ERIK M. WEISSBEI

ESSENTIALS OF Clinical Binocular Vision

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Foreword

Why another book about binocular vision? After all, there are currently available many excellent texts for the reader to choose from–Scheiman and Wick, Rutstein, Griffin, Rouse and Caloroso, etc.–each a fine text approaching the subject of binocular vision in a scholarly and comprehensive manner and differentiated by philosophy, style, and experiences of the authors. Although each is suitable for use by the student of binocular vision or the advanced clinician, none are designed to be readily available and approachable to a clinician with limited knowledge in the subject and a patient in his or her chair. This book, *Essentials of Clinical Binocular Vision*, fills that niche of a clinically useful and relevant tool in the management of the patient with binocular vision anomalies.

Optometry has undergone extraordinary progress and change during my 25+ years in pediatric optometry. When I was a student at the New England College of Optometry in the early 1970s, the diagnosis and treatment of eye disease was but a far-off dream of our profession. Clinical practice was concentrated on refractive care and binocular vision. My contemporaries all received extensive experience in these areas during our training and in our clinical practice. There were relatively few books available on binocular vision (actually few books written by optometrists at all), but there were lots of mimeographed manuals with extensive and explicit instructions on the diagnosis and treatment of binocular anomalies that were developed by our optometric faculty. We were comfortable with clinical care of patients with binocular problems, even as students.

This now seems like a long time ago (it was!). Our profession has changed tremendously. Disease happened in a far bigger way

than almost any of us could have imagined. The curriculum at all of the optometry schools transformed from a functional orientation to an intensively medical model. There was a veritable explosion of content in the basic sciences. Models of clinical care progressively emphasized more disease and less visual function. The strength that had formerly existed in the teaching and clinical care in binocular vision waned dramatically, to the point today that it is a small, narrow, and often underappreciated area of optometric education and clinical care.

However, starting a few years ago, a new trend in optometry developed. Led first by a group of optometrists who became convinced that the optometric care of the infant was the next frontier in our profession (i.e., Operation Bright Start and my text, *Eyecare for Infants & Young Children*), and now more recently through the nationwide efforts of the American Optometric Association and a number of individual state associations to mandate universal eye exams for all children entering school, children have been rediscovered by our profession. There is now active discussion about specialization and certification in pediatric optometry, and there already is in binocular vision through OEP and COVD.

Those of us who consider ourselves to be specialists in binocular vision and pediatric optometry are both pleased and a little amazed by this sudden re-awakening of interest in children. However, many of us are concerned as well. We know that within the profession as a whole, binocular vision and pediatric optometry have not been where the action was, and those optometrists educated in the last 15 or so years had relatively little didactic and clinical education in these areas. Of course this is not the case for those who have active practices in functional or behavioral optometry or those specializing in the care of the child. Nonetheless, probably a majority of practicing optometrists today have relatively little experience in binocular vision and pediatric optometry. However, as a result of the strenuous efforts of organized optometry to bring more children into our practices, this is going to change dramatically in the future. Based on manpower studies of practicing eye care professionals in the United States, it can be expected, if optometry is successful in legislating mandates for universal eye exams for children, that many optometrists will be seeing dozens, if not hundreds more children in their practices each year. Many of these children will

have binocular anomalies requiring diagnosis and treatment, and many of the adults that are currently being seen will be newly discovered to have binocular vision anomalies also requiring remediation. There is a need to increase our efforts to properly educate students and practitioners in their care.

Thus the reason for this manual. Simply stated, to provide an accessible, maneuverable, and ready means of aiding the student and clinician to care for patients with binocular vision anomalies. This manual is not intended to supplant the role of the comprehensive text in providing a proper foundation in binocular vision. As I said before, there are many excellent texts that do this well. There is not, however, a single source that can be used on a day-to-day basis to guide the student or practitioner actively engaged in clinical practice to best serve the patient. The format of this manual follows the style of the *Wills Eye Manual*, which has become the universal handbook of ocular disease: extremely accessible, clear, comprehensive, and perhaps most importantly, easy to use under the pressure of active clinical practice. It is my belief that this manual will fill a critical void in helping the clinical optometrist to care for patients of all ages with binocular anomalies, to make that care available to all ODs, even those with minimal experience or interest, and to make them comfortable in providing that care by using a simple and standardized methodology of diagnosis and treatment that is consistent with efficient and effective clinical care.

I am delighted to have participated in the genesis and production of this manual. For me, this has been emblematic of the reconstitution of binocular vision and pediatric optometry to what I believe is its central role in our profession, and specifically to the New England College of Optometry. There has been a dramatic development of this department since I returned to the College from the Boston Children's Hospital and the Harvard Medical School 6 years ago, where I practiced for the preceding 22 years. Senior faculty have been re-energized by the increasing emphasis on binocular vision and pediatric optometry within our institution and within the profession. Best of all has been the development of an active, talented, and confident group of young faculty with a strong commitment to providing a superior education and patient care in binocular vision and pediatric optometry. Watching the growth and development of this extraordinary group of young educators and clinicians has been

the most satisfying professional experience I have had. I see this same thing happening in many other programs around the country, and can't help but think that the next generation is in place to bring this area to the prominence within the profession that is essential for optometry's advancement. Erik Weissberg has led our colleagues at the New England College of Optometry in a wonderful effort to produce a very important source of clinical knowledge and utility for our profession. I expect that it will fill a void that many of us believe exists, and lead to the student and practitioner becoming more comfortable and effective in delivery care to their patients.

Bruce Moore

Preface

The motivation and concept for this book was spawned from my early experiences as an optometric student and later confirmed after several years as an optometric educator. Originally I observed classmates and now I am observing my students, who leave the exam room somewhat perplexed by a binocular vision problem they may have just encountered. If the condition was of the disease variety, there were several textbooks suitable as the first book off the shelf. However, in the area of binocular vision, there was no book to serve in this capacity. With a multitude of excellent and exhaustive textbooks that explain "why", there was an absence in the field of binocular vision of texts whose focus was simply to explain "what" to do. The purpose of this book is to fill that void. Used in conjunction with the more exhaustive types of texts, I believe that the knowledge needed to diagnose and manage patients with binocular vision problems will ultimately become more readily accessible to the clinician, leading to better and more comprehensive care for the patient.

The format and content of this book follows a unique approach to presenting clinically relevant information in this topic area. Directed at the primary care practitioner, it is suitable for anybody that has ever asked a question concerning binocular vision and wished that the person they asked would just give them the answer instead of all the background and theory. Special care has been taken to keep the text as succinct as possible while still providing the appropriate information and direction for the clinician.

The first part of the book presents specific clinical conditions organized by signs, symptoms, differential diagnosis, work-up, treatment, and follow-up. Each condition is written so it can

stand alone, but the potential to link with additional information is easily accomplished if needed. Specifically, if a clinical condition is mentioned as part of the differential diagnosis, a page number link is provided for the reader who would like more information on this topic. Furthermore, if a diagnostic or therapeutic technique is mentioned as part of the work-up or management, a page number link is provided to direct the reader who requires a step-by-step description of how to perform and interpret the technique. In this way, the original clinical condition sought after by the reader remains streamlined, with the pertinent information extracted quickly and easily.

The next major section of the book presents a step-by-step description of clinically relevant diagnostic techniques used in the work-up of binocular vision disorders. Care was taken to include those techniques that are easily performed in a primary care setting with minimal investment in special equipment. The clinical tools chosen are versatile and provide valuable information when dealing with patients of this variety.

