superior rectus muscle on attempted elevation.\textsuperscript{20} For this reason, unilateral anteriorization should be reserved for patients with appreciable hypertropia in upgaze that requires correction. However, others believe that this procedure may be considered for any SO paresis if the hypertropia in primary position is at least 25 PD.\textsuperscript{16, 21} Other IO weakening procedures may be considered in unilateral situations.

The effect of IO muscle surgery on excyclotorsion has only recently been quantitatively studied (Fig. 34–2).\textsuperscript{39} Anteriorization of the IO muscle affects excyclotorsion within the first 6 weeks after surgery. Thereafter, only eyes that have undergone anteriorization adjacent or anterior to the temporal insertion of the IR muscle continue to demonstrate benefit from the procedure with respect to torsion. Eyes that exhibit recurrent IO overaction after surgery also have shown recurrent excyclotorsion.

\section*{CONTRAINDICATIONS}

Patients with V-pattern strabismus but not IO muscle overaction should undergo horizontal rectus surgery with the appropriate transposition. An X pattern is seen in patients with long-standing exotropia causing an overshoot in upgaze, simulating IO overaction (and a downshoot in downgaze, simulating SO overaction), because the LR muscle functions as a leash in these positions. Apparent overaction may be observed in patients with craniofacial syndromes, but orbital and other anatomic abnormalities, evident radiographically, usually explain the observed IO pseudo-overaction (see Chapters 14 and 30).

IO muscle overaction generally is associated with excyclotorsion. Occasionally, however, complicated strabismus situations occur in which a patient may have coexisting incyclotorsion and IO muscle overaction. This may be noted after retinal detachment surgery when a restrictive strabismus develops. If the surgeon weakens the IO muscle, the incyclotorsion may worsen, especially if anterior transposition is performed. In this instance the surgeon should either avoid IO muscle surgery altogether, perform a 10-mm recession that has little or no effect on torsion, or combine IO muscle surgery with a different procedure that addresses incyclotorsion.\textsuperscript{30, 41}

\begin{figure}[h]
\centering
\includegraphics[width=\textwidth]{figure34-2.png}
\caption{A, Preoperative excyclotorsion of the fundus found in the case of a severely overactive inferior oblique muscle. B, Six weeks after anterior transposition, excyclotorsion of the fundus is reduced by 12 degrees.}
\end{figure}
Techniques of Surgery

CAPTURING THE INFERIOR OBLIQUE MUSCLE

For most extraocular muscles, the surgeon has a choice of conjunctival approaches. For the IO muscle, however, most strabismus surgeons make an incision in the bulbar conjunctiva and intermuscular septum in the inferotemporal quadrant 7 to 9 mm from the limbus, which provides an excellent approach to the muscle and causes minimal postoperative discomfort.

The inferotemporal limbus is grasped with toothed forceps by an assistant to rotate the globe in extreme elevation and adduction. The conjunctival incision is made approximately 8 mm from the limbus in the inferotemporal quadrant (Fig. 34–3). Tenon’s fascia is cut perpendicular to the conjunctival incision to expose bare sclera. The LR muscle is secured with a muscle hook (e.g., Green hook). The assistant then shifts his or her grasp to the LR hook, releases the hold on the limbus, and positions the cornea in a supronasal position.

A flat iris spatula is used under direct visualization to dissect the fascial attachments of the IO muscle to the globe. A muscle hook is positioned under the belly of the IO muscle (Fig. 34–4). The hook is placed under direct visualization to avoid injuring the vortex vein, which may cause extensive bleeding. The IO muscle is carefully freed from the surrounding periorbital fat with Westcott scissors, being aware of the nearby inferotemporal vortex vein (Fig. 34–5). A larger square hook or spring-loaded clamp (e.g., an Apt toothless muscle clamp) replaces the small hook, and the hook holding the LR muscle is removed. Connective tissue attachments to the LR muscle are freed. Adhesions between the insertion of the IO muscle and the sclera and adjacent tissue are severed to expose the whole width of the IO insertion.

When isolating the IO muscle insertion area, the surgeon must be aware of the possibility of multiple congenital insertions (Fig. 34–6). Failure to isolate and section these additional fibers may account for persistent or residual postoperative overaction of the muscle. "Bifid" insertions also may be iatrogenic when a muscle hook used to isolate the IO "splits" the muscle. This too may lead to persistent or residual overaction postoperatively if only part of the muscle is disinserted, recessed, or myotomized. Similarly, when performing a myectomy, the surgeon should be sure that a full section of the muscle has been removed and no residual
fibers remain connecting the two ends of the muscle. The use of traction testing on the IO muscle at the time of surgery may help detect incomplete disinsertion, myotomy, or myectomy of the muscle.

If the chosen procedure requires suturing of the IO muscle, a 6-0 absorbable suture is passed through the entire width of the tendon about 1 mm from the insertion (Fig. 34–7) or, if a clamp is used, through the cut end of the muscle immediately anterior to the clamp. Locking knots are placed at both sides. The IO muscle is then disinserted with Westcott scissors.

The IO muscle inserts in the area of the macula and the papillomacular fibers of the retina. The macula is a mere 1 to 2 mm posterior and superior to the posterior insertional tip of the IO muscle. Surgeons should, therefore, exercise extreme caution to avoid perforating the sclera in this area. The confined space around the IO muscle insertion can make needle placement challenging, especially when applying lock bites to the sides of the muscle before disinserting it. The surgeon should never point a needle toward the sclera in this area. Alternatively, a clamp such as an Apt clamp may be placed across the insertion and the muscle disinserted before the suture needle is inserted.

MYOTOMY

Today, myotomies are performed at the insertion of the muscle rather than at its origin, as was done in the earlier part of the 20th century. After the IO muscle is isolated, the surgeon simply incises the tendon at its insertion and allows the muscle to retract without placing muscle sutures. This procedure may be “self-adjusting,” because a very overactive IO muscle will be tight and therefore will contract more after disinsertion, reinserting itself farther from the original insertion than a less overactive (and presumably less tight) muscle. In an interesting study on primates, disinserting normal (but not overactive or tight) IO muscles resulted in the muscles reinserting over a very wide area.34

MYECTOMY

Most myectomies today are performed near the insertion of the IO muscle. After the muscle is isolated, two clamps are placed across the muscle 4 to 8 mm apart (Fig. 34–8). The muscle between the clamps is excised, and the muscle stumps on the clamps are cauterized to prevent hemorrhage or possible reattachment to the sclera after the clamps are released. The muscle should be inspected to be sure that no remaining strands connect the muscle ends. A unilateral myectomy will correct 5 to 20 PD of hypertropia. The proximal end of the muscle may be placed behind the opening in Tenon’s capsule and the opening sutured closed.

Proponents of this surgery also believe that it is somewhat “self-adjusting,” as described earlier.35 Overcorrections are not as common as might be thought. After myectomy, some weak connections between the residual IO muscle and the eye via attachments to the IR muscle and connective tissue remain. Reoperation on the muscle, if necessary, may be difficult after myectomy with cauterization, because relocating the muscle can be a problem.

RECESSION

In a true recession, the entire muscle insertion is receded along the anatomic course of the muscle and reattached to the sclera with one or more sutures. This definition is somewhat extended in the case of the IO muscle. A maximum recession (12 to 14 mm) of the IO muscle places its anterior border 4 mm behind the true temporal insertion site of the IR muscle and 5 mm behind the pseudo-insertion point (see Fig. 34–1). Care must be taken to avoid the inferior temporal vortex vein when performing 10- to 14-mm recessions of the IO muscle, because the intrascleral or extrascleral extensions of the vein, or both, are often close to the posterior reinsertion site of the muscle if the surgeon retains the full normal width of the insertion.

Graded recessions for greater or lesser degrees of IO muscle overaction are advocated by some ophthalmologists. Measurements derived from an anatomic study were recommended for performing graded recessions ranging from 6 to 12 mm (Table 34–2).1

The popular 10-mm IO muscle recession (8-mm recession of Parks) is in truth not a pure recession.1 When the anterior border of the recessed muscle is placed 3 mm posteriorly along the lateral border of the IR muscle and 2 mm laterally...
Table 34-2. Recommended Measurements for Graded Recession of the Inferior Oblique Muscle (in mm)

<table>
<thead>
<tr>
<th>Desired Amount of Recession</th>
<th>Measured Posteriorly (from Lateral Border of True IR Insertion)</th>
<th>Measured Superiorly (Perpendicular to Lateral Border of IR)</th>
</tr>
</thead>
<tbody>
<tr>
<td>6</td>
<td>5.0</td>
<td>6.4</td>
</tr>
<tr>
<td>7</td>
<td>5.0</td>
<td>5.4</td>
</tr>
<tr>
<td>8</td>
<td>4.0</td>
<td>4.4</td>
</tr>
<tr>
<td>9</td>
<td>4.0</td>
<td>3.4</td>
</tr>
<tr>
<td>10</td>
<td>4.0</td>
<td>2.4</td>
</tr>
<tr>
<td>11</td>
<td>4.0</td>
<td>1.4</td>
</tr>
<tr>
<td>12</td>
<td>4.0</td>
<td>0.4</td>
</tr>
</tbody>
</table>

IR, inferior rectus


(Scheie-Parks point), the IO muscle is anteriorized by 1 to 1.5 mm (Fig. 34–9; see also Fig. 34–1). Probably for this reason, long-term follow-up showed that the 10-mm recession actually is a more powerful weakening procedure than the 12- to 14-mm recession.41

One “recession” operation secures the insertion of the disinserted IO muscle to the sclera at a point 10 mm posterior to the IR muscle insertion along the lateral border of the IR muscle (Fig. 34–10). This point is usually indicated by the presence of a vortex vein. It should be realized that this surgery is not a true recession, because the posterior portion of the muscle is reinserted a number of millimeters posterior to its natural course. Over time, much of the initial weakening induced by this surgery reverts toward recurrent overaction.41

ANTERIOR TRANSPOSITION

The anteriorization procedure is achieved by placing the IO muscle insertion anywhere anterior to the normal course of the muscle. In practice, a rectus muscle is used for reference.

The disinserted IO muscle may be placed anteriorly to just below the lower border of the LR muscle (“pure anteriorposition”) (Fig. 34–11).28 This procedure is technically relatively easy because the IO muscle is normally inserted about 10 mm behind the LR muscle insertion. We have found the operation to be useful in a few cases of mild to moderate IO muscle overaction.

In performing the more common anteriorization operation, the IR muscle is secured with a muscle hook after the IO muscle is isolated. The anterior fibers of the IO muscle are anchored to the sclera at the desired position temporal to the IR insertion (Fig. 34–12). The posterior fibers then are bunched up toward the anterior fibers, using a needle pass through the sclera about 0.5 to 1.0 mm temporal to the first needle pass, in a mini-crossed-swords or parallel orientation. Alternatively, the muscle may be attached to the globe with a single scleral bite after suturing the anterior and posterior muscle fibers together while on the clamp after disinsertion.1 If the new insertion of the IO is spread out laterally, there is an increased likelihood of restricting elevation.29

Most strabismus specialists use the IR as the muscle of reference. Anteriorization would place the IO muscle insertion anterior to a point about 4 mm posterior to the true IR muscle insertion site—where the IO usually crosses the IR muscle (see Fig. 34–1). It is convenient to grade anteriorization from the IR insertion line; anteriorization to the line...
Figure 34–12. Placing the inferior oblique muscle (in red) at the insertion of the inferior rectus muscle creates an anteriorization and stretches the neurovascular bundle (in yellow). (Courtesy of David Stager, MD.)

could be considered the “zero” station (see Fig. 34–12). Placement anterior to the line would be denoted in “positive” millimeters, whereas posteriorly it would be “negative.” Thus, anteriorization to 2 mm anterior to the IR would be declared as “+2” or to the “+2 station,” whereas placement 1 mm posterior to the IR would be “−1” or to the “−1 station.”

Because the IO muscle is brought quite forward in an anteriorization, it is important to dissect away all attachments to the muscle. Otherwise the eyelid may appear thicker or “fuller.” This may be particularly noticeable after unilateral anteriorization.

NASAL MYECTOMY

The treatment of residual IO muscle overaction after anteriorization was, until recently, considered to be difficult. We have found that further anteriorization is either ineffective or results in restriction to elevation. Nasal myectomy is believed to be an effective supplementary procedure. In this procedure, the IO muscle is captured near the IR muscle where it was anteriorized. It is then dissected cleanly along its course to where it emerges nasally from Tenon’s capsule.

One clamp is placed across the muscle just anterior to Tenon’s capsule and a second about 5 mm more temporally (Fig. 34–13). The muscle tissue between the clamps is resected and cautery applied to the muscle stumps on the clamps before they are released. No suturing is necessary. On the underside of the temporal (originally anteriorized) part of the muscle, one finds the neurovascular bundle to the IO (Fig. 34–14).

The operation may succeed simply because one has performed a large myectomy. However, Stager believes that nasal myectomy after anteriorization capitalizes on the neurovascular bundle as the new origin of the IO muscle by virtue of its anatomic course, prominent fibrocollagenous capsule, and ligamentous nature. He demonstrated this in experiments on cadavers and living patients. A practical difference between these two hypotheses occurs in patients with coexisting residual IO muscle overaction and DVD. If this operation succeeds only through its myectomy effect, coexisting DVD should not improve after nasal myectomy. A neurovascular bundle origin of the myectomized muscle, however, would serve to decrease DVD either by active depression or resistance to elevation.

DENERVATION AND EXTIRPATION

In this procedure the surgeon isolates the IO muscle, cauterizes the neurovascular bundle for denervation and hemostasis, and performs a large myectomy of as much as possible of the IO muscle from the point where it emerges from Tenon’s capsule up to its insertion. The opening in
Tenon's capsule may be sutured to prevent any reattachment of the IO muscle stump to the sclera. The procedure was found to result in less undercorrection than when 12- to 14-mm recessions are performed in patients with marked (4+) overaction. This rather extreme surgery permits no further procedure on the IO muscle if overcorrection or undercorrection persists, because only a small stump of muscle remains posterior to Tenon's capsule. Even with no IO muscle attached to the globe, apparent overcorrection or undercorrection can occur.

With apparent undercorrection, the surgeon may perform orbital imaging to evaluate the rectus muscles for an unusual insertion site, as may occur with an elevated insertion of the medial rectus muscle. For apparent overcorrection, one cannot reverse the extirpation. This operation now is seldom performed because large recessions and anteriorization operations have proved effective.

Choice of Sutures and Needles

Although the selection of suture material is easy, the choice of needles is not. Because an absorbable suture is desirable, the surgeon may choose from among the synthetic glycolic acid polymer sutures—Ethicon's Vicryl (plain and coated), Davis and Geck's Dexon and Dexon II, and Alcon's Biosorb-C. The last suture is relatively new; it slides through tissues easily, is simple to tie, and lacks memory or resistance. The preferred suture sizes are 5-0 and 6-0.

The ideal needle for IO surgery is yet to be developed. To minimize the possibility of scleral perforation, the needle should be spatulated. The needle should also be rather short because only a short scleral tunnel is required to secure an IO muscle. Because the surgeon must often try to reinsert an IO muscle while working in a small space, a highly curved needle is desirable. Inasmuch as the ideal needle is not available, the surgeon must select the suture product and needle that most closely meet his or her needs.

Recommendations

Considering the wide variety of IO muscle weakening procedures available, strabismus surgeons will have preferences acquired from their training or personal experience. Having been through both of these adventures, we suggest the following.

When unilateral IO muscle overaction requires correction, the surgeon should focus on the amount of hypertropia in upgaze. Almost any IO muscle weakening procedure will correct unilateral elevation on adduction and reduce a small V pattern, but operations differ in their effects on the deviation in upgaze. If the hypertropia increases in upgaze by 5 PD or less, a 12- to 14-mm recession is appropriate. If the hypertropia increases in upgaze by 5 to 10 PD, a 10-mm recession will be helpful because it includes both a recession and a small anteriorization. With unilateral IO muscle overaction and a hypertropia that increases more than 10 PD in upgaze, the surgeon may consider an anteriorization procedure but should be mindful that this may decrease elevation and induce hypotropia on attempted upgaze.

When correcting bilateral IO muscle overaction, the degree of overaction determines the procedure of choice. A mild to moderate overaction will usually benefit from bilateral 12- to 14-mm recessions. Moderate overaction deserves a 10-mm recession (for the reason described previously), whereas moderate to marked overaction is best treated by anteriorization. The severity of the IO muscle overaction indicates the amount of anteriorization to be performed.

Nasal myectomies have been performed for residual overaction after anteriorization. In some hands they have proved valuable.

Denervation and extirpation of the IO muscle may be considered in unilateral situations in which a 10-mm recession or anteriorization has already been performed. Even in these situations, other options exist. Recessions may be further anteriorized, or an anteriorization may be supplemented by a nasal myectomy.

Complications

FAT ADHERENCE SYNDROME

If excessive adipose tissue is liberated by rupturing posterior Tenon's capsule when exposing the IO muscle, hemorrhage and subsequent scarring may occur in the inferotemporal quadrant. The scarring can, through traction, either pull the eye downward or restrict upgaze. The patient will then have unilateral hypotropia and be unable to fully elevate the eye. Some believe this complication to be more common after myectomy than other IO muscle weakening procedures. On the other hand, surgeons who perform myectomies claim that the adherence syndrome is rare.

Precautions should be taken to avoid this complication. In isolating the IO muscle the surgeon should be careful not to incise the fat pad that encompasses much of the muscle (Fig. 34-15). With the muscle captured on a muscle hook, the surgeon should pull any adipose tissue off the hook until the tip appears immediately adjacent to the muscle. Only then should scissors be used to incise the tissue on the hook.

Faced with this complication, one must operate to remove the scarred tissue. Unfortunately, scarring has a strong tendency to recur. Attempts to cover the sclera with a material such as Supramyd, followed by sub-Tenon's injection of a corticosteroid, are sometimes successful in preventing scarring.

Figure 34-15. The muscle hook has captured the inferior oblique muscle and some of the surrounding orbital fat.
EFFECTS ON THE LATERAL RECTUS MUSCLE

Because the IO muscle inserts 1 to 2 mm beneath the LR muscle, the surgeon, when attempting to isolate the IO, could erroneously capture both muscles on the same hook, place a single suture through both of them, and disinsert them. This error is more likely to occur if the LR muscle has previously been resected. Also, during a previous operation on the LR muscle the IO muscle might have been pulled forward so that part of it lies close to the inferior insertion of the LR muscle. This distortion of the course of the IO muscle has been termed the J-shaped deformity (Fig. 34–16). It may cause either hypertropia or hypotropia in primary gaze with limited elevation of the involved eye.

To be certain that one is operating only on the IO muscle and not the LR as well, one should examine what is thought to be the IO muscle to be sure that it has been properly isolated before proceeding with the operation. The body of the IO muscle has a unique shape, somewhat resembling a sausage. If there is excessive tissue at the insertion, the surgeon should consider the possibility that the LR muscle has also been captured. With inspection, one can observe the horizontal orientation of the LR muscle fibers, contrasting to the vertical orientation of the IO muscle.

Another complication that involves the LR muscle is postoperative paresis. We have seen two patients who had an acute esotropia postoperatively that resembled sixth cranial nerve palsy. The surgical procedure and anesthetics had been uneventful. In one case, the esotropia spontaneously resolved. The second patient, however, required surgery for persistent esotropia. The problem may have resulted from damage to the LR muscle by the black silk suture used to retract the LR muscle when isolating the oblique muscle. To avoid this complication, the authors advise using a muscle hook on the LR muscle instead of a suture when exposing the oblique muscle, and then removing it as soon as the muscle is captured.

PUPIL DILATION

Excessive trauma or traction to the IO muscle may cause temporary or permanent mydriasis by damaging the nerve to the ciliary ganglion that bears the parasympathetic fibers to the pupil. James and Elston noted mydriasis in the operated eyes in 17 of 19 strabismus patients. Pupil dilation was seen with both oblique and rectus muscle surgery and bore no relation to the number of muscles operated on. The authors hypothesized that mydriasis results from the release of neurotransmitters from tissues damaged during surgery. Often the pupil will return to normal size over a few months, but permanent dilation may occur. Permanent pupil dilation may also result from pressure on the optic nerve from hemorrhage or direct trauma during the operation. Care should be taken to handle the IO muscle asatraumatically as possible.

HEMORRHAGE FROM THE INFERIOR OBLIQUE MUSCLE

The IO muscle is a highly vascularized muscle. It actually may be seen to swell during surgery and, when perforated, can bleed profusely. If a myectomy is performed, clamps and cautery are applied to prevent bleeding. Ligation or cauterization of the neurovascular bundle should be performed before denervation and extirpation. Otherwise the muscle may bleed subconjunctivally and blood may extend beneath the eyelid to create ecchymosis or, rarely, it may bleed within the orbit and cause proptosis. Hemorrhage in the region of the optic nerve may cause pressure and necrosis with secondary optic atrophy. Another possible cause of hemorrhage is from perforation of the inferotemporal vortex vein.

UNDERCORRECTION

Persistent or residual postoperative overaction characteristically either worsens or stabilizes over time. Undercorrection, however, almost never improves spontaneously. The criteria for a second IO muscle operation should be similar to those established for the original surgery. It is prudent to wait at least 3 months before reoperating.

The choice of a second procedure generally is dictated by what was done previously. If the second operation is to be performed on both eyes, a recession should be converted to an anterior transposition. A previous anterior transposition may call for a myectomy on the nasal side of the IR muscle. In the case of myotomies and myectomies, it is difficult to be specific. The surgeon should determine the site of muscle reinsertion and gauge the tightness of the muscle before deciding on the second operative procedure.

If the second operation is to be done on one eye, one must be mindful that a unilateral anterior transposition may limit upgaze and result in hypotropia in primary position and upgaze. Therefore, after a previous 10- or 14-mm IO muscle recession, the surgeon may wish to choose a denervation-extirpation or myectomy as a second procedure.

Figure 34–16. The previously resected lateral rectus muscle has advanced part of the adjacent inferior oblique muscle, causing a J deformity in this diagram. (Courtesy of Ronald Price, MD.)
PSEUDO–INFERIOR OBLIQUE OVERACTION

Pseudo-IO overaction may occur after anteriorization more than 1 mm anterior to the insertion of the IR. The anteriorized IO causes a restriction that limits elevation in abduction, produces overerelevation in adduction, and mimics the appearance of IO overaction. This may cause a V or Y pattern and is believed to be more likely to occur if the insertion is spread out laterally at the time of anteriorization than if the IO muscle is bunched up adjacent to the IR muscle. To correct this problem, the surgeon should either convert the anteriorization(s) to 12- to 14-mm recession(s) or perform a denervation and extirpation procedure.

OVERCORRECTION

When an overcorrection occurs after an IO muscle weakening procedure, the eye depresses on attempted adduction and an A pattern is evident. If the surgical procedure was carefully chosen, as discussed earlier, overcorrections are uncommon. In our experience, undercorrections are more frequent than overcorrections, in a ratio exceeding 10:1.

After an obvious overcorrection it may be difficult to reverse or alter the previous IO muscle operation. An anterior transposition may be converted to a 10- or 14-mm recession. If secondary SO overaction develops, the surgeon may consider weakening the SO tendon. A full or partial (3/4 or 7/8) tenotomy, tenectomy, or recession of the SO tendon may be helpful.

REFERENCES

In 1898, Von Graefe considered surgery on the oblique muscles as a *noli me tangere* (touch me not). As the anatomy of the superior oblique (SO) muscle came to be better understood, various procedures were designed to weaken or strengthen it. Parks, in 1970, described directly visualizing the SO tendon using a temporal approach. This improved method of exposure renewed interest in SO surgery.

Several currently used SO procedures include tenotomy and tenectomy of the SO tendon, partial posterior tenectomy of the SO, use of an SO expander, anterior lateralization of the anterior fibers of the SO (Harada-Ito procedure), and SO tuck.

The reader is referred to Chapters 15 and 25 on SO palsy and Brown syndrome for salient features of anatomy and physiology.

**Superior Oblique Tenotomy and Tenectomy**

**HISTORICAL PERSPECTIVE**

Superior oblique tenotomy and tenectomy are the most commonly used weakening procedures. In 1946, Berke described tenectomy of the SO tendon using a blind sweep through a superonasal incision to engage the SO. This maneuver was associated with a high rate of complications, such as damage to the vortex vein, incomplete tenotomy, adherence syndrome, and ptosis.

The technique was improved using direct visualization. The initial incision was placed in the superotemporal quadrant; access to the superonasal area was gained by reflecting the conjunctiva.

**INDICATIONS**

Tenotomy or tenectomy of the SO tendon has been used to treat Brown syndrome with variable results. It is also indicated for treating A-pattern strabismus with overacting SO muscles, and SO myokymia. The procedure is able to correct 20 PD or more of A pattern in the primary position and up to 45 PD in downgaze, but the amount of correction achieved depends mostly on the size of the preoperative pattern and the amount of SO overaction. Change in horizontal alignment in the primary position may vary from zero to 10 to 15 PD. Caution is advised when performing this procedure in patients with bifoveal fixation, because torsional diplopia may occur and impede fusion. It should not be performed in patients with both SO and IO overaction.

**TECHNIQUE**

The conjunctiva is incised in the superonasal quadrant 8 mm posterior to the limbus. The incision is enlarged approximately 6 mm circumferentially and a muscle hook placed in the posterior lip of the incision to engage the SO tendon. The tendon is isolated on a small muscle hook under direct visualization of the tendon with as little surrounding Tenon’s capsule as possible. Blind sweeping in the superonasal quadrant should be avoided to not injure the vortex vein. Under direct visualization the SO tendon is divided (tenotomy), or a section is removed (tenectomy). The surgeon should make sure that the complete width of the tendon has been isolated by passing another small hook to isolate any remaining fibers. Incomplete tenotomy or tenectomy is a common cause of undercorrection. The tendon is allowed...
to retract. Forced duction testing is performed to confirm the release of restriction in Brown syndrome (see Chapters 3 and 4).22

Knapp (personal communication, 1975) described using a "chicken suture" to promote recovery of the tenotomized ends of the tendon should further surgery be needed. A nonabsorbable suture (e.g., Mersilene 5-0 or 6-0) is woven through the proximal end of the SO tendon before tenotomy. A large loop is created and a similar pass made 2 mm distal to the first suture (Fig. 35–1A). When a segment of the SO needs to be removed, the distance between the sutures can be increased. The tendon is cut between the 2 sutures, taking care not to disrupt the integrity of the loop (see Fig. 35–1B and C). The suture retracts in the subconjunctival space, within the surrounding Tenon's fascia (see Fig. 35–1D). The conjunctiva is closed with interrupted 8-0 Dexon sutures.

COMPLICATIONS

Iatrogenic SO palsy (overcorrection) has been described by several authors after SO tenotomy and tenectomy.11, 12, 15, 21, 45, 54–56, 67, 71, 92 It has been reported in 30% to 85% of cases and may appear up to 24 months after surgery.62 Patients present with limited depression in adduction and intractable cyclovertical diplopia (Fig. 35–2A). Simultaneous prophylactic recession of the ipsilateral IO has not entirely eliminated this problem.56 In some cases, SO palsy can be improved only by rejoining the cut ends of the tendon using a silicone spacer (Fig. 35–2B).24 If this procedure is contemplated, the surgeon should confirm that the tendon can in fact be located. High-resolution dynamic magnetic resonance imaging may provide crucial information79 (see also Chapters 6 and 41).

Undercorrection is usually due to incomplete tenotomy or

Figure 35–1. Superior oblique tenotomy. A. A nonabsorbable suture is woven through the proximal end of the superior oblique (SO) tendon and a similar pass is made 2 mm distal to the first suture creating a large loop. The superior rectus (SR) is isolated in a muscle hook. B. The tendon is cut between the two sutures with Westcott scissors, taking care not to cut the preplaced loop. C. The SO tendon has been tenotomized; the ends remain attached to the loop. The chicken suture (arrowhead) is held by a needle holder. D. The loop is allowed to retract in the subconjunctival space, within the surrounding fascia. (White arrowhead[s] point to nonabsorbable muscle suture; conjunctival opening is indicated by dashed lines; SR muscle is indicated by solid lines.)
SUPERIOR OBLIQUE PROCEDURES

Figure 35-2. A patient who had undergone two separate procedures to correct a left superior oblique palsy after superior oblique tenectomy was performed for Brown syndrome. Note the underaction of the left superior oblique and 10 PD of V-pattern esotropia. Also a left hypertropia increases on right gaze and downgaze. B, Six months after successful reanastomosis of the left superior oblique with a 240 silicone band as a bridge, improvement of left hypertropia on right gaze and improved rotations of left superior oblique can be observed. Some residual left superior oblique underaction remains.

ALTERNATIVE PROCEDURES

Different weakening procedures have been described for SO overaction and Brown syndrome, some of which have been abandoned because of unpredictable results. They in-

Figure 35-3. A patient who had undergone bilateral superior oblique procedures for bilateral Brown syndrome. Note bilateral ptosis, which is more evident on the right.
Figure 35–4. Posterior tenectomy of the superior oblique (SO). A, The conjunctival incision is fashioned 8 mm posterior to the limbus in the superior temporal bulbar quadrant. B, The incision (dotted lines) is extended radially 6 mm. C, The superior rectus (SR) muscle, isolated on a muscle hook, should be cleaned of the intermuscular septum on its lateral border. D, Desmarres retractor is placed in the posterior lip of the incision, and the SO tendon is isolated under direct visualization at its insertion. E, The SO is isolated on a muscle hook, including the posterior fibers. F, The SO fibers are cut at the scleral insertion from a posterior approach.
include Z-tenotomy, split tendon lengthening, and SO recession. SO recession is a technically more difficult procedure than tenotomy and tenectomy and has been associated with undercorrections. The results obtained by tenotomy and recession are similar.

Interest has arisen in performing trochlear luxation to lengthen (and thus weaken) the SO, using an external periosteal incision. For patients with overacting SO muscles and binocular fusion, torsional misalignment is a concern. Partial posterior tenectomy of the oblique fibers of the SO, preserving the anterior torsional fibers, allows SO weakening without affecting torsion. The SO expander is an alternative that is discussed later in this chapter.

**Posterior Tenectomy of the Superior Oblique**

**HISTORICAL PERSPECTIVE**

The anterior fibers of the SO tendon are responsible for torsion, whereas the posterior fibers are implicated in downward rotation and abduction. This procedure was described by Prieto-Diaz in 1976 to selectively weaken the abducting and downgaze functions of the SO while preserving its torsional properties.

There is minimal risk of inducing SO palsy, excyclotorsion, or anomalous head posture. The procedure may be performed in bifoveal fixators because negligible excyclotorsion is induced. Induced vertical deviation after the procedure is infrequent.

**INDICATIONS**

This surgery may serve to correct moderate A-pattern strabismus (up to 20 PD) with overacting SO muscles, as well as incomitant dissociated vertical deviation associated with an A pattern and an overacting SO (in conjunction with SR recession) (see Chapter 17).

**TECHNIQUE**

A lid speculum is inserted and the globe rotated inferiorly. An incision is fashioned in the temporal bulbar conjunctiva 8 mm posterior to the limbus (Fig. 35-4A) and extended...
Figure 35-5. A, Patient with moderate Brown syndrome, right eye. Note hypotropia on left gaze, downshoot in the right eye, and limitation of elevation in adduction. The right superior oblique rotation is normal (bottom right). B, After a superior oblique spacer procedure, the eyes are now straight in left gaze, without a downshoot. Rotation in elevation and adduction is improved but remains limited. Right superior oblique function is preserved.
radially for 6 mm (Fig. 35–4B). The SR muscle is isolated on a muscle hook and cleaned of intermuscular septal tissue on its lateral border (Fig. 35–4C). A Desmarres retractor then is placed in the posterior lip of the conjunctival incision, and the SO tendon is isolated under direct visualization at its insertion (Fig. 35–4D and E). For enhanced visualization posteriorly, the lid speculum may be removed. Care is taken to ensure that the posterior fibers of the SO insertion are included. The surgeon should be aware of anatomic variations in the insertion. Misidentification of the posterior fibers leads to incomplete tenotomy and undercorrection. The posterior portion may need to be followed nasally beneath the SR.

The SO fibers are cut at the scleral insertion from a posterior approach, leaving the anterior 1 to 2 mm of tendon in place. To avoid injuring the superotemporal vortex vein, the scissors tips should be directed away from the globe. A quadrilateral posterior tenectomy is completed (Fig. 35–4F and G). The remaining anterior fibers then are measured, making sure that no more than 1 to 2 mm remains attached (Fig. 35–4H). Finally, the conjunctiva is closed with interrupted 8-0 Dexon suture (Fig. 35–4I and J).

Unequal tenectomies in bilateral cases may lead to residual asymmetric SO overaction and induced vertical deviation. Ptosis, if present postoperatively, usually subsides within a few weeks.

**Superior Oblique Expander**

**HISTORICAL PERSPECTIVE**

SO tenotomy may result in significant overcorrection and excyclotorsion with bothersome diplopia. A graded weakening procedure described by Wright involves expanding the length of the tendon using a silicone spacer. The weakening effect is graded by varying the length of the silicone that bridges the gap between the cut ends of the tendon. The spacer lengthens the tendon without altering the muscle mechanics at its insertion. Both ends of the tendon are secured to the spacer, preventing scleral adhesions and allowing it to be localized if reoperation is necessary. Excyclotorsion is not induced, making the procedure beneficial for patients with bifoveal fusion.

**INDICATIONS**

This procedure is used to correct SO overaction in A-pattern strabismus, eliminating as much as 20 to 55 PD of deviation. and also SO overaction associated with skew deviations. The expander is beneficial in treating Brown syndrome (Fig. 35–5). We have used a silicone spacer to reanastomose tenotomized or tenectomized SO tendons for correcting iatrogenic SO palsy (Fig. 35–2).

**TECHNIQUE**

A lid speculum is inserted and the assistant rotates the eye inferiorly. The conjunctiva is incised in the superotemporal quadrant 8 mm posterior to the limbus, and the incision is extended circumferentially in a nasal direction for 6 mm (Fig. 35–6A). The SR muscle is isolated on a muscle hook, and the globe is rotated inferiorly. The opening in the conjunctiva is reflected over the muscle hook nasally to expose the intermuscular septum nasal to the SR. Tissue manipulation is minimized, and the SO tendon is identified near the nasal border of the SR (Fig. 35–6B). A buttonhole incision is fashioned in the intermuscular septum to expose the whole width of the tendon. As much as possible of the nasal intermuscular septum should be preserved so that a sheath of connective tissue will envelop the spacer, permitting free movement of the tendon beneath the SR. This also will prevent scarring to sclera and minimize the risk of implant extrusion.

With the SO tendon isolated on a small muscle hook, a double-armed 6-0 nonabsorbable suture is woven through the tendon and locked on each end 1 to 2 mm nasal to the SR border (Fig. 35–6C). A similar suture is woven 2 mm from the first (Fig. 35–6D), and the SO tendon is cut (tenotomized) between the two sutures (Fig. 35–6E). The surgeon should make sure that the entire width of the tendon has been severed and that the SR is not injured. A silicone band (size 240 or 40) used for retinal buckling procedures is soaked in antibiotic solution (e.g., gentamicin), rinsed, and trimmed according to the desired length. The length of the expander varies with the degree of SO overaction or the amount of A pattern (Table 35–1). A longer expander is preferred in cases in which the A pattern is disproportionately large for the degree of SO overaction. In Brown syndrome, a 6- to 7-mm expander is recommended.

Each nonabsorbable suture is passed flat through the thickness of the band along its width (Fig. 35–6F). The sutures are tied snugly on the surface of the spacer, securing the band to both ends of the SO (Fig. 35–6G).

An intraoperative forcedduction test is performed to verify that restriction has been released in cases of Brown syndrome (Fig. 35–6H). The buttonhole incision of the intermuscular septum is closed to prevent adhesion of silicone to the conjunctiva (Fig. 35–6I). The conjunctiva then is closed separately with Dexon 8-0 sutures (Fig. 35–6J).

**COMPLICATIONS**

If performed well, this procedure can be successful (see Fig. 35–5B) and cause very few complications. The most common problem is persistent SO overaction and undercorrection. The surgeon has the option of going back to the SO tendon and changing the length of the spacer to achieve more effect, although this has not been effective in our experience.

If too much dissection has been performed, postoperative inflammation leads to fibrosis, scarring, and restriction of the free sliding movement of the tendon beneath the SR.

<table>
<thead>
<tr>
<th>Table 35-1. Recommended Length of Silicone Spacer</th>
</tr>
</thead>
<tbody>
<tr>
<td>Superior Oblique Overaction</td>
</tr>
<tr>
<td>+1</td>
</tr>
<tr>
<td>+2</td>
</tr>
<tr>
<td>+3</td>
</tr>
<tr>
<td>+4</td>
</tr>
</tbody>
</table>
Figure 35–6. Superior oblique (SO) expander. A, A conjunctival incision fashioned in the superior temporal bulbar quadrant 8 mm posterior to the limbus is extended circumferentially for 6 mm. B, The superior rectus (SR) muscle is isolated on a muscle hook, and the temporal incision on the conjunctiva is reflected nasally. The SO tendon is identified near the nasal border of the SR. Tissue manipulation should be minimal. C, A double-armed nonabsorbable suture is woven through the SO tendon 2 mm nasal to the SR border. D, A similar suture is woven 2 mm nasal to the first. The two ends of the suture are retracted to show their position with respect to the SO tendon. The tendon is cut between the two preplaced sutures. E, The two cut ends of the SO tendon. F, The nonabsorbable sutures are passed through the thickness of a 240 or 40 silicone band (spacer), along its width.
Figure 35–6 Continued G. The sutures are tied snugly over the silicone band and secured to the two ends of the SO tendon. H. An intraoperative forced duction test is performed to verify that the restriction present in Brown syndrome has been released (arrow indicates direction of forced duction testing). I. The intermuscular septum is closed with interrupted 8-0 Dexon sutures (small white arrows) to prevent adhesions of the silicone band to the conjunctiva. J. The conjunctiva is closed with interrupted 8-0 Dexon sutures. (White arrowhead[s] point to nonabsorbable muscle suture; dashed lines indicate conjunctival opening; solid lines indicate SR muscle.)
Persistent restriction of downgaze with diplopia in the reading position has also been reported.\textsuperscript{93} Iatrogenic SO palsies may be caused by an excessive length of silicone. A patient may develop secondary IO overaction and associated V-pattern strabismus.\textsuperscript{96} If necessary, the surgeon can replace the spacer with a shorter silicone band. Discomfort in the superior fornix may be caused by movement of the expander against adjacent tissue. This usually disappears once the silicone has been encapsulated and can move freely under the SR. Implant extrusion has occurred infrequently.

**Superior Oblique Tuck**

**HISTORICAL PERSPECTIVE**

The efficacy of SO tuck for a paretic SO has been controversial. Some authors favor the procedure,\textsuperscript{20, 26, 37} whereas others prefer weakening of the IO—which commonly overacts as a result of SO underaction.\textsuperscript{20, 26, 36, 49, 84, 87} IO procedures are discussed in Chapter 34. The (strengthening) tuck procedure is recommended for an incompletely paralyzed SO muscle\textsuperscript{39, 44} but is technically challenging.\textsuperscript{3, 31, 87} An alternative resection procedure at the SO insertion has been described but undercorrections may result.\textsuperscript{1}

**INDICATIONS**

This procedure is appropriate for the patient having profound SO underaction without IO overaction who also has at least 4 degrees of excyclotorsion and little to no vertical deviation in lateral gaze out of the field of the involved SO (Fig. 35–7).\textsuperscript{20, 26, 37, 39, 61, 76, 84} The operation aims to eliminate hypertropia and cyclotropia in the primary position and downgaze, correct anomalous head posture, and restore and enlarge the binocular diplopia-free field of vision.\textsuperscript{27, 36, 72}

It corrects torsion in the primary position and up to 12 degrees in downgaze, while also correcting as much as 15 PD of vertical deviation.\textsuperscript{51} The precise amounts of torsion and vertical deviation corrected depend on the size of the tuck and the amount of residual SO function. This surgery is more effective in patients with unilateral acquired SO palsy than in bilateral cases.\textsuperscript{29, 30, 94} It works well in patients with lax SO tendons—commonly seen in congenital cases (see Chapter 15).\textsuperscript{28, 58} Bilateral SO tucks can correct 15 to 20 PD of esotropia in downgaze.\textsuperscript{51} The tuck should be used judiciously in patients without tendon laxity.

**TECHNIQUE**

A lid speculum is inserted and forced ductions for oblique muscles performed to assess tendon laxity (see Chapters 4 and 15).\textsuperscript{22, 58, 99} The SO tendon is exposed by a superotemporal conjunctival incision, similar to that described for SO expander. Adequate visualization is critical. With a Desmarres retractor on the posterior lip of the wound, the intermuscular septum on the lateral border of the SR muscle is cleaned (Fig. 35–8A). Care is taken to ensure that the whole width of the tendon is identified and included in the tuck.

We use a marking suture (e.g., 5-0 black nylon) to tag the entire SO tendon nasal to the SR. The suture is passed under the SO tendon nasal to the SR (Fig. 35–8B) and brought to the temporal side of the SR to visualize the entire insertion (Fig. 35–8C through E). The desired amount of tuck is measured with a caliper (Fig. 35–8F). Only half of the desired tuck should show on the caliper because the SO tendon is folded on itself. The tuck is performed temporal to the SR by imbricating the anterior and posterior halves of the tendon with interrupted 6-0 nonabsorbable sutures (Fig. 35–8F). Some surgeons use a tendon tucker or a muscle hook for this procedure (Fig. 35–8G). Forced duction testing is repeated to assess the adequacy of the tuck and detect any restriction. The ideal position is based on resistance to elevation in adduction; the inferior limbus should reach the horizontal meridian during passive traction testing (Fig. 35–8H).\textsuperscript{73} The final appearance of the tuck before conjuncti-
val closure is shown in Figure 35–8I. The conjunctiva is closed with interrupted 8-0 Dexon suture.

In cooperative adults we have used nonabsorbable interrupted sutures (e.g., Novafil 6-0) and left the conjunctiva open for postoperative adjustment in the office. Additional interrupted sutures are added, one nasally and one temporally, to the designated amount of tuck. A loop of Vicryl 6-0 suture is placed beneath the SO tendon to assist its identification during adjustment. Scleral “bucket handles” may be used to aid exposure (as used for adjustable recession or resection). The amount of tuck can be modified by releasing one interrupted suture at a time. Conjunctival closure is optional following adjustment.

**COMPLICATIONS**

Iatrogenic Brown syndrome is the most common complication following SO tuck, occurring in 17% of patients. This can be avoided if careful forcedduction testing is performed during surgery. If the restriction is mild, the patient usually improves over the next 4 to 6 weeks. Excessive restriction or intolerable torsional diplopia should be treated by releasing or reducing the tuck in the immediate postoperative period. The procedure is difficult to reverse if sufficient time is allowed for healing (see also Chapter 41).

Residual vertical deviation is found in cases of undercorrection, especially if there is a large preoperative vertical deviation in the primary position. In these cases it is preferable to combine the tuck with an appropriate vertical rectus muscle procedure or IO recession.

**Harada-Ito Procedure**

**HISTORICAL PERSPECTIVE**

Anterior lateralization of the anterior torsional fibers of the SO tendon was first described by Harada and Ito in 1964 as a treatment for excyclotropia. Originally, the procedure was performed by splitting the tendon in two and passing a suture 5 mm behind the insertion. The suture then was anchored to the sclera 5 mm anterior to the SO insertion. Since then the eponym Harada-Ito has been used for any strengthening procedure involving the anterior half of the SO tendon.

In 1974, Fells modified the technique, splitting the tendon in two and disinserting and advancing the anterior half anteriorly and laterally. The tendon is secured on the sclera just above and perpendicular to the superior border of the lateral rectus muscle. This moves the axis of the tendon closer to the anterior-posterior axis of the globe, thereby improving torsion. The procedure usually has no effect on vertical deviation and does not generally limit elevation in adduction (acquired Brown syndrome).

**INDICATIONS**

Torsion anomalies become symptomatic beyond 5 degrees of cyclodeviation. Excyclotropia is usually secondary to SO palsy, with trauma being the most common cause. Some cases have been associated with retinal detachment surgeries (see Chapter 22). The Harada-Ito procedure corrects up to 10 degrees of excyclotorsion in primary position and up to 15 to 20 degrees in downgaze. The procedure may be difficult to perform in patients who have had previous surgery on the SO tendon and resultant scarring.

**TECHNIQUE**

Access to the SO is gained through a superotemporal approach, as described for SO expander except that the incision is extended temporally in a circumferential direction. The globe is controlled with a muscle hook beneath the SR. With a Desmarres retractor on the posterior lip of the incision, the intermuscular septum on the lateral border of the SR is incised. The SO tendon is isolated under direct visualization at its insertion. Its attachments to the SR, especially on its undersurface, should be severed to allow easier transposition.

The tendon then is split into anterior and posterior halves up to the level of the midportion of the SR, using small Westcott scissors (Fig. 35–9A through C). A preplaced 6-0 Vicryl suture is woven through the anterior half of the tendon and locked on each end. If an adjustable technique will be performed, we prefer using a smooth nonabsorbable suture such as Prolene or Novafil. The anterior half of the tendon is disinserted (see Fig. 35–9D). The lateral rectus muscle is isolated on a muscle hook, exposure its superior border. The disinserted tendon is transposed anteriorly and laterally on the sclera 8 mm posterior to the lateral rectus insertion and just superior to the upper border of the lateral rectus muscle (see Fig. 35–9E and F). With the adjustable technique the conjunctiva is left open (see Fig. 35–9G); otherwise it is closed with interrupted absorbable sutures (e.g., 8-0 Dexon). Bucket handles and bolster sutures may be used, as described in Chapter 33.

An intraoperative adjustment has been described for use in children, observing the relationship of the fovea to the optic disc. The fovea is aligned with the junction of the upper two thirds and lower one third of the disc before the sutures are permanently tied.

**COMPLICATIONS**

Rarely, advancing the anterior half of the SO tendon can limit elevation in adduction on forcedduction testing (iatrogenic Brown syndrome). This can be prevented by splitting the tendon 15 mm longitudinally.

**ALTERNATIVE PROCEDURES**

Several procedures have been described to correct excyclotorsion (see also Chapter 4). Appropriate horizontal transposition of the vertical rectus muscles can be performed primarily or after failed or undercorrected Harada-Ito surgery for excyclotorsion. Nasal transposition of the inferior rectus of one full tendon width will correct approximately 7 degrees of excyclotorsion in the primary position and 10 to 15 degrees in downgaze. However, transposition of the inferior rectus may limit downward rotation. The surgery can be combined with temporal transposition of the SR muscle for greater effect, correcting more than 20 degrees of excyclotorsion in downgaze. Vertical transposition of the
Figure 35-8. Superior oblique (SO) tuck. A. The superior rectus (SR) muscle is isolated on a muscle hook, and the SO tendon is identified nasal to the SR. B. A 5-0 black silk suture is used to mark the SO. C, D, and E. With a series of maneuvers, the silk suture is passed beneath the SR and brought out temporal to the SR. With the marking suture on the temporal side, the entire width of the SO insertion is identified. F. left, The amount of tuck is measured with a caliper. (Only half of the desired tuck is shown on the caliper because the SO tendon is folded over.) F. right, The tuck is performed by imbricating the anterior and posterior fibers of the tendon with a nonabsorbable suture.
Figure 35-8 Continued G, The tucking procedure can also be performed using a tendon tucker. H, An intraoperative forced duction test is performed to verify if a restriction has been created with the tuck. The ideal position of the globe during the forced duction test is for the inferior limbus to reach the horizontal meridian. I, Appearance of a tucked SO tendon before it is allowed to retract beneath the SR. (White arrowhead[s] point to nonabsorbable muscle suture; dashed lines indicate conjunctival opening; solid lines indicate SR muscle.)
**Figure 35–9.** Harada-Ito procedure. A, The superior rectus (SR) muscle is isolated on a muscle hook. A Desmarres retractor is placed on the posterior lip of the wound to enhance visualization. The caliper is shown measuring half of the superior oblique (SO) tendon. The SO tendon insertion is identified under direct visualization. B, The tendon is split longitudinally using small Westcott scissors. C, The SO tendon is shown divided into an anterior and posterior portion. D, After the anterior half of the tendon has been secured with muscle sutures (e.g., Vicryl 6-0), the anterior half of the tendon is disinserted.
horizontal muscles is reportedly less effective than horizontal transposition of the vertical rectus muscles for correction of torsion. If excyclodeviation is associated with vertical deviation in primary gaze, an SO tuck may be effective. Slanting of the insertions of all four rectus muscles is discouraged due to the risk of anterior segment ischemia.

REFERENCES

84. Toosi SH, von Noorden GK: Effect of isolated inferior oblique muscle
The focus in this chapter is on selected surgical procedures for limited ocular rotations with minimal or no residual muscle function. We do not discuss transposition of the vertical rectus muscles for torsion or transposition of the horizontal rectus muscles for small vertical deviations or pattern strabismus. The reader is referred to the appropriate reference. The principles of vertical rectus muscle transposition for torsion are discussed in Chapter 4. The management of pattern strabismus with associated horizontal deviations is reviewed in Chapter 14. Anterior transposition of the inferior oblique is discussed in Chapter 34.

The earlier term used for muscle transposition was muscle transfer procedure. Because the insertion or belly of the muscle is not really transplanted but rather repositioned to alter its action upon the globe, the term muscle transposition is now preferred.

General Indications

The goals of most of the procedures described in this chapter are to (1) improve rotation, (2) expand binocular diplopia-free fields, and (3) shift any residual binocular diplopia-free field to more functional fields such as primary gaze and downgaze. The surgeon and patient should realize that to restore full ocular motility and relieve all restriction to full rotation are unrealistic expectations from muscle transposition surgery.

If a rectus muscle is not functioning, any strengthening procedure such as tucking, resection, or advancement will have only a temporary mechanical effect and will not enhance rotation in the field of the palsied muscle. Because a palsied muscle does not contract, transposition procedures are required to improve rotation in that field. Of necessity, transposed muscles must be functional to provide this force.

Thus, it is of paramount importance that residual or absent muscle function be precisely diagnosed. Our algorithm for determining the appropriate surgical strategy is shown in Figure 36–1. If a muscle can rotate beyond the midline, it is safe to assume that it generates sufficient force. If the eye rotates short of the midline, the crucial question to answer is whether the muscle generates force. This may be clarified by force generation testing.

In cases in which force generation testing yields weak or equivocal results, saccadic velocity analysis clarifies the degree of residual muscle function. In many situations, electro-oculography techniques will suffice. Scleral search coil techniques are more precise, especially if vertical saccades are to be analyzed. Twenty-degree saccades are sufficient for evaluating muscle function. For example, to assess lateral rectus (LR) function in abducens nerve palsy, saccades may be generated from 30 degrees of adduction to 10 degrees of adduction. Other authors variably define palsy as (1) a more than 30% reduction from normal saccadic velocities, (2) the amount of generated force less than 20 g, and (3) more than a 40% difference between agonist and antagonist saccades.

We have found that an LR saccadic velocity less than 100 degrees/sec for a 20-degree saccade is reliable for identifying patients who will require transposition procedures.

If preoperative evaluation discloses sufficient (moderate) force, a strengthening procedure such as resection is effective. If no significant muscle force generation is apparent, the paretic muscle should be left undisturbed to preserve the blood supply from its anterior ciliary arteries. A transposition procedure should be the initial surgical strategy. Both procedures are usually combined with weakening of the ipsilateral antagonist.

Theories

Initially, the efficacy of transposed muscles was thought to reflect the relearning of muscle function. It was hypothesized that a readjustment occurred so that the transposed muscle performed coordinated eye movements in a direction different from its original function.
With the advent of quantitative muscle force measurements, this theory was refuted. The postulated innervational plasticity did not occur. Rather than relearned function, the effects of a transposition procedure are based solely on a shift in available mechanical factors. Transposition surgery results in improved saccadic velocities, though not to normal levels. When vertical rectus muscle transposition was performed for LR palsy, the vertical muscles did not recruit on attempted abduction but merely fired at a low level. The change in the direction of vector forces was minimal. A full transposition of the vertical rectus muscles to the LR produces approximately 16 g of abducting force.

Recently, pulley systems have been identified by surface coil magnetic resonance imaging studies. The posterior two thirds of a transposed muscle belly moves less than expected relative to the orbital walls. That is, the transposed muscle does not follow the shortest path from its origin to its new insertion. Connective tissue sheaths and orbital fat serve as pulley systems that restrict free movement to the shortest path. These pulley systems are currently believed to be the explanation for the limited effect of transposition surgery, because they prevent shifting of the muscle belly toward the desired optimal direction.

### Procedures for Abduction Deficit

Of all the transposition procedures, those performed for abduction deficit have yielded the best results because only one muscle, the LR, is paralyzed. The procedure is also effective for absent LR function caused by a lost LR muscle after strabismus surgery, trauma to the LR, and congenital absence of the LR. Significant abduction deficit in Duane syndrome also has responded to this type of surgery (Fig. 36–2). Several vertical rectus transposition procedures have evolved, including partial tendon transposition, full tendon transposition, and the rectus muscle union procedure (Fig. 36–3).
Partial Tendon Transposition

Rectus Muscle Union

Full Tendon Transposition

PARTIAL TENDON TRANSPOSITION

In 1907, Hummelsheim\(^{34,35}\) described the first partial tendon transposition procedure for a paralytic LR muscle. The temporal halves of the superior rectus (SR) and inferior rectus (IR) were transposed to the LR insertion (see Fig. 36–3A). O’Connor\(^{61,62}\) performed the transposition procedure using the nasal halves of the SR and IR to decrease adducting power while enhancing abduction. Berens and Girard\(^3\) combined a horizontal rectus recession-resection with the transposition. Both vertical rectus muscles were disinserted and moved half a tendon width temporally. Only the temporal halves were sutured to the resected LR. All four rectus muscles are operated upon in this procedure. Wiener\(^{89,91}\) transected the LR muscle and split the proximal tendon to join the SR and IR. Carlson and Jampolsky\(^4\) described an adjustable partial tendon transposition that permits self-adjustment in the vertical axis of the united temporal halves of the transposed partial tendons of the vertical rectus muscle. This technique makes the abduction effect of the transposition adjustable (see Fig. 36–3B).

RECTUS MUSCLE UNION

Jensen\(^{18}\) described a rectus muscle union procedure for palsied muscles. In LR palsy, for example, the SR, IR, and LR are all split along their long axis. The lateral half of the SR is united with the superior half of the LR; similarly, the lateral half of the IR is united with the inferior half of the LR (see Fig. 36–3C). To be effective, the antagonist medial rectus (MR) is recessed. This procedure was originally thought to prevent anterior segment ischemia that complicates several of the transposition procedures. Because the muscles are not disinserted but only split, the ciliary vessels should be preserved. Unfortunately, experience over several decades has shown that the technique is prone to strangulate vessels from the three rectus muscles that are united.\(^{17,23-25}\) Kushner\(^{42}\) modified the Jensen procedure by using only the middle fourths of the LR in union with the temporal halves of the SR and the IR. If the anterior ciliary circulation cannot be distinctly identified on the surface of the LR, only the middle third of the vertical rectus muscles is included in the union with the LR. In this way the ciliary circulation from both vertical rectus muscles may be preserved.

FULL TENDON TRANSPOSITION

O’Connor\(^{61}\) initially performed a full tendon transposition of the SR and IR to the LR insertion, combined with a cinct procedure on the LR muscle. More abduction was obtained, but vertical imbalance was more frequent. In addition, because of MR contracture, the procedure needs to be combined with MR recession, increasing the risk of anterior segment ischemia. This led O’Connor to shift to the partial tendon transfer described earlier. To circumvent the risk of anterior segment ischemia, Hildreth\(^{33}\) joined the entire tendons of both vertical rectus muscles with nonabsorbable suture. Schillinger\(^{86}\) transposed the full tendon near the LR insertion parallel to the spiral of Tillaux. Uribe\(^{66}\) combined the Schillinger procedure with a horizontal recession-resection, again operating on four rectus muscles.

We described a full tendon vertical transposition procedure with adjustable sutures to correct surgically induced vertical deviations and consecutive exodeviations in the immediate postoperative period.\(^{44}\) The vertical rectus muscles are sutured to sclera near the LR insertion parallel to its long axis (see Fig. 36–3D). The procedure initially was combined with MR recession for maximal effect. Later, pharmacologic weakening on the antagonist MR was suggested.\(^{71}\) McManaway\(^{69}\) and associates described a similar procedure in which the transposition is performed parallel to the spiral of Tillaux. Recently, Foster\(^{31}\) described augmenting of the transposition by a lateral fixation suture joining the transposed muscle to sclera, with encouraging results (see Fig. 36–3E).

CURRENT TECHNIQUES

We describe the adjustable transposition technique in this section as well as present Foster’s modification. The first procedure is ideal for reluctant surgeons who fear induced vertical deviations and overcorrections. Lateral augmentation, on the other hand, is recommended if maximal transposition force is desired.

Adjustable Transposition

Passive forced duction testing is performed to assess secondary contracture of the antagonist MR.\(^{44}\) Traction sutures (e.g., Vicryl 6-0) are placed in the oblique quadrants between the LR and IR muscles and between the LR and SR muscles. The conjunctival opening is made 7 mm posterior to the limbus and extended circumferentially 6 mm between the LR and the vertical rectus muscle (Fig. 36–4A).

The SR is isolated and disinserted, with the surgeon taking care that its attachments to the superior oblique (SO) muscles are severed. The LR muscle is isolated on a hook. The SR is sutured to the sclera 1 mm above the superior border of the LR muscle (Fig. 36–4B) and aligned parallel to the long axis of the LR (see Fig. 36–3D). If an adjustable procedure is planned, the sutures are tied in a slip knot manner as in other adjustable rectus muscle procedures (see Chapter 33). A mirror-image technique is carried out on the IR muscle; it should be verified that capsulopalpebral attachments and the intermuscular septum are separated from the muscle (Fig. 36–5).

At the conclusion of surgery, the conjunctival incisions are left open to provide access to the adjustable sutures. Passive forced duction testing is repeated superiorly and inferiorly to detect iatrogenic vertical restriction. A sub-Tenon’s injection of corticosteroid is optional but is recommended to decrease inflammation on the first postoperative day, allowing easier adjustment.

On the day of adjustment, if there is an induced vertical deviation, the appropriate vertical rectus muscle is recessed using the adjustable sutures. If there is consecutive exodeviation, both vertical rectus muscles are recessed by the same amount. Closure of the conjunctiva (usually with 8-0 Dexon) after the adjustment is optional.

The procedure is commonly combined with ipsilateral MR weakening—either MR recession 4 to 6 months later\(^{20}\) or botulinum toxin injection of the MR several days after transposition surgery.\(^{71}\) If MR recession is chosen, this muscle may be placed on an adjustable suture, allowing centration...
of the binocular field.\textsuperscript{20} We prefer pharmacologic denervation 2 to 3 days after surgery.

**Lateral Augmentation with Posterior Fixation Suture**

In this technique, the same conjunctival opening is created 7 mm posterior to the limbus as just described (Fig. 36–6A).\textsuperscript{21} The transposition of the vertical rectus muscles is placed parallel to the spiral of Tillaux. The insertion of the rectus muscles serves as a landmark for suture placement (Fig. 36–6B). This initially creates a 45-degree orientation of the vertical rectus muscles and causes a gap between the LR and the transposed muscle. A nonabsorbable suture (e.g., 5-0 Dacron) is placed 7 to 8 mm posterior to the muscle insertion through the lateral 25% of the SR or IR (Fig. 36–6C) and secured to the sclera just superior or inferior to the border of the LR (Fig. 36–6D), respectively, closing the gap between the LR and the transposed muscle (Fig. 36–6E). Unlike the adjustable transposition procedure, weakening of the antagonist MR may not always be required. This procedure has shown encouraging initial results, enhancing abduction (Fig. 36–7) and also improving the size and centration of the binocular diplopia-free field.\textsuperscript{21}

**Procedures for Vertical Muscle Palsy**

**KNAPP PROCEDURE**

Knapp\textsuperscript{41} was the first to describe full tendon transposition for double elevator palsy, transposing both horizontal rectus muscles to the SR (Fig. 36–8A). The mean correction achieved ranges from 19 to 38 PD of hypotropia, and elevation improves by 20 to 45 degrees.\textsuperscript{6, 9, 41, 46} Full elevation is not obtained unless the SR retains moderate strength.\textsuperscript{21} In smaller vertical deviations, graded shifting of the horizontal rectus muscles is advised.\textsuperscript{16} The same procedure may be used to correct hypotropia due to SR palsy.

As in transposition procedures for abduction deficit, the procedure will prove inadequate in the presence of secondary IR contracture that restricts upward rotation.\textsuperscript{20, 18, 28, 46, 50, 52, 80} It may be necessary to stage the procedure by recessing the IR before the transposition.\textsuperscript{59} The IR recession may be performed using a vessel-sparing technique to decrease the risk of anterior segment ischemia.
SELECTED TRANSPOSITION PROCEDURES • 481

**Figure 36-6.** Posterior fixation suture augmentation of vertical muscle transposition. A, Conjunctival opening is started 7 mm posterior to the limbus in the oblique quadrant (superotemporal for access to the superior rectus [SR] and inferotemporal for access to the inferior rectus [IR]). B, Suture placement parallel to spiral of Tillaux. The original muscle insertion of the IR shown here (open arrowheads) serves as an important landmark. The nasal border of the transposed inferior rectus is sutured to the lateral edge of the IR insertional stump. Black arrow points to spatulated needle at the point of attachment. Illustration continued on following page

Successful results have been reported, but few long-term results are available. This may reflect the rarity of double elevator palsy.

**INVERSE KNAPP PROCEDURE**

The principles of the Knapp procedure were applied to congenital absence of the IR and have become known in common parlance as the inverse Knapp procedure. This surgery has also been effective in cases of double depressor palsy or underaction as well as of absent IR function due to a surgically lost IR or IR palsy after orbital floor fractures.

In contrast to double elevator palsy, secondary SR restriction is uncommon. The procedure can correct 15 to 25 PD of hyperdeviation in primary gaze and as much as 30 to 35 PD in downgaze. Binocular diplopia-free fields are improved.

In both the Knapp and inverse Knapp procedures, the horizontal rectus muscle may be transposed to straddle the SR or IR insertion, placed following the spiral of Tillaux, or sutured parallel to the long axis of the vertical rectus muscle.

**ALTERNATIVE PROCEDURES**

Callahan described a rectus muscle union procedure for elevator palsy resembling the Jensen procedure for LR palsy. Any of the transposition procedures may be performed by transposing or uniting the horizontal rectus muscles to the involved vertical rectus muscles. The Knapp (or inverse Knapp) procedure may also be combined with horizontal recession and resection surgery. If a muscle is recessed, the recession is measured from the SR (or IR) insertion.

Foster reports good results from posterior fixation suture augmentation for larger hypodeviations seen in double elevator palsy. We have successfully performed a full tendon transposition of the horizontal rectus muscles, combined with botulinum injection of the antagonist SR, in a patient with IR injury who had complete IR palsy. Surgical SR recession was not possible in this case because of the risk of anterior segment ischemia. The transposed horizontal rectus muscles may also be placed on adjustable sutures, as in the procedure described for LR palsy.

**Procedures for Adduction Deficit**

Complete third nerve palsy is surgically challenging. Enhancing the motility of four of the six extraocular muscles is required, yet only the SO and the LR muscles are functioning. The LR muscle is too anatomically distant from the MR muscle to take part in a transposition procedure for adduction deficit. Both surgeon and patient need to be aware that the main goal of surgery is to achieve cosmetic alignment in primary gaze. A useful area of binocular fusion is usually not achievable. Improved motility is not probable. Persistent postoperative diploia may be relieved by using an occluder contact lens.

In partial third nerve palsy, there are more surgical options. If the vertical rectus muscles are functional, they may be transposed medially to augment adduction. Resection of both transposed vertical rectus muscles is needed to enhance adduction by decreasing the slack created by transposing the muscles medially. Foster reports that posterior fixation augmentation is also beneficial and may eliminate the need for an antagonist weakening procedure. A sclera-augmented muscle-tendon transfer, using a strip of banked sclera to transfer the medial halves of the vertical rectus muscles to the insertion of the MR, has been described. The procedure is combined with recession of the antagonist LR.

In combined horizontal and vertical deviations due to partial third nerve palsy, assuming some residual function of the SR or IR is present, recession and/or resection of these muscles may be performed with the transposition. The mus-
Figure 36-6 Continued. C, left. The posterior (lateral) augmentation is placed 7 to 8 mm posterior to the muscle insertion and (C, right) incorporates approximately 25% of the muscle mass. Notice the 45-degree orientation of the IR after transposition following the spiral of Tillaux. D, The fixation suture is then anchored to sclera 7 to 8 mm posterior to and just below the lateral rectus (LR) insertion. E, Upon conclusion of the augmentation, the IR (left) and SR (right) orientation changes and parallels the long axis of the LR. The gap between the transposed IR and SR is markedly reduced.
Figure 36-7. Results after posterior suture augmentation of vertical rectus transposition in right sixth nerve palsy. A, Preoperative photographs of patient with right sixth nerve palsy fixing with the paretic eye. Large esotropia in primary gaze increases on right gaze and improves on left gaze. No abduction could be seen in the right eye. B, After vertical rectus muscle transposition to the lateral rectus with lateral augmentation suture was performed (and after botulinum toxin injection of the medial rectus), patient is now orthotropic in primary gaze with improved abduction of the right eye noted on right gaze. Only a mild restriction to adduction of the right eye is evident, as well as a mild reduction in downward rotation. Centration and expansion of the binocular diplopia-free field was achieved.
Knapp Procedure with Variations of Tendon Placement

Figure 36-8. Knapp procedure with variations of tendon placement. A, Classic Knapp procedure with the horizontal rectus muscles straddling the superior rectus insertion. B, Transposition following the spiral of Tillaux. C, Transposition parallel to the long axis of the superior rectus.

cles are recessed or resected and placed parallel to the spiral of Tillaux. In large exodeviations and hypodeviation with residual MR function, a large supermaximal recession-resection procedure has been recommended but this commonly leaves residual hypodeviation. Both horizontal muscles could be offset superiorly or inferiorly for vertical deviations up to 10 PD. For larger vertical deviations, a vertical recession and resection with ciliary vessel sparing may be required.

When a significant segment of the MR muscle is destroyed after trauma (in one of our cases, part of the MR was extirpated during endoscopic sinus surgery), the preferred procedure is vertical rectus muscle transposition. However, if for some reason the transposition surgery is contraindicated or cannot be completed, we have described a procedure in which a piece of temporalis fascia is spliced to the proximal segment of the MR to act as a medial “stay suture,” aligning the eyes in primary gaze. Access to the proximal segment of the MR is gained through a medial orbitotomy (Fig. 36–9).

SUPERIOR OBLIQUE TRANSPOSITION

Historical Perspective

In complete third nerve palsy, the only muscle that can provide adducting force is the SO. A large LR muscle recession is required for the procedure to be effective. Some authors advocate combining the procedure with MR resection to additionally restrict abduction.

Using the SO as an internal rotator in third nerve paralysis was first described in 1933. Fracture of the trochlea was often necessary to disengage the SO tendon from the trochlea before the transposition (Fig. 36–10A). The procedure was technically difficult, especially in older patients with a calcified trochlea, and risked severing the SO tendon. An alternative procedure was described to avoid problems with the trochlea. The SO tendon may be moved 2 mm anterior and nasal to the SR insertion without trochlear fracture (Fig. 36–10B).

By shortening (resecting) the SO and transposing the tendon to the MR insertion, the SO functions of depression, abduction, and incyclotorsion are converted to mild adduction. The tonic adducting force from the SO is used to prevent recurrent exotropia resulting from preserved LR tone. In addition, the SO tendon acts as a leash to restrict abduction. The SO tendon will supply 62% of the adduction vector in an eye with complete third nerve palsy.

Surgical Technique

Before surgery, forcedduction testing is done to confirm suspected LR contracture. If surgery is to be combined with

Superior Oblique Transposition


LR recession as recommended, that procedure is performed first.

For SO transposition, a traction suture (e.g., Vicryl 6-0) is placed in the superonasal limbus to assist in rotating the globe inferotemporally. A conjunctival incision is made in the superonasal fornix 8 to 10 mm from the limbus and extended 6 mm circumferentially.

The SR muscle is isolated on a muscle hook and cleaned of the intermuscular septum on its nasal border, where it is intimately attached to the SO tendon. The SO tendon can now be directly visualized (Fig. 36–11A). Surrounding intermuscular septum and fascia are cleared from the SO tendon. A nonabsorbable suture (e.g., Novafil 6-0) is woven through the width of the tendon at the level of the nasal border of the SR with interlocking knots at both ends. The SO tendon is tenotomized, leaving it attached to its functional origin (the trochlea) and the nonabsorbable suture (Fig. 36–11B).

The amount of SO tendon to be resected (usually 8 to 10 mm) is measured by caliper. Nonabsorbable sutures are placed 0.5 to 1 mm behind the intended area of resection. The shortened tendon is transposed superonasally as shown in Figure 36–10B. Temporary slip knots are used. Forcedduction testing then is repeated. The goal is to create a leash that will restrict abduction. Alignment under anesthesia should aim for orthotropia to a small-angle esotropia. The nonabsorbable sutures are tied permanently.

Results

Less adduction was documented after pure anteriorization of the SO tendon without trochlear fracture than after SO transposition with trochleotomy. In addition, inadequate horizontal alignment, hypertropia, and paradoxical ocular movements have been observed. However, good alignment in primary gaze has been documented. Often large LR recessions or, occasionally, horizontal recession-resection were needed to improve the results. SO transposition does not increase the range of movement. At best, only 10 degrees of binocular diplopia-free field may be achieved.

ALTERNATIVE PROCEDURE

Orbital Periosteal Flap Globe Stabilization

In collaboration with Robert Goldberg, M.D., we have developed a procedure to fixate and stabilize the globe by creating a periosteal flap. The flap is harvested from the orbital wall and then attached to the globe adjacent to the rectus muscle insertion desired (Fig. 36–12). If harvested from the medial orbital wall, lateral rotation can be prevented and the globe aligned in the primary position. When appropriate, flaps may also be dissected from the superior, medial, or inferior orbital walls. We have found this more effective and technically easier than SO transposition with fewer complications. We have used this procedure successfully in patients with complete third nerve palsy and orbital fibrosis syndrome (Fig. 36–13).

Common Pitfalls and Complications

FAILURE TO TEST FOR MUSCLE FUNCTION

If the supposedly weakened muscle, for example, the LR, is not truly palsied, a resection procedure is most effective.
Figure 36-12. Periosteal flap for static globe fixation. A, Using a conjunctival approach, a 1-cm wide medial periosteal flap is created by incising the periosteum at the posterior lacrimal crest and dissecting it off the medial orbital wall. The base of the flap is toward the orbital apex. B, Cut axial view showing the periosteal flap tunneled from the extraconal to the intraconal space, and attached to the medial rectus muscle insertion to stabilize and fixate the globe against lateral rotation. (Ant., anterior; a., artery; m., muscle.)

Figure 36-13. A, Patient with right third nerve palsy showing only lateral rectus abduction. Large exotropia seen in primary gaze. Patient can fuse in extreme right gaze (left face turn shown in left bottom frame). B, After orbital periosteal flap procedure, the patient is aligned in primary position. The flap creates a medial restriction to lateral rotation preventing recurrent exotropia.
Vertical rectus muscle transposition may lead to consecutive exodeviations because of the strong muscle force that is transposed in concert with the functioning LR.

Similarly, if the transposed muscles are palsied, as in involvement of the third cranial nerve the procedure will be ineffective. The muscles proposed for transposition must have at least some residual function for the surgery to be effective. The same is true for SO transposition—the trochlear nerve and SO muscle should be functioning. Relying entirely on observed ocular rotations is insufficient. These situations stress the need for thorough preoperative evaluation of muscle function. When the clinical findings are equivocal, laboratory confirmation of muscle function may be helpful.

**UNDERCORRECTIONS**

Undercorrections are more common than overcorrections. The many modifications of transposition techniques attest to the inadequacy of these procedures. Undercorrections are commonly observed after vertical rectus transposition to the MR insertion and supermaximal horizontal recession-resection procedure in cases of third cranial nerve palsy.

In complete third nerve palsies, supermaximal recession of the LR (as much as 17–20 mm) should be done to totally abolish LR rotation. This reduces the chance that residual LR function may ultimately lead to recurrent exotropia. Unopposed LR muscle force may result in LR contracture and act as an antagonist to the transposed muscles medially.

In vertical rectus muscle transposition to the MR insertion, a resection procedure of both vertical rectus muscles enhances the outcome. This is not necessary in vertical rectus transposition procedures performed to the LR, because the temporal movement of the vertical rectus muscles actually puts the vertical muscles on stretch (Fig. 36–14). Weakening of the ipsilateral antagonist is commonly required and, if overlooked, leads to undercorrections. As an alternative, matching the incomitance with a contralateral fadenopera
tion may improve binocular diplopia-free fields.

**LATE OVERCORRECTIONS**

Late overcorrections commonly present as consecutive exotropia after a transposition procedure for LR palsy. There are various reasons for this. Despite waiting a considerable period of time (usually 6 months) before operating, LR function may rarely return. Excessive antagonist recession (e.g., MR recession with transposition) leads to underaction of this muscle, favoring lateral abducting forces. Late slippage of the MR muscle should be recognized (see also Chapter 40). These problems can be prevented by complying with the recommended waiting period, testing muscle function adequately, waiting for stable alignment, and adhering to proper surgical technique.

The same phenomenon is observed after the Knapp procedure for double elevator palsy. With long-term follow-up, the effect of the procedure increases over time. Undercorrections are expected to improve, whereas overcorrections are expected to increase.

**INDUCED VERTICAL DEVIATIONS**

Vertical deviations follow vertical rectus muscle transposition to the LR combined with botulinum toxin in 20% to 32% of patients. Both surgical transposition and chemodenervation can cause a vertical deviation. With an adjustable transposition technique, the surgeon is able to eliminate the induced deviation a day after surgery. Complications after chemodenervation are discussed in Chapter 32.

**ANTERIOR SEGMENT ISCHEMIA**

Anterior segment ischemia has been reported after several variations of the transposition procedure in both children and adults. Pharmacologic denervation may have reduced the incidence, but anterior segment ischemia has been described in a high-risk individual. In adults, vasculopathic and hematopoietic diseases increase this risk. In children, risk factors include congenitally...
absent rectus muscles, severe nutritional deficiency, cicatricial retinopathy of prematurity, and optic nerve glioma. Fortunately, many cases of anterior segment ischemia resolve over time and have very little effect on vision. The risk is estimated at 1 in 13,300 operations. Avoiding surgery on the third muscle in a nonseeing or amблиopic eye may not be necessary. Choosing to perform surgery on the better eye may actually carry a higher risk of ocular damage.

Chemodenervation may be used as an alternative to surgical weakening of the ipsilateral antagonist. In Möbius syndrome, aside from bilateral involvement of the abducens nerve with deficient abduction and facial palsy, patients have gaze palsy and asymmetric fibrosis resembling congenital fibrosis. Because of the combined paretic and restrictive strabismus, transposition surgery may be problematic if restrictions are enhanced.

Some propose performing a vertical rectus transposition to the palsied LR bilaterally, because large MR resections will further weaken the adduction commonly associated with gaze palsy. Although a large MR recession and, secondarily, a vertical rectus transposition procedure can be done in stages, overcorrection is almost certain. We now recommend performing vertical rectus transposition as a first procedure. MR recession is reserved as a secondary surgery for residual esotropia (see also Chapter 26). The management of fibrosis syndromes of the extraocular muscles is discussed in Chapter 27.

REFERENCES

SELECTED TRANSPOSITION PROCEDURES


Historical Perspective

In the early 1970s, Cüppers described a new procedure for strabismus surgery that he called the fadenoperation. The operation involves suturing a rectus muscle to the sclera at some point behind the equator to create a new “functional” insertion (Fig. 37–1). This weakens the muscle in its primary field of action without appreciably altering alignment of the eyes in other gaze directions. This effect is distinctly different from standard rectus muscle procedures (i.e., recession/resection) that alter the basic relationship between the agonist and antagonist muscles, thereby changing the alignment of the eyes in all gaze positions. The fadenoperation may be combined with other procedures to further enhance their effect in a particular gaze position. This ability to change ocular alignment in only one direction of gaze is particularly advantageous in cases of complicated strabismus secondary to paresis, restriction, or innervational anomalies.

Terminology

Faden is the German word for thread or suture; the term fadenoperation refers to using a suture to attach muscle to sclera. Because sutures are also used in most other types of strabismus surgery, the term was believed by some authors to be vague and nondescriptive, especially since it had been used in the older German literature to describe traction sutures. Von Noorden has suggested two alternative terms: posterior fixation suture and retroequatorial myopexy. The latter term has not gained wide acceptance despite being the most accurate description of the procedure as it is performed today. Fadenoperation, with all its linguistic problems, remains the most popular term. In this chapter, posterior fixation suture will be used in reference to the suture, while the term fadenoperation will refer to the surgical procedure.

Mechanisms of Action

The effect of the posterior fixation suture on ocular alignment results from three physiologic effects.

First, the amount of ocular rotation that a rectus muscle can accomplish depends on a variety of factors. A major determinant is the point of attachment of the muscle to the eye (see Fig. 37–1). Theoretically, maximum rotation occurs when the rotational force vector of the muscle becomes perpendicular to the surface of the globe. At this point, the angle of the “lever arm” (see Fig. 37–1A, dotted line) becomes zero, and further tension on the muscle provides no additional rotating force. In reality, this point is never reached because existing structures limit the amount of actual rotation. The posterior fixation suture decreases maximum ocular rotation by firmly attaching the muscle to the eye at a point posterior to its normal insertion (see Fig. 37–1B, point A), creating a new reference point (see Fig. 37–1B, point A’) for the muscle’s rotating action. This does not change the normal resting position of the eye because muscle length and tension remain the same. As the muscle begins to contract, additional force is needed to rotate the eye because the lever arm is reduced. Because the “functional insertion” has moved posteriorly, the point of maximum rotation occurs much earlier (see Fig. 37–1B). The net result is that the muscle becomes limited in its field of action without changing the primary or other gaze positions. The muscle’s relationship with its antagonist remains unaltered. The farther behind the original insertion the muscle is sutured to the globe, the greater the force needed to rotate the eye and the more limited the movement.