The book concludes with two sections specially dedicated to the diagnosis and management of non-strabismic near point binocular vision disorders. These two sections were included because of the high prevalence of these conditions encountered in optometric practice. This is the area in which optometrists may be the most well suited to make the diagnosis and implement the management. First, a clinically relevant approach to diagnosing these problems is presented. The approach is specifically designed to be easily integrated into a primary care exam. The second section presents a general vision therapy program that is flexible enough to treat patients with any of the most common near point binocular conditions.

It is not a secret that optometry has slowly been shifting away from binocular vision while placing increased emphasis on ocular disease. Today's recent graduate has vast knowledge of pathological conditions, but may be very uncomfortable when it comes to diagnosing common binocular vision disorders. Combine this with the fast pace of clinical practice, and it is not surprising that binocular vision disorders often go overlooked. Clinicians repeatedly express that they just don't have the time to perform a binocular work-up as part of their examination. As educators, we often express our dismay and concern about this problem, but it is time to begin to do something about it. We

need to begin teaching in a way that will allow for the routine testing of binocular vision conditions in a primary care setting. We need to teach practical binocular vision for the primary care doctor as opposed to expecting everyone to be a specialist.

This book is something that I would have benefited from greatly during my early education. It is not trying to be something more than what it is: an easily referenced, concisely written manual to direct the primary care clinician in the diagnosis, management, and appropriate referral of patients with binocular vision disorders.

Erik M. Weissberg

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I would like to express my sincere gratitude to the library staff of the New England College of Optometry, especially Claire Rork for her patience, humor, and expertise.

I would be remiss if I didn't specifically thank several people that have been influential in my career thus far. Without the advice, guidance, inspiration, and opportunities afforded to me by Bruce Moore, Jack Richman, and Stacy Lyons, my career in academia may have never even begun.

Erik M. Weissberg

1

Non-Strabismic Binocular Vision Anomalies

RICHARD C. LAUDON AND ERIK M. WEISSBERG

1.1 Convergence Insufficiency

GENERAL INFORMATION

Convergence insufficiency (CI) is the most commonly recognized binocular dysfunction. This typically idiopathic and benign condition represents a mismatch between a patient's visual capabilities and near point demands. Presenting symptoms may be mild to severe with a gradual onset. This may occur in elementary school, in college, or on the job as a result of increased workload. The majority of CI patients will have an associated accommodative dysfunction.[1,2] In some cases, presenting symptoms may be coincident with the onset of presbyopia (reduction in accommodative function aggravating a preexisting fragile state of binocularity). CI has also been associated with reading difficulties and ADD.[3]

SYMPTOMS

Some or all of the following symptoms may be reported after or during extended periods of reading:

- Double vision at near
- Words moving around the page and/or words swimming
- Eye pulling or straining
- Eye fatigue and/or eye strain

- Headaches *above the eyes*, later in the day, associated with reading and/or computer use
- Blurred vision or focusing problems at near
- Limited visual attention/concentration
- Avoidance of near point tasks

SIGNS

Reliable measurements should be confirmed by repeating abnormal test findings or by documentation of multiple measurements confirming the same diagnosis.

- Receded near point of convergence is greater than 6 cm.[4] Immediate diplopia may be noted when the test is repeated while a red lens is held over one eye.
- Exophoria is present at near, typically greater than 6 to 8 prism diopters. Intermittent exotropia may be noted during near testing.
- Near lateral phoria testing may reveal higher exophoria than predicted by the cover test.
- Low positive fusional vergences based on Sheard's criterion. (Compensating vergences must be twice the phoria measurement.)
- Low AC/A ratio (norm is 4:1).
- *Accommodative facility:* Difficulty clearing plus lenses oculus uterque (OU).
- Low negative relative accommodation (NRA)/High positive relative accommodation (PRA) (norm for NRA is +2.50, PRA –3.50).
- *Accommodative dysfunction (p. 20):* Signs of an associated accommodative dysfunction are typically present. (Some reports suggest accommodative insufficiency is most common,[1] but it is the author's experience that accommodative excess is most common.) Specific findings (monocular facility, amplitude, and Monocular Estimation Method [MEM] or FCC) depend on the type of accommodative dysfunction present.

DIFFERENTIAL DIAGNOSIS

- *Uncorrected refractive error:* Hyperopia, myopia, astigmatism, and especially anisometropia may be obstacles to visual effi-

ciency. After a prescription has been determined, a progress evaluation should reassess the patient's near point function before one considers additional intervention.

- *Pseudoconvergence insufficiency:* The primary problem is an accommodative insufficiency that causes a secondary convergence insufficiency. Use of a plus add will improve the patient's near point of convergence. Vision therapy may still be needed to resolve any residual difficulties.
- *Basic exophoria:* Large exophoria, equal in deviation at distance and near. Occasionally an intermittent exotropia may be present. If the deviation is intermittent and the patient's condition is symptomatic, vision therapy is the preferred intervention. Prism spectacles may be a helpful alternative in certain cases.
- *Divergence excess (p. 58):* Exophoria or exotropia is more significant at distance than near.
- *Convergence palsy:* Typically an acute onset of severely limited convergence with associated neurological signs and symptoms.
- *Myasthenia gravis (p. 133):* May result in reduced convergence secondary to weakness of the medial rectus muscle. Ptosis, diplopia, blurred vision, and incomplete lid closure may also be present.
- *Vertical phoria:* A vertical imbalance from a CNIV paresis (p. 93) can cause a secondary binocular dysfunction. A head tilt indicates the possibility that this type of deficit is present.

WORK-UP

1. *History:* Are symptoms consistent with the functional etiology (onset, type, association with eye use)? Severity of symptoms (do they interfere with school or office work)? Associated neurological symptoms?
2. *Correct any refractive error:* If a significant refractive error is present, recommend a progress evaluation in 1 to 2 months to reassess binocular and accommodative function with spectacles.
3. *Distance binocular profile:* Measurement of distance phoria and compensating vergence range should be performed to rule out divergence excess (p. 58) and basic exophoria.
4. *Near point binocular vision work-up (p. 186):* Cover test, near lateral phoria, near point of convergence (NPC) (light, red glass and through +1.00), AC/A ratio, fusional vergence ranges,

accommodative facility, amplitude of accommodation, and monocular estimation method or FCC.

5. *Stereopsis:* Local (Wirt circles) and random-dot stereopsis should be assessed. Absence of or reduced stereopsis should raise one's suspicions about the presence of a strabismus.
6. *Cycloplegic refraction (p. 179):* Rule out latent refractive error when indicated.

TREATMENT

- *Home/Office-based vision therapy (p. 197):* Considered to be the treatment of choice for CI, vision therapy has been shown to be effective.[5-7] The duration of treatment may vary, but approximately 2 to 3 months with good compliance are usually adequate. One-hour office visits every 1 or 2 weeks to monitor progress and implement changes are required.
- *Pencil push-up therapy (Box 1.1-1):* An easier and quicker approach to the treatment of CI. This therapy requires minimal office visits, but the lack of patient compliance and motivation can be a major obstacle. This therapy has been shown to alleviate symptoms in some patients.[8]
- *Visual hygiene:* Patient education concerning multiple breaks of short duration during near point tasks. Fixation should be

BOX 1.1-1
Directions for Pencil Push-up Therapy

1. Hold and fixate on pencil tip at 40 cm.
2. Slowly move pencil tip toward the nose until it "doubles."
3. Move pencil tip 1 to 2 cm away from nose until it is "single" again.
4. With pencil tip held at the distance determined in step 3, shift fixation to a distance target (approximately 3 meters away).
5. Shift fixation between the pencil tip and distance target 10 times, holding fixation on each for 3 seconds.
6. Repeat steps 1 to 5 for a total of 3 to 5 minutes daily until symptoms are alleviated.