Second, increased innervation is necessary to rotate an eye after placement of a posterior fixation suture. If the suture is placed on a muscle of the fixing eye, increased effort will be required to move that eye. This will increase innervation to the corresponding yoke muscle of the fellow eye (Hering’s law). Because in most circumstances this is the weaker muscle, the increased innervation will help reduce...
misalignment in the desired direction. For example, in a left sixth nerve palsy, a posterior fixation suture may be placed on the right medial rectus (MR) to reduce lateral incomitance on left gaze. Because more innervation is now needed to rotate the right eye to the left, additional innervation will also go to the left lateral rectus (LR), thereby increasing its rotational force and abducting power (Fig. 37–2). In addition, Sherrington’s law of reciprocal innervation results in inhibition of the contralateral antagonist (the left MR), further assisting the paretic left LR. In patients who have undergone a transposition procedure, further improvement in abduction may be noted after a contralateral MR fadenopera
tion because of this inhibitory effect on the antagonist MR.

Third, after placement of a posterior fixation suture, the muscle fibers between the insertion and the scleral fixation suture (those from A to A’ in Fig. 37–1B) can no longer contract effectively. This results in an overall reduction in generated force and rotational ability, because part of the muscle has been rendered inactive.

Indications

The posterior fixation suture may be used to treat any ocular motility disorder that presents as asymmetric ocular rotations, excessive muscle action, or functionally abnormal head position (Table 37–1). For the posterior fixation suture procedure to be successful, the operated muscle must have reasonably good function and the contralateral yoke must have some function. Eyes that have complete muscle paralysis, severe restrictions, or excessively recessed muscles are not good candidates for this procedure.

INCOMITANT STRABISMUS

Because the primary benefit of the posterior fixation suture procedure is selective limitation of ocular rotation, the most useful application is to correct incomitant strabismus. This is especially true if primary-position alignment is satisfactory and change is required in only one direction of gaze. Unfortunately, in most circumstances this means operating on the “normal” eye to improve binocular vision and expand the binocular diplopia-free fields by creating matchingduction defects in both eyes. In some patients this may be advantageous, because it avoids the increased risks incurred with strabismus surgery in eyes with other ocular disease.

Vertical Incomitance (Inferior Rectus Paresis, Blow-Out Fractures, Thyroid Myopathy, After Scleral Buckling Surgery)

Patients with vertical diplopia on downgaze are particularly symptomatic because of the many functions performed in this position. If misalignment in the primary position is minimal, a posterior fixation suture on the better-depressing eye may improve or eliminate the disparity in downgaze. For example, patients who have sustained inferior rectus (IR) injury after a blow-out fracture are often orthotropic in the primary position but have both elevation and depression deficits. Standard recession-resection vertical muscle surgery is usually ineffective and worsens the duction defect. Weak-
Sixth Nerve Palsy
Posterior Fixation Suture

**Figure 37-2.** Effect of a posterior fixation suture on the contralateral medial rectus in a patient with a sixth nerve palsy. Before the operation, the nonparetic fixing eye rotates better laterally than the paretic eye. After placement of a posterior fixation suture, the nonparetic eye is limited and requires more innervation to rotate the eye. The additional innervation is also given to the paretic eye, thereby increasing its rotation. (Hering's law of equal innervation to yoke muscles.) The net result is that the eyes are more coordinated on lateral gaze.

Opening the contralateral IR may be quite effective in restoring downgaze function (Fig. 37–3). Similarly, patients who have restricted vertical ocular rotations after scleral buckling procedures or orbital scarring from previous surgery may be improved by avoiding manipulation of the offending eye and creating a matching “weakness” of the contralateral yoke muscle. The fadenoperation may also be used to eliminate or preventiatrogenic incomitant vertical strabismus after maximum contralateral IR muscle recession. Patients with restrictive thyroid myopathy often have tight, fibrotic muscles that function poorly after recessions. Postoperatively, these patients often have a downgaze deficit. Placing a posterior fixation suture on the normal IR muscle can balance the rotational forces and eliminate diplopia. Occasionally, the posterior fixation suture is placed on the contralateral IR in anticipation that a downgaze defect will occur postoperatively. This is especially helpful when performing an adjustable-suture IR muscle recession. Often the surgeon is faced with the dilemma of how much adjustable recession to perform to return the eye to the primary position while not weakening the muscle so much that downgaze is affected. It is much easier to decide where to leave the recessed muscle when the relatively normal contralateral IR is restricted by a posterior fixation suture at the time of the adjustment.

**Table 37–1.** Indications for Fadenoperation

<table>
<thead>
<tr>
<th>Incomitant strabismus</th>
<th>Inferior rectus paresis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sixth nerve palsy</td>
<td></td>
</tr>
<tr>
<td>Third nerve palsy</td>
<td></td>
</tr>
<tr>
<td>Restrictive strabismus</td>
<td></td>
</tr>
<tr>
<td>Thyroid myopathy</td>
<td></td>
</tr>
<tr>
<td>After scleral buckling surgery</td>
<td></td>
</tr>
<tr>
<td>Blow-out fracture</td>
<td></td>
</tr>
<tr>
<td>Reoperations</td>
<td></td>
</tr>
<tr>
<td>Internuclear ophthalmoplegia</td>
<td></td>
</tr>
<tr>
<td>Duane syndrome</td>
<td></td>
</tr>
<tr>
<td>Double-elevator palsy (monocular elevation deficiency)</td>
<td></td>
</tr>
<tr>
<td>Esotropia</td>
<td></td>
</tr>
<tr>
<td>High accommodative convergence/accommodation esotropia</td>
<td></td>
</tr>
<tr>
<td>Convergence excess</td>
<td></td>
</tr>
<tr>
<td>Nystagmus blockage syndrome</td>
<td></td>
</tr>
<tr>
<td>Sensory with cross fixation</td>
<td></td>
</tr>
<tr>
<td>Dissociated vertical deviation</td>
<td></td>
</tr>
<tr>
<td>Nystagmus (head position)</td>
<td></td>
</tr>
</tbody>
</table>

**Horizontal Incomitance (Sixth Nerve Palsy, Third Nerve Palsy, Blow-Out Fractures, Internuclear Ophthalmoplegia, After Strabismus Surgery, After Scleral Buckling Surgery)**

As with vertical deviations, the posterior fixation suture can be used with mild to moderate horizontal incomitance. The best example is a patient with sixth nerve palsy who has had an LR muscle–strengthening procedure and is improved in the primary position but still has diplopia on lateral gaze. A posterior fixation suture on the contralateral MR muscle, with or without a recession, can expand the window of single binocular vision without creating exodeviation on opposite gaze (Fig. 37–4). This same procedure may be done on patients who have a medial wall blow-out fracture and limited abduction from a restricted MR muscle.
Figure 37-3. A, A patient sustained a blow-out fracture of the right orbit. Preoperatively (left upper/lower), diplopia was noted only on downgaze. A posterior fixation suture was placed on the inferior rectus of the left eye. Postoperatively (right upper/lower), there was no change in the primary position and elimination of diplopia on downgaze. Note improved duction of the right inferior rectus due to Hering’s Law. B, Diplopia-field of patient in A. Preoperatively (top), diplopia (DIPL) was noted in downgaze with single binocular vision (SBV) elsewhere. Postoperatively (bottom), diplopia was eliminated until 40 degrees on downgaze. C, Posterior fixation suture (white solid arrows) on the inferior rectus muscle (IR). Note vortex vein (open arrow) just posterior to the suture. It usually prevents a more posterior placement.
Figure 37-4. A, Patient with left sixth nerve palsy after a transposition operation on the left eye. Note the head position necessary to achieve comfortable single vision. Right gaze (B) and left gaze (C). A posterior fixation suture was placed on the right medial rectus. D, Improvement in head position after surgery. E, Right gaze is unaffected. F, Left gaze is improved.
Medial-wall exploration usually does not relieve the restriction, and recessing the MR muscle only creates an exodeviation in the muscle's field of action. A posterior fixation suture on the contralateral MR improves lateral gaze without sacrificing normal function in other gaze positions (Fig. 37–5).

The LR muscle is less amenable to the posterior fixation suture technique. The long arc of contact this muscle makes with the globe necessitates placing the suture farther back from its insertion to achieve an effect, increasing the technical difficulty. In addition, suture placement is more hazardous because the macula and inferior oblique muscle insertion are located near the operative site.

**ESOTROPIA (HIGH ACCOMMODATIVE CONVERGENCE/ACCOMMODATION, CONVERGENCE EXCESS, SENSORY ESOTROPIA)**

The surgical management of esotropia that is significantly greater at near than at distance (high accommodative convergence/accommodation ratio) presents a therapeutic challenge. The fadenoperation, alone or combined with bilateral MR muscle recessions, has been used successfully for this purpose.\(^1,10,13,21,24\) Von Noorden\(^24\) performed a fadenoperation on 21 patients who previously had undergone maximal rectus recessions but still had esotropia greater at near than distance. He found the mean distance change to be 9 PD and the mean near change, 14 PD. Kushner\(^12\) compared the efficacy of bilateral MR recessions plus a fadenoperation with augmented bilateral MR recessions and found both approaches to be equally effective in reducing the near-distance disparity. Peterseim and Buckley\(^14\) used the fadenoperation alone in patients with little or no deviation at distance and a large esotropia at near and achieved a mean 25-PD reduction in near esotropia. The procedure had a small effect (4 PD) on the distance alignment.

Some patients will develop esotropia owing to loss of vision in one eye, especially if the loss occurs during infancy. This deviation is often quite variable, which makes surgical correction using recession-resection procedures difficult. Elsas\(^9\) has advocated using a posterior fixation suture on the poorly seeing eye to improve ocular alignment while reducing the risk of an unwanted overcorrection.

**NYSTAGMUS (NYSTAGMUS BLOCKAGE SYNDROME, HEAD POSITION)**

Certain forms of congenital nystagmus can be dampened, and visual function improved, by convergence efforts (nystagmus blockage syndrome). These patients become esotropic and fix with their eyes in adduction. Attempts by the patient to move the eyes to the primary or abducted position results in a jerky nystagmus and reduced visual performance. The esotropia is often intermittent and is manifest only when performing tasks that require good visual function. Bilateral MR posterior fixation sutures have produced good ocular alignment and improved visual function in many of these patients.\(^19,22\) Care must be exercised in combining MR recessions with the posterior fixation suture in this disorder, because overcorrection may occur. In a review of 12 patients having combined procedures, 7 (58%) were within 10 PD postoperatively, 1 remained esotropic, and 4 (33%) were converted to an exodeviation.\(^10\)

Unilateral MR fadenoperation may be helpful in patients having poor vision in one eye that results in manifest latent nystagmus in the good eye with a head turn. This type of jerk nystagmus is dampened by fixation in adduction because it obeys Alexander’s law (i.e., a jerk nystagmus dampens on gaze away from the direction of the fast component). Because the better-seeing eye is controlling the head position, an MR posterior fixation suture will limit the ocular rotation and thereby improve the head position.\(^9\)

Patients with congenital or acquired nystagmus often have a preferred gaze position (null position) that improves visual function. If the resultant head position is functionally unacceptable, surgery to limit movement of the eyes into that position is considered. The standard approach employs an Anderson-Kestenbaum-type procedure to shift the eyes away from the preferred gaze position. This may be enhanced or replaced by posterior fixation sutures on the yoke muscles used in that gaze position. For example, if right gaze is

---

**Figure 37–5.** A. Patient with a left medial wall blow-out fracture. Note restriction to abduction of the left eye. B. A posterior fixation suture was placed on the right medial rectus creating a matching duction defect and improving diplopia on left gaze.
preferred, left MR and right LR posterior fixation sutures could be used. Posterior fixation sutures are also useful for improving visual function in acquired vertical nystagmus with oscillopsia (such as downbeat nystagmus), to improve vertical head positions (i.e., chin tuck, chin elevation) and reduce the amplitude of pendular nystagmus (when placed on all four horizontal muscles).

**DISASSOCIATED VERTICAL DEVIATION**

The *faden* operation, alone or combined with a superior rectus (SR) recession, has been advocated as a treatment for dissociated vertical deviation (DVD).\textsuperscript{10, 20, 22} Because the position of the involved eye is often normal when the vertical deviation is not present, standard rectus muscle recessions, especially if unilateral, run the risk of creating a hypotropia or large secondary deviation. The posterior fixation suture can be used to “enhance” a smaller recession in an attempt to avoid this problem. Significant improvement has been reported in 60% to 95% of patients.\textsuperscript{10, 20, 22}

Achieving a significant reduction in large DVDs is difficult using the *faden* operation alone. The posterior fixation suture must be placed at least 20 mm, and preferably 23 to 25 mm from the limbus, which often is technically troublesome. A more effective initial procedure is a recession of the SR on a “hangback” suture or (in those cases with associated inferior oblique overaction) anterior placement of the inferior oblique. The *faden* operation can be very effective for small deviations (less than 10 mm), asymmetric deviations (either as an adjunct or as the sole procedure), or as a second procedure if a residual defect persists or if surgery on the IR or further recession of the SR is contraindicated. The *faden* operation does avoid lid fissure abnormalities, which may occur after larger vertical rectus muscle surgery.

**Contraindications**

Because the posterior fixation suture technique usually requires surgical manipulation of the “better” moving eye, it is important to select patients carefully. The muscle that is to be sutured to the globe must have reasonably good function. If it is already fibrotic, paretic, or otherwise compromised, the surgical result can be very unpredictable, and complete inhibition of function often results (see Fig. 37–5). Excessive scarring or multiple previous surgeries at the same site may also contribute to a poor outcome.

The yoke muscle must have some function to “match” a duction weakness. If there is a complete lack of movement secondary to paresis or restriction, it usually is not possible or desirable to create severe loss of function in the “normal” muscle.

Extensive dissection of tissue around the muscle is needed to gain exposure and properly place the fixation suture. This increases fibrosis and inflammation, making reoperation more difficult and success less predictable. Careful assessment of possible further benefit is in order before additional strabismus surgery is considered (especially a second posterior fixation suture) on a muscle that already has a posterior fixation suture in place.

### Surgical Technique

**TECHNICAL CONSIDERATIONS**

**Needle and Suture Material**

Using the proper needle and suture material can simplify the procedure. The site of suture placement is usually 15 to 20 mm posterior to the limbus (Table 37–2). Even with maximum exposure, the space is shallow and the horizontal distance available to make the scleral pass and retrieve the needle is limited. A short, small, shallow-curved, spatulated needle is highly desirable (Fig. 37–6). Nonabsorbable suture material is advisable to prevent late failure from poor muscle and sclera adhesion. Also, the suture is easier to locate in the event that posterior fixation needs to be released.

**Exposure**

To gain access to the optimum location for placing the posterior fixation suture, retraction of the overlying Tenon's capsule and other orbital tissues is necessary. A Desmarres, small ribbon, or Fison retractor is used (Fig. 37–7). The Fison retractor has several advantages for this procedure. Its blade is narrow at the base, allowing excellent elevation of the tissue and creating an A-shaped opening, contrasting to the more square-shaped opening made by the other retractors. The result is a higher opening, a real advantage when trying to pass a suture in a deep hole. The flat and deep blade helps to lift tissue over the suture site. For SR surgery, removing the lid speculum will also increase exposure.

The orbital tissues at this location can be floppy and tend to billow into and obscure the surgical field. This is especially true when extensive manipulation has taken place. Exposure can be enhanced by first placing a slightly stiffer material into the space and then retracting. Neurosurgical sponge (\(\frac{1}{4}\) inch) trimmed to an appropriate length is very useful for this purpose (Fig. 37–8). Exposure is also increased by retracting the globe as far as possible away from

![Figure 37–6. Spatulated needle (Alcon D-5) recommended for performing a fadenoperation. The shallow curve and short cord length make it ideal for this procedure.](image-url)
Figure 37-7. Fison (left) and Desmarres (right) lid retractors. The Fison retractor has a longer blade with a wide end that assists in providing exposure for the placement of a posterior fixation suture.

Figure 37-8. Surgical view of medial rectus (MR) posterior fixation suture. Note neurosurgical sponge (open arrow) holding back orbital tissue. Suture is placed approximately 18 mm from limbus (white solid arrow).

The muscle can be isolated using either a limbal or fornix-based incision (Fig. 37–9). Once it has been isolated, a lid retractor (Desmarres or Fison) is inserted into the conjunctival or Tenon’s opening to adequately expose the proposed posterior fixation site. Extensive dissection is required to clean check ligaments and other attachments off the muscle. A small neurosurgical sponge (¼ inch wide) may be placed under the retractor to hold back tissue and facilitate adequate exposure. A small muscle hook and/or forceps is used to lift the edge of the rectus muscle. The posterior fixation suture

(e.g., Ethicon 5-0 Dacron, D-5 needle) is attached to the sclera with the needle point emerging beneath the elevated muscle (see Fig. 37–9A). The needle is passed through the muscle at this site by relaxing tension on the muscle hook or forceps. About 25% of the muscle is incorporated in the suture, and the suture is tied firmly to attach the muscle edge to the sclera. This process is repeated on the other side. Care is needed not to incorporate the anterior ciliary arteries in the suture if possible. The site is inspected and checked for a firm attachment by passing a small muscle hook under the muscle at the suture location (see Fig. 37–9B).

**Posterior Fixation Suture With Double Side Stitch and Rectus Muscle Recession**

A retractor (Desmarres or Fison) is placed beneath the muscle surface to expose bare sclera (Fig. 37–10). A small neurosurgical sponge (¼ inch wide) may be placed under the retractor to facilitate exposure. Any remaining episcleral attachments are cleaned from the proposed posterior fixation site. The original insertion is grasped with forceps and, using a single-armed 5-0 Dacron suture, a tunnel is made for approximately 2 mm into the sclera at the desired posterior...
fixation point (see Fig. 37–10A). The suture should be placed at the point where the border of the rectus muscle will be located after the recession. A second suture should be placed on the opposite side. The needle is passed through the sclera so that the needle end of the suture is closest to the center of the rectus muscle. The suture ends are looped out of the way, and the muscle recession is continued.

The retractor is removed and the rectus muscle is re-attached at the proposed recession site (see Fig. 37–10B). Once the rectus muscle has been reattached, the retractor is reinserted to expose the posterior fixation site and locate the needle part of the posterior fixation sutures. With a small muscle hook or forceps, the edge of the rectus muscle is lifted and the previously placed posterior fixation suture needle is passed through the muscle at the point where the muscle crosses the scleral attachment site (see Fig. 37–10C). One should try to incorporate 2 to 3 mm of the muscle edge. The knot is tied firmly to secure the muscle to sclera. The process is then repeated on the other side.

**Posterior Fixation Suture with a Single Central Stitch and Rectus Muscle Recession**

After the muscle is disinserted, a lid retractor (Desmarres or Fison) is placed beneath the muscle to expose the sclera.
Any remaining episcleral attachments and tissue are cleaned off at the proposed posterior fixation suture site (see Fig. 37–11A, distance “a” from original insertion). The insertion is grasped with forceps and a double-armed 5-0 Dacron suture is attached to the sclera, tunneling for 2 to 3 mm (see Fig. 37–11A). The suture should be underneath the central part of the recessed rectus muscle.

The lid retractor is removed to free up the rectus muscle and reposition it to hold the remaining tissue out of the surgical field. Determining where the posterior fixation suture will attach to the rectus muscle is done by subtracting the proposed amount of recession (distance “b”) from the distance between where the fixation suture was attached to sclera and the original insertion (distance “a”). This will yield the location of the posterior fixation suture (a – b = c). The distance “c” is measured from the end of the severed rectus muscle, and both ends of the 5-0 Dacron suture are passed from the underside of the rectus muscle to the top side in the center of the muscle, separating the two stitches by approximately 3 mm (see Fig. 37–11B). One should tighten but not tie the suture permanently at this time.

The lid retractor is removed and the rectus muscle is reattached at the proposed recession site (distance “b”). Once the rectus muscle has been reattached, the posterior fixation suture is completed by permanently tying it over the rectus muscle (see Fig. 37–11C). The knot should be firmly tied to ensure that an adhesion forms between the muscle and sclera. The recession distance “b” and the posterior fixation distance “a” are normally measured from the original insertion (see Fig. 37–11D).

Adjustable Fadenoperation (Sutureless Fadenoperation)

One drawback of the standard posterior fixation procedure is that it cannot be combined with the adjustable suture technique, which allows postoperative manipulation of the muscle location and optimizes the chance of a successful outcome. A modification has been proposed that theoretically achieves this goal (Fig. 37–12).18

The fadenoperation limits ocular rotation by changing the “functional” insertion and preventing part of the muscle from contracting (see section on mechanism of action). If a piece of the muscle is removed and the muscle reattached where that end would have normally contacted the globe in the primary position, the result is identical to placing a posterior fixation suture at the same location. The amount of muscle resected corresponds to the distance behind the insertion that the posterior fixation suture would have been placed. The muscle then is recessed to this location on a “hangback” suture. Postoperatively, after assessing ocular alignment, this position may be advanced or further recessed. This technique is useful in situations when suturing the muscle to the globe is hazardous because of inadequate exposure or other anatomic constraints.

Common Pitfalls

As indicated earlier, the operation can be facilitated by good exposure, adequate lighting, and proper needle selection. There are several points that deserve special mention. A posterior fixation suture on the SR muscle must be placed at least 20 mm from the limbus to have a significant effect. Unfortunately the superior oblique tendon runs beneath the SR muscle and can be misdirected if it ends up behind the posterior fixation suture. This may be avoided by first placing the suture through the sclera and then through the superior oblique tendon where it would normally cross the suture site (Fig. 37–13). This maintains a normal superior oblique tendon configuration and direction.

In many cases the fadenoperation is combined with a muscle recession procedure. One should avoid performing larger than normal muscle recessions in this situation. The
Figure 37-13. Surgical view of a superior rectus (SR) fadenoperation. A, SR muscle is disinserted. B, Double-armed 5-0 Dacron suture is placed at the posterior fixation site. Note that the SR and superior oblique (SO) are retracted out of the surgical area. C, Needle in the sclera. D, SO tendon is allowed to assume normal position. E, Suture is placed through SO tendon. F, SR is reattached.
posterior fixation suture will enhance the recession’s effect, so that a smaller than normal amount is needed to achieve the desired change in ocular alignment. Excessive muscle recession combined with placement of a posterior fixation suture can result in a severe duction defect (Fig. 37–14). This is especially important in patients who have had previous strabismus or other types of extraocular surgery (e.g., scleral buckle, orbital decompression) in the planned surgical site.

Esotropic patients who are having MR muscle surgery as well as a posterior fixation suture are also at risk for overcorrection. Because of its shorter arc of contact with the globe compared with the other rectus muscles, the MR can easily be overweakened by the fadenoperation. The posterior fixation suture should not be placed farther than 20 mm from the limbus on the MR muscle (see Table 37–2).

Alternative Procedures
MULTIPLE MUSCLE RECESSIONS—ONE EYE

To achieve a similar effect on ocular motility without using a posterior fixation suture, surgery on more than one muscle is usually required. The major advantage of the fadenoperation is its ability to limit ocular rotation in one direction without altering the muscle’s relationship with its antagonist in other directions. The only other way to weaken a muscle is to recess it, which shifts the balance of forces in the direction of the antagonist. A simultaneous recession of the antagonist muscle is required to retain the previous relationship. Therefore, a recession of both the muscle to be weakened and its antagonist muscle can substitute for a posterior fixation suture.

MULTIPLE MUSCLE RECESSIONS/RESECTIONS—BOTH EYES

Another way to achieve the same effect as a posterior fixation suture is to recess and/or resect multiple muscles in both eyes. For example, in a patient with a left sixth nerve palsy, a posterior fixation suture could be placed on the right MR muscle to match the left LR duction defect (see Fig. 37–2). An alternative would be to recess the left MR muscle or resect the paralytic left LR. This results in an exodeviation in right gaze (where there was no deviation), which could
be improved by recessing the right LR. That, however, would strengthen the right MR (the yoke to the paralytic left lateral), which in turn would have to be recessed to avoid worsening the esodeviation on left gaze. Thus, a recession of the antagonist to the paralytic muscle plus bilateral recessions of the horizontal muscles in the contralateral eye can have an effect similar to that of a posterior fixation suture on the contralateral yoke muscle.

### LARGE MUSCLE RECESSIONS

Larger than normal rectus muscle recessions may be used in lieu of a posterior fixation suture in patients with high accommodative convergence/accommodation esotropia and DVD. Bilateral augmented MR recessions have been shown to be as effective as the fadenoperation in reducing near–distance disparity in esotropic patients. By performing an MR recession in an amount calculated to correct the larger near deviation, the distance–near disparity can be eliminated. Overcorrections at distance do not seem to occur. This method is less useful in patients who have only a near deviation. Likewise, in patients with DVD, bilateral large SR muscle “hangback” recessions are comparable to the fadenoperation in reducing the tendency for the eyes to drift vertically. Use of the hangback technique avoids the hazards of trying to place a scleral suture deep in the superior fornix.

### Complications

#### UNDERCORRECTION AND/OR OVERCORRECTION

The most common postoperative complication is undercorrection secondary to inadequate posterior suture placement, loose sutures, or anterior migration of sutures. The use of absorbable sutures can result in a late loss of effect if the muscle fails to adhere firmly to the globe before the suture dissolves.

If the suture is placed too far posteriorly, a severe duction deficit may occur. The risk of significantly limited duction is also increased by previous surgery, especially if there is scarring in the posterior Tenon’s area (see Fig. 37–14). The MR is the easiest muscle in which to limit ocular rotation because of the short arc of contact that it makes with the globe. Placing the suture farther than 20 mm from the limbus will cripple the MR muscle. The risk is further increased if a simultaneous recession is also performed. Care must be exercised when using an MR posterior fixation suture in patients with preexisting convergence insufficiency, because this condition may be worsened.

### SCLERAL PUNCTURE

Puncturing the sclera with the suture needle while attempting to attach the rectus muscle to the eye may result from a needle passed deep into the sclera, inadequate exposure, failure to grasp the sclera firmly with forceps, an extremely thin sclera, and a host of other unforeseen circumstances. Algo reported on the fundus findings in a large series of patients who received posterior fixation sutures and noted a scleral perforation rate of 15%. No serious complications occurred, and many authors have found even lower perforation rates. Proper technique, adequate exposure, and use of a small, flat needle can minimize this risk.

### COMPLICATED REOPERATIONS

The placement of a posterior fixation suture causes a firm adhesion to form between the sclera and muscle at the site of the suture. The muscle tissue anterior to the fixation suture tends to atrophy and become a thin band of fibrous tissue. This can make reoperating on this muscle more difficult, because the muscle anterior to the suture tends to lose its function over time. If reoperation is contemplated, it should be performed within 4 to 6 weeks after the original surgery.

When undoing a fadenoperation, the surgeon needs to keep in mind the changes that may have taken place in front of the posterior fixation suture. The longer the interval, the more likely that strong scleral adhesions will have formed. The limbal approach is preferred, because the “blind” muscle hooking necessary when using the fornix approach may result in disorientation. Knowledge of the location of the muscle insertion (i.e., has it been recessed?) is very helpful in guiding the procedure.

The initial approach is to carefully dissect conjunctiva and Tenon’s tissue off the sclera to the level of the muscle insertion. With the use of a small muscle hook (superior oblique tenotomy hook), the edge of the muscle usually can be identified. More often than not, it is impossible to pass a larger hook under the muscle because of adhesions. Occasionally, the only portion of the muscle that can be hooked is that posterior to the fixation suture. It is important to recognize when this occurs in order to proceed safely. The small hook can be used as a guide to further isolate the muscle. It may have to be switched to the other border to facilitate dissection. The fixation suture(s) should be identified and removed. If the hook was passed under the muscle posterior to the fixation suture, the anterior portion may need to be dissected from the sclera. Placing a suture in the muscle (which could be used to reattach the muscle to the eye) before any further dissection will avoid a potential lost muscle if inadvertent disinsertion subsequently occurs.

Regardless of where the muscle was originally hooked, adhesions from the fixation suture to the insertion can usually be freed using Westcott scissors. Most of the time it is not possible to free the muscle all the way to its actual insertion, and this needs to be taken into account. Where a fadenoperation is being “undone” because of a duction lag, advancement and even resection may be necessary. The functional insertion was at the site of the fixation suture, not the insertion itself. Removing the suture will have no effect on the primary deviation and only a small effect on the

<table>
<thead>
<tr>
<th>Muscle</th>
<th>Amount (mm)</th>
<th>Special Concerns</th>
</tr>
</thead>
<tbody>
<tr>
<td>Medial</td>
<td>15–20</td>
<td>Easiest to overcorrect</td>
</tr>
<tr>
<td>Inferior</td>
<td>15–18</td>
<td>Convergence insufficiency</td>
</tr>
<tr>
<td>Lateral</td>
<td>18–24</td>
<td>Vortex veins (see Fig. 37–3C)</td>
</tr>
<tr>
<td>Superior</td>
<td>20–24</td>
<td>Medial oblique insertion</td>
</tr>
</tbody>
</table>

**Table 37-2. Posterior Fixation Suture Amounts**
duction lag. When more correction is necessary, it should be made as if the muscle were attached to the most anterior site of muscle adhesion released. This is usually anterior to the fixation suture and posterior to the “real” insertion. Care is needed not to over-recess in this situation.

OTHER COMPLICATIONS

Other unusual and rare complications include choroidal ischemia, persistent mydriasis, optic atrophy, macular edema, vitreous hemorrhage, and choroidal detachments.

REFERENCES

Reoperation may be expected in 5% to 10% of patients who undergo strabismus surgery, despite the most meticulous efforts to perform a careful preoperative evaluation and determine the appropriate surgical strategy.\(^7\) \(^6\) Reoperations occur more often in cases of complicated strabismus such as superior oblique (SO) palsy, sixth nerve palsy, Duane syndrome, Möbius syndrome, and Brown syndrome.\(^7\) Furthermore, any reoperation introduces a 33% probability of yet another procedure that may be needed to achieve satisfactory results.\(^7\) With persistence, however, 80% to 90% of patients can ultimately expect satisfactory results.\(^7\)

The principles for any strabismus reoperation are the same as in traditional surgery.\(^4\) However, mechanical and muscle tone imbalance may be greater than expected from the usual strabismus operations. The surgeon must be more attentive to anatomy, innervation, and mechanical imbalance. Cooper’s\(^5\) dictum that “patients should be treated as a new case on the basis of current findings” should be interpreted with caution because of anatomic alterations.\(^7\)

Preoperative Evaluation

HISTORY

As stressed in the first chapter of this textbook, history taking forms an integral part of the strabismus evaluation. Obviously, one needs to elicit any history of childhood strabismus. Dissociated vertical deviation, for instance, may be expected after infantile esotropia regardless of the timing of surgery. Is there an area of suppression scotoma that may affect treatment goals and the choice of surgical strategy? When was strabismus first documented? How long did the patient’s eyes remain straight after surgery? How many previous surgeries took place? What was the postoperative course? The surgical goals will be based on a thorough understanding of the patient’s strabismus history.

Previous operative notes detailing the amount of surgery performed in a recession or resection aids the ophthalmologist in the evaluation. The findings from these reports are correlated with an objective assessment of ocular rotations. In addition, the surgeon will be able to determine preoperatively whether the former procedure involved a limbal approach, with or without concurrent conjunctival recession, or a fornix approach.

Potential risk factors for anterior segment ischemia should be identified, especially in the elderly. These include vasculopathies such as diabetes, hypertension, and atherosclerosis; hematopoietic diseases; and cerebrovascular accidents. Any associated neurologic disease such as myasthenia gravis can masquerade as almost any form of strabismus.

CLINICAL ASSESSMENT

Evaluation of Ocular Rotations

Ocular rotations are examined by both duction and version movements (see Chapter 1 for details). If mechanical restriction or muscle weakness is suspected, tests of muscle function—forced duction testing, force generation, and saccadic velocity analysis—should be performed (see Chapter 3). The results will help the surgeon choose the appropriate surgical strategy.

An apparent underaction may in reality be due to a secondarily contracted antagonist muscle. This should be suggested by forced duction and force generation testing. If a muscle is tight, a recession procedure of this muscle is usually required to improve ocular rotations and minimize incomitance. Resection of the antagonist will limit full rotation, worsen the incomitance, and restrict binocular fields. If a muscle is truly underacting, surgery that further weakens this muscle should be avoided. The possibility of a slipped muscle should also be suspected. Instead of resecting an antagonist, one should consider an advancement procedure.
or a weakening operation on the contralateral yoke such as a fadenoperation.

**Anterior Segment Evaluation**

If previous operative reports are not available, the location of conjunctival scars is noted. For horizontal strabismus, the surgeon should note whether the plica is drawn up into the medial bulbar scar. A search should be made for signs of fat prolapse, and subtle signs of anterior segment ischemia identified such as focal iris atrophy, an irregularly shaped pupil, or low-grade anterior segment inflammation.

**LABORATORY EVALUATION**

Although rarely required, laboratory studies of muscle function may be indicated if lost and/or slipped muscles are suspected and in certain cases of muscle paresis or palsy. In high-risk situations, anterior segment imaging studies may be required. The results may lead one to change the surgical strategy. For example, an 81-year-old man with esotropia, right hypertropia, and left excyclotorsion had undergone prior strabismus surgeries. They included medial rectus (MR) recession of both eyes with inferior rectus (IR) recession of the left eye and a Harada-Itô procedure of the left SO. Previous surgery in the right eye involved only the MR. Imaging studies were obtained because of the patient’s age and the preoperative plan of right lateral rectus (LR) and superior rectus (SR) recession (that would have meant surgery on three rectus muscles for the right eye) and left SR transposition for torsion. Normally, the entire iris circulation fills by 30 seconds. The photograph made at 52 seconds showed delayed filling and persistence of temporal and inferior filling defects (Fig. 38–1). The surgeon abandoned the preoperative plan of recessing the SR in the right eye to preserve any anterior ciliary vessels supplied by this muscle. The hyperdeviation was addressed by re-recessing the con-

**Common Problems**

There will be occasions when the surgeon will not have the benefit of knowing the details of prior strabismus procedures. The ophthalmologist can ask about what type of strabismus was present before surgery. Old photographs may help the patient or a parent remember. From this information, one can make an educated guess as to what surgical procedures may have been performed and correlate this with the scars observed on the conjunctiva. In this setting, one may need to explore at least one rectus muscle to develop a good surgical plan.

Most reoperations are best performed no earlier than 2 months after the initial surgery to permit complete healing and resolution of postoperative edema and also to allow more stable and reliable measurements of the deviation. Immediate reoperation is rarely indicated except in the following situations:

1. Significantly limited rotations in the field of an operated muscle with marked slowing of saccades. This suggests the possibility of a lost muscle.
2. Large overcorrections after an SO tuck. A tuck is best relieved on the first postoperative day. After this time, adhesions and healing impede successful reversal of the procedure.
3. Large vertical deviations induced by muscle transposition procedures. These are most easily corrected by repositioning the transposed muscle on the first or second postoperative day.

In the immediate postoperative period, it is technically easier to go back within the first few days; there are fewer adhesions, and neovascular proliferation is minimal. Surgical planes can still be clearly identified. Often the surgeon can use the same incision to gain access to the operative field. The muscle suture can still be easily identified and retrieved in cases of lost or slipped muscles. Postoperative edema and neovascular proliferation are maximal 2 to 4 weeks after surgery.

Mechanical restrictions to ocular rotation are primarily found at three anatomic sites:

1. The conjunctiva may be foreshortened after long-standing deviations or inappropriate conjunctival closure from prior surgical procedures (Fig. 38–2). The latter is commonly observed after retinal detachment repair in which the conjunctiva is reattached to the limbus despite the addition of a bulky exoplant. In these cases, a conjunctival recession may prevent the development of restrictions.
2. Muscle restriction follows secondary contracture or excessive resections.
3. Scar tissue formation may lead to a “surgical leash” that restricts ocular rotation.

Weakened muscle force is observed after excessive recessions, late slippage of a rectus muscle, and lost muscles. If
recession has been maximal and is confirmed by limited ocular rotations, an advancement procedure may be required. Slipped and lost muscles are discussed in Chapter 40. We have observed muscles that dehisce at the mid-belly level. Management of this problem can be a technical challenge (see Chapter 41).

Common Reoperations

Common reoperation scenarios include undercorrection, overcorrection, slipped or lost muscles, restrictions after a prior ophthalmic surgical procedure, and the development of a new strabismus problem. Undercorrection occurs in recurrent esotropia or exotropia, and overcorrection occurs in consecutive or secondary exotropia or esotropia. A similar situation can occur after vertical strabismus surgery. Restrictions usually develop after excessive resections or scar tissue formation. The latter may follow intraconal fat violation or previous ophthalmologic operations such as retinal detachment repair, implantation of a glaucoma device, repair of orbital trauma, or excision of a pterygium.

The development of new problems, such as dissociated vertical deviation in infantile esotropia or the unmasking of a vertical deviation after a horizontal strabismus surgical procedure, may be treated as new cases because the muscles that will be involved in a subsequent surgery act in a different plane and have not previously been operated upon. The reader is referred to the appropriate chapter discussing the management of these problems.

HORIZONTAL STRABISMUS

Adults may be happier if an existing strabismus is undercorrected rather than overcorrected. Patients at greater risk of overcorrection and subsequent reoperation after horizontal strabismus surgeries are those with high hyperopia (more than +5.0 D), A-pattern strabismus, and amblyopia. The nuances of decision-making and surgical strategy in some of these common reoperative situations are discussed in this section.

Recurrent or Residual Esotropia

Medial and lateral rotations are carefully assessed. If the MR has limited adduction, the surgeon must be wary of further weakening this muscle (Fig. 38–3). The MR is an unforgiving muscle because of the relatively short arc of contact between the muscle and the globe. This is especially true in very young children and hyperopes with small globes. There is more leeway in myopes because of the longer axial length. A previous LR resection may also cause limited adduction, which may be confirmed by forced duction testing.

Distance and near deviation measurements can assist one in arriving at the appropriate surgical strategy. If the distance deviation is more than the near deviation, the MR should not be recessed further. This is an indirect sign that maximal MR recession has already been performed. If the near deviation exceeds the distance deviation, the MR may be recessed...
further. An additional 2- to 3-mm recession of the MR when its insertion is already close to the equator can result in a larger correction than if this was the initial procedure.

**Secondary Exotropia**

Small overcorrections can be treated with minus lenses immediately postoperatively. Up to 25% of cases may be salvaged with this technique and reoperation avoided.

If both MR muscles have been recessed previously, careful assessment of adduction is imperative. If it is markedly limited, a slipped MR should be considered (Fig. 38–4). This suspicion is confirmed by forced duction testing at the time of surgery or saccadic velocity analysis preoperatively. If only mildly limited adduction is observed, the previous MR recession may already be maximal and the secondary deviation due to and/or aggravated by secondary contracture of the LR muscle. If the exotropia at distance is greater than the near deviation, one should consider LR recession (Fig. 38–5).

If the previous procedure was a horizontal recession-resection, the careful evaluation of any limitation of adduction is required. Excessive LR resection and a slipped MR will present as secondary exotropia. Forced duction testing and saccadic velocity analysis will confirm the diagnosis.

In either situation, MR advancement is always a consideration. If the MR slipped, it is essential to excise the pseudodendon and advance the muscle. The slipped MR is usually tight, owing to secondary contracture, and the proper amount to advance is difficult to determine. We usually advance to a point where restriction to forced duction in abduction is noted. Temporary slip knots are used so that the forced duction can be performed and the position of the insertion changed if required. If the MR muscle has not slipped, the effect of an advancement is very unpredictable, and it usually does not suffice to fully correct the esodeviation after over-corrected esotropia.