Goal: Move pencil tip 2 to 3 cm from nose and retain single and comfortable vision.
Patients can be further instructed to note that when they look at the pencil tip, the distance target should appear double and vice versa. This phenomenon is normal and serves as a suppression check during this exercise. If this observation is not appreciated by patients, they should be encouraged to pursue a complete vision therapy program (p. 197).

upon a distance target during the frequent rests of 1 to 2 minutes. The patient's attention can be drawn to the importance of maintaining an adequate and consistent working distance (40 cm or Harmon's distance) throughout near point work. Harmon's distance is the length between the elbow and the middle knuckle of the index finger.

- *Spectacles:* Although not the treatment of choice, lenses can improve visual efficiency and adequately alleviate symptoms in certain cases.
  - *Addition lenses:* The use of plus lenses for reading can benefit those CI patients with an associated accommodative insufficiency. A proper binocular work-up (p. 186) will clearly identify those patients who may fall under this category. The power of the lenses falls typically between +.75 and +1.25, with the optimal prescription determined through trial framing and the repetition of abnormal binocular tests through the addition lens (i.e., NPC, NRA/PRA, facility testing, MEM).
  - *Prism:* The use of a base-in prism in combination with plus lenses or by itself may benefit certain CI patients. First perform a prism adaptation test (p. 172). Only consider a prism prescription if the patient is not a prism adapter. Magnitude typically falls between 2 to 6 total prism diopters, split between the two eyes (e.g., 3 prism diopters would be prescribed as 1.5 pd base-in OD and 1.5 pd base-in OS). Satisfy Sheard's criterion to determine the starting prism power (Box 1.1-2). Refine power through trial framing for comfort. Patients who are going to benefit from a prism typically have a positive reaction during trial framing.

FOLLOW-UP

Because CI, with the exception of symptoms, is considered a benign condition, cases can be followed depending on treatment.

- *Vision therapy:* Followed every 1 to 2 weeks until completion
- *Spectacle intervention:* Initially followed in 1 to 2 months to evaluate effectiveness of glasses
- *Education or visual hygiene only:* Followed yearly or as needed if symptoms increase

BOX 1.1-2
Sheard's Criterion and Determination of Prism Spectacles for CI Patients

1. According to Sheard's criterion, the compensating vergence should be twice the phoria. If the criterion is not met, the patient is likely to have symptoms.[9] If the criterion is satisfied, symptoms are likely to be relieved.[10]
2. The least amount of prism that satisfies Sheard's criterion should be the starting point for the trial framing of a prism prescription.
3. A patient with an exophoria of 12 and a base-out break point of 18 is likely to be symptomatic, because Sheard's criterion has not been met.
4. Prescribing 3 prism diopters base-in would reduce the exophoria to 9 and increase the base-out break point to 21. This prescription would satisfy Sheard's criterion and should be used as the starting point for trial framing.
5. If satisfying Sheard's criterion necessitates the use of greater than 10 to 12 total prism diopters, a prism prescription is not likely to prove successful. The patient should be strongly encouraged to pursue a complete vision therapy program (p. 197).

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1.2 Convergence Excess

GENERAL INFORMATION

Convergence excess (CE) is a common binocular dysfunction[1] that can cause significant visual symptoms. Symptom onset is typically experienced during or in association with extended near point tasks, such as reading or computer work. It is not uncommon to find an associated accommodative dysfunction (p. 20) when convergence excess is present. Symptoms resulting from CE can often be alleviated by the use of plus lenses for reading, or sometimes by vision therapy.[2]

SYMPTOMS/BEHAVIORS

Symptoms typically experienced after or during prolonged near point tasks include the following:

- Print swimming or moving around, and/or words running together
- Diplopia
- Eye strain/fatigue
- Pulling sensation around the eyes
- Headaches above the eyes late in the day
- "Over-focusing" problem
- Blurred vision (may be reported at distance or near)
- Avoidance of near point activities

The following behaviors may be observed in CE patients:

- Holding reading material close
- Tendency to close an eye
- Possible head tilt after visual fatigue (may be noticed at the end of the exam)

SIGNS

The magnitude of esophoria has been shown to correlate with the presence of symptoms.[3-4]

- Esophoria is greater at near than at distance.
- High AC/A ratio (greater than 5:1).
- Low divergence ranges (BI) in comparison with the phoria and/or highly elevated convergence ranges (BO) at near.
- *Near point of convergence (NPC):* To the nose.
- *Accommodative facility:* Difficulty or inability clearing minus lenses OU.
- *High NRA/Low PRA (less than 1.25):* May be associated with convergence excess but does not exclude other possible binocular vision disorders.[5]
- *Accommodative dysfunction (p. 20):* Signs of an associated accommodative dysfunction are typically present (most commonly accommodative excess). Specific findings (monocular facility, amplitude, and MEM or FCC) depend on the type of accommodative dysfunction present.

DIFFERENTIAL DIAGNOSIS

- *Refractive error:* An uncorrected refractive error (typically hyperopia or overcorrected myopes) can mimic a convergence excess. Symptoms and signs will typically be alleviated once the refractive error is corrected. Cycloplegic refraction should be performed.
- *Basic esophoria:* The magnitude of the esophoria is similar at both distance and near. Treatment should be focused on eliminating the esophoria through lenses or vision therapy.
- *Divergence insufficiency (p. 53):* Esophoria is greater at distance than at near. Occasional diplopia in the distance may be reported. This is a rare condition; suspect lateral rectus weakness and consider a neurological consult.
- *Pharmacological causes:* Certain drugs can cause overconvergence and an accommodative spasm (e.g., Eserine, Pilocarpine).[1] Careful patient history should elicit a connection between the onset of symptoms and the beginning of or change in the dosage of the medication.
- *Convergence spasm:* Characterized by intermittent episodes of esophoria at near, accommodative spasm, and pupil constriction. The spasm may result from a serious underlying pathologic cause, from local inflammation, or more commonly as a result of a "hysteric" or neurotic personality.[6] Tubular fields may be present in the latter scenario.

WORK-UP

1. *History:* Are symptoms consistent with the functional etiology (onset, type, association with eye use)? Severity of symptoms (do they interfere with school or office work)? Recent change in medications?
2. *Correct any refractive error:* If a significant refractive error is present (especially hyperopia), recommend a progress evaluation in 1 to 2 months to reassess binocular and accommodative function with spectacles.
3. *Cycloplegic refraction (p. 179):* Needed in cases in which latent hyperopia is suspected or refractive error is fluctuating. Esophoria at near may be a sign of latent hyperopia.
4. *Distance binocular profile:* Distance phoria testing should be performed and compensating vergence range measured to rule out divergence insufficiency (p. 53) and basic esophoria.
5. *Near point binocular vision work-up (see p. 186):* Cover test, near lateral phoria, NPC (light, red glass, and through +1.00), AC/A ratio, fusional vergence ranges, accommodative facility, amplitude of accommodation, and monocular estimation method or FCC.
6. *Stereopsis:* Local (Wirt circles) and random-dot stereopsis should be assessed. Absence of or reduced stereopsis should raise one's suspicions about the presence of a strabismus.
7. *Pupils:* Rule out miosis associated with a convergence spasm.
8. *Visual fields testing:* Performed in cases when a "hysterical" cause is suspected.

TREATMENT

- *Plus reading lenses for near:* Treatment with plus reading lenses is considered to be the most effective. The magnitude of the esophoria at near has been directly correlated to the likelihood of symptoms, and as such the effort to alleviate symptoms should be focused on eliminating the esophoria.[3-4] A typical near prescription ranges from +.75 to +1.25 if hyperopia is not present. (This may be higher if the patient is hyperopic.) Repeat the modified Thorington (p. 153) or cover test to ensure elimination or adequate reduction of esophoria at near while the patient is wearing the spectacles. Perform a trial framing of the near prescription to ensure the patient's comfort and recheck near visual acuity (VA). The response to near point

lenses is critical. Patients should report that the print is larger and that their eyes feel more comfortable, and they should be able to hold books farther away. Signs that a patient is unable to accept the plus lens at near are minimal elimination of the esophoria, blurring of near VA, and a negative reaction to the glasses during trial framing.
  - Single-vision spectacles exclusively for near vision can be prescribed if the patient has symptoms only during or after prolonged near tasks.
  - *Bifocal:* Consider prescribing a bifocal if a patient's visual demands require continual alternation between distance and near fixation (e.g., looking from board to desk in school), or if esophoria is present in the distance with hyperopia. Flat top 28 with the segment set 3 mm below the middle of the pupil is usually an adequate lens design. Glasses can be worn during all symptom-producing tasks.
- *Home/Office-based vision therapy (p. 197):* Although less effective than convergence therapy, divergence therapy can be an important adjunct in certain cases. It is used when the patient is adverse to lenses or unable to accept plus lenses at near, or when lenses result in marginal improvement. Duration of the treatment may vary, but approximately 2 to 3 months with good compliance are usually adequate. One-hour office visits every 1 or 2 weeks to monitor progress and implement changes are required. Besides the emphasis on fusional ranges, accommodative therapy is also important to improve the patient's plus acceptance at near. Within a short period of time (2 to 6 weeks) lens therapy may be reconsidered.
- *Visual hygiene:* Patient education concerning multiple breaks of short duration during near point tasks. Fixation should be upon a distance target during the frequent rests of 1 to 2 minutes. The patient's attention can be drawn to the importance of maintaining an adequate and consistent working distance (40 cm or Harmon's distance) throughout near point work. Harmon's distance is the length between the elbow and the middle knuckle of the index finger.

FOLLOW-UP

Because CE, with the exception of symptoms, is considered a benign condition, cases can be followed depending on treatment.

- *Vision therapy:* Followed every 1 to 2 weeks until completion
- *Spectacle intervention: Initially followed* 1 to 2 months to evaluate the effectiveness of the glasses
- *Visual hygiene only or asymptomatic case:* Followed yearly or, if symptoms increase, as needed

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1.3 Fusional Vergence Dysfunction

GENERAL INFORMATION

Fusional vergence dysfunction (FVD) is an often overlooked binocular vision disorder characterized by abnormal binocular findings that do not fit into a specific pattern, or by a mismatch between the patient's symptoms and marginal findings during the binocular work-up. This may make the condition more difficult to diagnose than a convergence insufficiency or convergence excess. Although typically found in school-age children, this profile may appear in adults as a result of their attempt to compensate for a long-standing binocular dysfunction, such as a convergence insufficiency. Among other names, FVD has also been referred to as a general skills case, binocular instability, or a sensory fusional deficiency.[1-3]

SYMPTOMS

- Blurred vision at distance and/or near
- General discomfort associated with near tasks

- Symptoms increase over time or occur later in the day
- Loss of attention or concentration during extended near point visual tasks
- Avoidance of sustained near point tasks
- Double vision or a sensation that words are moving around the page
- Vision-related complaints not explained by an initial binocular work-up (i.e., cover test, near point of convergence, and stereopsis)
- *Headaches above the eyes:* Experienced at the end of the day, school- or work-related, associated with near work.

SIGNS

Not all findings need be present to diagnose fusional vergence dysfunction. Findings may also fluctuate from visit to visit or be greatly influenced by appointment time (morning vs. evening, weekday vs. weekend).

- *Normal or marginally abnormal phoria measurement*: Phoria measurements may be different at the beginning of the exam and at the end.
- *Reduced negative and/or positive fusional vergences*: An especially poor recovery may be noted during vergence testing.
- *AC/A ratio:* Typically normal. (Normative value is 4:1.)
- *Near point of convergence:* Marginally receded (greater than 6 cm) or normal.
- Patients may have reduced sensory fusion at near.
- Negative relative accommodation (NRA) and positive relative accommodation (PRA) may both be reduced. (Norm for NRA is +2.50, PRA -3.50.)
- *Accommodative facility testing (+2/–2):* Greater difficulty clearing both plus and minus when the testing is done binocularly than when it is done monocularly.
- *Vergence facility (3BI/12BO) (p. 158):* Reduced ability (less than 15 cycles per minute).[4]

DIFFERENTIAL DIAGNOSIS

- *Uncorrected refractive error:* Hyperopia, myopia, astigmatism, and especially anisometropia may be obstacles to visual efficiency. After a prescription has been determined, a progress

evaluation should reassess the patient's near point function before one considers additional intervention.

- *Aniseikonia:* A rare condition in which the difference in perceived image size is an obstacle to fusion. Aniseikonia is more commonly found in patients with high anisometropia or after surgery (retinal detachment, cataracts). Perform a thorough work-up to rule out other binocular dysfunctions, carefully assess refractive error, and refer the patient to a specialist if aniseikonia is still suspected. Small modifications in lens design (base curve, index, thickness, vertex distance) may be used to treat some cases.
- *Vertical phoria:* A vertical imbalance (i.e., CNIV paresis *[p. 93]*) may result in an obstacle to fusion and a secondary binocular vision dysfunction. Head tilt may indicate the possibility of this type of deficit. A vertical prism is typically successful in correcting it.
- *Myasthenia gravis (p. 133):* May mimic any near point binocular problem in both symptoms and signs. This condition is characterized by fluctuation of findings from visit to visit, with no specific pattern to the binocular dysfunction. Ptosis, diplopia, blurred vision, and incomplete lid closure may also be present. An increase in symptoms and signs may be noted during repetitive tasks.

WORK-UP

Time of day may greatly impact the outcome of the binocular findings (e.g., morning evaluation may be inconclusive especially if symptoms are occurring in the late afternoon).

1. *History:* Are symptoms consistent with functional etiology (onset, type, association with eye use)? Severity of symptoms (do they interfere with school or office work)? Associated neurological symptoms?
2. *Externals:* Ptosis may be sign of myasthenia gravis. Head tilt may indicate vertical deviation.
3. *Correct any refractive error:* If a significant refractive error is present, recommend a progress evaluation in 1 to 2 months to reassess binocular and accommodative function with spectacles.
4. *Distance binocular profile:* Phoria testing should be performed and compensating vergence range measured if patient has distance symptoms.

5. *Near point binocular vision work-up (see p. 186):* Cover test, near lateral phoria, NPC (light, red glass, and through +1.00), AC/A ratio, fusional vergence ranges, accommodative facility, amplitude of accommodation, monocular estimation method or FCC, and vergence facility. NPC and/or facility testing may be repeated at the end of the exam to investigate the effect of fatigue.
6. *Stereopsis:* Local (Wirt circles) and random-dot stereopsis should be assessed. Absence of or reduced stereopsis should raise one's suspicions about the presence of strabismus.
7. *Cycloplegic refraction* (p. 179): Rule out latent refractive error when indicated.