**Recurrent or Residual Exotropia**

This is probably the most common reoperative scenario. Prisms and minus lenses may be tried for small-angle deviations.

If a previous LR recession has been performed, careful evaluation of abduction rotation and measurements in lateral gaze are indicated. If abduction is full, more LR recession may be considered. If abduction is decreased and/or marked lateral incomitance is noted, further LR recession may cause an esotropia in right and left gaze. MR resection should be considered in this situation. If a true or pseudodivergence excess pattern of exotropia is observed, it is best to re-recess the LR rather than resect both MR muscles. If a convergence insufficiency pattern is noted, bilateral MR resection is beneficial. If horizontal recession-resection surgery has been performed, more predictable results can be obtained if the residual exotropia is corrected by surgery on the unoperated eye.

**Secondary Esotropia**

If the child is younger than age 5 years, base-out prisms should be used if the esotropia persists longer than 2 weeks to reestablish bifoveal fixation and avoid the development of monofixational esotropia with amblyopia. If the patient is hyperopic, full correction or the maximum tolerated hyperopic spectacles should be prescribed.

If the previous procedure involved LR recession in both eyes, abduction should be carefully evaluated. If it is significantly limited or asymmetric, a slipped LR should be considered (Fig. 38–6). Although less frequent than a slipped MR, the LR muscle can slip and cause secondary esodeviation. If abduction is not markedly limited, distance and near measurements will help determine the appropriate surgical strategy. If the near deviation is more than the distance measurements, both MR muscles may be resected. If the distance is more than near deviation, one should consider advancing both LR muscles.

If the prior procedure was a horizontal recession-resection,

![Figure 38-4. Secondary exotropia after left medial rectus recession. Note the exodeviation in primary gaze that increases on right gaze. The left medial rectus can only move minimally past the midline. The reoperation strategy should consider a slipped medial rectus muscle and should be managed accordingly.](image-url)
Figure 38-5. Secondary exotropia after bilateral medial rectus muscle recession. Note the mild limitation of adduction in both eyes. Good saccadic velocities were generated in adduction so that slipped medial rectus muscles were unlikely. Both medial rectus muscles have been recessed maximally. The patient was managed with bilateral lateral rectus recession.

Weak abduction may be due to a restriction caused by MR resection (Fig. 38–7). This may be confirmed by forcedduction testing, saccadic velocity analysis, and cover test measurements showing lateral incomitance. If this is the case, unilateral or bilateral MR recession is the preferred procedure, depending on the size of the esodeviation. If, however, the esodeviation is less than 10 PD at near, it is best to advance the LR in the operated eye, adding a resection of the LR in the unoperated eye if the distance deviation is large.

VERTICAL STRABISMUS

Previous operative notes are even more important in reoperations for vertical strabismus. These are generally more difficult reoperations, and the surgical scars may not reveal which of the cyclovertical muscles were operated on previously. Torsion measurements are crucial. This may be a factor inhibiting fusion, as opposed to the more obvious vertical deviation. Comitance of the deviation and the evaluation of vertical rotations are essential.

Many different scenarios can occur in reoperations for vertical strabismus. It is beyond the scope of this chapter to discuss each specific case. Our comments are limited to general principles and some of the more common clinical situations.

Forcedduction and force generation testing provide essential and critical information for developing a surgical plan. If there is significant mechanical restriction to vertical rotation, this probably needs to be relieved if the globe cannot be rotated more than 10 to 15 degrees in a specific field of gaze. If the eye can rotate at least 25 degrees into a particular gaze field, the surgeon may elect to ignore the restriction.

If surgical overcorrection has occurred and the IR has

Figure 38-6. Incomitant esotropia due to slipped left lateral rectus muscle after a scleral buckling procedure. Note the profound limitation of abduction and widening of palpebral fissure on abduction of the left eye.
been recessed, the surgeon should carefully evaluate infra-
duction and lid retraction in the eye where the IR has been
recessed. Even with residual infuduction, lid retraction can
occur; one should consider a slipped IR (a common occur-
rence) or excessive recession. The IR muscle is also an
unforgiving muscle because of its short arc of contact.

If surgical overcorrection is the problem and the IR has
been resected, the surgeon should carefully evaluate upward
globe rotation. If this is limited, a tight IR due to excessive
resection is the explanation. This IR muscle needs to be
recessed to relieve the restriction and address the overcorrec-
tion.

Incomitance is evaluated by cover testing in upgaze and
downgaze. If a residual deviation that is small in downgaze
is much larger in upgaze, surgery should be directed at the
muscles responsible for upward rotation—the SR and the
inferior oblique. The IR and SO should probably not be
involved in the reoperation. The reverse is true if the devia-
tion is larger in downgaze and minimal in upgaze. In second-
ary contracture of a vertical muscle (e.g., the SR in thyroid
ophthalmopathy and long-standing dissociated vertical devi-
ation), the deviation may appear greater in downgaze. The
diagnosis is confirmed by forced duction testing. The tight
rectus muscle needs to be recessed. Otherwise, the decision
to operate on the oblique or rectus muscle depends on
comitance or incomitance in horizontal gaze fields as well
as torsion measurements.

In residual cyclo deviations, vertical rectus transposition
may be considered if a Harada-Ito procedure for excyclotor-
sion has led to undercorrection.27 For incyclotorision, either
tenectomy of the anterior torsion fibers of the SO at its
insertion or vertical rectus transposition should be consid-
ered. To correct excyclotorision, the SR is transposed tempo-
 rally or the IR is transposed nasally. For incyclotorision, the
SR is transposed nasally and the IR is transposed tempo-
rally.27

**Surgical Approach**

Strabismus reoperations aim to achieve stable alignment,
full ocular rotations with no incomitance, and good cosme-
sis with the fewest surgical procedures.14 Favorable cosmesis
usually requires a white, noninflamed eye with normal, sym-
metric lid fissures. The latter is especially relevant in exces-
sive IR recession, which may cause widened lid fissures,
and excessive resections which cause narrowing.

The importance of intraoperative forced duction testing
cannot be overemphasized. It confirms the surgeon’s preop-
erative diagnosis and demonstrates whether surgery has ef-
effectively relieved restrictions. A characteristic string or in-
dentation sign14, 21, 22 is observed when the conjunctiva
contributes to a restriction (see Fig. 38–2). Forced duction
testing needs to be repeated several times during the surgical
procedure to determine whether a restriction has been ade-
quately relieved.

The use of traction sutures is important for orientation
and to improve exposure. Traction sutures are placed at the
6- and 12-o’clock positions for horizontal rectus surgery and
at the 3- and 9-o’clock positions for vertical muscle surgery.

**CONJUNCTIVAL OPENING**

The conjunctiva may contribute to problems associated
with strabismus reoperation by causing restriction and a poor
cosmetic outcome. The conjunctiva may be tight and restrict
rotation in the opposite field (leash effect). If associated with
scar tissue and adhesions, it may limit ocular rotations in the
same field (reverse leash).15 Poor cosmesis is observed when
the conjunctiva is scarred, has an irregular external surface,
or appears thickened. The latter commonly occurs medi ally
with fat incarceration or when the plica is pulled laterally.

The best way to adequately evaluate and explore compli-
cated strabismus problems is with a large surgical field. For
this reason, a limbal incision—which provides the widest
exposure—is preferred. One should free a large apron of
conjunctiva to create a large operative field. The conjunctiva
is usually scarred down at the area near the original muscle
insertion and may have to be freed with sharp dissection.
Radial incisions are performed in the oblique axes. If de-
sired, marking sutures may be placed on the anterior edge of
the conjunctiva.
THE MUSCLE

In isolating the muscle, small spreading movements are made with blunt-tipped Westcott scissors to create a buttonhole in Tenon’s fascia (Fig. 38–8). Once a portion of sclera is exposed, the scissors are applied to bare sclera, starting from a distance greater than the expected edge of the muscle and tunneling under the muscle as far back as necessary to get completely around it. In some situations, this maneuver will need to be done without visual control, keeping the scissors tip tangential to the globe at all times. The muscle should not be approached from the external aspect of the scar tissue. Dissection in layers until the muscle is exposed is difficult in reoperations because tissue planes are difficult to identify. It is safer to identify the true muscle by approaching from a lateral and posterior direction.

The muscle hook should glide gently behind the belly of the muscle. Once the path beneath the muscle is cleared with blunt-tipped scissors, the muscle hook is passed between the blades of the same scissors, which remain beneath the extraocular muscle to serve as a guide. The surgery proceeds slowly until the entire muscle is hooked.

The Kowal squint hook, with 10 × 2-mm blades and a 0.5-mm thickness, is much thinner than standard muscle hooks and is easier to insert beneath a tight muscle (Kowal L, personal communication, 1997). In addition, it has a central 1-mm groove between its blades, allowing the surgeon to preplace muscle sutures or perform a tenotomy while protecting the globe (Fig. 38–9). After the whole width of the muscle has been identified, control is aided by applying gentle traction with the hook.

The intermuscular septum is then severed as far back as necessary to isolate the muscle. In inferior oblique reoperations, injury to the vortex vein and the neurovascular bundle must be avoided to prevent iatrogenic mydriasis and bleeding. In SO reoperations, the muscle plan may be difficult to identify. The dissection should begin at the undersurface of the SR where the SO and SR are intimately attached.

If the muscle still does not slide freely, posterior footplates may be present and require excision. With the blades of the scissors closed, the surgeon can identify the area of the muscle where the SO tendon is scarred to the nasal border of the SR insertion.

In any reoperated muscle, careful attention should be paid to the insertional edge of the muscle once it is detached. A cicatricial expansion of 1 to 1.5 mm between the muscle and the sclera is common. This scar tissue is recognized by its more anterior insertion on the globe than the actual muscle insertion (identified while the muscle hook is applied). This tissue should be excised to ensure maximal function of the muscle postoperatively (Fig. 38–10). To better expose this tissue, the muscle hook is rotated 180 degrees away from the limbus. With fine scissors and forceps, small snips are taken until the muscle is reached to avoid accidentally disinserting the muscle. If sutures are inadvertently placed in this tissue, the muscle will not function optimally after surgery. This will cause undercorrection despite an apparently appropriate surgical strategy.

If the muscle still does not slide freely, posterior footplates may be present and require excision. With the blades of the scissors closed, the surgeon can identify the area of the
posterior adhesions by following the curve of the globe in the muscle's path. Once the area of restriction has been identified, the blades are opened slightly and small snips made until the restriction is freed.

**SCAR TISSUE**

Excess scar tissue is excised from the surface of the muscle until the muscle can be clearly identified. Some scar tissue on the top surface of the muscle may be left without any adverse effect on alignment or cosmesis. However, the undersurface of the muscle should be clean enough to allow its free movement and reattachment to the globe. For the same reason, adhesions and scar tissue in the muscle's path also need to be excised. Scar tissue in the exposed area of the globe should be removed to achieve a "white eye" effect. Bumpy red scar tissue that is left exposed will be cosmetically objectionable. Bare white sclera can epithelialize well, giving the appearance of a white conjunctiva. One must remain wary of ill-defined surgical planes. The scleral lamella, for instance, may be mistaken for scar tissue. The surgeon must not be overzealous in freeing all restrictions. In some cases, not all restrictions can be relieved. Scar tissue should be excised judiciously, removing just enough to achieve good cosmesis and function. Posterior restrictions cannot be relieved by standard strabismus techniques without endangering other vital structures such as the optic nerve, ciliary arteries, and nerves.

In repeat resections, regardless of the surgeon's preference in unoperated cases, the amount of recession should be measured from the limbus. In adults, it takes 12 to 15 mm of LR recession from the limbus to create a -1.5 limitation in abduction and approximately 11 to 12 mm recession of the MR from the limbus to cause the same limitation in adduction. Some surgeons would rather perform a Z-myotomy procedure, but novice surgeons have difficulty in quantifying the postoperative effect. Reoperating on a thinned-out muscle after Z-myotomy can lead to an increased risk of lost muscles.

In advancement procedures, the insertion site may be identified from prior operative notes or measured from the limbus to the spiral of Tillaux based on normal tables. A previously recessed muscle takes up its own slack, producing time-related shortening. Changes in eye muscle sarcomeres based on eye position have been described. In addition, suture placement in the first recession procedure, as well as in the reoperation, may have a small resection effect. All these factors contribute to the inability to advance a previously recessed muscle to its original insertion without causing a restriction. It is helpful to release the traction...
suture and rotate the globe toward the muscle being advanced to relieve tension. Temporary sutures should be placed and the forced duction test performed to avoid iatrogenic restriction and postoperative incomitance.

CONJUNCTIVAL CLOSURE

Appropriate management of the conjunctiva contributes significantly to both the success and stability of postoperative alignment after reoperation. If the conjunctiva contributes to recurrent strabismus and mechanical restriction, simple closure will not suffice in most cases.

Often the conjunctiva needs to be recessed to avoid future restrictions. One must be sure that the conjunctiva is attached securely to sclera by taking firm episcleral bites to prevent anterior creep of the tissue. T-closure has also been advocated to relieve restrictions. In some cases, excision of a scarred conjunctiva with large bare scleral closure is an excellent alternative. Unfortunately, this technique is used infrequently. Too often, conjunctival scarring leads to a recurrence of the original strabismus problem. Large areas of conjunctiva may be completely excised from limbus to fornix. Rarely, bare muscle may even be left exposed, especially superiorly and inferiorly where the lids can act as a cover. Thickened conjunctiva may be thinned by excising superficial fibrous tissues and/or deep scarring. If the medial plica has been pulled laterally, plicaplasty may be performed. If necessary, conjunctival autografts may be placed.

For severely obliterated conjunctiva, fornix reconstruction may be performed with the assistance of an ophthalmic plastic and reconstructive surgeon, using buccal mucosa grafting and plastic or Teflon implants (as used in the management of the anophthalmic socket). Forced duction testing should be done after conjunctival closure or reconstruction to ensure that mechanical restriction has been relieved and not recreated.

IMMEDIATE POSTOPERATIVE MANAGEMENT

If adjustable sutures are used, postoperative drugs such as meperidine (Demerol) and scopolamine, which affect muscle tone, should be avoided. We prefer to adjust 24 hours after surgery to ensure maximal alertness. Permanent adhesions have not been a problem. Postoperative edema and chemosis are minimized by sub-Tenon's injection of corticosteroid. Prolonged recuperation after surgery is expected and may entail the extended use of topical corticosteroid.

Conclusions

Despite the best preoperative evaluation and surgical technique, reoperations will continue to be a part of strabismus management. This is not surprising considering that mechanical manipulations are used for what may in reality be a complex interplay between cortical dysfunction, neuromuscular function, and sensory systems. Mild overcorrections and undercorrections may be treated nonsurgically by means of prisms and orthoptics and by manipulating spectacle correction (such as the use of overminus lenses for exotropia). In cases that warrant reoperation, successful alignment and functional results can be achieved.

REFERENCES

ANTERIOR CILIARY VESSEL SPARING PROCEDURE

CRAIG A. McKEOWN, MD

Historical Perspective

Anterior segment ischemia (ASI) is a rare but potentially serious complication of strabismus surgery. Considerable effort has been devoted to understanding the mechanisms responsible for the development of ASI and preventing its occurrence.

ASI was initially recognized as a consequence of systemic disease in individuals who had not undergone surgery. Schmidt is credited with the first clinical description of anterior segment changes caused by ischemia in 1874. Most of the disorders causing ASI directly or indirectly affect the circulation, particularly to the head, orbit, or eye. Anterior segment ischemic changes have been described in association with aortic arch syndrome, carotid artery obstruction, ophthalmic artery obstruction, carotid-cavernous fistula, sickle cell disease, herpes zoster ophthalmicus, rubella, acute angle closure glaucoma, high myopia, irradiation, chronic forms of leukemia, and a variety of other disorders.

The first descriptions of ASI after ocular surgery were in experimental animals. In 1941, Leinfelder and Black reported anterior segment changes in nonhuman primates undergoing strabismus surgery, although the authors did not recognize that the changes were caused by ASI. In 1954, Chamberlain described irregular, dilated pupils and iritis after rectus muscle transposition in monkeys.

ASI was first described as a complication of strabismus surgery in humans in 1955, in a 76-year old having retinal detachment repair that included disinserting the lateral rectus. Later reports cited a variety of circumstances in which ASI developed in patients undergoing ocular procedures, such as retinal detachment repair, cyclocryotherapy, and laser photocoagulation. Certain coexisting systemic disorders, such as systemic lupus erythematosus and the SS and SC hemoglobinopathies, increase the risk of ASI. In addition, there is a single report of ASI and neovascular glaucoma after implantation of a posterior chamber intraocular lens, with deep erosion of a Prolene loop into the ciliary body.

ASI was first reported as a complication of strabismus surgery in humans by Stucchi and Bianchi in 1957. Initial descriptions involved surgery on three or four rectus muscles, whereas later reports described the unusual occurrence of ASI in patients having surgery on as few as two rectus muscles. Most were adults with one or more concurrent problems, including chronic lymphocytic leukemia, thyroid-related orbitopathy, hypertension, and other disorders. However, ASI was also noted in a 4-year-old child with cicatricial retinopathy of prematurity who underwent a horizontal rectus muscle recession and resection.

Anatomy and Physiology

ANTERIOR CILIARY VESSEL ANATOMY

Leber’s classic description of the vascular anatomy of the eye in 1903 has served as the foundation of teaching ophthalmic anatomy for more than 90 years. He described two anterior ciliary arteries on each of the four rectus muscles except the lateral rectus, which had only one. Later reports cited a variety of circumstances in which ASI developed in patients undergoing ocular procedures, such as retinal detachment repair, cyclocryotherapy, and laser photocoagulation. Certain coexisting systemic disorders, such as systemic lupus erythematosus and the SS and SC hemoglobinopathies, increase the risk of ASI. In addition, there is a single report of ASI and neovascular glaucoma after implantation of a posterior chamber intraocular lens, with deep erosion of a Prolene loop into the ciliary body.
the check ligaments and intermuscular septum have been severed (Fig. 39–1).

After they emerge from within the rectus muscles, the anterior ciliary vessels travel along the surface of the muscle and tendon, serving as a conduit for blood flow between the deeper orbital region and the globe. In their journey on the surface of the muscle and tendon, the vessels may travel alone or may be bundled together in groups of two, three, or even four vessels. This is referred to as an anterior ciliary vessel group (Figs. 39–1 and 39–2). The vessels follow a somewhat serpentine course, creating considerable slack or redundancy in the overall length of the vascular system. Relatively few branch vessels leave the anterior ciliary vessels overlying the tendon, which may reflect the relatively low metabolic demands of tendon tissue. On occasion, anomalous vessels may be found exiting directly from Tenon’s capsule overlying the muscle. These vessels then enter the muscle capsule and course anteriorly with the other anterior ciliary vessels.

Although the anterior ciliary vessels are quite delicate, they are relatively sturdy when compared with the surrounding connective tissue of the muscle capsule. As the vessels reach the tendon insertion they continue to travel a short distance on the surface of the sclera before becoming firmly attached to, or penetrating, the sclera (Fig. 39–3; see also Fig. 39–1). The surface location of the vessels as well as their envelopment by relatively loose connective tissue provide a potential plane of surgical dissection between the vessels and the underlying muscle, tendon, and sclera. In addition, the redundancy in vessel length (slack) and tensile strength of the anterior ciliary vessels provide the anatomic characteristics needed to successfully dissect and preserve them during rectus muscle surgery.99–103

**ANTERIOR SEGMENT BLOOD FLOW**

The arterial blood supply to the anterior segment of the eye originates from the ophthalmic artery and is carried to the eye almost exclusively by the anterior ciliary arteries and the long posterior ciliary arteries. The seven or so anterior ciliary arteries follow the course of the four rectus muscles, whereas the two long posterior ciliary arteries pursue an intrascleral course and are located medially and temporally, beneath the medial and lateral rectus muscles.104 The anterior ciliary vessels are disrupted by conventional full-tendon strabismus procedures. The two long posterior ciliary vessels are in an intrascleral position where they may be damaged during strabismus surgery involving the horizontal rectus muscles as well as other procedures, particularly retinal detachment repair.

The anterior ciliary arteries and long posterior ciliary arteries contribute to several collateral circulatory systems including the episcleral limbal plexus, the intramuscular circulation within the ciliary body, and the major arterial circle in the iris root.24, 66, 91, 93, 104 This robust, multichannel blood supply to the anterior segment probably has much to do with the relative rarity of ASI after strabismus surgery.
Anterior segment vascular anatomy and blood flow have been investigated in unoperated as well as operated animals and humans using a variety of techniques. They include postmortem injection studies as well as in vivo fluorescein angiography with either conventional photography or low-dose video techniques that provide a large number of viewing frames per second.8, 20, 33–35, 45, 48, 54, 63–66, 69, 71, 93, 96 The information provided in these reports has occasionally been contradictory, probably because of fundamental differences in the techniques used as well as varying age, species, and living or nonliving status at the time of the study.

A majority of anterior segment blood flow is normally supplied by the anterior ciliary vessels. Occlusion of the long posterior ciliary arteries alone usually does not produce ischemic changes. Under normal circumstances the long posterior ciliary arteries are thought to contribute roughly 30% of blood flow to the anterior segment of the eye.33, 82, 91, 94 Several reports have commented on the relative importance of the medial versus the lateral long posterior ciliary artery in patients having horizontal rectus surgery combined with vertical rectus surgery; the results occasionally conflict.34, 69, 73, 82, 94

The blood supply to the iris appears to be sectoral, and iris blood flow may serve as an important indicator of the status of the anterior ciliary vessels in each quadrant. Iris fluorescein angiography has proved to be particularly useful for this purpose, although the heavy pigmentation of brown irides requires special modifications.8, 33–35, 42, 54, 56, 63, 64, 66, 68, 69, 71, 73, 91, 93 Recently, Rosenbaum (personal communication, 1998) has employed indocyanine green videoangiography with promising results.

Abnormalities detectable by iris angiography after full-width tenotomy of rectus muscles include a delay or absence of iris-vessel filling in the quadrant corresponding to the tenotomized muscle as well as late leakage of contrast material. Normally, the iris vessels begin to fill 12 to 20 seconds after the intravenous injection of 10% to 20% sodium fluorescein. Complete vessel filling, from the time of the first appearance of fluorescein in the iris to the completion of filling at the pupillary border, usually takes less than 10 seconds, although there is rather wide variation between individuals.68, 69 Surgically induced iris-vessel filling abnormalities are probably best evaluated by comparing preoperative to postoperative iris angiograms for delay in vessel filling, the size of the sector with delayed filling, and late leakage.

In normal individuals, disrupting all of the anterior ciliary vessels on the medial rectus muscle and/or the lateral rectus muscle usually does not alter iris blood flow in a way that is detectable by iris fluorescein angiography. This presumably is explained by collateral blood flow that comes predominantly from the underlying long posterior ciliary arteries. In contrast, humans as well as nonhuman primates commonly develop iris perfusion abnormalities after tenotomy of one or both of the vertical rectus muscles, provided that no surgery has previously been performed on these muscles.34, 36, 68, 69, 73, 91 In one study, 89% of adult eyes showed angiographic evidence of iris sector perfusion defects after detachment of one or two previously unoperated vertical rectus muscles.69 Detachment of both vertical rectus muscles or the inferior rectus and medial rectus muscles had additive effects, resulting in a greater mean delay in iris vessel filling, a larger sector of delayed filling, and more severe clinical signs of ASI. Interestingly, the abnormalities were more severe when the surgery was performed on two contiguous rectus muscles (inferior and medial) rather than on the two vertical rectus muscles alone. The effects of surgery on other contiguous combinations were not evaluated in this study.

There is experimental as well as clinical evidence that the anterior ciliary vessels generally do not recanalize after primary rectus muscle surgery. When a vertical rectus muscle...
is subjected to a second procedure, sector hypoperfusion does not occur. Blood flow from the anterior ciliary arteries disrupted by the first procedure presumably is compensated for by collateral flow. Experimental microvascular casting studies confirm that the anterior ciliary arteries do not revascularize after primary rectus muscle surgery.

Blood flow within the anterior ciliary vessels is bidirectional; some vessels flow toward the limbus, whereas others flow toward the orbit. Blood flow in some thin-walled vessels may occasionally exhibit flow reversal at the time of surgery. It generally is not possible to distinguish between arteries and veins by clinical observation alone at the time of surgery.

**Anterior Segment Ischemia**

**RISK FACTORS**

The overall risk of developing ASI after routine strabismus surgery is extremely low. This is probably due, at least in part, to the large number of one- or two-muscle horizontal strabismus procedures that are performed on the eyes of healthy children as well as certain somewhat arbitrary, although quite effective, guidelines that most surgeons follow to reduce the risk of ASI.

Estimates of the actual risk of ASI depend on the population being studied and how the question is asked. In a 1984 membership survey of the American Association for Pediatric Ophthalmology and Strabismus, only 30 cases of documented ASI were reported in an estimated 400,000 strabismus procedures. This represents less than one case of ASI for every 13,000 strabismus operations. The study suffers from several limitations, including the use of a retrospective survey relying solely on surgeon recall and what probably represents highly variable ASI detection techniques, particularly with respect to subtle ASI. When the question is approached using a different population and methodology, the risk of developing ASI becomes exceptionally high. In a report of patients having three-rectus-muscle surgery on the two vertical rectus muscles as well as the medial rectus, all five adults aged 34 to 65 years developed clinically detectable ASI. Three underwent simultaneous surgery, whereas two had staged procedures. In the same study, none of four pediatric patients 1 to 8 years of age developed ASI.

The two major risk factors for ASI are individual patient susceptibility and the extent of strabismus surgery. Advancing age appears to be the most important single risk factor. There are only a few well-documented cases of ASI in children, all occurring in unusual clinical settings such as cicatricial retinopathy of prematurity, prior orbital surgery, congenital ocular malformations, or malnutrition. Age becomes particularly important when coupled with certain systemic, orbital, or ocular disorders. These include abnormalities affecting blood flow, such as leukemia, homocystinuria, and hemoglobinopathies (hemoglobin SS and SC), as well as small- and large-vessel abnormalities secondary to diabetes mellitus, hypertension, atherosclerosis, carotid artery disease, carotid-cavernous fistula, or other disorders. In addition, certain orbital and ocular abnormalities, including prior uveitis and thyroid-related orbitopathy, may increase susceptibility to ASI.

The extent of strabismus surgery is the second major risk factor; it includes the proposed procedure as well as prior eye surgery, especially surgery on the rectus muscles (not the obliques), a scleral buckle, diathermy, cryotherapy, or other procedures that may affect anterior segment blood flow. Most reported cases of ASI have followed surgery on three or four rectus muscles. Significant exceptions have been described, particularly with advancing age, a concurrent orbital disorder, or systemic disease. Mild ASI has even been observed in susceptible patients after surgery on a single vertical rectus muscle.

**PATHOPHYSIOLOGY, CLINICAL MANIFESTATIONS, AND TREATMENT**

Although the exact mechanism by which ASI develops is unknown, it is generally thought to occur when surgery and/or disease causes a disturbance of anterior segment blood flow that is sufficient to cause ischemia. The spectrum of ASI ranges from subtle abnormalities detectable only by iris angiography to severe ischemic changes resulting in significant anatomic alterations and visual loss.

Most clinically detectable cases of ASI are probably characterized by mild, self-limited iritis, which can easily be missed by the patient as well as the examiner in the setting of expected postoperative discomfort—unless specifically looked for. More severe ASI is not easily missed; it results in significant iritis with pupillary abnormalities and keratopathy. The sequelae of ASI include iris atrophy, corectopia, immobile pupil, corneal clouding, cataract, glaucoma, hypotony, and phthisis bulbi.

Treatment of the acute stages of ASI usually includes cycloplegic agents and topical corticosteroids. Systemic corticosteroids have also been advocated, and hyperbaric oxygen has been used. Efforts are made to control intraocular pressure if it is elevated. Potentially reversible surgical procedures, such as the Jensen procedure, may be taken down; in theory, this may improve anterior ciliary blood flow by releasing pressure on otherwise intact anterior ciliary vessels.

**REDUCING THE RISK OF ASI**

**Limiting the Number of Rectus Muscles Operated On**

Ophthalmic surgeons usually limit the number of rectus muscles tenotomized at one time in an effort to reduce the risk of ASI. Although recommendations vary widely, the majority of ophthalmologists would probably not tenotomize four rectus muscles in one surgical session, even in a healthy child in whom the risk of ASI may not be particularly high. Many would avoid tenotomizing three rectus muscles at one sitting in an adult, particularly a middle-aged or elderly patient with concurrent disorders known to increase the risk of ASI.

**Staging the Procedures**

Staging surgery reduces (but does not eliminate) the risk of ASI, probably by allowing collateral flow to develop. The
premiere mechanism by which staging surgery limits the risk is unknown. There is compelling evidence that the anterior ciliary arteries generally do not recanalize after surgery. A second operation on a previously tenotomized vertical rectus muscle does not result in sectoral iris perfusion defects, as may occur with the initial procedure.69, 70

The recommended waiting period between procedures ranges from about 1 month to 6 months.29, 69, 84, 87 Most surgeons probably wait several months before performing additional strabismus surgery when staging is employed to reduce the risk of ASI. However, patients have developed ASI with much greater intervals between operations, even as long as 20 years.62, 82, 84 Iris fluorescein angiography usually shows quite rapid recovery of iris filling times in humans as well as experimental animals having rectus tenotomies.34, 66, 91 In one study the majority of iris angiograms returned nearly to normal within 2 weeks of surgery. However, some patients with more severe ischemic changes required up to 3 months to recover. In all patients with iris sector hypoperfusion the last sector to fill was the hypoperfused one, even in patients evaluated 1 year after surgery.69

Choice of Surgical Incision

The surgical incision site may have some influence on anterior segment blood flow, although ASI has been reported after strabismus surgery using either limbal or fornix conjunctival incisions. The limbal incision disrupts the perilimbal conjunctival-episceral collateral network and theoretically may increase the risk of ASI in susceptible patients. In a primate model with tenotomies of three or four rectus muscles, using limbal and fornix conjunctival incisions, eyes receiving limbal incisions showed more severe ischemic changes than those having fornix incisions. The authors hypothesized that the fornix approach preserves the perilimbal conjunctiva-Tenon’s circulation, which provides collateral blood flow to the iris and anterior segment.24 The fornix approach may therefore be preferable in patients who are considered to be at risk for developing ASI.24, 82

Botulinum Toxin

Botulinum toxin may be used to reduce the total number of rectus muscles operated on by serving as an alternative to rectus muscle recession in patients believed to be at risk of developing ASI. Botulinum is commonly used to treat acute sixth cranial nerve palsy and may be injected into a contracted medial rectus muscle before, during, or after vertical rectus muscle transposition to treat a persisting sixth cranial nerve palsy.3, 25, 47 Using botulinum in these circumstances may eliminate the need for detachting three rectus muscles from the globe.76, 77, 82, 83 Nonetheless, ASI has been reported in an elderly patient receiving a botulinum injection in the medial rectus in addition to vertical rectus muscle transpositions. It is not known whether the botulinum injection itself or other factors led to ASI.47

Anterior Ciliary Vessel Sparing

INDICATIONS

Although the risk factors for ASI are well known, there are no reliable clinical or laboratory studies from which to determine the precise risk of ASI in a given patient. At times, the ideal procedure from a mechanical perspective creates a potentially significant risk for the development of ASI. An example would be an elderly adult with a longstanding sixth nerve palsy and documented small- and large-vessel disease affecting blood flow to the head. Intractable diplopia caused by the palsy has been unsuccessfully treated by a recession and resection of the horizontal rectus muscles of the paretic eye. For many surgeons the next procedure would be a full tendon transposition of the superior and inferior rectus muscles. However, this may create a significant risk of ASI, because it represents surgery on the third and fourth rectus muscles in an individual with several important risk factors. Anterior ciliary vessel sparing procedures may be indicated in this and similar situations.

Iris fluorescein angiography can be performed on blue-eyed patients and may provide useful information preoperatively, as well as furnish indirect evidence of anterior ciliary vessel patency postoperatively. Indocyanine green videoangiography to image the iris vessels and filling patterns may be used in patients with dark-colored irides (Rosenbaum AL, personal communication, 1998). Vessel-sparing procedures may be contraindicated or require modification if preoperative iris angiography shows abnormal vessel filling in certain sectors. In the example illustrated, the delayed filling identified preoperatively was taken as evidence of increased susceptibility to ASI (Fig. 39–4). The patient underwent a full tendon transposition of the superior and inferior rectus to the appropriate borders of the lateral rectus insertion, with preservation of three clinically apparent anterior ciliary vessel groups on each muscle. Postoperative iris fluorescein angiography, performed on day 5, showed no change in the filling pattern of the iris vessels (Fig. 39–5). This contrasts
Figure 39–5. Postoperative iris fluorescein angiogram of patient shown in Figure 39–4 on fifth day after full-tendon transposition of the superior and inferior rectus muscles to the appropriate borders of the lateral rectus insertion. The iris vessels are dilated with simultaneous filling of the superior, inferior, and temporal vessels, which is unchanged from the preoperative study. Dilation of the conjunctival vessels is also apparent with delayed filling of a single superficial perilimbal vessel in the superior temporal region. Picture was taken 14.3 seconds after fluorescein injection. (From McKeown CA, Lambert HM, Shore JIV: Preservation of the anterior ciliary vessels during extraocular muscle surgery. Ophthalmology 1989;96:498–506)

Anterior ciliary vessel sparing procedures should be considered when the proposed surgery creates a potentially significant risk of ASI. The surgeon must consider the patient’s age, general health, ocular health, and prior ocular surgery in the context of the proposed procedure. Table 39–1 lists a number of known risk factors for ASI. The presence of one or more of these risk factors may lead the surgeon to consider anterior ciliary vessel sparing. Appropriately performed vessel sparing procedures do not appear to significantly alter the effectiveness of recessions, resections, or transpositions.

CONTRAINDICATIONS

A nearly absolute contraindication to performing successful anterior ciliary vessel sparing is prior surgery on the same muscle. Rectus recessions, resections, and full tendon transpositions disrupt all of the anterior ciliary vessels on the operated muscle(s). Because the anterior ciliary vessels generally do not recanalize, vessel sparing is not possible during reoperation. Vessel sparing may also be extremely difficult or impossible when the dissection from prior surgery on an adjacent muscle extends over the muscle in which vessel sparing is planned. Scar tissue and adhesions from the original procedure may preclude successful dissection and preservation of the anterior ciliary vessels. In addition, operations that spare part of the anterior ciliary circulation on the operated muscle, such as the Hummelsheim and Jensen procedures, often result in significant scarring on the unoperated half of the muscle and, therefore, may not be amenable to vessel sparing when attempting to convert these procedures to full tendon transpositions. Ocular trauma may also damage the anterior ciliary vessels or cause scarring that makes vessel sparing impossible.

It is common for patients with strabismus to be unaware that they had ocular surgery in childhood. This is especially true of procedures performed during infancy and early childhood. Some parents do not inform their children of the surgery, or the adult patient may simply have forgotten about eye surgery done early in life. The prospective surgeon should evaluate the eyes carefully for signs of ocular trauma or prior surgery, looking for scar tissue involving the conjunctiva, Tenon’s capsule, or tendon insertion as well as the absence of anterior ciliary vessels in the quadrants corresponding to each of the four rectus muscles.

SURGICAL TECHNIQUE

Dissection and preservation of the anterior ciliary vessels may be carried out during conventional full-width procedures on the rectus muscles, including recessions, resections, and transpositions. The surgeon should be as certain as possible that the technique employed reliably spares the anterior ciliary vessels. It is wise for the novice surgeon to develop vessel-sparing skills in situations in which the risk of ASI is very low. Small portions of the anterior ciliary vessel sparing procedure may be incorporated into routine strabismus surgery without altering the operation, even if the

Table 39–1. Risk Factors for Anterior Segment Ischemia That May Influence Consideration for Vessel Sparing

| Increasing age: adults, particularly the elderly |
| Systemic disorders affecting blood flow |
| Hemoglobinopathies (particularly hemoglobin SS and SC) |
| Certain metabolic disorders (homocystinuria) |
| Leukemia or polycythemia |
| Large or small blood vessel disease |
| Atherosclerosis |
| Carotid artery stenosis |
| Hypertension |
| Diabetes mellitus |
| Others |
| Carotid-cavernous sinus fistula |
| Others |
| Orbital and ocular disorders |
| Thyroid-related orbitopathy |
| Orbital disorders affecting blood flow |
| Uveitis |
| Congenital anomalies including the absence of selected rectus muscles |
| Others |
| Defects on iris fluorescein or indocyanine green angiography |
| Prior ocular surgery |
| Potentially compromised long posterior ciliary blood flow from retinal surgery |
| Prior strabismus surgery on the rectus muscles (not the oblique muscles) |
| Other ocular procedures that damage anterior ciliary or long posterior ciliary vessels |
| Proposed surgery |
| Increasing number of rectus muscles on one eye |
| Three muscles (particularly both vertical recti and the medial rectus) |
| Four muscles |
| Surgery on multiple rectus muscles in a single stage |
| Limbal conjunctival incision |
vessels are ultimately severed. There is probably little value to routinely sparing the vessels, unless it is thought that additional procedures may be required in the future that would place the eye at increased risk of ASI.

The procedure is usually the most technically difficult to perform on the medial rectus and most straightforward on the two vertical rectus muscles. Interestingly, the relative difficulty of dissection and preservation of the anterior ciliary vessels appears to be well matched with the most crucial requirements for anterior segment blood flow (the vertical rectus muscles are most important). Unless other indications exist, it is generally only necessary to spare the anterior ciliary vessels on the vertical rectus muscles, even when horizontal muscle surgery also is performed.

The first known reference to attempted dissection and preservation of anterior ciliary vessels during strabismus surgery was by Carlson and Jampolsky in 1979. Both microscopic and loupe techniques have been described for sparing the anterior ciliary vessels. The microscopic technique probably allows more precise vessel dissection as well as better evaluation of blood flow intraoperatively. The rate of unplanned vessel destruction appears to be lower when using the microscope. Operating time in a teaching hospital is roughly 45 minutes to 1 hour for each muscle on which the anterior ciliary vessels are to be dissected and preserved. The dissection time and the risk of unplanned vessel destruction are reduced when an experienced surgical team is at hand.

**Surgical Equipment**

The surgical equipment for anterior ciliary vessel dissection and preservation includes standard strabismus instruments as well as supplemental equipment for the microvascular dissection. The standard strabismus equipment is listed by name and Storz identification number in Parks' Atlas of Strabismus Surgery. The supplemental microvascular equipment includes Grieshaber 70-degree angled microscissors with 3.1-mm blades, a Michels 90-degree vitrectorial hook or Sinsky hook, a jeweler microtip bipolar cautery, and straight and angled 0.3-mm-tip microvascular forceps (S & T Microsurgical, Accurate Surgical & Scientific Instrument Corporation, Westbury, NY). In addition, a Jameson muscle hook is modified by cutting off all but about 1 inch of the handle, through which a hole is drilled in the stump of the handle for passage of a 4-0 silk traction suture. The suture is clamped to the surgical drapes to hold the eye stable during the vessel-sparing procedure.

**Conjunctival Incision**

The fornix approach is recommended, because it requires less dissection anterior to the tendon insertion. This reduces the likelihood of damaging the anterior ciliary vessels in front of the tendon insertion and preserves the perilimbal conjunctival contribution to anterior segment blood flow. There is some controversy about the significance of the superficial contribution to anterior segment blood flow, particularly with respect to single-quadrant incisions. However, Fishman's study in nonhuman primates supports a potential contribution of the superficial marginal plexus at the limbus. This may be important when surgery involves multiple quadrants, a common occurrence in patients with complex strabismus.