TREATMENT

- *Home/Office-based vision therapy (see p. 197):* A broadly based vision therapy program concentrating on both vergence and accommodation is the treatment of choice. In this case, vision therapy may take on a diagnostic value as well as a therapeutic value. The patient's progress during the course of training will often make clear the strengths and weaknesses of the patient's specific visual skills. The duration of treatment may vary, but approximately 2 to 3 months with good compliance are usually adequate. One-hour office visits every 1 or 2 weeks to monitor progress and implement changes are required.
- *Addition lenses:* Although not the treatment of choice, plus lenses for near can improve visual efficiency and alleviate symptoms when an accommodative dysfunction (p. 20) coexists. Results of the accommodative work-up should be carefully interpreted for signs of plus acceptance. A high NRA and/or low PRA, a high lag of accommodation on the monocular estimation method (p. 159) or fused cross-cylinder test, and difficulty clearing minus lenses during accommodative facility testing are all signs of plus acceptance. The power of the lenses typically falls between +.75 and +1.25, with the optimal prescription to be determined by trial framing and the repetition of abnormal binocular tests through the addition lens.
- *Visual hygiene:* Patient education concerning multiple breaks of short duration during near point tasks. Fixation should be upon a distance target during the frequent rests of 1 to 2 minutes. The patient's attention can be drawn to the importance

of maintaining an adequate and consistent working distance (40 cm or Harmon's distance) throughout near point work. Harmon's distance is the length between the elbow and the middle knuckle of the index finger.

FOLLOW-UP

Because FVD, with the exception of symptoms, is considered a benign condition, cases can be followed depending on treatment

- *Vision therapy:* Followed every 1 to 2 weeks until completion. If symptoms have not resolved, seriously reconsider the initial diagnosis.
- *Spectacle intervention:* Initially follow in 1 to 2 months to evaluate the effectiveness of glasses.
- *Education or visual hygiene only:* Followed yearly or, if symptoms increase, as needed.

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1.4 Functional Ocular Motor Dysfunction

GENERAL INFORMATION

Ocular motor dysfunction is a common type of visual skills deficit found in children with learning difficulties.[1] It may become more apparent in children between the ages of 5 and 13 years because of increased reading demands and symptoms noted during the act of reading. A functional ocular motor disorder typically affects pursuits, saccades, and fixations; it does not exhibit an asymmetry (i.e., there is no gaze-dependent difference); and it has a gradual onset. This disorder must be differentiated from a pathologic etiology that may affect only one

of the major eye movements (pursuits *or* saccades), that can exhibit asymmetry, and that typically has a more sudden onset. Children with ocular motor disorder will often be referred because of a "tracking" problem, and they may have associated binocular and accommodative anomalies.[2]

SYMPTOMS

Symptoms typically experienced during the act of reading include the following:

- Loss of place
- Frequent and repeated rereading of lines
- Excessive head movements when reading
- Skipping words or sentences
- Difficulty copying from "whiteboard" to paper
- Problems with accuracy when filling out "scantron" type tests
- Eye-hand coordination problems

SIGNS

The following behaviors may be observed during clinical testing:

- Poor fixation during the cover test
- Using a finger to direct fixation during acuity testing
- Erratic fixations and apparent lack of concentration during version testing

Performance-based signs of ocular motor dysfunction include the following:

- Below-normal performance on visual-verbal ocular motor tests, such as the developmental eye movement test (DEM)[3] (p. 167)
- Below-normal performance on observational ocular motor tests, such as the NSUCO (p. 164)[4]
- Below normal-performance on electrophysiological ocular motor tests, such as the Visagraph II

DIFFERENTIAL DIAGNOSIS

- *Uncorrected refractive error:* Hyperopia, myopia, astigmatism, and especially anisometropia may be obstacles to visual efficiency. After a prescription has been determined, a progress evaluation should reassess the patient's near point function before one considers additional intervention.

- *Strabismus:* May mimic or result in symptoms consistent with an ocular motor dysfunction. This phenomenon is especially true in cases of alternating or intermittent strabismus, because of unstable or poor fixation patterns. Intervention should focus on the treatment of the strabismus.
- *Vergence/Accommodative dysfunctions:* May be present with symptoms similar to those of an ocular motor dysfunction. A complete binocular work-up (p. 186) will determine the primary problem.
- *Medication:* Several medications may impact eye movements (e.g., antidepressants and anticonvulsants).
- *Gaze disturbances:* Horizontal and vertical gaze disturbances may result in "tracking" problems. These may show marked asymmetry and are typically easily differentiated from a functional ocular motor dysfunction. A pathologic etiology should be suspected if saccades are affected while pursuits are normal, or vice versa. Testing both saccades and pursuits and making a note of undershooting or overshooting in one particular gaze, will aid in the differential diagnosis.

WORK-UP

1. *History:* Are symptoms consistent with the functional etiology (onset, type, association with eye use)? Severity of symptoms (do they interfere with school or office work)? Associated neurological symptoms?
2. *Correct any significant refractive error:* If a significant refractive error is present, recommend a progress evaluation in 1 to 2 months to reassess ocular motor function and symptoms with spectacles.
3. *Near point binocular vision work-up (see p. 186):* Cover test, near lateral phoria, NPC (light, red glass, and through +1.00), AC/A ratio, fusional vergence ranges, accommodative facility, amplitude of accommodation, and monocular estimation method or FCC. These tests are essential for the differential diagnosis and the detection of associated vergence/accommodative dysfunctions.
4. *Developmental eye movement test (DEM) (p. 167):* A visual-verbal test of ocular motility, specifically designed to differentiate between functional ocular motor disorders and visual-perceptual problems. The test can be administered by

ancillary staff. It is designed to test eye movements associated specifically with reading.

5. *NSUCO ocular motor test (p. 164):* Standardized observation method of assessing pursuits and saccades. This test requires no special equipment and is easily administered. It specifically separates out pursuit from saccadic testing.
6. *Stereopsis:* Local (Wirt circles) and random-dot stereopsis should be assessed. Absence of or reduced stereopsis should raise one's suspicions about the presence of a strabismus.
7. *Cycloplegic refraction (p. 179):* Rule out latent refractive error when indicated.

ADDITIONAL TESTING

- Consider a visual-perceptual work-up for children with considerable school- or learning-related problems, exceedingly poor handwriting, and developmental delays.
- *Electrooculography (i.e., Visagraph):* Provides objective information regarding fixations, regressions, duration of fixations, reading efficiency, and reading level.

TREATMENT

Regardless of the treatment approach, communication of findings to teachers, occupational therapists, and any other school personnel involved in the child's education is essential.

- *Vision therapy (see p. 197):* The treatment of choice for a functional eye movement dysfunction. Didactic and clinical research has supported the idea that eye movements can be modified and that ocular motor vision therapy results in improved eye movements associated with reading.[5-7] Since an ocular motor disorder will typically be part of a triad of deficits, the training regimen may also include accommodative and binocular techniques. The patient and/or parent should become aware of improved accuracy in saccadic and pursuit functions, as well as a decrease in subjective symptoms, such as loss of place or the need to reread sentences.
- *Compensatory strategies:* In cases in which patients are unwilling or unable to follow a vision therapy program, certain strategies, such as using a finger to follow along during reading or a ruler to underline the sentences, may be beneficial and offer some relief.

- *Addition lenses:* The use of low amounts of plus correction is recommended by some as an initial approach. Unless there is a concurrent accommodative problem, this strategy may be of little use and in fact may further degrade function when prescribed needlessly.[8]

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2

Disorders of Accommodation

RICHARD C. LAUDON AND ERIK M. WEISSBERG

2.1 Accommodative Dysfunction

GENERAL INFORMATION

Accommodative dysfunction is a general term incorporating accommodative excess, accommodative insufficiency, and accommodative infacility. Although the more specific condition can often be identified, it is not uncommon to find overlapping signs and symptoms. Accommodative dysfunction may be present in isolation or in combination with other binocular disorders (e.g., convergence insufficiency). As a result of the intricate involvement of accommodation and vergence, a deficit or excess in one system may trigger an overaction or underaction in the other. Recommended management of this condition (spectacles or vision therapy) depends on the presence or absence of an associated binocular problem and the specific type of accommodative dysfunction.