**Isolating the Rectus Muscle**

As the rectus insertion is exposed, care is taken to avoid damaging the anterior ciliary vessels on the surface of the tendon or the vessels and their branches on the surface of the sclera. The modified Jameson muscle hook is placed beneath the insertion, and the globe is secured in position by clamping the attached silk suture to the surgical drapes. In addition, 6-0 silk traction sutures are used to retract the conjunctiva and Tenon's capsule to expose the anterior ciliary vessels. These maneuvers free the assistant's hands to aid the microvascular dissection.

The intermuscular septum is incised at the border of the rectus muscle and tendon. The fibrous tissue connections between the muscle capsule and Tenon's capsule (check ligaments) are dissected. At all times, care is taken to avoid damaging the anterior ciliary vessels. Excessive traction may elevate the vessels off the surface of the muscle and tendon, exposing them to inadvertent damage as the check ligaments and intermuscular septum are cut.

**Ciliary Vessel Dissection and Preservation**

After the tendon and muscle are exposed, all clinically apparent anterior ciliary vessels are identified for dissection and preservation. This includes all single vessels or groups of vessels of significant size that travel from muscle to sclera, regardless of the direction of blood flow (see Fig. 39–1). No attempt is made to distinguish arteries from veins. A surgical dissection plane is created beneath each vessel or vessel group, using microvascular forceps to grasp the connective tissue adjacent to the vessel and making spreading motions with the Grieshaber scissors (Fig. 39–6). Alternatively, Vannas scissors may be used for this purpose, but the tips of the Grieshaber scissors are finer and the blades can be rotated 360 degrees. It is usually easier to lift the vessels off the tendon 2 to 4 mm posterior to the tendon insertion. It is important to maintain tension on the tendon during this process to avoid including tendon fibers in the vessel dissection. With a combination of sharp and blunt dissection, with traction and countertraction, dissection proceeds posteriorly to the point where the vessels emerge from the muscle and anteriorly onto the scleral surface, just in front of the tendon insertion (Fig. 39–7; see also Fig. 39–6). Blunt dissection generally is used more than sharp dissection. The anterior ciliary vessels are never directly grasped with the forceps during the dissection; rather, traction is accomplished by grasping the surrounding connective tissue or by lifting the vessels with a blunt right-angled hook or a smooth cycloidalysis spatula. Thin-walled silicone tubing has also been used to lift the vessels.

After completing the vessel dissection, a 5-0 Mersilene suture can be placed in the sclera adjacent to each border of the tendon insertion. The Mersilene sutures are used for traction on the globe after the tendon or muscle is cut. If traction sutures are used, they should be placed in the sclera at a site that does not damage the anterior ciliary vessels or their branches.
**Assessing Vessel Patency**

Vessel patency may be documented by three techniques: direct observation at the time of surgery, iris angiography, and direct observation at reoperation. A simple but effective intraocular means of determining whether vessels are functioning at the time of surgery is by directly visualizing vessel refilling. The column of blood present in the lumen of most anterior ciliary vessels can easily be seen under the operating microscope, particularly when the preserved vessels are silhouetted against the sclera after the tenotomy is completed and the tendon has been allowed to retract. The direction of blood flow can usually be determined by observing the anterior ciliary vessel refilling with blood after gentle peristaltic compression of the vessel against the sclera using a Stevens muscle hook. Consistent with fluorescein studies, some vessels show flow directed from inside the eye toward the orbit, whereas others demonstrate flow directed from the orbit toward the globe. In some vessels the rate of flow is too rapid to determine flow direction by direct observation at the time of surgery. Pulsating vessels are relatively common and may have thicker walls and very rapid flow rates. Abnormal blood flow detected at the time of vessel sparing may warrant modification of the rest of the procedure or may preclude continuing the surgery.

**Completing the Rectus Muscle Procedure**

A standard, double-armed 6-0 Vicryl suture is passed through the tendon or muscle in the appropriate position for recession, resection, or transposition (Figs. 39–7 through 39–9). The S-29 needle (Ethicon) has a very thin wire diameter and is well suited for this purpose. As the needles are passed through the tendon or muscle, the dissected anterior ciliary vessels are gently “combed” in the appropriate direction to avoid injury by the needle or suture. Vicryl has a tendency to adhere to and “grab” surrounding soft tissues.

---

**Figure 39–6.** A, Initiation of dissection by creating a surgical plane beneath the anterior ciliary vessels using Grieshaber scissors. B, Microvascular forceps grasp connective tissue and lift vessel to expose plane of dissection between vessel and tendon. A small branch vessel is also shown. C, Right-angled blunt retinal hook is used to lift partially dissected vessel. Note branch vessel that has been cut after cauterization. (Modified from McKeown CA, Lambert HM, Shore JW. Preservation of the anterior ciliary vessels during extraocular muscle surgery. Ophthalmology 1989; 96:498–506.)

**Figure 39–7.** Diagram of sagittal section of rectus muscle with completed dissection of the anterior ciliary vessel. The vessel can be seen to emerge from deep in the muscle on the left and lies suspended on the retinal hook on the right. The “slack” in the vessel and the continuation of the dissection onto the scleral surface provides adequate space for suture passage adjacent to the insertion. Dashed line indicates the proposed tenotomy site. (Modified from McKeown CA, Lambert HM, Shore JW. Preservation of the anterior ciliary vessels during extraocular muscle surgery. Ophthalmology 1989; 96:498–506.)

**Figure 39–8.** Diagram of partially completed tenotomy with suture in place and one vessel suspended on a retinal hook. Dashed line marks the proposed site of completion of the tenotomy. (Modified from McKeown CA, Lambert HM, Shore JW. Preservation of the anterior ciliary vessels during extraocular muscle surgery. Ophthalmology 1989; 96:498–506.)
including dissected vessels. As the suture is pulled through the tendon, muscle, or sclera, care is needed to avoid damage to the dissected vessels.

After the Vicryl suture has been passed through the tendon or muscle, the silk suture holding the modified Jameson hook is released. The hook is held by the assistant or surgeon for controlling tension during the tenotomy. In the case of a recession or full tendon transposition, the tenotomy may be accomplished using the relatively fine tips of sharp Westcott scissors, which can be positioned to avoid damage to the dissected anterior ciliary vessels. As with passage of the Vicryl suture, the vessels are “combed” in the appropriate direction to avoid injury by the scissors during the tenotomy. Traction on the globe should be reduced as the tenotomy nears completion. This avoids abrupt tendon “snap back,” which may cause inadvertent vessel damage. A partially completed tenotomy is shown in Figure 39–8.

After completion of the tenotomy, the Jameson hook is removed and the previously placed Mersilene sutures (or forceps) are used to position the globe. Each arm of the Vicryl suture that is attached to tendon or muscle is passed in the sclera in the appropriate position for recession, resection, or full tendon transposition (Figs. 39–9 through 39–13). Scleral suture passage with recessions is similar to any standard recession—the dissected vessels usually do not present a significant problem. With resections, single-suture techniques are preferred to reduce the risk of inadvertent vessel dragging by the Vicryl sutures. As the resected tendon or muscle is advanced to the original insertion site, the excess length and location of the preserved vessels makes inadvertent vessel dragging more likely than with recessions. If a transposition is to be done, it is often helpful to pass the Vicryl suture from the most distal pole beneath the tendon before the tenotomy. Once the tenotomy is completed, it may be difficult to pass the distal pole suture beneath the dissected vessels that now are pulled firmly against the sclera by the retracted muscle.

**COMPLICATIONS**

Patient selection is crucial for successful vessel sparing. A comprehensive ophthalmologic examination, including a careful history, will usually uncover evidence of prior ocular trauma or surgery, including strabismus procedures. Regardless of the history, the conjunctiva and anterior ciliary vessels in each quadrant should be inspected by slit lamp before surgery. The absence of anterior ciliary vessels, or the presence of scar tissue over the vessels, may have a direct and
significant impact on the risk of ASI as well as the feasibility of vessel sparing. In some patients, the risk of developing ASI is extremely high and failure to effectively spare the anterior ciliary vessels may lead to severe ischemic complications.

Surgical technique determines the success or failure of vessel sparing. Failures can be minimized if the surgeon develops vessel-sparing skills in situations in which successful preservation of the anterior ciliary vessels is not critical. Inadvertent anterior ciliary vessel destruction may be recognized at the time of surgery or may go unrecognized. The latter form of vessel destruction may be more dangerous, because the surgeon has no opportunity to alter or terminate the procedure. Anterior ciliary vessel sparing procedures associated with recognized, inadvertent vessel destruction or unrecognized vessel destruction, or both, may place the patient at significant risk for developing ASI.

The rate of inadvertent vessel destruction recognized at the time of surgery is roughly 10%. Although unplanned vessel destruction occurs during vessel dissection, it is surprising how often it occurs during other phases of the procedure. The Vicryl suture may snag and disrupt dissected anterior ciliary vessels as it is pulled through the tendon or muscle, or when locking bites are taken at the borders of the tendon or muscle. Successfully spared vessels may be destroyed when the last few fibers of the tendon are cut and the muscle hook beneath the tendon insertion pulls forward as the muscle abruptly snaps posteriorly. The vessels may also be disrupted when the sutures snag the preserved vessels as the tendon or muscle is moved into the proper position for recession, resection, or transposition. Vessel destruction that is recognized at the time of surgery often provides the surgeon with an opportunity to continue, modify, or terminate the surgical procedure if the risk of ASI is high.

Of greater concern are anterior ciliary vessels that appear to be spared at the time of surgery but fail to function or have significantly reduced flow postoperatively. Damage to the vessels during surgery or excessive traction from large recessions or transpositions may result in reduced or absent blood flow. Thrombosis could, in theory, result from damage to the vascular endothelium at the time of surgery or from reduced flow caused by excessive tension or external pressure on the preserved vessels after the completion of surgery. It is also possible for the surgeon to preserve a cord of connective tissue that resembles an anterior ciliary vessel group but contains no functioning vessels.
ALTERNATIVE SURGICAL PROCEDURES

The surgeon has quite a number of alternatives when faced with the need to operate on multiple rectus muscles in the face of known or suspected ASI risk factors. Careful surgical planning as well as modifications of conventional strabismus surgery techniques can significantly reduce the risk of ASI in vulnerable patients. Techniques for preserving all or a portion of the anterior ciliary vessels have been available since the beginning of the 20th century, well before the first description of ASI complicating ocular surgery in humans in 1955. They include muscle-splitting transposition procedures such as the techniques described by Hummelsheim in 1908, Jensen in 1964, and numerous modifications, summarized by Helveston.

Muscle splitting procedures can effectively create the new force vectors required in transposition procedures while sparing some or all of the anterior ciliary vessels. In theory, the Jensen procedure can spare all of the anterior ciliary vessels on the operated muscles, whereas the Hummelsheim and related procedures can spare the anterior ciliary vessels on the halves of the muscles and tendons that are not transposed. Care must be taken in selecting patients and performing these operations, because ASI has been reported after both the Hummelsheim and Jensen procedures (see also Chapter 36).

Other techniques have evolved, including muscle tightening procedures using muscle or tendon plication. Marginal myectomy that results in plication of the central region of the tendon or muscle has been described. This potentially leaves the central anterior ciliary vessels intact. The O'Connor cinch shortens a palsied muscle, plicating the full width of the tendon. However, it is likely that the anterior ciliary vessels will be strangled by this procedure. A modified rectus tuck that specifically attempts to preserve anterior ciliary blood flow without dissecting the anterior ciliary vessels has also been proposed (Fig. 39–14). The ability of this procedure to spare functioning anterior ciliary vessels was demonstrated in an animal model.

Surgery may also be performed on the opposite eye to reduce the number of operated muscles on the eye at risk for ASI. Transposition of the vertical rectus muscles in the parietic eye may be combined with recession of the medial rectus muscle in the contralateral normal eye in patients with sixth cranial nerve palsy. Recession or resection may be combined with vertical or horizontal transposition procedures to reduce the total number of operated muscles.

Figure 39–14. Wright's modified rectus tuck attempts to preserve the anterior ciliary vessels without the need for vessel dissection. A, Nonabsorbable suture is passed through half-thickness muscle and tendon, deep to the anterior ciliary vessels, in the appropriate measured position for the tuck. Locking bites secure each pole while avoiding the anterior ciliary vessels. Each arm of the suture is passed parallel to and just in front of the tendon insertion, sparing the anterior ciliary vessels on the scleral surface. B, Posteriorly placed sutures are advanced to the scleral suture site by pulling on the sutures, creating a fold or tuck of redundant tissue while leaving the anterior ciliary vessels intact. (From Wright KW. Rectus strengthening procedures. In: Color Atlas of Ophthalmic Surgery: Strabismus. Philadelphia, JB Lippincott, 1991. Reprinted with permission.)

REFERENCES

76. Safran AB, Janin Y, Roth A: Bitemporal palsy of the pupillary spincterם.
The slipped rectus muscle and the lost rectus muscle are recognized complications of strabismus surgery and retinal surgery and also may result from trauma. The tendon of the slipped muscle may break away from its scleral attachment and recoil within its capsule toward its penetration through Tenon’s capsule. Although weakened, the slipped horizontal or vertical muscle continues to deliver some degree of duction up to or past the midline. Or, the tendon and its capsule may have been severed from their connections to the sclera but retain some motor function. The tendon-capsule complex develops connections to the intermuscular septa and check ligaments, usually achieving a new scleral attachment. Because the muscle may recoil minimally in its capsule, it would probably be better to call this complication a disinserted or severed muscle rather than a slipped muscle.

Unlike the slipped muscle within its capsule, the disinserted or severed muscle usually attaches to sclera at an exaggerated recession site, providing some degree of limited duction in its field of action. The lost muscle, on the other hand, recoils through Tenon’s capsule back into the posterior orbit and delivers little or no duction when stimulated.

The surgical findings, management, and prognosis differ for each of these complications. The anatomy pertinent to these complications is reviewed first.

**Anatomy**

Knowledge of the anatomic features of the muscle and its tendon, the musculotendinous capsule, Tenon’s capsule, intermuscular septum, and check ligaments is important for understanding the nature of these complications.

**MUSCLE CAPSULE**

The muscle and tendon fibers of the extraocular muscles are enveloped by a glistening, avascular capsule with a smooth external surface. Its internal surface is attached by elastic connective tissue to the vascular muscle fibers, the avascular tendon fibers, and the anterior ciliary vessels that course forward on the anterior surface of the tendon. A break in the capsule causes immediate and profuse bleeding into both the surrounding surgical field and the capsule, producing an intramuscular hematoma. The elastic connective tissue interface between the capsule and muscle/tendon fibers allows the muscle fibers to move back and forth within the capsule during contraction and relaxation.

**TENON’S CAPSULE**

Tenon’s capsule is a relatively avascular fascial layer surrounding the eye from limbus to optic nerve. The anterior portion of the rectus muscles are covered by Tenon’s capsule because they penetrate the capsule posterior to the equator of the globe (Fig. 40–1). At the site of penetration, the muscle capsules are firmly attached by sleeves of dense elastic connective tissue to Tenon’s capsule. The sleeve permits to-and-fro movement of the muscle through the penetration site as it contracts and relaxes. This arrangement also creates a watertight sub-Tenon’s space. The four rectus muscle penetrations divide Tenon’s capsule into anterior and posterior portions. The anterior Tenon’s capsule is a barrier between the sub-Tenon’s space and the extraconal fat, which comes to within 10 mm of the limbus. The posterior Tenon’s capsule is a barrier between the intraconal fat and posterior sclera. The inner surface of Tenon’s capsule is smooth and glistening, providing an ideal surface for maximal motion of one fascial plane on the other during rotation of the globe.

The external surface of Tenon’s capsule is rough because of the multitude of elastic connective tissue septa within the extraconal and intraconal fat that attach to it. The elastic tissue septa are very vascular, causing the fat to bleed profusely when cut.

A rent in Tenon’s capsule more than 10 mm posterior from the limbus results in the prolapse of orbital fat into the sub-Tenon’s space, exciting an inflammatory response in the vascularized elastic tissue septa. These septa form scar tissue in the sub-Tenon’s smooth tissue planes, which become...
inelastic and restrict freedom of rotation of the globe. Through scarring, the eye is displaced from its center of rotation. The rent in Tenon’s capsule is the cause of the adherence syndrome.

**INTERMUSCULAR SEPTUM**

Retrobulbar to Tenon’s capsule, the intermuscular septum joins the four rectus muscle capsules together to form the muscle cone. Similarly, between anterior Tenon’s capsule and sclera, the intermuscular septum unites the extracocular muscles and tendon capsules. Here it is a thin, avascular, transparent fascial sheet, fusing with Tenon’s capsule approximately 3 mm peripheral to the limbus. It is very prominent in young patients but gradually degenerates in the elderly, becoming almost unrecognizable.

In the sub-Tenon’s space, the intermuscular septum also joins the lateral and inferior rectus muscles to the inferior oblique muscle and joins the superior rectus muscle to the reflected tendon of the superior oblique muscle. These unions are strong enough to hold the severed rectus muscle forward, preventing it from passing through the anatomic rent in Tenon’s capsule to become a lost rectus muscle. Unfortunately, the medial rectus muscle capsule is attached to neither the oblique muscle nor its tendon, making it likelier than other rectus muscles to be lost.

**CHECK LIGAMENTS**

The space between Tenon’s capsule and the rectus muscle capsule is bridged by sheets of avascular elastic connective tissue called check ligaments. These extend radially between Tenon’s capsule and the muscle capsule as two or three falciform folds, arranged lengthwise along the terminal 10 mm of the horizontal rectus muscles. They are best visualized as Tenon’s capsule is retracted away from the muscle. The check ligaments for the vertical rectus muscles are sheets of avascular tissue coming off the inner surface of Tenon’s capsule and attaching transversely across the muscle capsule 6 to 7 mm anterior to the muscle penetration site of the capsule.

**SLIPPED MUSCLE**

Slipped muscle occurs most frequently as a complication of strabismus surgery. Although unsubstantiated, evidence suggests that recent refinements in strabismus surgery, including improved needles, suture material, and instruments, are related to the apparent increase in this complication. It is now possible using fine suture materials and needles to preplace the suture only in the muscle/tendon capsule, rather than deeply into the muscle fibers, before disinserting the muscle (Fig. 40–2). When disinserted, the muscle/tendon begins to slip posteriorly within the capsule (Fig. 40–3). If unrecognized, the surgeon inadvertently sutures only the capsule, not the muscle/tendon unit, to the sclera. As the muscle receives stimuli to contract postoperatively, recoiling of the muscle/tendon continues to pull the disinserted end posterior to the site of penetration through Tenon’s capsule (Fig. 40–4). The muscle capsule fuses with Tenon’s capsule at this site, which holds the disinserted end in place.

Not all slipped muscles recoil to this level. Some remain entrapped within the capsule, unattached to the sclera, after slipping only a few millimeters. However, the pathologic aspect of this complication is that the muscle/tendon fibers are not attached to sclera. The duction power of the involved muscle is therefore translated through its connections to the Tenon’s capsule penetration site and the empty capsule distal to the disinserted muscle/tendon end.
Disinserted or Severed Muscle

At present, this is an infrequent complication of strabismus surgery. Historically, it was the invariable complication associated with the first surgical technique introduced for strabismus in the mid-19th century. The surgeon merely hooked the rectus muscle and cut it off the sclera, the so-called free tenotomy. The tendon and its capsule were disinserted en bloc. No suturing was done. The muscle/tendon and its capsule recoiled according to the limit permitted by the intermuscular septum, where the tendon eventually attached to the sclera—similar to today's hang-back rectus muscle weakening technique. Overcorrections of the strabismus angle were common. Uncontrolled retraction of the tenotomized muscle caused an exaggerated recession effect, producing a consecutive tropia with compromised duction in the tenotomized muscle's field of action. Because the muscle/tendon capsule was not sutured to sclera, no slippage of the muscle/tendon occurred within its capsule.

Therefore, the complication technically is not a slipped muscle.

Today there are many causes of a severedor disinserted muscle. One is inadvertent myotomy or myectomy of the inferior rectus or lateral rectus when the intended goal is to operate on the overacting inferior oblique. Similarly, the superior rectus has been myectomized rather than performing a tenectomy on the overacting superior oblique. These surgical misadventures all result from blindly sweeping the muscle hook in hope of engaging the oblique muscle/tendon but instead hooking the neighboring rectus muscle.

Another cause of a severed muscle is the "pulled-in-two syndrome," to which Greenwald (personal communication) gave the descriptive acronym PITS. It usually occurs when the surgeon or assistant pulls excessively on the rectus muscle during dissection. Because tissues in the elderly are more friable, they are prone to suffer this complication. The weakest site in the rectus muscle is at its musculotendinous junction, where most cases of PITS occur.

Severed muscles can also complicate retinal detachment surgery. They tend to occur late, after an encircling band or exoplant has created pressure necrosis and eroded through the overriding rectus muscle. Stab wounds of the orbit, whether sharp or blunt, can sever a rectus muscle anywhere along its course from its insertion to deep in the orbit.

Lost Muscle

This complication may appear during extraocular muscle surgery for strabismus, during retinal detachment surgery, or when managing an injured muscle. Usually the surgeon will have dissected the muscle away from its surrounding intermuscular septa and check ligaments. A mishap occurs:
Table 40–1. Twenty-Year Experience and the Incidence of Slipped, Lost, and Disinserted Rectus Muscle

<table>
<thead>
<tr>
<th>Type of Muscle</th>
<th>Slipped</th>
<th>Lost</th>
<th>Disinserted</th>
</tr>
</thead>
<tbody>
<tr>
<td>Medial rectus</td>
<td>87*</td>
<td>19*</td>
<td>2</td>
</tr>
<tr>
<td>Lateral rectus</td>
<td>16</td>
<td>3</td>
<td>2</td>
</tr>
<tr>
<td>Superior rectus</td>
<td>8</td>
<td>1</td>
<td>2</td>
</tr>
<tr>
<td>Inferior rectus</td>
<td>18</td>
<td>1</td>
<td>2</td>
</tr>
<tr>
<td>Totals</td>
<td>129</td>
<td>24</td>
<td>8</td>
</tr>
</tbody>
</table>

*Includes two patients each with a slipped muscle that was lost during an attempt to pull a tight muscle forward and to suture it to the sclera.

the PITS complication; the clamp on a tight, previously operated muscle is accidentally removed; or a suture breaks. The muscle then springs backward, no longer restricted by the intermuscular septa and check ligaments, and enters the tunnel through which it penetrates Tenon’s capsule.

Lost muscles may go unrecognized after the completion of surgery. The suture may have been weakened or partially cut with an instrument or passed superficially in the sclera. The knots may have unraveled, or some mishap may have occurred during postoperative adjustment of the operated muscle. Any of these possible causes may go unrecognized at the time it occurs. Postoperatively the signs of a lost muscle become prominent. However, the muscle cannot be declared lost until a careful search is done and an attempt made to grasp it.

Incidence

The frequency of slipped, lost, and disinserted rectus muscles is unknown. However, some perspective regarding the relative incidence of these complications may be gained from my experience with surgically confirmed cases seen over a 20-year span (1977 through 1996) (Table 40–1).

Two patients were listed in both the slipped and lost columns in Table 40–1 who were surgically proved to have a slipped muscle that was lost during an attempt to pull a very tight muscle forward and suture it to the sclera.

Clinical Diagnosis

The slipped muscle, severed or disinserted muscle, and lost muscle share similar clinical features. A postoperative strabismic deviation occurs opposite to the preoperative deviation. Also, theduction produced by the involved muscle is deficient. The vertical dimension of the palpebral fissure is usually larger in the involved eye and enlarges further on attempting duction into the field of action of the involved muscle (Fig. 40–5). Duction into this field is crippled more with a lost muscle than with a slipped and/or severed muscle. Saccadic velocity is similarly reduced according to the degree of limited duction. In time, the direct antagonist of the involved muscle undergoes secondary tightening because it is never fully stretched. This further impedes movement of the eye into the field of action of the involved muscle. As a result, the intraocular pressure should rise as the eye is moved in the field of action of the involved muscle. Moreover, traction testing reveals increased resistance to passively rotating the eye into the field of action of the involved muscle.

Imaging studies, particularly magnetic resonance imaging (MRI), are usually not helpful in differentiating between a slipped, disinserted, and lost muscle. MRI usually reveals a mass of tissue in the area that already is suspected from clinical observation, but it does not precisely localize the insertional edge of the muscle. MRI does not preclude the need for surgical exploration to confirm the diagnosis.

Surgical Diagnosis

The final diagnosis of this ocular motility disorder, characterized by overcorrection combined with limited duction in the field of action of a rectus muscle, is confirmed only

Figure 40–5. A patient with consecutive exotropia after recession of the medial rectus muscles. The large-angle exotropia, widened left palpebral fissure, and limited adduction of the left eye with further widening of the palpebral fissure suggest a clinical diagnosis of either slipped, disinserted, or lost left medial rectus muscle.
during surgery. Once the clinical diagnosis is made and postoperative inflammation has subsided, surgical exploration is indicated to confirm the diagnosis. An exception occurs when the surgeon loses the rectus muscle and, after an unsuccessful search, closes and refers the case to a more experienced surgeon. In this instance, the secondary surgery should be performed with minimal delay.

The best lighting and instrumentation are essential, and a surgeon’s head lamp is helpful. Thought should be given as to how the eye will be examined under anesthesia before surgery. The eye should not be forced into the opposite field of action of the suspect slipped, disinserted, or lost muscle. The resultant traction may be the final force that disrupts attachments of the muscle or its capsule to the sclera or forces the impending lost muscle through its penetration site in Tenon’s capsule.

The surgeon must give thought to selecting the incision that will provide maximal exposure for viewing the previous surgery while minimally disturbing the tissues (Fig. 40–6). First, attention should be directed to the scleral insertion site where the muscle in question was attached. One must avoid positioning the eye or doing the traction test in a manner that puts stress on the tissue overlying the involved muscle. For example, if the medial rectus is believed to have slipped, the surgeon should not begin by placing forces on the eye and doing a thorough traction test. That test should be delayed and, instead, the eye should be abducted just enough to gain access to the medial conjunctival tissue and make the incision of choice. After Tenon’s capsule is incised, a careful attempt is made to visualize first the normal scleral insertion site and then the site of the intended recession.

**SLIPPED MUSCLE**

If dissection confirms a slipped muscle, an empty muscle capsule will be apparent, but only if the index of suspicion is high. The empty capsule will be attached at the scleral site to which it was sutured (Fig. 40–7). Pink tissue will probably be seen attached to sclera. It is usually a friable, wispy scar tissue or fibrotic thickening. A small Steven muscle hook is passed posteriorly under the tissue to prove that the empty capsule is free from the sclera beyond its site of attachment. The small hook is changed to a larger muscle hook, and a test is done gingerly to determine how firmly the capsule is attached to the sclera. Some of the fibrotic tissue overlying the empty muscle capsule may have to be partially dissected before it becomes apparent that the capsule is on the hook. An empty capsule is translucent, allowing the hook to be visible through it, whereas tendon or muscle is opaque so that a hook placed beneath it is not visualized (Fig. 40–8).

Once the empty capsule is identified, dissection continues posteriorly to the penetration site of the slipped rectus muscle through Tenon’s capsule (Fig. 40–9). One should avoid pulling firmly on the empty capsule to try and gain a better view of where the empty capsule leads. Angled toothed forceps can damage the sclera and clutter the small operative field. The best way to gain maximal exposure, as the dissection continues posteriorly, is to place a 6-0 scleral anchor suture as far posteriorly as possible. By pulling on this suture, the globe is brought forward in the orbit and abducted (Fig. 40–10). Now the most crucial stage of the surgery is at hand. The surgeon, using sharp dissection, incises through the sleeve of Tenon’s capsule that attaches to the empty muscle capsule. For the first time the thick, vascular muscle mass begins to appear in the site where it normally penetrates Tenon’s capsule. The muscle end presenting through the small opening in Tenon’s capsule is grasped with a small clamp (Hartmann clamp, Storz E-3915) (Fig. 40–11). Multiple passes of 6-0 synthetic absorbable suture are locked into the clamped muscle end. The muscle is freed from its surrounding attachments to the Tenon’s capsule sleeve tissue and then pulled forward through Tenon’s capsule (Fig. 40–12). Invariably, the muscle is tight because it has not been stretched since the time it slipped in its capsule.

What has been described is the maximal degree of slippage that can occur. Some muscles will slip only a few millimeters within the capsule and fortunately are found far anterior to the Tenon’s capsule penetration site. In this situation, dissection after the empty capsule is found is less daunting. Regardless of the degree of slippage within the
capsule, to be considered a slipped muscle the muscle ends must not be attached to sclera. Itsduction power therefore is rendered through the empty muscle capsule, which is attached to the sclera, and through the muscle’s attachment to Tenon’s capsule.

**DISINSERTED MUSCLE**

Once dissection discloses that the muscle is not attached at the expected scleral insertion site, the surgeon inspects the area looking for where the muscle might be. Unlike the slipped muscle with its friable empty capsule, the disinserted muscle is associated with thick fibrotic tissue attached to the sclera. After the muscle in its capsule is hooked and further dissection frees it from its fibrosed intermuscular tissue and check ligaments, one can note that the tissue on the hook is opaque (similar to Fig. 40–8B), unlike a slipped muscle (see also Figs. 40–8A and 40–3). The tendon or muscle is clearly identified as being attached firmly to sclera. How far from its normal insertion the disinserted muscle gained scleral attachment determines how tight it will be.

In dissecting a disinserted superior rectus, one can expect it to be held forward in the sub-Tenon’s space by its attachments through the intermuscular septum to the capsule of the superior oblique tendon. The tendon should first be identified to separate it from the disinserted superior rectus. Both the disinserted lateral rectus and inferior rectus muscles also are expected to be held forward in the sub-Tenon’s space by their intermuscular septal connections to the inferior oblique.

**LOST MUSCLE**

Unplanned escape of a disinserted muscle during surgery demands the immediate release of all instrumentation and

---

**Figure 40–8.** A, Small hook visible through translucent empty muscle capsule. B, Small hook invisible through opaque muscle/tendon fibers. After disinserting the muscle, inspection while the muscle is suspended on the double-armed preplaced suture ensures that there is no slippage of the muscle/tendon fibers within the capsule.

**Figure 40–9.** Empty capsule of slipped muscle traced posteriorly through its penetration site in Tenon’s capsule leads to the true muscle fibers.

**Figure 40–10.** Anchor suture for retracting globe eliminates need for introducing instrumentation to accomplish excellent exposure of the site where rectus muscle penetrates Tenon’s capsule. (Illustration by PR Mitchell, MD.)
SLIPPED, DISINSERTED OR SEVERED, AND LOST MUSCLES • 535

Fig. 40-11. The muscle end where it penetrates Tenon’s capsule is grasped with a small clamp.

removal of the surgeon’s and assistant’s hands from the operative field. The anesthesiologist is informed of the situation and must make sure that anesthesia is deep. Think! Realize that during deep anesthesia, the atonic disinserted muscle is probably within the sub-Tenon’s space. As long as the muscle does not receive a stimulus to contract, and the surgeon does not desperately jerk the eye with forceps in the opposite direction in hope of exposing the involved muscle, or grab it before it recoils back to the orbit, there is a good chance the muscle will still be anterior to its penetration site through Tenon’s capsule. If the surgeon is not wearing an operative head light, he or she should request one to be brought into the operating room, put it on, and adjust it. The lid speculum is reinserted. All bleeding sites should be touched up with light cautery. The eye should not be rotated with the intent to gain optimal exposure of the escaped rectus, in the hope of visualizing it deep around the surface of the globe. This maneuver might just force the muscle through Tenon’s capsule.

Instead, an attempt is made to visualize the muscle before it has passed posteriorly through Tenon’s capsule. First, the eye is displaced against the opposite orbital wall and retroplaced into the orbit. Firm iris repositor or a Bard-Parker scalpel handle are good instruments with which to retract the tissue away from the globe with one hand and displace the eye against the opposite orbital wall with the other (Fig. 40–13). If this maneuver fails, at least one “all-out” attempt to locate the muscle is necessary. The surgeon should displace the eye opposite to the escaped muscle. Tenon’s capsule is retracted from the globe with a Desmarres lid retractor (size 11 mm wide, Storz E-980), and the area of Tenon’s capsule is inspected where the muscle is expected to penetrate. If still unable to locate the penetration site, one should refrain from incising or lacerating Tenon’s capsule. Extraconal fat prolapsing into the sub-Tenon’s space through rents in the Tenon’s capsule can result in an adherence syndrome.

If the rectus muscle is suspected of having been lost weeks or months before but this has not been proved, the surgeon should proceed directly to obtaining maximal exposure of Tenon’s capsule as just described. However, the integrity of Tenon’s capsule should not be violated in searching for a lost muscle. It is better to declare the rectus muscle lost and proceed with the logical steps for improving alignment and motility.

Surgical Management

The operative methods used to treat slipped muscle, disinserted muscle, and lost muscle differ. Therefore, a surgical

Fig. 40-12. Recovered slipped medial rectus muscle is invariably tight as it is brought out of its penetration site in Tenon’s capsule.

Fig. 40-13. Gaining access to a lost rectus muscle. Intraoperatively, the unexpected detached muscle has just disappeared from the surgeon’s view (upper left). Do not (upper right) pull the Tenon’s capsule forward or (lower left) maximally rotate the eye in the direction opposite to the involved muscle. Instead (lower right), compress the eye against the opposite orbital wall, retroplace it in the orbit, and reflect the loose Tenon’s capsule aside. Pulling the Tenon’s capsule forward and/or strongly rotating the eye in the hope of gaining a better view of the “lost muscle” are prone to cause the detached muscle to penetrate the Tenon’s capsule posteriorly. (Illustration by PR Mitchell, MD.)
diagnosis, or confirmation of a clinical diagnosis, is mandatory before proceeding to surgical management.

**SLIPPED MUSCLE**

The discussion of surgically diagnosing a slipped muscle ended with the muscle being brought forward using a preplaced 6-0 synthetic suture. At this point the problem of a tight slipped muscle is most apparent. Generally, the muscle is too tight to be brought to its original insertion, although that should be the objective. Also, the direct antagonist of the slipped muscle has become tight secondary to limited duction in the field of action of the slipped muscle. In some cases, the direct antagonist is so tight that it must first be recessed before the slipped muscle can be usefully advanced.

Each case is unique. There is no set of rules about how much advancement of the slipped muscle or recession of the antagonist is appropriate. The best approach, I think, consists of two steps in deciding where to attach the slipped muscle. First the eye is held by forceps in the primary position while the slipped muscle is brought forward against the sclera by a preplaced double-armed suture (Fig. 40–14). The sclera is marked where the end of the muscle reaches it, and here it is sutured and tied down with a bow knot. Alignment under anesthesia is appraised by the Krimsky light reflex method. If further correction is deemed necessary, the knot is untied; the sutures with their needles are removed from the sclera; and a new scleral site is chosen for suturing the slipped muscle in place. The Krimsky evaluation then is repeated. When the surgeon is satisfied that the best alignment has been achieved, the bow knot is converted to a square knot and no further adjustments are made (Fig. 40–15).

Care must be exercised when the slipped muscle is sutured to sclera. The tight slipped horizontal rectus muscle should be accurately centered and properly oriented circumferentially. Very slight suprplacement or infraplacement will cause a vertical deviation in the primary position. The tight horizontal rectus, if infraplaced, will depress the eye to produce hypotropia, whereas a suprplaced muscle produces hypertropia. The only way to correct these induced vertical deviations is to center the muscle at a second operation.

This raises concern about using an adjustable suture technique for managing slipped muscles. The muscle is so tight in most cases that it is a challenge, even under general anesthesia, to tie the muscle down to sclera before it slips a bit. I would presume that adjustable surgery using a hang-back suture, without the muscle being firmly sewn to the sclera and centered on its insertion, will allow vertical slipping one way or another and dispose to vertical deviation. Furthermore, at adjustment the muscle will be so tight that it is almost impossible to pull it up any further. If an attempt is made to release the muscle posteriorly to reduce the correction, it would be a real challenge to prevent a greater recoil of the muscle than is intended.

**DISINSERTED MUSCLE**

The disinserted muscle, when confirmed by surgical exploration, is ready to be sutured to the sclera at a new site. Essentially the operation consists of an advancement procedure. How far recessed the disinserted muscle was before reattaching itself to the globe will determine the degree of tightness. This, in turn, helps in deciding how far the muscle needs to be advanced to correct the deviation and improve duction in its field of action. The tighter the muscle, the greater the degree of correction obtained by advancing it.

Limited duction in the disinserted muscle’s field of action, caused by progressive tightening of the direct antagonist, is a secondary problem that increases the angle of deviation. Tightening of the direct antagonist must be reckoned with when formulating a surgical strategy to correct the misalignment. It may have to be recessed so that advancement of the disinserted muscle can manifest its full potential benefit.

**LOST MUSCLE**

After having declared the rectus muscle lost as a result of failure to find it without violating Tenon’s capsule, definitive surgical correction must be decided on. The direct antagonist is undoubtedly tightened, and therefore must be recessed by the maximal amount that still permits full duction in its field of action. The ultimate goal is to ensure orthotropia in the
primary position. This will require a transposition procedure on the neighboring rectus muscles, creating force by attaching either the entire neighboring muscles or at least their nearest divided halves to the normal scleral insertion site of the lost muscle.

A full muscle transposition carries a risk of reducingduction in the muscle’s field of action. In the case of a lost medial rectus, for example, where the entire superior and inferior rectus muscles are transposed to the scleral position of the medial rectus insertion, the vertical rectus muscles would be at a disadvantage when exerting their vertical function. This undesirable outcome is partially prevented by transposing only half of each vertical rectus muscle. Moreover, the vascular supply to the anterior segment is less disrupted. If the entire vertical rectus muscles are transposed (and the antagonist horizontal rectus muscle recessed, while the other is involved as a lost muscle), blood supplied by the anterior ciliary arteries of all four rectus muscles will be eliminated. The anterior ciliary circulation of the direct antagonist could be spared by injecting botulinum toxin into the muscle rather than recessing it. However, if the direct antagonist has undergone tightening, botulinum toxin probably will not weaken the muscle sufficiently. Transposing only half the neighboring rectus muscles, known as the modified Hummelsheim procedure, was introduced as the appropriate procedure for lost rectus muscle by Hill in 1955 (Fig. 40–16).

In performing the Hummelsheim procedure, the surgeon should be aware that the maximal effect in countering the function of the direct antagonist is achieved by attaching the halves of the neighboring rectus muscles at the normal insertion site of the lost muscle. The surgeon might reason that the power of the procedure would be enhanced by transposing the slips posterior to this site. However, the fact is that the tautness of the transposed slips is maximal when they are placed at the normal insertion site. This location offers the maximum possible wraparound distance on the globe. If attached posterior to the insertion line of the lost muscle, the wraparound distance on the globe is reduced, making the transposed slips less taut.

**Prevention**

The best treatment is prevention. Adopting certain precautions when performing muscle surgery can reduce the incidence of these complications.

**SLIPPED MUSCLE**

The slipped muscle complication can be virtually eliminated by:

1. Ensuring that the preplaced suture needle passes at a depth that is in the tendon or muscle fibers, not merely in the capsular tissue encasing the muscle/tendon unit.
2. Directing the needle perpendicularly through the muscle/tendon for the suture locking bites, rather than tangential to the muscle tendon (Fig. 40–17). This ensures that the preplaced suture will include muscle/tendon fibers. Otherwise only capsule may be included in the preplaced suture.
3. Inspecting the disinserted muscle/tendon by holding it up into the operative field by its preplaced suture to check the adequacy of the locking bites while maintaining muscle width and ensuring that the opaque muscle/tendon extends to the preplaced suture (Fig. 40–18).