SYMPTOMS[1]

- *Decreased distance acuity:* Typically fluctuates throughout the day (worse in the afternoon, after looking up from a near point task, or upon driving home from work at night)
- *Decreased near acuity:* Typically after prolonged reading

- Tendency to fall asleep or lose concentration when reading
- Eye strain and/or fatigue reported when reading
- Headaches above the eyes associated with eye use (may occur only during school or work days, and be absent on the weekends)
- Light sensitivity

Parents may note or children may report the following:

- Words going in and out of focus
- Inability to see small print
- Tendency to hold books very close
- Tendency to miscall words ("visual guesser")

SIGNS

General signs of all accommodative dysfunctions include the following:

- Acuity fluctuates during the visual exam, especially at near or at the end of the exam (e.g., patient's visual acuity is 20/20 through a particular prescription but when retested is unable to achieve the same level of acuity).
- Patient exhibits fluctuations in pupillary response upon reading the acuity chart.
- Inconsistencies exist between entering acuity and refractive error.
- Refractive error during retinoscopy is fluctuating.
- End point is indistinct, or the patient is unable to achieve a clear and stable visual acuity of 20/20 during subjective refraction.
- The presence of reduced vergence ranges and/or clinically significant phoria may be detected. (Abnormal binocular function may be the cause or result of accommodative dysfunction.)

Specific characteristics of accommodative dysfunctions are shown in Table 2.1-1.

Signs of an accommodative insufficiency include the following:

- Reduced accommodative amplitudes based on Hofstetter's minimum (15-.25 [age]).
- High lag of accommodation on monocular estimation method (MEM) (see p. 159) and/or fused cross-cylinder test (greater than +1.00).
- Inability or reduced ability to clear minus lenses during binocular and monocular accommodative facility testing.

TABLE 2.1-1
Common Accommodative Dysfunctions and Expected Findings

Type	MEM or FCC	Facility	Amps	NRA/PRA
Accommodative Excess (difficulty relaxing accommodation)	Lead	Fails + OU, OD, OS	Normal	Low NRA High/ Normal PRA
Accommodative Insufficiency (difficulty stimulating accommodation)	Higher than normal lag	Fails – OU, OD, OS	Reduced	High NRA/ Low PRA
Accommodative Infacility (difficulty sustaining accommodation)	Fluctuations or normal response	Difficulty with + and – that increases toward end of task	Normal	Normal, or both reduced

- Elevated negative relative accommodation (NRA) and/or reduced positive relative accommodation (PRA). (norm for NRA is +2.50, PRA –3.50).

Signs of accommodative excess include the following:

- Lead of accommodation during MEM (p. 159) and/or fused cross-cylinder test (= plano).
- Inability or reduced ability to clear plus lenses during binocular and monocular accommodative facility testing.
- Reduced NRA and/or elevated PRA.
- *Pseudomyopia:* Myopia (usually less than 1.00 diopter) that may be found on subjective refraction and that results from an overaccommodation during testing. Pseudomyopia typically decreases after careful delayed subjective (p. 181) or cycloplegic refraction (p. 179).

Signs of accommodative infacility include the following:

- Fluctuations may be detected during MEM (p. 159) or the fused cross-cylinder test.
- Patient demonstrates an inability or reduced ability to clear plus and minus lenses during accommodative facility testing, especially toward the end of the task.
- NRA and PRA may both be reduced.

DIFFERENTIAL DIAGNOSIS

- *"Emerging" myopia:* Newly myopic children have been shown to have an insufficient accommodative response to blur.[2] This phenomenon may result in clinical findings resembling an accommodative dysfunction and lead the clinician to suspect pseudomyopia. An appropriate cycloplegic refraction (p. 179) will aid in the final diagnosis.
- *Spasm of accommodative/near reflex:* Similar to accommodative excess but a more severe degree of impairment. The entire near triad, including convergence and miosis, is usually involved. Visual acuity may be dramatically reduced; limitation of abduction and myopia may also be present.[3]
- *Accommodative paralysis:* A rare condition that has a nonfunctional etiology. Onset is usually associated with illness, lead poisoning, or trauma (especially head trauma). It can be present in one or both eyes. A significant unilateral accommodative paralysis may be a sign of a more serious underlying etiology.
- *Pseudoconvergence insufficiency:* Clinical findings indicate a convergence insufficiency, but the binocular dysfunction is secondary to a primary accommodative insufficiency.[4] With the use of appropriate near point lenses the convergence insufficiency is resolved.
- *Binocular dysfunctions:* Convergence insufficiency or excess can cause a secondary accommodative dysfunction. With the resolution of the primary binocular dysfunction, the accommodative dysfunction becomes less a factor.
- *Uncorrected refractive errors:* Uncorrected refractive errors—especially hyperopia, or low amounts of myopia and/or astigmatism—may mimic the symptoms of accommodative dysfunction.
- *Streff nonmalingering syndrome[6]:* May be confused with accommodative dysfunction. This syndrome is characterized by reduced bilateral acuity, minimal refractive error, and an initial decrease in acuity that ultimately improves to 20/20 without any corrective lenses or with the use of low amounts of hyperopic correction. It is unclear whether this condition differs from "psychogenic amblyopia."[7]

WORK-UP

1. *History:* Are symptoms consistent with the functional etiology (onset, type, association with eye use)? When does blurring

occur, and is it consistent or fluctuating? Severity of symptoms (do they interfere with school or office work)? Associated neurological symptoms?

2. *Correct any refractive error:* If a significant refractive error is present, recommend a progress evaluation in 1 to 2 months to reassess binocular and accommodative function with spectacles.
3. If myopia is suspected, care should be taken to ensure that it is not the result of overaccommodation (pseudomyopia). Tests such as delayed subjective refraction (p. 181), cycloplegic refraction (p. 179), and NRA/PRA are often helpful. Furthermore, reported distance blur that is present in the morning and remains consistent throughout the day is likely refractive in origin. Conversely, distance blur that is only present in the afternoon or that gets worse throughout the day is strongly suggestive of an accommodative etiology (pseudomyopia).
4. *Distance binocular profile:* Phoria testing should be performed and the compensating vergence range measured if patient has distance symptoms.
5. *Near point binocular vision work-up (p. 186):* Cover test, near lateral phoria, NPC (light, red glass, and through +1.00), AC/A ratio, fusional vergence ranges, accommodative facility, amplitude of accommodation, and monocular estimation method or FCC. NPC and/or accommodative facility testing may be repeated at the end of the exam to determine whether fatigue creates greater disruption in the patient's binocular dysfunction.
6. *Stereopsis:* Local (Wirt circles) and random-dot stereopsis should be assessed. Absence of or reduced stereopsis should raise one's suspicions about the presence of a strabismus.
7. *Cycloplegic refraction (p. 179):* Rule out latent refractive error (especially hyperopia) when indicated.

TREATMENT

The preferred treatment approach (Table 2.1-2) depends on the specific type of accommodative dysfunction, the presence or absence of an associated binocular dysfunction, the severity of the symptoms, and the patient's level of motivation.