**DISINSERTED MUSCLE**

The incidence of disinserted muscle caused by pulling away from the intended scleral insertion site can be reduced by passing the needle into the scleral fibers, not just into the superficial scleral tissue. Some surgeons are so fearful of perforating the sclera that they pass the needle too superficially. In performing the resection, the sutures should be passed intrasclerally rather than through the tendon stump left on the sclera after disinsertion. On resumption of muscle contraction after surgery, the suture can easily pull out of fibers in the tendon stump, but not out of the scleral lamellae.
To reduce recoil of the disinserted muscle after surgery, it would be best to have performed minimal dissection of the intermuscular septa on either side of the muscle and its check ligaments. Admittedly, certain muscle procedures require rather complete dissection of these tissues away from the muscle being operated on before disinsertion. For most procedures, however, such as initial recession on any of the rectus muscles except the inferior rectus, essentially no benefit is gained by dissecting the surrounding intermuscular septa or its check ligaments.

Another detail that may diminish the risk of disinsertion involves the type of knot used to tie the suture. Square knots (also called surgeon’s knots) are tighter than granny knots, as any sailor knows. Tying down a muscle is as important as securing a boat to a mooring. All knots tend to loosen with time, so the tighter the knot, the less chance the muscle will pull away from its sutured site in the immediate postoperative period.

Cautery and sutures are not ideal bedfellows. Bringing the two together in a small operative field is risky. Partial thermal damage to the suture may not be recognized intraoperatively, but later the damaged suture may be disrupted.

The severed rectus muscle that results from blindly sweeping the muscle hook (in the hope of engaging the inferior oblique muscle or superior oblique tendon but instead hooking a neighboring rectus muscle) must be avoided by directly visualizing the oblique muscle/tendon. The eroded rectus muscle caused by an encircling element or explant used in retinal surgery will, it is hoped, disappear as retinal surgeons use vitrectomy and internal tamponade to reattach the retina.

Traumatic disinsertion and avulsion of a rectus muscle is one complication over which we have no control. The surgeon can, however, prevent the “pulled-in-two” syndrome. Both the surgeon and assistant must be aware, particularly in the elderly, of how hard the other is pulling on the rectus muscle during dissection.

**LOST MUSCLE**

Preventing a lost muscle entails a number of steps in the surgical dissection routine. For example, one should never depend on an instrument alone to retain control of a disinserted muscle but always have a preplaced suture in the muscle before its disinsertion. The instrument may be defective, or it may be inadvertently released by the assistant or surgeon.

Prevention of a slipped muscle is also an important aspect of preventing a lost muscle. Repairing a slipped muscle that has been far posterior for some time and has become tight carries a high risk of converting it to a lost muscle.

**REFERENCES**

Several minor consequences result from strabismus surgery but only rarely are severe or major complications encountered. The ophthalmic surgeon and patient should discuss the probable postoperative status before surgery is undertaken. For instance, postoperative diplopia occurs predictably after uncomplicated eye muscle surgery. Therefore, postoperative diplopia—expected, understood, and anticipated by the surgeon—should not surprise the patient. Other “complications” perceived by the patient are considered by the surgeon to be usual consequences of strabismus surgery: temporary postoperative conjunctival imperfections, sensitivity to exogenous irritants for weeks or months, and a noticeable site of previous insertion of a now recessed extraocular muscle.

Other problems such as subconjunctival cysts or even operating on the “wrong” muscle(s) may be considered serious complications in that further surgery may be required, although eventual vision and alignment usually are satisfactory. Serious complications of strabismus surgery are those that result in visual loss, require complicated or expensive treatment, or cause prolonged morbidity. Disastrous complications from eye muscle surgery, such as permanent visual loss from retinal detachment or endophthalmitis, do occur. Fortunately, serious complications are rare and usually can be prevented through careful preoperative evaluation and by adhering to accepted and proven surgical techniques and principles.

Minor complications and consequences of strabismus surgery may be characterized as transient postoperative problems that require only reassurance or minor treatment rather than further surgical intervention. These problems do not add significantly to the patient’s inconvenience or expense. It is possible, however, for a neglected minor complication to become a major problem. Postoperative nausea and vomiting, for example, if neglected or inadequately treated, can lead to electrolyte imbalance, dehydration, cardiovascular collapse, and major systemic complications, particularly in patients who are very young or elderly.

Minor Complications

POSTOPERATIVE NAUSEA AND VOMITING

Incidence

Owing to technologic improvements in monitoring equipment and pharmacologic advances, serious complications of general anesthesia have become rare. Reported mortality rates are in the order of 1 in 100,000. As the number and complexity of surgical procedures performed on an outpatient basis have increased, so has the demand for anesthesia causing low morbidity and minimal residual postoperative drug effects. Prompt and safe release of patients from outpatient surgery is facilitated when postoperative morbidity is infrequent. Nausea and vomiting remain troublesome aftereffects of anesthesia given for strabismus surgery.

The incidence of postoperative nausea and/or vomiting after eye muscle surgery has been reported as 17% to 88%. This variability is due in part to individual patient risk factors. Women are three times more likely than men to have postoperative vomiting. The difference may be hormonally mediated because postoperative vomiting is not noted with the same frequency in prepubertal or elderly female patient populations and is most marked during the luteal phase of the menstrual cycle. Children are at least twice as likely to have nausea and vomiting after general anesthesia as adults, whereas infants (birth to 12 months of age) are at low risk. Among patients undergoing strabismus surgery, children are at the highest risk of postoperative vomiting; the complication is noted in up to 88% of cases. The incidence of vomiting after all types of surgery requiring general anesthesia is 20% in children 1 to 5 years of age and peaks in preadolescence (11–14 years) at 51%. Other patient-dependent risk factors for postoperative nausea and vomiting include obesity, a history of motion sickness, high preoperative anxiety, and gastroparesis (e.g., due to diabetes, chronic cholecystitis, or pregnancy).
Anesthesia-related factors predisposing to vomiting include stomach distention during mask ventilation and a longer duration of anesthesia. Anesthetic agents implicated in postoperative nausea and vomiting include opioids given for analgesia as well as inhalational anesthetics used to induce and maintain anesthesia. Older opioids are well known to cause nausea, and those in current use such as fentanyl have been shown to increase postoperative vomiting. Intravenous propofol, now a mainstay of anesthesia for outpatient surgery, is less emetogenic and may even have antiemetic properties. Current inhalational anesthetics such as halothane, enflurane, and isoflurane are emetogenic, although less so than older agents such as cyclopropane. Nitrous oxide probably has some emetogenic activity, but recent studies have failed to show increased nausea and vomiting when it is used along with halogenated inhalational agents.

Surgical and postoperative factors may also cause nausea and vomiting. Certain types of surgery more commonly cause vomiting; strabismus surgery is notoriously emetogenic. The more eye muscles are included in the surgery, the more likely is postoperative nausea and vomiting, independent of the duration of surgery. Postoperative factors inducing emesis include pain, hypovolemia, early ambulation, and oral intake in the first few postoperative hours.

Prospective studies to determine the incidence of postoperative nausea and vomiting in patients receiving either general or retrobulbar anesthesia showed a slightly (but statistically insignificant) lower incidence in patients given retrobulbar anesthesia. To our knowledge, studies comparing the incidence of nausea and vomiting seen with topical anesthesia to that occurring with general anesthesia in eye muscle surgery do not exist. In the experience of one of the authors (FDE), nausea and vomiting after strabismus surgery using topical anesthesia alone is exceedingly rare. Whether this lack of side effects is due to more delicate handling of the tissues, required when using topical anesthesia, or results from the absence of emetogenic anesthetics is not clear.

Prevention

Preventing postoperative nausea and vomiting is important for several reasons. These events occasionally result in serious complications such as aspiration, atelectasis, pneumonia, dehydration, electrolyte imbalance, cardiovascular collapse, and major systemic problems. Bleeding and wound disruption induced by retching or vomiting are possible but unlikely after strabismus surgery. More commonly, patient discomfort and delayed discharge from the hospital are the only sequelae. Although physicians may be desensitized to the significance of postoperative nausea and vomiting owing to its relative low morbidity, most patients who are dissatisfied with their ambulatory surgical experience cite postoperative nausea and vomiting as the reason. Routine pharmacologic prophylaxis may not be indicated, however, because of side effects from the drugs used. Special precautions are in order for patients at high risk.

Nonpharmacologic preventive measures may be equally important. Active efforts to lower a patient’s anxiety level, avoid inefficient mask ventilation, and reduce trauma from intubation can lessen the risk of postoperative nausea and vomiting. Appropriate communication between the surgeon and anesthesiologist can minimize unnecessary anesthesia time at the beginning and end of each procedure. Careful attention to the patient’s volume status optimizes the benefits of perioperative intravenous rehydration. Transportation of the patient should be restricted if possible in the early postoperative period. Postoperative oral intake should be instituted gradually.

Treatment

The same agents are used to prevent and treat postoperative nausea and vomiting. Antiemetic medications block at least one of the receptor types in the chemoreceptor trigger zone of the medulla: dopaminergic (D2), histamine (H1), serotonin (5HT3), and muscarinic cholinergic. Phenothiazines, butyrophenones, antihistamines, and anticholinergics all are commonly used medications. These agents, with few exceptions, produce varying degrees of side effects.

Prochlorperazine is a widely used phenothiazine that may cause sedation and extrapyramidal signs. Metoclopramide, a benzamide, acts centrally to block dopaminergic receptors but has additional direct effects on the intestinal tract, increasing gastric emptying time and decreasing gastroesophageal reflux. Intravenous metoclopramide is known to cause hypotension, sedation, and extrapyramidal reactions, especially in children. A commonly used butyrophenone, droperidol, blocks dopaminergic receptors. Even in the higher dose range this drug is only partially effective in children undergoing strabismus surgery and causes prolonged sedation and extrapyramidal signs in a significant proportion of patients. Cyclazine, an antihistamine and effective antiemetic, causes somnolence and dry mouth but no extrapyramidal effects. Atropine, an anticholinergic, has antiemetic action and is commonly used in combination with a cholinesterase inhibitor (neostigmine) during reversal of anesthesia. Finally, ondansetron, a serotonin (5HT3) receptor antagonist, is effective in preventing and treating nausea without producing sedation, extrapyramidal effects, or cardiovascular changes.

Prolonged or Excessive Ocular Irritation or Pain

Most strabismus surgery results in irritation and minor discomfort confined to the eye or the periorbital area. Pain in excess of what is expected must be investigated because of the possibility of endophthalmitis. However, the usual cause of such discomfort is an eyelash, suture, dellen formation, filamentary keratitis, or cellulitis. Strabismus surgeons should warn parents and patients that increasing inflammation and discomfort are reasons to contact the surgeon. Generally, the operated eye is more swollen and red the first morning after surgery than at any other time. Postoperative discomfort tends to be irritation and aggravation rather than a deeper aching pain that becomes progressively more intense and signifies a more serious problem.

Wide variations exist in recommendations for the timing of postoperative visits. Some surgeons see the patient on the first postoperative day, others in 2 or 3 days, and some a week after surgery. The rapidity with which endophthalmitis develops is more likely due to the virulence of the organism and to the host’s susceptibility rather than being predictable. The incidence of endophthalmitis is so low that any scheme
Patients who have undergone eye muscle surgery are given standard over-the-counter analgesic medications for minor discomfort. Narcotics or prescription drugs seldom are required for pain relief. Local treatment with cool compresses periodically applied to the periorbital area suffices in the immediate postoperative period.

Topical antibiotics and antibiotic/corticosteroid combinations generally are prescribed for the first postoperative week. Some surgeons favor routine systemic or topical antibiotics preoperatively; some administer antibiotics by intraoperative subconjunctival injection; others use topical and/or systemic antibiotics postoperatively; and a few use no antibiotics at all. Patients with known risk factors should be treated appropriately.

VISIBLE LINE OF PREVIOUS MUSCLE INSERTION

When a muscle is recessed, the former site of the original muscle insertion becomes visible through the conjunctiva. Thinner sclera located behind a normal muscle insertion contrasts sharply with the surrounding thicker sclera. This contrast does not change significantly in its appearance over time after eye muscle surgery, nor does it occur at the site of surgical attachment of a muscle when that muscle is moved again. The site of previous muscle insertion can easily be seen when the patient looks away from the recessed muscle. The patient seldom sees this line, but an observant parent often will ask about the “scar” (Fig. 41-1). Reassurance and explanation are all that is required.

REDUNDANT OR IRREGULAR CONJUNCTIVA AT THE SITE OF SURGERY

Conjunctival closure should be meticulous with accurate apposition of the tissues. One of the more difficult tasks for the beginning ophthalmic surgeon is to distinguish clearly between conjunctiva and Tenon’s capsule at the end of an operation. Tenon’s capsule (in contrast to conjunctiva) becomes edematous and becomes more white in color when irrigated with balanced salt solution. Marking sutures at the edge of the conjunctival incision may be helpful to the novice surgeon. Failure to replace conjunctiva accurately can result in an unattractive appearance, externalization of Tenon’s capsule, dellen formation, and corneal irritation. Conjunctival incisions should be made with care and should not extend through the plica semilunaris.

Resection and advancement of a muscle may result in excessive or redundant conjunctival tissue in the area of the advancement. Time will improve this situation but may not eliminate the redundant appearance. There is no substitute for meticulous wound closure. Redundant or excessive conjunctiva may be trimmed appropriately. Care must be taken to avoid creating a restriction by excessively短ening the conjunctiva, leading to a leash or a reverse-leash effect (Fig. 41-2).

Dellen formation

Dellen (pits or depressions) develop when the ocular surface is inadequately lubricated by the distribution of tears across the cornea. After strabismus surgery this may result from elevation of the pericorneal tissues, conjunctiva, and Tenon’s capsule. The patient will complain of more pain and irritation than expected. Examination will reveal a depression in the cornea where wetting does not occur. Fluorescein will pool in the area and may stain the corneal surface if epithelial cells are damaged. This process may predispose the patient to corneal infection and ulceration.

Careful conjunctival closure should prevent most cases of dellen formation after strabismus surgery. Occasionally, postoperative edema will produce enough conjunctival swelling to induce dellen formation. If dellen develop, treatment should be directed toward removing the cause. The ocular surface should be lubricated adequately. Lubricants, temporary patching, and removal of the causative agent should lead to prompt resolution.

INCREASED VASCULARITY OF THE CONJUNCTIVA

Increased vascularity of the conjunctiva may be expected for 6 to 12 months after extraocular muscle surgery. This
erythematous appearance is not constant but will recur with exposure to irritants such as chlorine in a swimming pool, smoke, and allergens. The involved eye may develop more erythema at the operative site than the fellow eye when exposed to irritants several months after surgery. This is less a problem for children than for adults who are concerned about their appearance. A history of multiple prior operations also increases the probability of postoperative conjunctival injection with each subsequent operation. Adults whose work or social obligations make these matters important should be prepared to deal with this for at least a few weeks postoperatively (Fig. 41–3).

Reassurance and explanation are comforting to the patient who sometimes thinks the eye has been permanently sensitized by surgery. Topical vasoconstrictor agents may be helpful if used appropriately. Topical corticosteroids, given for 1 or 2 weeks postoperatively, reduce the immediate inflammatory response but have no effect on increased sensitivity over the longer term.

**FILAMENTARY KERATITIS**

Filamentary keratitis has many causes; it has been reported in association with dry eyes, topical ocular medications, and corneal sutures such as those used in cataract surgery. Other cases have occurred after retrobulbar botulinum injections and strabismus surgery. After strabismus surgery, the altered tear film occasionally leads to filament formation. Meticulous conjunctival closure and the appropriate use of topical medications such as artificial tears and lubricants will prevent or alleviate most cases of filamentary keratitis. Some patients require debridement.

**ALLERGIC REACTIONS TO TOPICAL OR SYSTEMIC MEDICATIONS**

Any topical or systemic medication has the potential to induce an allergic reaction (Fig. 41–4). Allergic ocular reactions are manifested by itching, a clear stringy discharge, and a papillary conjunctival reaction (Fig. 41–5). The conjunctiva appears pale and edematous.

Any medication in current use should be discontinued. Substitutes may be considered but should be added one at a time; no medication at all may be the wisest choice. Additional therapy may include cold compresses, artificial tears without preservatives, topical and/or oral antihistamines, vasoconstrictors, and mast cell inhibitors. Topical corticosteroids may be required for a short time.

**Serious Complications**

**SYSTEMIC COMPLICATION: STEVENS-JOHNSON SYNDROME**

Stevens-Johnson syndrome is a rare, chronic and recurrent, life-threatening disorder of mucosal and epidermal structures. It appears to be a part of a spectrum that includes erythema exudativum multiforme major and toxic epidermal necrolysis. It is not quite clear how these disorders may be related, because different patterns of histologic change are observed. Furthermore, overlap occurs in their clinical classification. Ocular involvement includes symblepharon formation, entropion, trichiasis, keratinization, corneal ulceration, and blindness. Rarely, orbital cysts have been reported as late complications.

This uncommon but serious complication is not a result...
of surgery itself but of the drugs used to prevent more common complications such as infection, pain, or discomfort due to the surgery. Medications reported to cause Stevens-Johnson syndrome are acetylsalicylic acid, sulfonamides, erythromycin, cotrimoxazole, valproic acid, paracetamol, and cephalosporins. Topical sulfonamides remain a popular antibacterial medication, and sulfonamide derivatives such as acetazolamide (Diamox) frequently are prescribed by ophthalmologists. Because this disorder may recur, an adequate history of prior drug reactions and/or Stevens-Johnson syndrome should be obtained before prescribing these drugs. A history of recent infection (i.e., herpes simplex) is frequently obtained from patients who have developed Stevens-Johnson syndrome.

Systemic complications and the potential for them to occur usually require that the patient be managed in a hospital environment. If ocular involvement is mild, treatment with artificial tears or topical corticosteroids may be all that is required. If symblepharon formation is significant, a symblepharon ring and frequent lysis of adhesions may be needed (Fig. 41–6). If keratinization, entropion formation, trichiasis, or corneal ulceration develop, ectropion surgery, mucous membrane grafts, or corneal transplantation might be necessary. The timing of surgery is important, because intervention too early in the course of the disease may worsen final scarring.

ANTERIOR SEGMENT COMPLICATIONS

Anterior Segment Ischemia

This complication is discussed in Chapter 39.

Subconjunctival Cyst Formation

Subconjunctival cysts have been reported to occur as long as 35 years after strabismus surgery. In some cases the cyst has been described as sudoriferous. In others, it reportedly arises between the anterior edge of the muscle and the site to which the muscle has been sutured. In these cases, the muscle attaches to the posterior wall of the cyst and not to the sclera. The precise origin of the cysts remains unclear, but they generally are attributed to faulty surgical technique.

MUSCLE AND STRABISMUS COMPLICATIONS

Slipped or Lost Muscles

Operations on the “Wrong” Muscle

Wrong patient, wrong eye, and wrong muscle operations all have occurred. Extraocular muscles are not always located at the expected anatomic positions. Eye muscles may be absent congenitally. The globe may be rotated into an abnormal position (usually but not invariably because of congenital craniofacial abnormalities or trauma), or the surgeon may be confused by local anatomic peculiarities. When under tension, a strip of the superior rectus (SR) or a fold in Tenon’s capsule may be mistaken for the superior...
oblique (SO) tendon. Both the inferior rectus (IR) muscle and the lateral rectus (LR) muscle have been misidentified as the inferior oblique (IO). A portion of the levator muscle mistakenly has been attached to the globe in place of the SR muscle.

It is difficult to say why these errors take place, but they seem to occur more often when surgery is performed by neophyte surgeons or by the occasional strabismus surgeon. Such mistakes should not occur if surgery is carefully planned and carried out. Operating on the correct muscle is the surgeon’s responsibility. Assessing previous procedures is important, particularly if muscles have been placed in other than the usual positions. Stay sutures, traction tests, identification of the muscles through the conjunctiva before making an incision, and frequent intraoperative checks should prevent errors.

In an eye with no history of previous surgery, ciliary vessels are identifiable through the conjunctiva and may serve as a guide to the location of muscles. These vessels are a virtual road map to the anterior borders of the rectus muscles. The single ciliary vessel of the LR muscle usually is located in the lower third of the muscle. The plica semilunaris is a reliable landmark to the location of the medial rectus. Both the SO and the IO muscles are located inferior to their respective vertical rectus muscles. If the IR and LR muscles are identified and secured on muscle hooks or are adherent, the IO muscle can easily be found coursing between them. The SO tendon may be found at its insertion by securing and observing the temporal border of the SR muscle. Once located, the SO tendon may be followed beneath the SR muscle, where it can be resecured, if desired, on the nasal side of the SR muscle. Occasionally, the SO tendon is absent or is inserted anomalously.  

Muscle Rupture

Iatrogenic rupture of an extraocular muscle may occur at the insertion of a muscle or along its belly. Although a structurally normal muscle can rupture during surgery if excessive force is applied, rupture is more likely when muscle fibers are pathologically altered. A muscle that has undergone previous surgery may be weakly attached to the sclera. An intrinsically diseased muscle is more vulnerable to forceful intraoperative manipulation, as is often required in patients with infiltrative myopathy secondary to Graves disease, for example. Increased age may raise the risk of extraocular muscle rupture during surgery. Other risk factors for intraoperative muscle rupture include prior muscle trauma, orbital myositis, orbital fibrosis syndrome, botulimum toxin injection, long-standing paresis, a past history of orbital infection, and muscle entrapment by an orbital fracture.

The force transferred to an extraocular muscle through a muscle hook held by the surgeon may be well beyond the physiologic range that the muscle can withstand. An inattentive or inexperienced surgeon or assistant easily can generate enough tension on a muscle to cause rupture. The strain placed on a muscle by a muscle hook or traction suture should be the minimum needed to position the eye appropriately. Continuous awareness of the force applied to an extraocular muscle is essential, even though the surgeon may focus temporarily on another site in the surgical field.

More importantly, if any of the risk factors just mentioned exist, the surgeon should take particular care with intraoperative maneuvers that place traction on an extraocular muscle.

Repair of a ruptured muscle is not unlike repairing a slipped muscle (see Chapter 40). If a previously operated muscle is ruptured, its repair may be more difficult, owing to the disruption of normal anatomic landmarks such as check ligaments and intermuscular membrane. Imaging (computed tomography or magnetic resonance) may be helpful. If unsatisfactory attachment at the new muscle insertion is noted during surgery, it should be repaired at the time it is recognized.

Inferior Oblique Inclusion

Intermuscular membrane extends from the inferior edge and underbelly of the LR to the insertional head of the IO. Inclusion of the IO in the insertion of the LR can occur after surgery on the LR if these membranous connections are not severed. This complication reportedly has been found in about one third of cases of LR recession or resection when reoperating for other reasons. Resection of the LR is more likely to result in IO inclusion than recession. The resulting motility abnormality most often is limited elevation in abduction, but a variable restrictive disturbance may occur. The diagnosis of IO inclusion often depends on the clinician’s level of suspicion and investigation (Fig. 41–9).

IO inclusion is to be differentiated from IO adherence, a result of breaches posterior Tenon’s capsule during an IO weakening procedure. In these cases, dense scarring and fat adherence in the inferotemporal quadrant cause hypotropia of the involved eye in the primary position and severely limited elevation in adduction (Fig. 41–10).

Adequate visualization with avoidance of blind, posterior sweeps with a muscle hook when isolating the LR will prevent inadvertent hooking of the IO. During dissection of the intermuscular membrane surrounding the LR, care must be taken to sever the connections between this muscle and the IO. Good exposure and a thorough understanding of IO anatomy are essential. During LR recession or resection, a final inspection of the underside of the disinserted muscle to identify connections to the IO is prudent before reattaching the LR to the sclera.

Figure 41–9. Inferior oblique inclusion beneath right lateral rectus.
Figure 41-10. Inferior oblique adherence simulating Brown syndrome. A, Patient looking to the right and up shows limited elevation in adduction in the left eye. B, In looking to the right and down, mild superior oblique overaction is observed.

Treatment of IO inclusion involves surgical reexploration with freeing of the oblique from the inferior border of the LR. Unfortunately, adhesions commonly recur in this area. Injections of corticosteroid into the area of adherence at the time of surgery may be helpful.

Iatrogenic Superior Oblique Palsy

Superior oblique weakening procedures usually are done for primary SO overaction. A-pattern strabismus with SO overaction, or Brown syndrome, and they are effective for these conditions. Reported complications of such procedures include inadvertent transection of the SR. Not including the posterior fibers of the oblique tendon during attempted tenotomy will result in failure of the procedure. Intractable diplopa may result from these procedures, especially if they are performed on visually mature patients who have excellent stereoscopic capabilities.

The most common complication of a successfully performed SO weakening procedure is iatrogenic SO palsy. Tenectomy, tenotomy, recession, and silicone spacers all are used to weaken the SO. Any of these procedures may produce an SO palsy.

Despite efforts to refine SO surgery, the surgical results remain less predictable than with other types of strabismus surgery. Furthermore, the risk-to-benefit ratio often is higher than from other strabismus procedures. Patients with Brown syndrome and primary SO overaction often are bifixators. Surgical overcorrection in younger patients can induce loss of fusion and in older patients may cause symptomatic complications such as vertical and torsional diplopia and torticollis. Surgical candidates must be selected judiciously, and the procedure performed carefully to avoid inadvertently producing a symptomatic SO palsy. Tenectomy of the SO involves removing a portion of the SO tendon either temporal or nasal to the SR muscle. Nasal weakening is the most effective weakening procedure, and the most likely to cause an SO palsy. Because of the high probability of overcorrection, tenectomy has largely been abandoned for fusing patients and other procedures substituted.

Nasal tenotomy with preservation of the tendon "sheath" is less likely to cause palsy of the SO. In theory, the preserved tendon "sheath" maintains a physical connection between the cut ends of the tendon so that some muscle tension can be transferred to the distal cut end and therefore to the globe postoperatively. The procedure initially was performed through a superonasal conjunctival incision. Disruption of intermuscular membrane nasal to the SR, with subsequent fat adherence, is a potential pitfall of this approach. Accessing the nasal tendon through a superotemporal conjunctival incision better preserves fascial planes and is less likely to result in scarring and fat adherence. Some titration of surgical effect may be possible: the closer to the trochlea the tendon is severed, the more the SO will be weakened.

Several surgical procedures exist that less readily cause postoperative palsy of the SO muscle. Because of the attachments of the SO to the underbelly of the SR, a complete tenotomy performed temporal to the SR preserves significant SO function. Some authors have espoused tenotomy only of the posterior part of the SO insertion to further guard against postoperative SO palsy, reasoning that this procedure selectively weakens the muscle's inductive action while retaining incyclotorsion. SO recession has been used as a graded weakening procedure. Placement of a silicone tendon spacer may provide a more controlled means of treating SO overaction and Brown syndrome than nasal tenotomy, but paretic overcorrection may follow this procedure. In patients who are undergoing horizontal rectus surgery, vertically shifting the horizontal muscles is a relatively safe way to correct A- and V-pattern strabismus with little or no oblique dysfunction.

Postoperative SO palsy may take as long as 24 months to become symptomatic or clinically significant. This delay may be due to changes in the antagonist IO, which occur with long-standing SO overaction or Brown syndrome. Reexploration of the SO after a tenotomy is fraught with difficulty and may lead to orbital fat adherence and scarring. Postoperative bilateral SO underaction with a V pattern and excyclotorsion may necessitate reuniting the cut ends of the tendon but is a potentially difficult task. Iatrogenic SO palsy is best dealt with by surgery on other extraocular muscles. Because of the high rate of overcorrection with SO weakening, some surgeons perform ipsilateral IO myectomy simultaneously. However, IO weakening may not relieve a vertical deviation in downgaze after an SO tenotomy because the SO has been significantly and permanently weakened in...
that gaze field. If the induced vertical deviation is greatest in downgaze, a contralateral IR recession is often necessary.

Iatrogenic Brown Syndrome

Brown syndrome is a limitation of elevation in adduction caused by a congenitally anomalous SO tendon complex. The term Brown syndrome often is broadly applied to a myriad of disorders that cause a Brown syndrome–like motility pattern. Such disorders, better termed simulated Brown syndrome, have been reported from a wide variety of causes. They include peribulbar injection,5–9 glaucoma drainage implants, sinus surgery,10 blepharoplasty,10 orbital trauma,14 autoimmune inflammatory conditions, and tumors in the region of the trochlea.15 This discussion is limited to acquired, simulated Brown syndrome occurring secondary to procedures for SO palsy.

Extraocular muscle surgery cannot restore normal function to a paretic muscle. Often, the best achievable result is obtained by shifting the globe’s reduced range of motion into a less symptomatic or a more useful position. Thus, in treating SO palsy, there may be a trade-off between strengthening the SO and creating a restrictive limitation to elevation of the eye in adduction. Of the various techniques used to strengthen a weak SO, an SO tuck will most readily cause simulated Brown syndrome; in fact, some degree of Brown syndrome is inevitable. The surgeon must decide what degree of limited elevation is acceptable while trying to achieve the desired amount of SO strengthening.1,8

Intraoperative traction testing of the SO can differentiate a normal from a lax tendon. If a normal tendon is detected, one should avoid tucking that tendon, because a significant Brown syndrome likely will result. Procedures other than tendon tucks that are less likely to produce a postoperative Brown syndrome have been used. Advancement of the anterior part of the distal SO tendon (Harada-Ito procedure) corrects exotropia with little effect on vertical deviation and is unlikely to cause Brown syndrome.5 Anterolateral transposition of the entire tendon has been recommended as a means of strengthening the SO without causing restriction of elevation.5

If a tucked tendon creates a significant Brown syndrome, the tuck must be taken down. This is not always a simple matter, because scar tissue may form and a residual Brown syndrome will persist after the tuck is released. In some cases the entire area of the tuck must be excised (re-creating the original lax SO tendon). Secondary procedures such as recession of the contralateral IR muscle must then be considered.

**LID COMPLICATIONS**

**Blepharoptosis**

Minor changes in lid position are common after strabismus surgery (see later), and, occasionally, significant ptosis of the upper lid is encountered. This may result from direct surgical trauma to the levator muscle or from injury to the nerve of the levator muscle. Blepharoptosis may result from postoperative eyelid edema with resultant thinning or dehiscence of fibers of the levator aponeurosis. This particular complication may be more common after surgery performed beneath the upper lid, such as SO and SR procedures.

Extraocular muscle procedures on the superior aspect of the globe must be confined to the tissues immediately adjacent to the globe. The surgeon must not “wander” up into the lid tissues, or the levator will be encountered and may be weakened in the process.

Surgical procedures such as posterior fixation sutures require extensive dissection and exposure of large areas of extraocular muscles. When such procedures are performed on the SR muscle the surgeon must be aware of the usual anatomic site of the nerve supply to the levator muscle. This nerve traverses the SR muscle on its way to the levator. Its location should be at least 26 mm from the insertion of the SR muscle, and it should not be injured by adequate exposure for SR posterior fixation extraocular muscle surgery. Placing a muscle hook too far posteriorly along the superior aspect of the SR muscle and excessive traction on the levator muscle with curved or bent retractors used to gain exposure in the posterior orbit may disrupt this nerve supply.

Injury to the levator muscle usually resolves in time. Postoperative edema or sutures beneath the lid can masquerade as levator injury but also will improve with time. Reoperation on an injured levator muscle is difficult in the immediate postinjury period. Three months after the injury, however, a damaged levator muscle may be repaired. If the damage cannot be repaired primarily (i.e., dehiscence repair), levator resection or frontalis suspension of the lid might be required.

**Lid Fissure Anomalies**

Examples of conditions in which fissure and lid abnormalities and asymmetries are seen before surgery are summarized in Table 41–1. Postoperative conditions associated with lid fissure anomalies (Fig. 41–11) are listed in Table 41–2.

In a retrospective analysis of the frequency of lid-level change after strabismus surgery, 21 of 23 patients (91%) undergoing SR recession and 18 of 19 patients (94%) having IR recession had lid retraction as a result of the surgery.54 Furthermore, all 7 patients had lower-lid ptosis (advancement or elevation of the lower lid) after IR resection. Strict criteria were used by these authors to judge lid fissure changes; any change in lid position greater than 0.1 mm was counted. The actual clinical significance of these changes is much less. Nevertheless, blepharoptosis or lid retraction after surgery on vertical rectus muscles is a common occurrence, and the ophthalmologist should take steps to avoid it.

**Table 41–1. Preoperative Lid and Fissure Abnormalities and Asymmetries**

<table>
<thead>
<tr>
<th>Preoperative Condition</th>
<th>Lid Anomaly</th>
</tr>
</thead>
<tbody>
<tr>
<td>Duane syndrome</td>
<td>Narrowing on adduction</td>
</tr>
<tr>
<td>Thyroid eye disease</td>
<td>Lid retraction, ptosis</td>
</tr>
<tr>
<td>Craniofacial syndrome</td>
<td>Mongoloid and antimongoloid fissures; ptosis</td>
</tr>
<tr>
<td>Paresis with secondary deviation</td>
<td>Widening</td>
</tr>
<tr>
<td>Third nerve palsy with and without aberrant regeneration</td>
<td>Narrowing secondary to ptosis; widening secondary to aberrant regeneration</td>
</tr>
</tbody>
</table>
The anatomy of the upper and lower lid retractors and their relationship to the SR and IR muscles explain lid retraction or ptosis complicating vertical rectus surgery. In the lower lid, the capsulopalpebral fascia arises from the underside of the IR about 5 mm posterior to the rectus insertion as a collection of fibrous tissue. The capsulopalpebral head then splits to surround the IO muscle and once again condenses anterior to this muscle to form the lower eyelid retractors. These fan out to insert into the inferior conjunctival cul-de-sac, the inferior edge of the lower tarsal plate, and the eyelid skin through the orbicularis layer. Because of this arrangement, IR surgery may have a direct and proportional effect on the postoperative lower lid position. A connection between the levator palpebrae superioris and the SR muscle is less substantial; they consist of weak fibrous strands that originate from Whitnall’s ligament, extend through thelevator, and attach to the anterior part of the SR muscle. Further posteriorly the two muscle sheaths are anatomically distinct and not interconnected. Theoretically, SR muscle surgery should affect eyelid position less than IR muscle surgery; in fact, it should have little, if any, effect on upper-lid position. However, some degree of lid retraction occurs commonly with SR recession, although the amount of lid retraction is not related to the size of the recession.

Patients with thyroid-related eye disease who undergo strabismus surgery require special consideration with respect to lid fissure changes. These patients are at high risk for tear-film abnormalities and corneal exposure problems. Lid retraction is often present before surgery; it can produce a widened palpebral fissure or may be secondary to coexisting proptosis. Orbital adhesions and fullness may augment the usual effects of eye muscle surgery on lid position. Hypotropia, which is usually treated by an IR recession, may mask preexisting lower lid retraction. Detailed preoperative patient counseling and operative planning are necessary when undertaking strabismus surgery on these patients (Fig. 41–12).

Although eyelid position is most readily affected by IR muscle surgery, lid fissure anomalies also can occur after horizontal rectus surgery, usually because of globe retraction or proptosis secondary to changes in extraocular muscle mechanics. MR or LR recessions may induce widening of the palpebral fissure. This effect is particularly pronounced after recessing both horizontal rectus muscles, as in a four-muscle recession for nystagmus. On the other hand, large resections may produce retraction of the globe and, consequently, narrowing of the palpebral fissure of the operated eye.

Several operative techniques have been described to prevent lower lid retraction after IR recession. Severing the check ligaments and fascial connections of the vertical rectus muscles as far posteriorly as the vortex veins has been recommended to avoid lid-position changes. However, extensive dissection did not eliminate lower lid retraction in a prospective study of nine patients undergoing IR recession. Other authors have suggested advancing and suturing the lower lid retractors to the IR muscle after recessing that muscle. A technique of IR muscle recession, using adjustable sutures to suspend both the IR muscle and the capsulopalpebral head from the IR insertion, has been described. After the IR is adjusted and secured, the second adjustable suture is used to set the level of the lower lid.

Lysis of the infratarsal portion of the capsulopalpebral fascia at the same time as IR recession has been suggested to avoid lower lid retraction. In addition to these methods, some surgeons elect to use a Frost suture (a traction suture...
placed through the gray line of the lower lid and taped to the forehead in the early postoperative period). No technique has proved successful if there is significant propotis.

In contrast to lid retraction after IR recession, little is known of how to prevent lid position changes after other types of strabismus surgery. Lower-lid ptosis and flattening of the lid contour may follow resection of the IR despite attempts to separate fascial connections between the capsulopalpebral head and the IR sheath.53 No reliable method of prevention has been reported. Likewise, information on preventing lid-level changes after SR surgery is scarce. Most reports deal with after-the-fact surgical treatment of such complications.

The best attempts to prevent unwanted lid-fissure changes after strabismus surgery may minimize, but will not eliminate, these changes. Even with advancement and suturing of the capsulopalpebral head, patients undergoing large IR recessions had up to 2.5 mm of lower lid retraction.36 And, although adjustable techniques yielded good results in a nonthyroid patient population, these probably would not be as effective as the larger recessions patients with Graves ophthalmopathy often require or if a significant degree of propotis is present.53

Fortunately, a number of surgical techniques exist to correct undesirable postoperative lid changes. Methods of treating eyelid retraction include lid retractors resection with or without a spacer such as sclera, hard palate mucosa, fascia lata, or synthetic material.46 Subtarsal lid retractor lysis also is effective.46 Horizontal lid tightening (e.g., with a lateral tarsal strip) commonly is performed in combination with one of these procedures.19 Müllercurectomy can reduce upper-lid retraction of 2 mm or less. Larger amounts of upper lid retraction may respond to temporalis muscle transfer, lid weighting with gold, levator recession, or levator extirpation with frontalis upper lid suspension.19, 24, 76 If part of the lid retraction is thought to be due to a hyperthyroid state, surgical intervention should be delayed. Normalization of thyroid hormone levels may relieve lid retraction in certain cases.36

POSTERIOR SEGMENT COMPLICATION: GLOBE PERFORATION

Inadvertent perforation of the globe during eye muscle surgery seldom is followed by serious consequences. It rarely is clinically significant, meaning that no treatment is required and adverse effects are seldom seen. Nevertheless, when visual loss occurs after eye muscle surgery, previously unrecognized perforation of the globe usually has occurred.

The true incidence of inadvertent perforation of the globe during strabismus surgery is unknown but probably is higher than is reported in surveys of individual physicians. The incidence has been reported to be 2.8% on a per-case basis and 1% on a per-muscle basis.11 Other sources note that the incidence of perforation has apparently decreased from a range of 8% to 12% to 1% or less.11 This decrease is attributed to better instrumentation.43 Recent surveys of pediatric ophthalmologists in the United States and Canada suggest that globe perforation may be recognized in slightly more than 1 in 1000 patients undergoing eye muscle surgery. Infrequent serious consequences of perforation include visual loss.66 Others have reported endophthalmitis, but with a good final visual outcome.77

Controlled passage of a spatula needle through superficial scleral fibers is the only way to prevent inadvertent globe perforation. These fibers are strong enough to hold the muscle in position. The muscle should be pulled to the entrance site of the needle tunnel into the sclera and the sutures tied just tightly enough to hold the muscle in apposition to this entry site without compressing scleral fibers. If sutures are tied too tightly (the suture attachment to the muscle should be tight), necrosis of the roof of the scleral tunnel may occur, producing delayed slippage of the muscle. Deep passage of the needle should be avoided, but, if it occurs, inadvertent retinal perforation should be ruled out before the conclusion of the surgical procedure by indirect ophthalmoscopy with the pupil dilated.