- *Plus reading lenses for near:* Conventional treatment for all accommodative dysfunctions, but especially effective in cases

TABLE 2.1-2
Recommended Treatment According to Specific Diagnosis

Diagnosis	Recommended Treatment
Accommodative Insufficiency	Addition lenses
Accommodative Excess with plus acceptance	Addition lenses
Accommodative Excess without plus acceptance	Orthoptics
Accommodative Infacility with plus acceptance	Addition lenses
Accommodative Infacility without plus acceptance	Orthoptics

of accommodative insufficiency. Typical near prescription ranges from +.75 to +1.25 if hyperopia is not present. (Prescription may be higher if the patient is hyperopic). Perform a trial framing of the near prescription to ensure the patient's comfort and recheck near acuity; repeat abnormal findings on binocular work-up through spectacles. The response to near point lenses is critical. Patients should report that the print is larger and that their eyes feel more comfortable, and they should be able to hold books farther away. Signs that a patient is unable to accept the plus correction are a blurring of near visual acuity and a negative reaction to the glasses during trial framing.

- Single-vision spectacles: Can be prescribed exclusively for near vision if the patient has symptoms only during or after prolonged near tasks.
- Bifocal: Consider prescribing a bifocal if the patient's visual demands require continual alternation between distance and near fixation (e.g., looking from board to desk in school). Flat top 28 with the segment set 3 mm below the middle of the pupil is usually an adequate lens design. Glasses can be worn during all symptom-producing tasks.

- *Vision therapy (p. 197):* May be used for the treatment of all accommodative dysfunctions,[5,8] but it is typically reserved for accommodative excess and accommodative infacility in which plus lenses for near are not accepted. It also plays an important role in treating accommodative dysfunctions with an associated binocular dysfunction. Vision therapy may be used to increase the flexibility of the accommodative system to allow for plus acceptance, or it may be the sole treatment.

- *Visual hygiene:* Patient education concerning multiple breaks of short duration during near point tasks. Fixation should be upon a distance target during the frequent rests of 1 to 2 minutes. The patient's attention can be drawn to the importance of maintaining an adequate and consistent working distance (40 cm or Harmon's distance) throughout near point work. Harmon's distance is the length between the elbow and the middle knuckle of the index finger.

FOLLOW-UP

Because accommodative dysfunction is considered a benign condition with the exception of symptoms, cases can be followed depending on the treatment.

- *Spectacle intervention:* 1 to 2 months to evaluate the effectiveness of glasses and repeat binocular work-up
- *Vision therapy:* Followed every 1 to 2 weeks until completion
- *Visual hygiene only, or asymptomatic case:* Followed yearly or as needed if symptoms increase

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3

Esodeviations

STACY AYN LYONS

3.1 Infantile Esotropia

GENERAL INFORMATION

Infantile esotropia (IE), also referred to as congenital esotropia, is considered a common form of esotropia.[1,2] Onset occurs typically during the first 6 months of life, with the cause being unknown. Even with timely and appropriate intervention, the prognosis for developing normal binocular vision with stereopsis is not good.[3]

SYMPTOMS

Infants: No symptoms are reported by patients because of their young age, but parents will typically have concerns about the appearance of the eyes, and family history may be positive for strabismus.

Adults: Patients are most commonly concerned with cosmesis. However, patients with long standing IE and early surgery may notice a change in the direction and magnitude of the strabismus, with possible asthenopia and infrequently intermittent diplopia later in life.

SIGNS

Characteristics of esotropia include the following:

- Magnitude is large (approximately 50 prism diopters [PD]), equal at distance and near.
- Always constant.

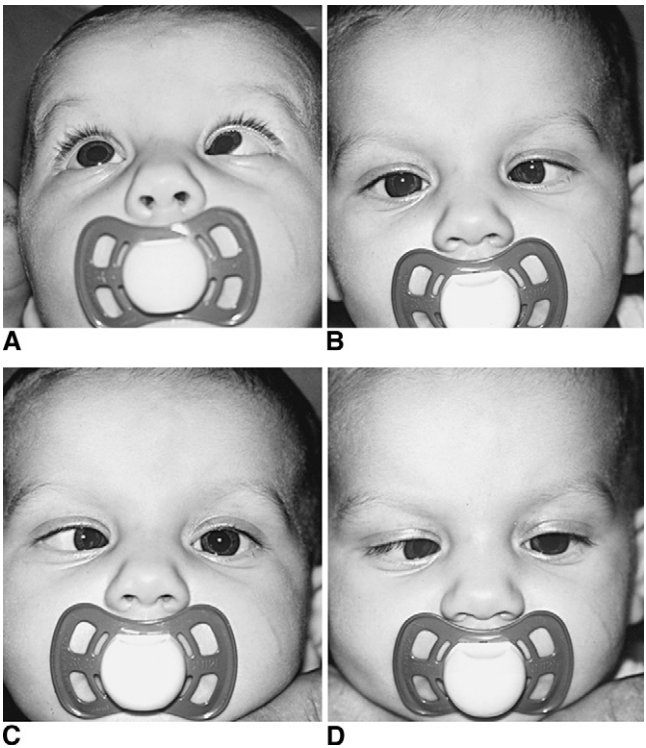

FIG. 3-1 Classic infantile esotropia demonstrated by a large angle alternating strabismus that is concomitant. **A,** Upgaze. **B,** Right eye fixating. **C,** Left eye fixating. **D,** Downgaze. (From Rosenbaum AL, Santiago AP: Clinical strabismus management, Philadelphia, 1999, WB Saunders.)

- Usually alternating (Fig. 3-1), but the patient may show a fixation preference.

Associated signs of IE include the following:

- *Overactive inferior oblique (OIO):* This symptom is unilateral or bilateral, characterized by hyperdeviation when the eye is adducted. It may not occur until 2 or 3 years of age.
- *Dissociated vertical deviation (DVD):* Double hyperdeviation is observed during cover test: both eyes move downward

and out upon removal of occlusion. This symptom may not occur until 2 or 3 years of age.
- *Latent and/or manifest nystagmus:* Nystagmus is observed in one eye when the other eye is occluded.
- *Refractive error:* Significant refractive error may or may not be present.
- *Amblyopia:* This may be present if the esotropia is unilateral, if a strong fixation preference exists, or if there is associated amblyogenic refractive error (anisometropia).
- There is an absence of underlying disease.

DIFFERENTIAL DIAGNOSIS

- *Pseudoesotropia:* This condition is often secondary to facial asymmetry and commonly confused with IE. Since the eyes are aligned, pseudoesotropia can be differentiated by use of the cover test, Hirschberg test, and Brüchner test.
- *Accommodative esotropia (p. 32):* The mean age of onset is approximately 2½ years of age, but early onset accommodative esotropia can be seen as early as 6 months of age. The presence of an esotropia that is larger at near than at distance and moderate hyperopic refractive error are key signs of accommodative esotropia. A hyperopic prescription will eliminate or appreciably reduce the amount of esotropia.
- *Abducens nerve palsy (p. 100):* Esotropia will be nonconcomitant with an abduction deficit in the affected eye. Version testing would confirm an esotropia in primary position, one which increases on gaze in the direction of the involved side. Neurological evaluation is recommended.
- *Duane retraction syndrome (p. 77):* An abduction deficit is present, coupled with retraction of the globe and narrowing of the palpebral fissure on adduction of the affected eye.
- *Nystagmus blockage syndrome:* This condition is associated with nystagmus in abduction.
- *Esotropia associated with neurological conditions:* Among patients with Down syndrome, cerebral palsy, and meningiocele there is a high incidence of strabismus. Strabismus detected in a member of this high-risk population warrants referral to a specialist due to the likelihood of associated ocular problems.