Patients with thin sclerae, such as those having multiple operations, retinal detachment surgery, Marfan syndrome, or high myopia, can be managed by avoiding penetration of the sclera with the suture needles. The so-called hang-back or hang-loose techniques have been shown to be safe and to yield predictable results.8, 9, 81, 63

Recognition of a perforation is important not so much to treat it locally as to determine the need for intravenous or oral systemic antibiotics if this is not part of the surgeon’s routine. The relatively high number of perforations reported and the relatively low number of complications after strabismus surgery suggest that direct treatment of the retinal hole(s) with the laser or cryotherapy is not necessary unless a known predisposition to retinal detachment is present.

INFECTION AND INFLAMMATION

Conjunctivitis

Infectious conjunctivitis is a rare complication of strabismus surgery, although its true incidence is unknown.37 It may be related to the local skin flora and is usually susceptible to commonly used postoperative antibiotics. Staphylococcus aureus is the most common pathogen cultured from patients with postoperative periocular infection but is found in cases of suspected orbital cellulitis, not in less severe conjunctivitis.37 Routine use of perioperative antibiotics renders culture for suspected bacterial conjunctivitis unreliable and makes it difficult to differentiate infectious conjunctivitis from a ster-
ile but exaggerated postoperative inflammatory response (Fig. 41–13).

Ligneous conjunctivitis is a rare form of membranous conjunctivitis that has been reported after strabismus surgery. In general, it is most common in young females and is usually bilateral and asymmetric. The cause is unknown. The condition presents as recurrent exudative conjunctival membranes that respond poorly to topical antibiotics and corticosteroids. Topical cyclosporine 2% is reportedly effective (Figs. 41–14 and 41–15).

Scleritis

Necrotizing scleritis has been reported after strabismus surgery, but more often following cataract surgery. It is common in patients with underlying systemic or autoimmune diseases such as diabetes, rheumatoid arthritis, systemic lupus erythematosus, Wegener’s granulomatosis, polyarteritis nodosa, and inflammatory bowel disease. Surgically induced necrotizing scleritis has been associated with trauma, Herpes zoster ophthalmicus, and Proteus infection. The precise relationships are unclear, but these varied conditions all may incite an autoimmune reaction (Fig. 41–16).

Patients with scleritis present after surgery with headache, eye or orbital pain, and diffuse conjunctival injection. The lids may be edematous, and proptosis may be present. Anterior uveitis may be seen in some cases. B-scan ultrasonography may reveal choroidal thickening. A transient myopic shift may occur in association with scleritis after strabismus surgery.

The differential diagnosis of scleritis includes anterior segment ischemia, endophthalmitis, and orbital cellulitis, but these generally present in the first few days after strabismus surgery. Necrotizing scleritis presents several weeks later. Treatment should be instituted as soon as the diagnosis is made and includes topical and systemic corticosteroids. Cytotoxic agents, nonsteroidal anti-inflammatory agents, and cycloplegic agents have been used as adjuncts or when corticosteroids are contraindicated. The inflammatory process may be resistant to treatment and may recur years later. Scleral grafting or enucleation may be necessary.

Orbital Cellulitis

Orbital cellulitis is a sight-threatening and even life-threatening complication of strabismus surgery. The incidence has been reported as one in 1900 cases. It often begins in the first 24 to 96 hours. Visual acuity may be difficult to assess in the young, sick patient. There may be eyelid edema,
conjunctival injection, decreased motility, proptosis, fever, and leukocytosis. Bacteremia may be documented. The diagnostic workup should include conjunctival and blood cultures and computed tomography. A computed tomographic scan is extremely helpful for differentiating preseptal cellulitis from orbital cellulitis. Observation of ocular rotations can help identify the site of inflammation. Significantly limited ocular rotations suggest orbital rather than preseptal involvement. Lumbar puncture should be considered in the very ill infant. *Staphylococcus aureus* is the most common pathogen cultured from the conjunctiva in these patients, but *Streptococcus* and *Proteus* have also been reported. No *Haemophilus influenzae* organisms were cultured from cases of postoperative cellulitis, even those presenting before the onset of vaccination against *H. influenzae* type b. This contrasts to the prevaccination frequency of spontaneous orbital cellulitis, which was commonly caused by *H. influenzae* type b.

Treatment with intravenous antibiotics is necessary and usually is successful.

**Endophthalmitis**

Only a few cases of endophthalmitis have been reported after strabismus surgery. This is a potentially blinding complication that generally is associated with inadvertent scleral perforation. The incidence of scleral perforation has been reported to be as high as 9.2%, whereas the incidence of endophthalmitis after strabismus surgery has been reported as 1 in 30,000. Clearly, most scleral perforations do not result in endophthalmitis. Cultures have yielded *Staphylococcus epidermidis* and *Streptococcus pneumoniae* in reported cases. Treatment requires prompt pars plana vitrectomy; vitreous tap and culture; and subconjunctival, intravitreal, and systemic antibiotics. Final visual acuity has ranged from 20/20 to no light perception. Possible sources of infection in postoperative strabismus cases have been discussed. Contamination at the time of surgery is possible, but bilateral cases are rare. Self-contamination by a patient with poor hygiene may contribute to postoperative infection. Corticosteroid use has also been implicated, but there is no direct evidence to support that suspicion. Spontaneous orbital or lid cellulitis or upper respiratory tract infection also may be sources, but the frequency of postoperative infection is low.

Three policies—preoperative or postoperative use of topical and/or oral antibiotics, preoperative povidone-iodine (Betadine)/saline, and no antibiotic at all—have exhibited no statistically significant differences in the risk of postoperative infection. In addition, no method yields complete protection against postoperative infection. It is difficult to draw conclusions regarding the best means of preventing these infections.

Parents should be advised about postoperative signs and symptoms and should follow-up with the physician when suspicion arises. Because most postoperative infections occur variably in the first 4 days after strabismus surgery, an early postoperative visit may miss infection.

**REFERENCES**

Published surgical dose tables should serve only as a guide. They may be modified according to actual results obtained by the individual surgeon. Differences in dose response may be due to variations in surgical technique and desired postoperative alignment.

Recommended numbers for esotropia, exotropia, and vertical deviations are listed in the following pages. (Chapter 31 includes a surgical dose table for nystagmus.) These numbers are recommended for normal-sized eyes (average axial length of 22 mm) having comitant deviations without restrictions and lacking associated pathologic processes (e.g., Graves disease, myasthenia gravis, or muscle palsy). Numbers may be modified based in part on (1) clinical diagnosis, (2) findings on tests for muscle function (e.g., forced duction test, force generation testing, saccadic velocity analysis), and (3) anatomic factors. Each patient should be treated on an individual basis.

Our technique of recession involves measuring the site of the rectus insertion before disinserting the muscle. The desired recession is then added to this number, and the new insertion is measured from the limbus. For recessions that exceed 10 mm we recommend using the curved Scott ruler for normal-sized eyes. The technique of resection involves use of a spring-loaded clamp and measuring the amount of resection from the muscle insertion to the site of suture placement. The numbers are designed to achieve a desired postoperative orthotropic alignment unless otherwise stated.
# Surgery for Esotropia

Table A-1. Medial Rectus Recession of Both Eyes

<table>
<thead>
<tr>
<th>Esotropia (PD)</th>
<th>Recession (mm)</th>
</tr>
</thead>
<tbody>
<tr>
<td>15</td>
<td>3.0</td>
</tr>
<tr>
<td>20</td>
<td>4.0</td>
</tr>
<tr>
<td>25</td>
<td>4.5</td>
</tr>
<tr>
<td>30</td>
<td>5.0</td>
</tr>
<tr>
<td>35</td>
<td>5.5</td>
</tr>
<tr>
<td>40</td>
<td>6.0</td>
</tr>
<tr>
<td>45</td>
<td>6.5</td>
</tr>
<tr>
<td>50</td>
<td>7.0</td>
</tr>
</tbody>
</table>

Table A-2. Lateral Rectus Resection of Both Eyes

<table>
<thead>
<tr>
<th>Esotropia (PD)</th>
<th>Recession (mm)</th>
</tr>
</thead>
<tbody>
<tr>
<td>15</td>
<td>3.5</td>
</tr>
<tr>
<td>20</td>
<td>4.5</td>
</tr>
<tr>
<td>25</td>
<td>5.5</td>
</tr>
<tr>
<td>30</td>
<td>6.0</td>
</tr>
<tr>
<td>35</td>
<td>6.5</td>
</tr>
<tr>
<td>40</td>
<td>7.5</td>
</tr>
<tr>
<td>45</td>
<td>8.5</td>
</tr>
<tr>
<td>50</td>
<td>9.0</td>
</tr>
</tbody>
</table>

Table A-3. Medial Rectus (MR) Recession and Lateral Rectus (LR) Resection (R & R)

<table>
<thead>
<tr>
<th>Esotropia (PD)</th>
<th>MR Recession (mm)</th>
<th>LR Recession (mm)</th>
</tr>
</thead>
<tbody>
<tr>
<td>15</td>
<td>3.0</td>
<td>4.0</td>
</tr>
<tr>
<td>20</td>
<td>3.5</td>
<td>4.5</td>
</tr>
<tr>
<td>25</td>
<td>4.5</td>
<td>5.0</td>
</tr>
<tr>
<td>30</td>
<td>5.0</td>
<td>5.5</td>
</tr>
<tr>
<td>35</td>
<td>5.5</td>
<td>6.5</td>
</tr>
<tr>
<td>40</td>
<td>6.0</td>
<td>7.5</td>
</tr>
<tr>
<td>45</td>
<td>6.0</td>
<td>8.0</td>
</tr>
</tbody>
</table>
## Surgery for Exotropia

### Table A-4. Lateral Rectus Recession of Both Eyes*

<table>
<thead>
<tr>
<th>Exotropia (PD)</th>
<th>Recession (mm)</th>
</tr>
</thead>
<tbody>
<tr>
<td>15</td>
<td>4.5</td>
</tr>
<tr>
<td>20</td>
<td>5.5</td>
</tr>
<tr>
<td>25</td>
<td>6.0</td>
</tr>
<tr>
<td>30</td>
<td>7.0</td>
</tr>
<tr>
<td>35</td>
<td>8.0</td>
</tr>
<tr>
<td>40</td>
<td>9.0</td>
</tr>
<tr>
<td>45</td>
<td>9.5</td>
</tr>
<tr>
<td>50</td>
<td>10.0</td>
</tr>
</tbody>
</table>

*Designed for 5 to 10 PD of esotropia in the early postoperative period for patients with intermittent exotropia.

### Table A-5. Medial Rectus Resection of Both Eyes

<table>
<thead>
<tr>
<th>Exotropia (PD)</th>
<th>Resection (mm)</th>
</tr>
</thead>
<tbody>
<tr>
<td>15</td>
<td>3.0</td>
</tr>
<tr>
<td>20</td>
<td>4.0</td>
</tr>
<tr>
<td>25</td>
<td>4.5</td>
</tr>
<tr>
<td>30</td>
<td>5.0</td>
</tr>
<tr>
<td>35</td>
<td>5.5</td>
</tr>
</tbody>
</table>

### Table A-6. Lateral Rectus (LR) Recession and Medial Rectus (MR) Resection (R & R)

<table>
<thead>
<tr>
<th>Exotropia (PD)</th>
<th>LR Recession (mm)</th>
<th>MR Resection (mm)</th>
</tr>
</thead>
<tbody>
<tr>
<td>15</td>
<td>4.0</td>
<td>3.0</td>
</tr>
<tr>
<td>20</td>
<td>4.0</td>
<td>4.0</td>
</tr>
<tr>
<td>25</td>
<td>6.0</td>
<td>4.5</td>
</tr>
<tr>
<td>30</td>
<td>6.5</td>
<td>5.0</td>
</tr>
<tr>
<td>35</td>
<td>7.5</td>
<td>5.5</td>
</tr>
</tbody>
</table>

### Table A-7. Lateral Rectus (LR) Recession and Medial Rectus (MR) Resection (R & R) for Sensory Exotropia*

<table>
<thead>
<tr>
<th>Exotropia (PD)</th>
<th>LR Recession (mm)</th>
<th>MR Resection (mm)</th>
</tr>
</thead>
<tbody>
<tr>
<td>40</td>
<td>8.0</td>
<td>6.0</td>
</tr>
<tr>
<td>50</td>
<td>9.0</td>
<td>6.0</td>
</tr>
<tr>
<td>60</td>
<td>10.0</td>
<td>6.0</td>
</tr>
<tr>
<td>70</td>
<td>10.0</td>
<td>7.0</td>
</tr>
</tbody>
</table>

*An example of sensory exotropia would be profound amblyopia in one eye.
**Surgery for Vertical Deviations**

Table A-8. Hypertropia

<table>
<thead>
<tr>
<th>Hypertropia (PD)</th>
<th>Operating on Ipsilateral (Hypertropic) Eye</th>
<th>Operating on Contralateral (Hypotropic) Eye</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>SR Recess (mm)</td>
<td>IR Resect (mm)</td>
</tr>
<tr>
<td>5</td>
<td>3.0</td>
<td>3.0</td>
</tr>
<tr>
<td>10</td>
<td>4.0</td>
<td>4.0</td>
</tr>
<tr>
<td>15</td>
<td>5.0</td>
<td>5.0</td>
</tr>
<tr>
<td>20</td>
<td>6.5</td>
<td>3.0</td>
</tr>
<tr>
<td>25</td>
<td>5.0</td>
<td>5.0</td>
</tr>
<tr>
<td>30</td>
<td>6.0</td>
<td>5.0</td>
</tr>
</tbody>
</table>

SR, superior rectus muscle; IR, inferior rectus muscle.

Table A-9. Dissociated Vertical Deviation (DVD) of Both Eyes

<table>
<thead>
<tr>
<th>DVD (PD)</th>
<th>SR Recession (mm)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Less than 10</td>
<td>6.0</td>
</tr>
<tr>
<td>10</td>
<td>7.0</td>
</tr>
<tr>
<td>15</td>
<td>8.0</td>
</tr>
<tr>
<td>20</td>
<td>9.0</td>
</tr>
<tr>
<td>25</td>
<td>10.0</td>
</tr>
</tbody>
</table>

SR, superior rectus.
INDEX

Note: Page numbers in *italics* refer to illustrations; page numbers followed by *t* refer to tables.

Abducens nerve. See Cranial nerve(s), 6th.

Abducens nerve palsy. See Cranial nerve palsy, 6th.

Abduction, simultaneous, in Duane syndrome, 329, 338–339

Abduction deficit, after scleral buckling, 264, 300

differential diagnosis of, 262–265
in CFEM, 364
in internuclear ophthalmoplegia, 387, 388
in Miotics syndrome, 360
in progressive external ophthalmoplegia, 384
in recurrent orbital myositis, 265
transposition procedures for, 477, 478

AC/A ratio, 75–76

Adduction deficit, transposition procedures for, 477, 478

Adductors, dissection/lysis of, in strabismus

Accommodation, control of, in measurement,

AC/A ratio, 75–76

Accommodation, control of, in measurement,

AC/A ratio, 75-76

Adduction deficit, transposition procedures for, 477, 478

Adductors, dissection/lysis of, in strabismus

Accommodation, control of, in measurement,

AC/A ratio, 75-76

Adhesions, dissection/lysis of, in strabismus after scleral buckling, 302

in strabismus after glaucoma implant, 305

Adjustable suture kit, 441, 447

Adjustable suture technique, 435–447
advantages of, 435–436

conventional vs, comparison of muscle creep
in, 436

disadvantages of, 436

historical perspective on, 435

in strabismus after scleral buckling, 302

indications for, 436–437

noose, 441, 442–443, 444–445

posterior fixation and, 501

postoperative adjustment in, 441–445

complications of, 445–447

pitfalls of, 445

principles of, 441

technique of, 441–445

preoperative considerations in, 437

slip knot, 437–441, 444

Age, and response to surgery in intermittent exotropia, 169

Alternate prism cover test (APCT), 14, 31, 74

Alternate-letter suppression testing, in intermittent exotropia, 166

Amblyopia, 24, 30
detection of, in intermittent exotropia, 166
in craniosynostosis syndromes, 396–398
in Duane syndrome, 327

Amblyopia (Continued)
in infantile exotropia, occlusion for, 132–133
in monofixation syndrome, 183

in nystagmus, treatment of, 410–411

surgical outcome and, 77
Anesthesia. See Chemodenervation, 430

Anectine. See Succinylcholine.

Anterior segment ischemia, 519–520

Anterior segment bloodflow, 517–520

Anterior segment, evaluation of, for

Anterior chamber angle, 110, 112

Asthenopia, as chief complaint, 3

Astrocytoma, cerebellar, comitant exotropia in, 155

Axial length, and corneal diameter, estimated location of equator underneath medial rectus muscle based on, 76
surgical outcome and, 77–81

Baerveldt device, 305

Bagolivieri test, 34, 62

Bell’s phenomenon, in MED, 276

lagophthalmos from, 398

Bielschowsky’s phenomenon, 240

Bifocals, dispensing of, 19

Binocular alignment, measurements of, computational modeling and, 103–103

postoperative, in intermittent exotropia, 169

simulated, after chordoma/strabismus surgery, 110, 112

Anterior segment ischemia (Continued)
after transposition procedures, 487–488

in pattern strabismus, 213

in 6th cranial nerve palsy, 268
pathophysiologic, clinical manifestations, and treatment of, 519
reducing risk of, 519–520

risk factors for, 519, 521

risk of, after scleral buckling, 302

A-pattern exotropia, 202

from nasal displacement of SR/superior displacement of LR muscles, 95

simulated, from aberrant regeneration of 3rd cranial nerve, 208

A-pattern exotropia, 203

after surgery for Graves ophthalmopathy strabismus, 294

from superior placement of LR muscles, 94

treatment of, 212

A-pattern strabismus, alignment in, simulated, 108

prevalence of, 206

APCT. See Alternate prism cover test (APCT).

Apert syndrome, 396

forehead advancement in, 398

ARC. See Anomalous retinal correspondence (ARC).

Arnold-Chiari type I malformation, comitant exotropia in, 153–154

ASH. See Anterior segment ischemia.

Asthenopia, as chief complaint, 3

in intermittent exotropia, surgery and, 168

Astrocytoma, cerebellar, comitant exotropia in, 155

Axial length, and corneal diameter, estimated location of equator underneath medial rectus muscle based on, 76
surgical outcome and, 77–81

Baerveldt device, 305

Bagolivieri test, 34, 62

Bell’s phenomenon, in MED, 276

lagophthalmos from, 398

Bielschowsky’s phenomenon, 240

Bifocals, dispensing of, 19

Binocular alignment, measurements of, computer modeling and, 103–103

postoperative, in intermittent exotropia, 169

simulated, after chordoma/strabismus surgery, 110, 112

Anterior segment ischemia (Continued)
after transposition procedures, 487–488

in pattern strabismus, 213

in 6th cranial nerve palsy, 268
pathophysiologic, clinical manifestations, and treatment of, 519
reducing risk of, 519–520

risk factors for, 519, 521

risk of, after scleral buckling, 302

A-pattern exotropia, 202

from nasal displacement of SR/superior displacement of LR muscles, 95
simulated, from aberrant regeneration of 3rd cranial nerve, 208

A-pattern exotropia, 203

after surgery for Graves ophthalmopathy strabismus, 294

from superior placement of LR muscles, 94

treatment of, 212

A-pattern strabismus, alignment in, simulated, 108

prevalence of, 206

APCT. See Alternate prism cover test (APCT).

Apert syndrome, 396

forehead advancement in, 398

ARC. See Anomalous retinal correspondence (ARC).

Arnold-Chiari type I malformation, comitant exotropia in, 153–154

ASH. See Anterior segment ischemia.

Asthenopia, as chief complaint, 3

in intermittent exotropia, surgery and, 168

Astrocytoma, cerebellar, comitant exotropia in, 155

Axial length, and corneal diameter, estimated location of equator underneath medial rectus muscle based on, 76
surgical outcome and, 77–81

Baerveldt device, 305

Bagolivieri test, 34, 62

Bell’s phenomenon, in MED, 276

lagophthalmos from, 398

Bielschowsky’s phenomenon, 240

Bifocals, dispensing of, 19

Binocular alignment, measurements of, computer modeling and, 103–103

postoperative, in intermittent exotropia, 169

simulated, after chordoma/strabismus surgery, 110, 112
Binocular alignment (Continued)
in A-pattern strabismus, 543
in SO palsy, 106, 107
normal, 106
Biofeedback, for nystagmus, 413
Bitemporal hemianopia, hemifield sliding and
abnormal binocular phenomena in, 191
postfixation blind area in, 192
Blepharospasm, postoperative, 546
Blind spot mapping, measurement of anatomic
torsion by, 59
Blindness, infantile uniconial, with bilateral
nystagmus, 407
postfixation, in bitemporal hemianopia, 192
Blood chemistries, in Graves ophthalmopathy, 290
Botulinum toxin. See also Chemodenervation.
chemistry of, 423–424
dosage of, in chemodenervation, 430
mechanism of action of, in strabismus, 424
technique for injection of, 430
Brain stem, cerebrovascular accident in, skew
deviation after, 39
in infantile esotropia, 119
Brain tumors, comitant esotropia in, 154–156
Brown syndrome, 347–356
acquired, 349
causes of, 349
from infection/inflammation, 349
from orbital fracture, 311
from trauma, 349
iatrogenic, 349
simulation of, after scleral buckling, 299
bilateral, 352
classification of, 350
clinical characteristics of, 349–350
clinical examination in, 350–351
congenital, 348–349
diagnosis of, 350–352
differential diagnosis of, 352
epidemiology of, 347
etiology of, 347–349
from cist in SO tendon, 97
from excessive tuck of SO tendon, 227–228
heredity in, 347
historical perspective on, 347
iatrogenic, 546
IO after, 350
natural history of, 349–350
ophthalmoscopic view of fundus in, 69
SO expander for, 464
SO tenectomy for, 461
strabismus evaluation in, 351–352
torsion analysis in, 68
treatment of, 352–355
nonsurgical, 352–353
surgical, 353–355
complications of, 354–355
indications for, 353
options for, 353–354
SO palsy after, 354
undercorrections in, 354–355
Brückner test, 11–12
asymmetry of red reflex in, 13
Burian classification (intermittent esotropia), 165t
Cataract, occlusion by, disorders caused by, 372–373
preexisting heterotropia/heterophoria masked by, 371–372
sensory esotropia from, 193
Cataract surgery, hypertropia after, Lancaster red-green plot of, 70
Cataract surgery (Continued)
hypotropia after, Lancaster red-green plot of, 71
vs MED, 278t
postoperative optical aberrations after, 375–376
strabismus after, 371–378
chemodenervation in, 429
clinical characteristics of, 373–376
diagnosis of, 376–377
etiology of, 371–373
fundus photograph in, 71
Lancaster red-green plot of, 70, 71
prevention of, 377–378
treatment of, 377
trauma/anesthetic toxicity from, strabismus from, 373
Cellulitis, orbital, postoperative, 548, 549–550
Central fusion, normal, 22–24
Central fusion disruption syndrome (CFDS).
28–29
partial field occlusion for, 29
Cerebellar astrocytoma, comitant esotropia in, 155
Cerebellar tumors, vs MED, 278t
Cerebral palsies, chemodenervation in, 428
Cerebrovascular tumors, vs MED, 278t
Cerebrovascular accident, brain stem, skew
deviation after, 389
Complications of 354–355
undercorrections in, 354–355
indications for, 353
SO palsy after, 354
options for, 353–354
Cerebellar tumors, vs MED, 278t
Central fusion, normal, 22–24
Central fusion disruption syndrome (CFDS).
28–29
fibrosis of:
paralysis from, 93
Cerebellar tumors, vs MED, 278t
Central fusion disruption syndrome (CFDS).
intracranial vessels from, 40
intrinsic binocular phenomena from, 191
in strabismus after, 389
intrinsic muscle disorders, 429
in strabismus after scleral buckling, 301
indications for, 429
intraocular pressure, 429
injection technique in, 430
intrinsic muscle disorders from, in strabismus, 424
preparation for, 430
surgery for, 429
scleral buckling, 301
SO palsy after, 354
undercorrections in, 354–355
Chief complaint, 3–4
Chordoma, enveloping abducens nerve, LR
paralysis from, 93
Chordoma (Continued)
surgery for, simulated alignment after, 110
Clement Clark EZL, 71
Co-contraction syndrome, 40
Coloboma, congenital, of upper lid, 399
Computed tomography (CT), orbital, 87. See
also Orbital imaging.
in orbital fracture, 313–315
Computer models, 99–111
binocular alignment measurements and, 102–103
biomechanical, 100
in diagnosis, 103–105
in treatment planning, 103
clinical applications of, 103–111
expert systems in, 99
extraocular imaging and, 101
homeomorphic, 100
immunohistochemistry/electron microscopy and, 101–102
in teaching, 105–111
orientation to, 99–100
physiologic basis of, 100–103
rectus muscle pulleys in, 102
surgical simulation by, 103–105
Conjunctivitis, postoperative, 548–549
Contact lenses, for nystagmus, 412
Contact ligaments, 530
Conjunctival incision, for anterior ciliary vessel
aberrant regeneration in, 253
Conjunctival incision, for anterior ciliary vessel
sparing, 522
in craniosynostosis syndromes, 401
in reoperation, 512
Conjunctival recessions, in strabismus after
scleral buckling, 302
Conjunctivitis, postoperative, 548–549
Contact lenses, for nystagmus, 412
Contour circle test, 9
Convergence, insufficiency of, in intermittent
esotropia, 165
measurement of, in intermittent esotropia, 167
Comen diameter, and axial length, estimated
location of equator underneath medial
rectus muscle based on, 76t
Cover tests, 13–14, 30–31
in Duane syndrome, 327
in MED, 276
in 3rd cranial nerve palsy, 254
responses to, as indicator of fusion in
monofocal horizontal esotropia, 145t
Cover/uncover test, 13–14
Convergent, in infantile esotropia, 119
2nd. See Optic nerve.
6th, chordoma enveloping, LR paralysis from, 93
Cranial nerve(s), in infantile esotropia, 119
saccadic velocity testing in, 48t
in 3rd cranial nerve palsy, 254
aberrant regeneration in, 253, 254
lid fissure widening from, 547
surgical and, 256
adult-onset, causes of, 251t
CFEOM, chemodenervation in, 426
childhood-onset, causes of, 251t
clinical characteristics of, 252–253
clinical evaluation in, 253–255
complications of, 257
diagnosis of, 253–255
differential diagnosis in, 255
INDEX •

Cranial nerve palsy (Continued)
etiology of 251
genetics of 251–252
in combination with other cranial nerve
involvement, 253

isolated muscle palsy in, 253
laboratory evaluation in, 255
orbital periosteal flap procedure in, 485,
486

partial, 252-253
surgery for, 256
strabismus evaluation in, 254–255
total, 252

surgery for, 256

Crouzon syndrome (Continued)
V-pattern exotropia in, 211
CT. See Computed tomography (CT).
Cyclodeviations, after scleral buckling, 299
surgical procedures for, 303
Cyclopentolate, 18
Cycloplegic refraction, dispensing, guidelines
for, 18–19

testing of 18–19
Cyst(s), in SO tendon, Brown syndrome from,
97

inclusion, restricted elevation of eye from, 97
orbital, vs Graves ophthalmopathy, 291
subconjunctival, postoperative, 543

treatment of 255–256

559

Double elevator palsy. See Monocular elevation
deficiency (MED).
Double Maddox rod test, 60

Douse target test, 32
Downbeat nystagmus, 408
Downshoots, in Brown syndrome, 350, 351
in Duane syndrome, 329, 335-336
Drugs, effect of, on results of forced duction
testing, 39–40
on results of saccadic velocity testing, 46
topical/systemic, postoperative allergic reac
tions to, 542

DS. See Duane syndrome.
Duane syndrome, 325–342
abduction deficit in, transposition of vertical
rectus muscles for, 477

nonsurgical, 255
surgical, 255–256

bilateral, management of 341–342
Dellen. 541

with fusion, 329

4th, chemodenervation in, 426. See also Supe
rior oblique muscle(s), palsy of

Denervation, pharmacologic. See

6th, 259-268

Depolarizing agents, effect of, on results of
forced duction testing, 39–40

management of 341–342
without fusion, management of 342
Y/lambda pattern, management of 342
clinical examination in, 326–330

DHD. See Dissociated horizontal deviation
(DHD).

sequence of 327–329
esotropic. See Duane syndrome, unilateral,

Digitopalpebral sign, in CFEOM, 367
Diplopia, analysis of 32
as chief complaint, 3

exotropic, 329
paradoxical results of forced duction testing

vs MED. 278t

bilateral, differential diagnosis of, 262t
saccadic velocity analysis in, 262
vs divergence paralysis, 160t
CFEOM simulating, 366
chemodenervation in, 424–426
clinical characteristics of 259-260

complications of 268
diagnosis of, 260–265
clinical, 260–262
differential. 262-265

laboratory, 262
epidemiology and risk factors for, 259
genetics of 259
in adults, 259, 260t
in children, 259
causes of, 260t
overcorrection in, 268

posterior fixation suture in, 493, 495
recovery in, 260
recurrence in. 260

treatment of 265–268

nonsurgical, 265
pitfalls in, 268
surgical, 265–266
botulinum toxin in, 266
results of, 267–268

true, paresis vs. 261–262
undercorrection in, 268

unilateral, differential diagnosis of 262t
vertical rectus transposition in, with poste
rior fixation suture augmentation, 483
Craniofacial syndrome, infantile exotropia in,
177

Craniofacial synostosis, V-pattern exotropia in,
395

Craniostenosis, congenital, 394
Craniosynostosis syndromes, clinical
characteristics of 395–396
fusion in 400

measurement of deviation in, 398-400
strabismus in 393-403

diagnosis of 396–400
clinical, 396

laboratory, 400
mechanisms of 393-395
treatment of, 400-402

complications of, 402-403
nonsurgical, 400–401
surgical, 400-402
approaches in, 401–402
timing of 400
V-pattern, 393-395
torsion in, 400

Crouzon syndrome, 394, 397
subluxation of globe in, 398

Chemodenervation.

in adults, 27–28

in esotropia with NRC, 28
in intermittent exotropia, surgery and, 168
in pattern strabismus, 213
intractable, after cataract surgery, 372
Dissociated horizontal deviation (DHD), 245,
246

in infantile esotropia, 131–132
Dissociated vertical deviation (DVD), 237–247,
555t

classification of 239
clinical evaluation of 239–240

esotropic.

in, 40, 42

patterns of, bilateral, 327
management of 331–342
diagnostic factors necessary for, 330t
unilateral, 326

saccadic velocity testing in, 48–49
terminology relating to, 325–326
treatment of 330–342

surgical indications/contraindications for,
330–331

unilateral, esotropic, management of 332–
336

clinical presentation of 238-239
comitant, surgical procedures for, 242-243
complications of 245–247

with both anomalous LR and (sub)nor

definition of 237

with upshoot/downshoot in opposite

diagnosis of 239–241
differential diagnosis of 241
epidemiology and risk factors for, 238
genetics of 238
historical perspective on, 237
in infantile esotropia, 131–132
incomitant, 240

surgical procedures for, 243–245
laboratory evaluation of 240-241
measurement of 239–240

posterior fixation suture for 497
synonyms for, 237t
theories of 237–238
tilt measurements in, 13

torsion analysis in, 66–67
treatment of 241–245

nonsurgical, 241–242
surgical, 241–242
indications for, 242

issues related to, 242

Distance stereoacuity, testing of 7–8
Divergence, artificial, surgery for, in
nystagmus, 417
true, excess of, in intermittent exotropia, 165
Divergence paralysis, 159–161
clinical characteristics of 159–161
clinical examination in, 159

diagnosis of, 159–160
laboratory evaluation in, 159–160
treatment of, nonsurgical, 160–161
surgical, 161
Double depressor palsy. See Monocular
depressor deficiency (MDD).

mal LR activity, management of,
334-335

gaze, management of 335-336

exotropic, management of 337–338
management of 332–341
with fixation with DS eye, management
of 339–340

with relatively immobile globe, manage
ment of 336–337

with simultaneous abduction of each eye,
management of 338–339
with subnormal LR innervation, manage
ment of 340–341

Y/lambda pattern, management of 340
Duction testing, 16–17
forced, 37–40
clinical indications for, 37t

in Duane syndrome, 329–330
in Graves ophthalmopathy, 290
in MED, 277
interpretation of results of, 38t, 39–40

pitfalls in, 39–40
intraoperative, 38–39
in strabismus after scleral buckling, 302
of oblique muscles, 39
office, 37–38

qualitative assessment of, 39
quantitative assessment of 39
DVD. See Dissociated vertical deviation

(DVD).

Eccentric fixation, 24, 30
in measurement of deviation, 76


Electro-oculography (EOG), in DVD, 241
Electromyography (EMG), in DVD, 241
Endophthalmitis, postoperative, 550
Exaggerated traction test, 54–55
Exotropia (Continued) motor alignment in, 183
motor tests in, 184
sensory characteristics of, 183
symptoms of, 182–183
treatment of, nonsurgical, 185
surgical, 185–186
in CFEOM, 365
in craniofacial dysostosis, 394
in Pfeiffer syndrome, 397
in progressive external ophthalmoplegia, 384, 385
incomitant, after glaucoma implant, 305
after scleral buckling, 298
in 6th cranial nerve palsy, 261
infantile, 132, 176–181
causes of, 177–178
classification of, 176
clinical characteristics of, 178
complications of, 180–181
definition of, 176
diagnosis of, 178–179
differential diagnosis in, 179
genetic considerations in, 177
history in, 178
incidence of, 176–177
neurophysiologic studies of, 177–178
ocular assessment in, 179
systemic evaluation in, 179
treatment of, 180
intertemporal, 163–173
amblyopia and refractive error in, 166
chemodenervation in, 428
classification of, 164–165
Burian vs Kushner, 165
clinical characteristics of, 163–165
complications of, 170–173
diagnosis of, 176–177
epidemiology and risk factors for, 163
incidence of, 176–177
in Pfeiffer syndrome, 397
in craniofacial dysostosis, 394
in 6th cranial nerve palsy, 261
infantile, 132, 176–181
causes of, 177–178
classification of, 176
clinical characteristics of, 178
complications of, 180–181
definition of, 176
diagnosis of, 178–179
differential diagnosis in, 179
genetic considerations in, 177
history in, 178
incidence of, 176–177
neurophysiologic studies of, 177–178
ocular assessment in, 179
systemic evaluation in, 179
surgical, 173
risk factors for, 172
PAT in, 167
temporal hemiretinal suppression scotoma in, 25
treatment of, nonsurgical, 167–168
surgical, 168–170
pitfalls in, 170
sensory criteria for success of, 169
undercorrection in, 170–171
clinical presentation of, 170–171
management of, 171
risk factors for, 170
recurrent, 170
residual, 170
surgical, 172
risk factors for, 172
sensory, 193–199
causes of, 193–194
classification of, 193
in adults, 193–194
in children, 193
clinical characteristics of, 195–196
complications of, 198–199

Electromyography (EMG), 37
in Graves ophthalmopathy, 289
Electron microscopy, computer modeling and, 101–102
Electro-oculography (EOG), in DVD, 241
saccadic velocity analysis by, 44, 45
Electroretinography, in nystagmus, 410
EMG. See Electromyography (EMG).
Endophthalmitis, postoperative, 550
Enophthalmos, after orbital floor fracture, 315
EOG. See Electro-oculography (EOG).
EOM. See Extraocular muscles.
Esotropia (Continued)
saccadic velocity analysis by, 44, 45
in Graves ophthalmopathy, 289
acquired, chemodenervation in, 427–428
decompensated, 143–144
clinical characteristics and diagnosis of, 141
clinical features of, 153–157
diagnosis of, 156–157
treatment of, 157
consecutive, 149–150
clinical characteristics and diagnosis of, 149
management plan for, 150
surgical, 150
 cyclic (periodic), 148
decompensated monofixational, 144–145
clinical characteristics and diagnosis of, 144–145
cover test responses as indicators of fusion in, 145
treatment of, 145
difficult, management of, 139–150
D-shaped scotoma in, 25
in CFEOM, 364
in Graves ophthalmopathy, 288
in Möbius syndrome, 360, 362
incomitant, after advancement and recovery of lost MR, 512
after scleral buckling, 300
due to slipped LR after scleral buckling, 511
due to 6th nerve palsy, 261
infantile, 117–135
abnormalities of cortical development predisposing to, 123
accommodative esotropia after correction of, 141
causation of, 117–123
brain stem vergence motoneurons in, 119
cranial nerves in, 119
extraocular muscles in, 119
historical perspective on, 117, 118
nature vs nurture in, 117–118
subcortical visual pathway in, 119
visual cortex in, 118–119
visual vs motor pathways in, 118
chemodenervation in, 426, 427
classic presentation of, 123–125
clinical characteristics of, 123–126
diagnosis of, 126–132
DVD and DHD in, 131–132
facial and abduction deficit in, 129–130, 131
functional deficits in, 121
functional-structural correlation in, 122
latent fixation nystagmus in, 127–129, 130
motion VEP asymmetry in, 129, 130, 133
neuroanatomic findings in, 121–123
pursuit asymmetry in, 127, 128
spectrum of clinical presentations of, 125–126
treatment of, 132–135
nonsurgical, 132–133
surgical, 133–135
anesthetic and operative considerations in, 135
follow-up regimen after, 135
preoperative counseling for, 134–135
rationale for early, 133–134
timing and preoperative measurements in, 134
visual cortical mechanisms in, 119–121
intermittent, 139–140
clinical characteristics and diagnosis of, 139–140
treatment of, 140
overcorrected, 146–148
clinical characteristics and diagnosis of, 146–147
treatment of, 147–148
posterior fixation suture in, 496
recurrent, 146–148
reoperation for, 151
residual, after hyperopic correction, 19
undercorrected, 145–146
clinical characteristics and diagnosis of, 145–146
treatment of, 146
V-pattern, 204
from IO overaction, 208, 209
from nasal displacement of LR muscles, 95
surgical, 185–186
afibrillatory motor response in, 186
amblyopia and refractive error in, 166
camptometer in, 187
chemodenervation in, 428
classification of, 164–165
Burian vs Kushner, 165
clinical characteristics of, 163–165
complications of, 170–173
diagnosis of, 176–177
epidemiology and risk factors for, 163
incidence of, 176–177
historical perspective on, 163
lateral incomitance and ocular rotations in, 166–167
measuring AC/A ratio in, 166
nonsurgical, 172–173
surgical, 173
risk factors for, 172
PAT in, 167
temporal hemiretinal suppression scotoma in, 25
treatment of, nonsurgical, 167–168
surgical, 168–170
pitfalls in, 170
sensory criteria for success of, 169
undercorrection in, 170–171
clinical presentation of, 170–171
management of, 171
surgical, 173
residual, 170
reoperation for, 150
secondary, after MR recession, 510
sensory criteria for success of, 169
undercorrection in, 170–171
clinical presentation of, 170–171
management of, 171
risk factors for, 170
recurrent, 170
sensory, 193–199
causes of, 193–194
classification of, 193
in adults, 193–194
in children, 193
clinical characteristics of, 195–196
complications of, 198–199

Esotropia (Continued)
Extraocular muscles (Continued)

tests of, 37–49
trauma to, in scleral buckling, 297
underaction of, in ocular myasthenia, 381
Eye, anterior segment of, blood flow in,
517–519
evaluation of, for reoperation, 508
in nystagmus, 409
ischemia of, 519–520
after transposition procedures, 487–488
in pattern strabismus, 213
in 6th cranial nerve palsy, 268
pathophysiology, clinical manifestations,
and treatment of, 519
reduction of risk of, 519–520
risk factors for, 519, 521
risk of, after scleral buckling, 302
postoperative complications in, 543
posterior segment of, evaluation of, in nystagmus, 409–410
postoperative complications in, 548
Eye movements, abnormal, as chief complaint, 3–4
falling or rising, in Duane syndrome, 329
recordings of, in nystagmus, 410, 411
waveforms of, in congenital nystagmus, 405
Eyelid, antimongoloid position of, V-pattern
strabismus with, 206
fissure anomalies of, 546–547
postoperative, 547
lower, retraction of, after surgery for Graves ophthalmopathy strabismus, 293–294
paradoxical retraction of, after orbital fracture, 317
postoperative complications in, 546–548
upper, congenital coloboma of, 399
residual hemiagiomata of, ptosis from, 147
retraction of, in ocular myasthenia, 381
Face turn, in infantile esotropia, 129–130, 131
Facultative inhibition, 24. See also Suppression.
Fadenoperation, 491–505. See also Posterior fixation surgery.
Force generation testing (Continued)
quantitative assessment in, 42–43
Force generation testing, (Continued)
quantitative assessment in, 42–43
Forced augmentation test, in Duane syndrome, 330
Forcedduction testing, 37–40
clinical indications for, 371
in Duane syndrome, 329–330
in Graves ophthalmopathy, 290
in MED, 277
interpretation of results of, 38t, 39–40
pitfalls in, 39–40
intraoperative, 38–39
in strabismus after scleral buckling, 302
of oblique muscles, 39
office, 37–38
qualitative assessment of, 39
quantitative assessment of, 39
Forced-choice preferential looking (FPL), in assessment of preverbal children, compared to other methods, 5
Forehead advancement, complete bilateral mechanical ptosis after, 399
in Apert syndrome, 398
Foures–PD base-out test for foveal suppression, 10–11
response to, 12
Fovea, position at, 53
time-averaged, 24
Foveal suppression, 4–PD test for, 10–11
response to, 12
FPL. See Forced-choice preferential looking (FPL).
Fresnel membrane prisms, for divergence paralysis, 161
Frontal plane position, 75
Frosted glass, blurring of second image by, in strabismus after scleral buckling, 301
Fundus, examination of, 59
excyclophoria of, in severe IO overaction, 451
ophthalmoscopic view of, 58
photography of, measurement of anatomic torsion by, 55–59
Fusion, assessment of, in craniosynostosis syndromes, 400
in Duane syndrome, 327
breakdown of, after scleral buckling, 298
central, disruption of, in adults, 28–29, 194
normal, 22–24
degrees of, 21
encouragement of, in DVD, 241–242
near, tacituous post-surgical, in intermittent exotropia, 169
peripheral, normal, 24
sensory, in monofixation syndrome, 183
torsional motor, 63
Fusional amplitudes, testing of, 11, 32–34
Gene therapy, for Brown syndrome, 347
in CF, 347
in CFEOM, 363
in DVD, 238
in Graves ophthalmopathy, 285–286
in infantile exotropia, 177
in intermittent exotropia, 163
in Möbius syndrome, 358
in nystagmus, 404–405
in 3rd cranial nerve palsy, 251–252
in 6th cranial nerve palsy, 259
Glaucma drainage devices, dimensions of, 304
of, 304
Glaucma implant, strabismus after, 304–306
adhesions and scar tissue formation in, 305
Glaucoma implant (Continued)
diagnosis of, 305–306
historical perspective on, 304
incidence of, 304
mass effect in, 305
mechanisms for, 304–305
muscle factors in, 305
prevention of, 306
treatment of, 306
Globe size, surgical outcome and, 77–81
Globe, 3-D reconstruction of, 89
immobile, in Duane syndrome, management of, 336–337
perforation of, during surgery, 548
static fixation of, peristomal flap for, 485, 486
subluxation of, in Crouzon syndrome, 398
Globe size, surgical outcome and, 77–81
Goniometer, 408
measurement of anomalous head posture with, 19
Graves ophthalmopathy, 285–294
after cataract surgery, 272
chemodenervation in, 290–300
dissociated, 245, 246
in infantile esotropia, 131–132
posterior fixation suture for, 492–496
reoperation for, 509–510
Horror fusionis, 27, 194
Hemianopia, bitemporal, hemifield sliding and
mass effect in, 305
Hemangioma, upper lid, ptosis from, 147
Hemifield, surgical correction of, in
inflammatory pseudotumor, 315
Hemorrhage, postoperative, 548–550
Heterotropia. See Strabismus.
Hering’s law, 16
Hess-Lancaster screens, pre- and postoperative, in
skew deviation, 390–391
Heterotropia. See Strabismus.
Hirschberg corneal light reflex test, 12, 74
Horizontal deviation, 117–214. See also
Esotropia; Exotropia.
Hypertelorism, repair of, amblyopia after, 399
Hypertropia, after cataract surgery, 376
Lancaster red-green plot of, 70
after glaucoma implant, 305
after scleral buckling, 298
from orbital floor fracture, 312, 317, 318
in Graves ophthalmopathy, 288
in IOP, 232, 235, 236
in progressive external ophthalmoplegia, 384
in 3rd cranial nerve palsy, 256
Hypoglobus, after orbital floor fracture, 315
Hypotropia, after cataract surgery, 372
Lancaster red-green plot of, 71
after orbital floor fracture, 311
after scleral buckling, 299, 300
after sinus surgery, 321
in Brown syndrome, 350, 351, 355
in CFEOM, 364, 365, 366
in Graves ophthalmopathy, 280
in IOP, 231
in MED, 275, 277, 279
in 3rd cranial nerve palsy, 252, 254
Image distortion, after scleral buckling, 298
Immunochemistry, computer modeling and,
(hARc), 25, 101–102
Head posture, anomalous, after scleral buckling, 301
after SO tenotomy for Brown syndrome, 336
assessment of, 19–20
horizontal, surgical correction of, dosage for, 415
in nystagmus, 414–415
in DVD, 240, 242
in nystagmus, 405–406
prisms for, 412
surgically induced, 418
in pattern strabismus, 213
measurement of, in nystagmus, 408–409
with goniometer, 19
nonocular causes of, 20

Inferior oblique muscle(s) (Continued)
palsy of, 230–236, 253
bilateral, treatment of, 235, 236
classic appearance of, 230, 231
clinical examination in, 232–233
clinical presentations of, 230–232
diagnosis of, 232–234
differential diagnosis of, 233–234
epidemiology and risk factors for, 230
historical perspective on, 230
treatment of, 234–235
nonsurgical, 234
surgical, 234–235
with comitant deviation, treatment of, 235
with incomitant deviation, complications of, 235–236
postoperative hemorrhage of, 457
recession of, for cyclotorsion after scleral buckling, 303
surgical anatomy of, 449, 450
underaction of, grading scheme for, 17
in V-pattern strabismus, 18
weakening of, 449–458
anterior transposition in, 454–455
contraindications to, 451
denervation and extirpation, 455–456
for treatment of pattern strabismus, 212
historical perspective on, 449–450
indications for, 450–451
mucosectomy in, 452–453
myotomy in, 453
nasal myectomy in, 455
recession in, 453–454
nonsurgical, 234
technique for, 452–456
Inferior rectus muscle(s), avulsion of, 96
chemodenervation of, injection technique in, 431
ciliary vessels of, 517
entrapment of, in orbital fracture, 96
fibrosis of, 368
late slippage of, after surgery for Graves ophthalmopathy strabismus, 293, 294
nasal displacement of, V-pattern strabismus from, 94
normal and parietic, saccadic velocity analysis of, 313
palsy of, 253
in orbital fracture, surgical management of, 317–318
pseudo-overaction of, in orbital fracture, 312
recession of, excessive, in posterior fixation suture, 593
in skew deviation, results of, 391
resection of, in DVD, 243
restriction of, in orbital fracture, 310, 311
surgical management of, 317–319
tight, thyroid myopathy with, ophthalmoscopic view of fundus in, 69
transposition of, temporalward, in Duane syndrome, 334
Inflammation, postoperative, 548–550
in Graves ophthalmopathy, 293
Infected scleral reflection technique, 44
Innervation, abberant, in forced duction testing, 40
Intermuscular septum, 530
avoidance of disruption to, in SO tenotomy for Brown syndrome, 355
Intracranial ophthalmoplegia, 387–389
asymmetric bilateral, 387
clinical characteristics of, 387–388
diagnosis of, 388
INDEX • 563

Internuclear ophthalmoplegia (Continued) saccadic velocity testing in, 48 strabismus diagnosis in, 388 treatment of, 388–389 wall-eyed bilateral, 389

Intorsion, 52. See also Torsion.

Intraocular pressure (IOP), differential, estimation of generated muscle forces by, 40

IO muscles. See Inferior oblique muscle(s).

IOP. See Intraocular pressure (IOP).

IR muscle(s). See Inferior rectus muscle(s).

Keratitis, filamentary, postoperative, 542

Kestenbaum-Anderson procedure, 414 variations of, 414–415

Knapp procedure, 480–481, 484

in stereoscopic examination, 441

Kown squint hook, 513

Kriskmy test, 12, 74

Kushner classification (intermittent exotropia), 165t

Lagophthalmos, from poor Bell’s phenomenon, 397

Labyrinthine disorders, vs MED, 278t

Kushner classification (intermittent exotropia), 165t

Lancaster red-green test, 60–62

Lang I stereoacuity test, 8

Lambda pattern strabismus, 205

Latent fixation nystagmus, 196, 197

Lenses for strabismus patient, 478–479

Lateral rectus muscle(s). See Inferior rectus muscle(s).

Lateral rectus muscle(s), anomalous actions/ recruitment of, in Duane syndrome, 328 decreased abduction of, in orbital myositis, 265
effect of IO weakening on, 457 inferior displacement of, in myopic strabismus, 95 innervation of, anomalous, in Duane syndrome, 326, 332 subnormal, in Duane syndrome, 340–341 paralysis of, cross-sectional area in, 92 from chordoma enveloping abducens nerve, 93 posterior fixation suture of, in Duane syndrome, 336 recession of, in Duane syndrome, 335–336 in intermittent exotropia, 169–170 overcorrection of, 172 residual esotropia after, 509 slipped, after scleral buckling procedure, incomitant esotropia due to, 511 superior displacement of, with nasal displacement of SR muscles, A-pattern esotropia from, 95 superior placement of, A-pattern esotropia from, 94 tight, syndrome of, 195 after treatment of intermittent exotropia, 170 in intermittent exotropia, 170, 171 Leash phenomenon, 397 in strabismus after glaucoma implant, 304–305 Lid retractors, for posterior fixation suture, 498 Light, bright, monocular eye closure in, intermittent exotropia and, 164 Light reflex tests, 11–12, 74 Ligneous conjunctivitis, postoperative, 549 Limbus, arc measurements from, cord (caliper) and, difference between, 81 LR muscle(s). See Lateral rectus muscle(s).

Macular differentiation, abnormal, in congenital nystagmus, 409 Macular ectopia, after scleral buckling, 298 Maddox rod test, double, 60 Maddox rods, 15 Magnetic resonance imaging (MRI), in DVD, 241 in orbital fracture, 313–315 of rectus and SO muscles, optimal patient and surface coil positioning for, 85 orbital, 86–87 parameters for, 876 Malar hypoplasia, V-pattern strabismus with, 206 Masses, orbital imaging for, 96–97 MDD. See Monocular depressor deficiency (MDD).

MED. See Monocular elevation deficiency (MED).


Motion visual evoked potential (VEP), asymmetry of, in infantile esotropia, 129, 130, 131


MRI. See Magnetic resonance imaging (MRI).

Muscle capsule, extraocular, 529

Muscle function, tests of 37–49

after transposition procedures, 485–487 in strabismus after scleral buckling, 301–302


inferior rectus, 253 in orbital fracture, surgical management of, 317–318 lateral rectus, cross-sectional area in, 92 from chordoma enveloping abducens nerve, 93 medial rectus, 253 superior oblique, 219–228 acquired, 221 after SO tenectomy for Brown syndrome, 461
Muscle palsy (Continued)

bilateral, 222, 468

Lancaster red-green plot in, 61

treatment of, 227
classification of, 219–221
clinical characteristics of, 221–222
complications of, 227–228
congenital, 219–220
cross-sectional area in, 620
diagnosis of, 222–226
clinical, 222
differential, 222–223
laboratory, 222
etiology of, 219
iatrogenic, 545–546
after treatment of Brown syndrome, 354
Lancaster red-green plot in, 61
ophthalmoscopic view of fundus in, 65
overcorrection in, 227
signs of, 221–222
simulated alignment in, 106, 107
symptoms of, 221
torsion analysis in, 64–66
treatment of, 225–227
algorithm for, 226–227
nonsurgical, 225
surgical, 225–227
superior rectus, 253
vertical rectus, torsion analysis in, 66
transposition procedures for, 480–481

Muscle pulleys, heterotopic, and pattern strabismus, computer modeling of, 107
rectus, 102, 104
average positions of, 89
Myasthenia gravis, 380–384
adult-onset, 381
clinical characteristics of, 381–382
comitant esotropia in, 156
congenital, 381
epidemiology of, 380
examination in, 382
historical perspective on, 380
history in, 382
juvenile, 381
pharmacologic testing in, 382–383
saccadic velocity testing in, 46–47, 48t
strabismus diagnosis in, 382–383
differential, 383
laboratory, 383
strabismus management in, 383–384, 385
complications of, 384
subtypes of, 383–381
treatment of, 383
vs MED, 278t
Myoclonus, oculopalatal, 408
Myopia, comitant convergent strabismus associated with (Bielschowsky type), 153
Myopic strabismus fixus, 95
Myotonia, in SO weakening, 453
marginal, in strabismus after scleral buckling, 305

Nasal myectomy, in IO weakening, 455
Nausea, postoperative, 539–540
incidence of, 539–540
prevention of, 540
nonsurgical treatment of, 540
Near stereacuity, testing of, 7
Needle(s), for IO weakening, 456
for posterior fixation suture, 497
for resecting adjustable suture, 447

Nerve palsy, cranial. See Cranial nerve palsy.
Neuroimaging, in nystagmus, 410
Neurologic disorders, presenting as comitant esotropia, 152–157
Neurologic disorders (Continued)
saccadic velocity testing in, 47–48
strabismus in, 235–236
Neuronomatous, ocular, 391–392
treatment of, 392
Nodular scleritis, postoperative, 549
Nose technique (adjustable sutures), 441, 442–443, 446
postoperative adjustment in, 444–445
Nystagmus, 404–418
acquired, 407–408
pharmacologic denervation in, 413
amblyopia in, treatment of, 410–411
bilateral, infantile uniconal blindness with, 407
chemodenervation in, 429
clinical characteristics of, 405–408
clinical examination in, 408–410
congenital, 405–407
clinical findings in, 405–406
etiology of, 406–407
pharmacologic denervation in, 413
diagnosis of, 407–408

genes of, 404–405
heterotropia in, modifications of surgical procedure for, 415
surgically induced, 418
historical perspective on, 404
laboratory examination in, 410
latent fixation, 407
in infantile esotropia, 127–129
optokinetic, in assessment of preverbal children, compared to other methods, 5
in saccadic velocity testing, 45
periodic alternating, 407–408
posterior fixation suture for, 496–497
spectacle correction in, 411–412
treatment of, 410–418
nonsurgical, 410–413
pharmacologic, 413
surgical, 413–418
complications of, 418
upbeat and downbeat, 408
vestibular, 408
vision assessment in, 6–7
Nystagmus compensation syndrome, 407
surgery for, 417

Oblique muscle(s), forced duction testing of, 39, 54–55
inclination/rotation of, after scleral buckling, 298
inferior, chemodenervation of, injection technique in, 431
inclination of, 544–545
overaction of, grading scheme for, 89
incomitant DVD from, 243, 244
primary, vs SO palsy, 223
pseudo-, 458
V-pattern esotropia from, 208, 209
palsy of, 230–236, 253
bilateral, treatment of, 235, 236
classic appearance of, 230, 231
clinical examination in, 232–233
clinical presentations of, 230–232
diagnosis of, 232–234
differential diagnosis of, 233–234
epidemiology and risk factors for, 230
historical perspectives on, 230
nonsurgical treatment of, 234–235
surgical, 234–235
with comitant deviation, treatment of, 235

Oblique muscle(s) (Continued)
with incomitant deviation, complications of, 235–236
postoperative hemorrhage of, 457
recession of, for cyclotorsion after scleral buckling, 303
surgical anatomy of, 34, 190, 449
underaction of, grading scheme for, 89
in V-pattern exotropia, 90
weakening of, 449–458
anterior transposition in, 454–455
complications of, 456–458
contraindications to, 451
denervation and extirpation, 455–456
for treatment of pattern strabismus, 212
historical perspective on, 449–450
indications for, 450–451
postoperative hemorrhage of, 457
nonsurgical treatment of, 458
muscle capture in, 452–453
myectomy in, 453
myotonia in, 453
nasal myectomy in, 455
overcorrection in, 455
recession in, 453–454
measurements for, 454
technique for, 452–456
undercorrection in, 457
insertions of, sagittalization of, pattern strabismus from, 204–206
overaction of, emerging, in pattern strabismus, 213
failure to diagnose/treat, in pattern strabismus, 213
primary, theoretical origin of, 54
torsion analysis in, 66
overaction/underaction of, pattern strabismus from, 204–206
superior, avulsion of, from ethmoidectomy, 96
chemodenervation of, injection technique in, 431
cross-sectional area of, contractile change in, 91
exaggerated traction test for, 56–57
functioning of, mechanical disadvantage of, from shallow medial orbital wall, 200
in craniosynostosis, 395
MRI scan of, optimal patient and surface coil positioning for, 85
myokymia of, 228–229
clinical features and diagnosis of, 228
complications of, 228
for treatment of, 228
overaction of, grading scheme for, 89
in Brown syndrome, 351
palsy of, 219–228
acquired, 221
after SO tenectomy for Brown syn
bilateral, 222, 468
Lancaster red-green plot in, 61
treatment of, 227
classification of, 219–221
clinical characteristics of, 221–222
complications of, 227–228
congenital, 219–220
cross-sectional area in, 92
contractile change in, 92
diagnosis of, 222–226
clinical, 222
differential, 222–223
laboratory, 222
etiology of, 219
iatrogenic, 545–546
Lancaster red-green plot in, 61
overcorrection in, 227
Index • 565

Oblique muscle(s) (Continued)
signs of, 221–222
symptoms of, 221
torsion analysis in, 64–66
treatment of, 225–227
algorithm for, 226–227
nonsurgical, 225
results of, 227
surgical, 225–227
posterior tenectomy of, 462, 463–465
historical perspective on, 463
indications for, 463
technique of, 463–465
tenotomy/tenectomy of, 459–465
historical perspective on, 459
in Brown syndrome, 353, 354
anomalous head posture after, 355
avoidance of intermuscular septum disruption in, 355
SO palsy after, 354
technique of, 459–460
transposition of, 484–485
historical perspective on, 484
mechanism of, 484
results of, 485
surgical technique in, 485
underaction of, grading scheme for, 89
weakening of, for cycloversion after scleral buckling, 303
for treatment of pattern strabismus, 211, 212
Occlusion, for infantile esotropia, 132–133
for 3rd cranial nerve palsy, 255
monocular, measurement of divergence by, 74
vision assessment by, 6
partial field, for CFDS, 29
for nystagmus, 411–412
Ocular dominance columns, 121
Ocular irritation/pain, postoperative, 540–541
Ocular myasthenia, 381. See also Myasthenia gravis.
Ocular neuromyotonia, 391–392
Ocular rotations, assessment of, 16–17, 37–40
for reoperation, 507–508
in Duane syndrome, 327–329
in intermittent exotropia, 166–167
in pattern strabismus, 208–209
in 3rd cranial nerve palsy, 254–255
restriction of, absolute, 39
algorithm for determining surgical strategy in, 477
conjunctiva contributing to, string or indentation by, 39
differential diagnosis of slowed saccadic velocities in, 43t
posterior, 40, 41
uniform, 39
Oculomotor nerve palsy. See Cranial nerve palsy, 3rd
Oculopatellar myelonus, 408
OKN. See Optokinetic nystagmus (OKN).
One-and-a-half syndrome, 388
Ophthalmoplegia, internuclear, 387–389
asymmetric bilateral, 587
clinical characteristics of, 387–388
diagnosis of, 388
saccadic velocity testing in, 48
strabismus diagnosis in, 388
treatment of, 388–389
wall-eyed bilateral, 589
progressive external, chronic, 384–387
clinical characteristics of, 385
diagnosis of, 385
differential diagnosis of, 386
Ophthalmoplegia (Continued)
ptosis management in, 386
saccadic velocity testing in, 47–48
strabismus diagnosis in, 385–386
strabismus management in, 386–387
vs MED, 278t
Ophthalmoscope, calibrated, 30
Ophthalmoscopic, indirect, measurement of
anatomic torsion by, 52, 58
grading system for, 57
Ophthalmotomograph, and string. 100
Ophthalmoplegia, 3-D reconstruction of, 89
Optic nerve head–foveal angle, determination of
by indirect ophthalmoscopy, 58
Optokinetic nystagmus (OKN), in assessment of
preverbal children, compared to other methods, 5
in saccadic velocity testing, 45
Ocularis oculi muscle, ragged red fibers in, in
progressive external ophthalmoplegia, 385
 Orbit, normal, MRI images of, 88
system/ disease involving, vs Graves ophthalmopathy, 290–291
Ocular cellitis, postoperative, 549–550
vs MED, 278t
Ocular connective tissue, computer
reconstruction of, 103
Ocular cysts, vs Graves ophthalmopathy, 291
Ocular disease, strabismus in, 76–77
Ocular fibrosis syndrome. See Extraocular muscles, congenital fibrosis of.
Ocular fracture, blow-out, 310, 311, 312
entrapment of IR muscle in, 97
posterior fixation suture in, 494, 496
vs MED, 278t
classification of, 309–310
result of, 315–317
early, surgical considerations in, 316
indications for, 315–316
late, surgical considerations in, 316–317
protocol for, 316
strabismus after, 309–319
clinical characteristics of, 310–311
clinical evaluation of, 311–312
diagnosis of, 311–312
diagnostic differential of, 311
incidence of, 309
laboratory evaluation of, 312
mechanisms of, 310
radiographic evaluation of, 313–315
frontal translation of, 317–319
surgical, 317–319
Ocular imaging, 84–98
clinical applications of, 90–98
tomography (CT), 87
computer modeling and, 101
general principles of, 84–86
history of, 84
in Graves ophthalmopathy, 287–288
in orbital fracture, 313–315
magnetic resonance (MRI), 86–87
parameters for, 87t
Ocular implant, 316
Orbital inflammatory disease, vs Graves ophthalmopathy, 290
vs MED, 278t
Orbital myositis, 291
decreased LR abduction in, 265
Orbital neoplasms, vs Graves ophthalmopathy, 291
Orbital periosteal flaps, 485, 486
Orbital ultrasonography, in Graves ophthalmopathy, 288–289
Orbital vascular lesions, vs Graves ophthalmopathy, 291
Orbitotomy, medial, approach to MR muscle by, 484
Orthoptics, for intermittent exotropia, 168
Oscillopsia, in congenital nystagmus, 406
Overcorrection, in esotropia, 146–148
in intermittent exotropia, 171–173
in IO weakening, 458
in nystagmus, 418
in posterior fixation suture, 504
in SO palsy, 227
in surgery for Graves ophthalmopathy strabismus, 293
in 6th cranial nerve palsy, 268
late, in transposition procedures, 487
Paradoxical anomalous retinal correspondence, 25, 26
Paralytic strabismus. See also Cranial nerve palsy: Muscle palsy.
chemodenervation in, 424–426
saccadic velocity testing in, 46
Partial field occlusion, for CFDS, 29
PAT. See Prism adaptation test (PAT).
Patient, apprehensive, interpretation of forced duction testing on, 39
fatigued, interpretation of saccadic velocity analysis on, 46
Patient position, optimal, in MRI scanner, for rectus and SO muscles, 85
Pattern strabismus, 202–214. See also specific pattern.
anatomic factors in, 206
classification of, 202–203
complications of, 213
definition of, 202
diagnosis of, 207–209
etiology of, 204–207
examination of ocular rotations in, 208–209
from muscle overaction/underaction, 204–206
from sensory deprivation, 206
heterotopic muscle pulleys and, computer modeling of, 107
historical perspective on, 202
motor testing in, 207–208
prevalence of, 203
sensory testing in, 209
treatment of, 209–210
algorithm for, 212–213
surgical strategy in, 210
technical considerations in, 210–212
Periosteal flap, static globe fixation by, 485, 486
Pfeiffer syndrome, 397
Phenylephrine, 18
Plagiocephaly, extorsion of orbit in, 53
Pontine glioma, comitantesotropiain, 155
Posterior fixation suture, adjustable, 501
alternative procedures to, 503–504
amounts of, 504t
augmentation of vertical rectus muscle transposition with, 362, 480, 481–482
computer modeling of, 107–111
complicated reoperations in, 504–505
complications from, 504–505
contraindications to, 497
exposure for, 497–498
for DVD, 497
for esotropia, 496
for incomitant strabismus, 492–496
for nystagmus, 496–497
for strabismus after scleral buckling, 305
Posterior fossa neoplasm, skew deviation due to Prentice position, 75
Pseudo-Brown syndrome, 329
Ptosis prop, 384
Postoperative adjustment, chemodenervation
Posterior segment, evaluation of, nystagmus, 416–417
Postoperative adjustment, chemodenervation
Pseudo-exotropia, 179
Pseudoptosis, in CFEOM, 366
Ptosis, from residual hemangioma of upper eyelid and exotropia, 147
in CFEOM, 365
in MED, 277, 279
in myasthenia gravis, 380, 381
in ocular myasthenia, 381, 382
in progressive external ophthalmoplegia, 385, 386
management of, 386
in 3rd cranial nerve palsy, 252, 254
mechanical, complete bilateral, after forehead advancement, 399
Ptosis prop, 384
PUCT. See Prism under cover test (PUCT).
Pupil dilation, after IO weakening, 457
Pursuit asymmetry, in infantile exotropia, 127, 128
Radiation therapy, for Graves ophthalmopathy, 292
Random dot stereoaucity test, 7
Random dot tumbler E test, 8
Receptive fields, characteristics of, cyclofusion and, 63

Rectus muscle(s), anatomy of, 529–530
3-D reconstruction of, 89
disinserted, 531
clinical diagnosis of, 532
incidence of, 532
prevention of, 536
surgical diagnosis of, 534
horizontal, overaction of, pattern strabismus from, 204
recession of, in nystagmus, 416–417
repositioning of, in treatment of pattern strabismus, 211
transposition of, in treatment of A and V patterns, 210–211
vertical transposition of, for hyperdeviations after scleral buckling, 305
inferior, avulsion of, 96
chemodenervation of, injection technique in, 431
combined paresis and restriction of, in orbital surgery, 318
entrapment of, in blow-out fracture of orbit, 306
fibrosis of, 368
late slippage of, after surgery for Graves ophthalmopathy strabismus, 293, 294
nasal displacement of, V-pattern exotropia from, 94
palsy of, 253
in orbital surgery, nerve disinsertion of, by slipknot technique, 531
pseudo-overaction of, in orbital fracture, 312
recession of, in skew deviation, results of, 391
restriction of, in orbital fracture, 310, 311
surgical management of, 317–319
tight, thyroid myopathy with, ophthalmoscopic view of fundus in, 69
transposition of, in Duane syndrome, 334
insertion of, visible line of previous, after surgery, 541
isolation of, for anterior ciliary vessel sparing, in reoperation, 513–514
lateral, anomalous actions/recruitment of, in Duane syndrome, 328
dehased abduction of, in orbital myositis, 265
effect of IO weakening on, 457
inferior displacement of, in myopic strabismus fixus, 95
innervation of, anomalous, in Duane syndrome, 326, 332
subnormal, in Duane syndrome, 340–341
paralysis of, cross-sectional area in, 92
from chordoma enveloping abducens nerve, 93
recession of, in Duane syndrome, 335–336
residual exotropia after, 509
superior displacement of, with nasal displacement of, SR muscles, A-pattern exotropia from, 95
tight, syndrome of, 195
after treatment of intermittent exotropia, 170
in intermittent exotropia, 170, 171
length-tension properties of, surgical outcome and, 81
lost, 531–532
clinical diagnosis of, 532

Rectus muscle(s) (Continued)
incidence of, 532
prevention of, 536
surgical diagnosis of, 534–535
surgical management of, 536–537
medial, anterior, computer reconstruction of, 102
avulsion of, from ethmoidectomy, 96
correction of, after retrobulbar anesthesia, 276
effect of posterior fixation suture on contralateral, in 6th cranial nerve palsy, 493
entrainment/restriction of, in orbital fracture, surgical management of, 319
equator underneath, estimated location of, based on axial length and corneal diameter, 76 t
medial orbitotomy approach to, 484
palsy of, 253
posterior fixation suture of, 498
recession of, in Duane syndrome, 333–334, 335
residual esotropia after, 509
secondary exotropia after, 387, 510
retraction of, anterior displacement due to, after surgery, 82
transsected, exotropia from, 95
MRI scan of, optimal patient and surface coil positioning for, 85
paresis of, coexisting with restricted rotation, 40
position and slibslip of, 90
pulley models of, 102, 104
pulleys of, average positions of, 89
recession of, adjustable, by slip knot technique, 437–439
large, 504
multiple, in both eyes, 503–504
in one eye, 503
posterior fixation suture with, 499–501
recession/resection/transposition of, in anterior ciliary vessel sparing, 523–524, 525
resection of, adjustable, by slip knot technique, 440–441, 445
severed, 531
slipped, 530, 531
clinical diagnosis of, 532
incidence of, 532
prevention of, 537
surgical diagnosis of, 533–534
surgical management of, 536
superior, chemodenervation of, injection technique in, 431
nasal displacement of, with superior displacement of, LR muscles, A-pattern exotropia from, 95
orientation of, after retrobulbar anesthesia, 376
incomitant DVD from, 246
palsy of, 253
posterior fixation suture of, 502–503
recession of, in DVD, 242–243, 245
transposition of, temporalward, in Duane syndrome, 334
undercorrection in, 487
tendon transposition in, full, 479
partial, 479
tenotomized, limiting number of, to reduce risk of ASI, 519
uck of, Wright’s modified, to preserve ciliary vessels, 526
union of, transposition procedure for, 479
vertical, horizontal transposition of, for cycloversion after scleral buckling, 303
palsy of, torsion analysis in, 66
transposition procedures for, 480–481
Strabismus (Continued)
surgically induced, 418
incatant, posterior fixation suture for, 492–496
lambda pattern, 205
in Duane syndrome, 329, 340, 342
treatment of, 213
latent, effect of, on motor measurements, 15
neurogenic, 380–392
differentiating features of, 381
paralytic. See also Cranial nerve palsy; Muscle
cpalsy.
chemodenervation in, 424–426
saccadic velocity testing in, 46
pattern, 202–214
anatomic factors in, 206
classification of, 202–203
complications of, 213
definition of, 202
diagnosis of, 207–209
etiology of, 204–207
examination of ocular rotations in, 208–
from muscle overaction/underaction, 204–
sensory deprivation, 206
heterotropic muscle pulleys and, computer
modeling of, 107
historical perspective on, 202
motor testing in, 207–208
prevalence of, 203
surgical technique in, 210–212
prevalence of 206
anomalous head posture after, 355
historical perspective on, 465
indications for, 468
symptoms of, 221
surgical, 225–227
torsion analysis in, 64–66
surgical, 225–227
algorithm for, 226–227
treatment of, 225–227
two-stage, 225–227
undercorrection in, 487
neurosurgical, 225–227
Among the Latent, 213
bilateral, 222, 468
Lancaster red-green plot in, 61
bilateral, 222, 468
classification of, 221–222
clinical characteristics of, 221–222
complications of, 227–228
congenital, 219–220
diagnosis of, 222–226
clinical, 222
differential, 222–223
laboratory, 222
etiology of, 219
iatrogenic, 545–546
after treatment of Brown syndrome, 354
Lancaster red-green plot in, 61
ophthalmoscopic view of fundus in, 65
overcorrection in, 227
signs of, 221–222
saccadic velocity testing in, 46
algorithm for, 226–227
surgical, 225–227
posterior tenectomy of, 462, 463–465
historical perspective on, 463
indications for, 463
techique of, 463–465
tenotomy/tenectomy of 459–465
alternative procedures to, 461–463
complications of, 460–461
historical perspective on, 459
in Brown syndrome, 353, 354
anomalous head posture after, 355
avoidance of intermuscular septum disrup-
tion in, 355
SO palsy after, 354
techique of, 459–460
transposition of, 484–485
historical perspective on, 484
mechanism of, 484
results of, 485
surgical technique in, 484–485
undercorrection of, grading scheme for, 17
weakening of, for cyclotorsion after scleral
buckling, 303
for treatment of pattern strabismus, 211, 212
Superior oblique tendon, absent, in SO palsy, 227
anterior displacement of, after scleral buck-
ling, 299
exaggerated traction test for, 56–57
functioning of, mechanical disadvantage of,
from shallow medial orbital wall, in
craniosynostosis, 395
MRI scan of, optimal patient and surface coil
positioning for, 85
myokymia of, 228–229
clinical features and diagnosis of, 228
complications of, 228
treatment of, 228
operation of, grading scheme for, 17
in Brown syndrome, 351
Superior oblique tendon (Continued)
diagrams for, 468
vtechnique of, 468–469, 470–471
Superior rectus muscle(s), chemodenervation
of, injection technique in, 431
nasal displacement of, with superior displace-
ment of LR muscles, A-pattern esotropia
from, 95
overcorrection of, after retrobulbar anesthesia,
379
incomitant DVD from, 246
palsy of, 253
monocular elevation deficiency from, 273
posterior fixation suture of, 502–503
recession of, in DVD, 242–243, 245
transposition of, temporalward, in Duane syn-
drome, 334
undercorrection in, 487
Suppression, 24–25
in intermittent exotropia, surgery and, 168
in strabismus after scleral buckling, 301
Surgery, complications of, 539–550
minor, 539–542
serious, 542–550
evaluation of, saccadic velocity testing in, 49
response to, anatomic factors in, 77–81
in intermittent exotropia, 169
measurement factors in, 73–82
method of testing and, 74
patient factors in, 73–81
surgical artifacts and, 81
surgical factors in, 81–82
testing factors in, 73–75
staging, to reduce risk of ASI, 519–520
timing of, 81
in infantile esotropia, 134
Surgical dose tables, for esotropia, 553
for exotropia, 554
for horizontal anomalous head posture, 415
for vertical deviations, 555
Surgical incision, choice of, and reducing risk
of ASI, 520
Surgical leash, in strabismus after glaucoma
implant, 304–305
Surgical techniques. See also specific
technique.
adjustable and nonadjustable, 437–447
Suture material, for IO weakening, 456
for posterior fixation suture, 497
Symblepharon, secondary to Stevens-Johnson
syndrome, 543
Synoptophore, 27, 32
Teller acuity cards, 6
Tenon’s capsule, 529–530
Thalamic disease, comitant esotropia in, 156
Three-step test, 15–16
in SO palsy, 222
Thyroid myopathy, with tight IR muscle,
ophthalmoscopic view of fundus in, 69
Thyroid-related ophthalmopathy. See Graves
ophthalmopathy.
Tight lateral rectus syndrome, 195
after treatment of intermittent exotropia, 170
in intermittent exotropia, 170, 171
Time of day, effect of, on results of saccadic
velocity testing, 46
Torsion, 52–72
anatomic, 52
measurement of, 52–60
assessment of, clinical applications of, 64–71
in craniosynostosis syndromes, 400
in SO palsy, 222
definitions relating to, 52
Vertical deviation (Continued)
after scleral buckling, 298–299
concomitant, in intermittent exotropia, 170
dissociated, 237–247
classification of, 239
clinical evaluation of, 239–240
clinical presentation of, 238–239
comitant, surgical procedures for, 242–243
complications of, 245–247
definition of, 237
diagnosis of, 239–241
differential diagnosis of, 241
epidemiology and risk factors for, 238
genes of, 238
historical perspective on, 237
in infantile esotropia, 131–132
incomitant, 240
surgical procedures for, 243–245
laboratory evaluation of, 240–241
measurement of, 239–240
posterior fixation suture for, 497
surgical dose table for, 555t
synonyms for, 237t
theories of, 237–238
torsion analysis in, 66–67
treatment of, 241–245
nonsurgical, 241–242
surgical, 241–242
indications for, 242
issues related to, 242
from orbital floor fracture, 312
induced, after transposition procedures, 487
in 6th cranial nerve palsy, 186
reoperation for, 511–512
surgical dose tables for, 555t
Vestibular nystagmus, 408
Vestibulo-ocular pathway, brain stem, in
infantile esotropia, 119
Vestibulo-ocular reflex, abnormalities of, in
congenital nystagmus, 406
Vision assessment, 4–7
fixation patterns and, 4–5
in nystagmus, 6–7
in preverbal children, alternate methods of,
5–6
in verbal children, 6
visual milestones and, 4
Visual acuity, in nystagmus, 405
measurement of, 408
prisms to improve, 412
surgery to improve, 416–417
normal, 22
Visual confusion, as chief complaint, 3
in adults, 27–28
in intermittent exotropia, surgery and, 168
Visual cortex, in infantile esotropia, 118–119
Visual evoked potential (VEP), in assessment of
treatment of preverbal children, compared to other
methods, 5
in DVD, 241
motion, asymmetry of, in infantile esotropia,
129, 130, 133
Visual field defects (Continued)
bilateral homonymous, 186–188
clinical characteristics of, 188
genetic and early-onset, 186–188
diagnosis of, 189
incidence of, 186
Visual mechanisms, cerebral, in infantile
esotropia, 119–121
Visual milestones, 4
Visual pathways, in infantile esotropia,
subcortical, 118
vs motor, 118
Vomiting, postoperative, 539–540
incidence of, 539–540
prevention of, 540
treatment of, 540
V-pattern exotropia, 204
from inferior oblique overaction, 208, 209
from nasal displacement of IR muscle, 94
treatment of, 212
V-pattern exotropia, 204
craniofacial synostosis associated with, 395, 397
in Brown’s syndrome, 348, 350
inferior oblique overaction in, 18
IO overaction in, 18
lax tendon in, 211
treatment of, 212–213
V-pattern strabismus, 206
in craniostenosis syndromes, 393–395
management of, 401
prevalence of, 206t
Wall-eyed bilateral internuclear
ophthalmoplegia, 389
W4D test, See Worth four-dot (W4D) test.
WEBINO, See Internuclear ophthalmoplegia,
wall-eyed bilateral.
Worth four-dot (W4D) test, 8–9, 32
X(T). See Exotropia, intermittent.
X-pattern exotropia, 205
in Möbius syndrome, 360
Y-pattern exotropia, 205
Y-pattern strabismus, in Duane syndrome, 329, 340, 342
treatment of, 213