WORK-UP

1. *History:* Age of onset? Laterality: which eye looks turned, or does the turn alternate between the two? Frequency: do the eyes always look crossed? Developmental and neurological history? Family history of strabismus?
2. *Assessment of deviation:* Measure magnitude at distance versus near, laterality, concomitancy, and frequency:
   - *Infants and toddlers:* Cover test, Hirschberg and Brüchner tests (p. 150), versions.
   - *Older children/Adults:* Cover test, Hirschberg test, modified Thorington (p. 153).
3. *Visual acuity:* Assess monocular acuities to check for amblyopia:
   - *Infants*: Preferential Looking method, such as Teller Acuity cards.
   - *Preschool-aged children:* Lea symbols, HOTV, or Broken Wheel cards.
   - *School-aged children*: Snellen chart or Log MAR.
   - *For all ages:* A behavioral assessment of acuity can be obtained by observing the patient's monocular/binocular fixation pattern (p. 147).
4. *Versions/Ductions:* Check for restrictions, oblique overactions, and/or nystagmus.
5. *Evaluating the effects of plus (+) at near:* AC/A in the young child can be indirectly evaluated by comparing the results of a cover test at near with those from a cover test with a +2.00 D flipper. A reduction in the size of the strabismus through the plus lenses indicates an accommodative (or at least partially accommodative) component to the esotropia.
6. *Cycloplegic refraction (p. 179):* This is mandatory in all cases in which strabismus, amblyopia, or significant refractive error is suspected. Recommended administration is as follows: 2 gtt of 1% cyclopentolate separated by 5 minutes. 1 gtt of 1% cyclopentolate can be used in infants under 1 year of age. Onset of maximum effect comes approximately 30 minutes after instillation. The duration of action is approximately 6 to 12 hours.
7. *Evaluation of the anterior and posterior health of the eye to rule out a sensory cause for strabismus (p. 66):* A dilated fundus evaluation is mandatory for any young child with strabismus.

TREATMENT AND FOLLOW-UP

Surgical Intervention

Surgery is the usual treatment for this condition; therefore, referral to an experienced strabismic surgeon is warranted. To maximize chances of fusional and/or cosmetic success, the recommended age for surgical intervention is between 18 and 24 months.[5] Multiple surgeries are often needed.[5] The desired outcome is subnormal binocular vision and the absence of a cosmetically apparent strabismus (e.g., microtropia, p. 47). After making a referral for a confirmation of the diagnosis, you may comanage the patient with the surgeon.

Comanagement

Written communication between comanaging practitioners is suggested to minimize unnecessary and excessive testing. It is especially important to communicate in the report what the characteristics of the strabismus are and whether significant refractive error was detected during your evaluation.

- The patient should be monitored for amblyopia (p. 140):
  1. *Repeated visual acuity testing:* The patient should be taught to respond to pediatric recognition visual acuity tests (e.g., LEA or HOTV) as soon as possible. A photocopy of the symbols for home instruction may facilitate this process.
  2. The strabismus should be checked for changes in laterality (switching from alternating to unilateral) or development of a strong fixation preference, which in turn may lead to amblyopia.
  3. *Assessment of refractive error:* Check the patient for amblyogenic anisometropia and hyperopia.
- Monitor the patient for stability of the strabismus.
  1. Development of an exotropia (p. 72) may indicate the need for additional surgery and/or orthoptics.
  2. Development of an increasing esotropia may indicate uncorrected hyperopia and/or an accommodative component to the strabismus.[7] If this is detected, assess the effect of the prescription on the strabismus.
  3. Ocular motility disorders such as DVD and OIO may become apparent around 2 to 3 years of age.

- Repeated assessment of the integrity of the visual system (pupils, acuity, confrontation visual fields if possible, retina, and optic nerve) is necessary.

ADDITIONAL MANAGEMENT OPTIONS

The prescription of significant amounts of hyperopia, together with the use of addition lenses, prism, and orthoptics, has been advocated by some practitioners as the sole treatment or as a treatment in combination with surgery.[1,8] If patients are resistant to surgery or are interested in this approach, referral to an eye care practitioner specializing in this area may be warranted.

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3.2 Accommodative Esotropia

GENERAL INFORMATION

Accommodative esotropia (Acc ET) is associated with uncorrected hyperopia, a high AC/A ratio, or a combination of both. Acc ET is a common cause of strabismus in children and becomes manifest typically between the ages of 2 and 3 years.[1] However, onset as early as 6 months and as late as 7 years has

been reported.[2,3] Prognosis for achieving normal binocularity is good, provided the appropriate treatment is given in a timely manner.[4] Acc ET can be divided into two different categories.

1. *Refractive Acc ET:* Abnormal amounts of hyperopia are the causative factor. The esotropia is eliminated with the use of a hyperopic prescription (Fig. 3-2).
2. *Nonrefractive Acc ET:* An abnormally high AC/A is the causative factor. Hyperopic refractive error may be present but in normal amounts. The use of an addition lens is typically required to reduce the esotropia at near.

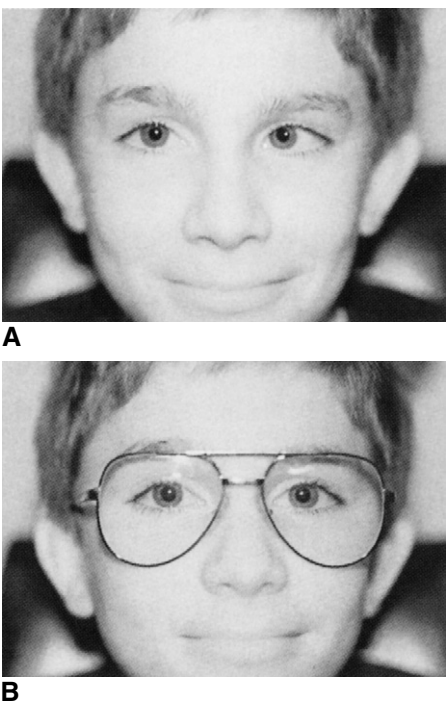

FIG. 3-2 A, Increased accommodative demand resulting from uncorrected hyperopia results in left esotropia when patient fixates on an object. **B,** Esotropia is eliminated when patient wears hyperopic correction. (From Evans B, Doshi S: Binocular vision & orthoptics: investigation and management, Oxford, 2001, Butterworth-Heinemann.)

NOTE: *Partially accommodative esotropia* mimics the refractive and/or non-refractive varieties with the exception that the use of plus lenses will reduce but not eliminate the esotropia. Partially accommodative esotropia commonly results from untreated and long-standing refractive or nonrefractive Acc ET.

SYMPTOMS

- Eye rubbing or eye closure is commonly observed coincident with or prior to onset of the strabismus.
- Parents may report the onset of esotropia after a febrile episode. Although the relationship is unclear, the esotropia is likely coincidental or results from additional stress caused by the fever.[3]
- The parent or caregiver typically has concerns about the appearance of the child's eyes.

SIGNS

The following are characteristics of the esotropia:

- Typically unilateral, concomitant esotropia is present, larger at near than at distance.
- *Magnitude:* 10 to 45 PD and subject to fluctuations during measurement due to changes in accommodation.
- *Frequency:* The condition is initially intermittent but may become constant if treatment is delayed.

The following are additional signs of Acc ET:

- *Refractive error and AC/A ratio:* Differences exist between the refractive and nonrefractive variety. In both cases, it is not uncommon to find anisometropia, with the strabismic eye being more hyperopic.
- *Refractive Acc ET:* Hyperopia ranges from +2 to +6 D; AC/A ratio is normal.
- *Nonrefractive Acc ET:* Normal to moderate amounts of hyperopia are present, averaging +2.25 D. An elevated AC/A ratio (greater than 5/1) is present.
- *Sensory adaptations:* Suppression and anomalous retinal correspondence may develop if treatment is delayed and the strabismus is constant.
- *Amblyopia:* This condition rarely results from esotropia because of the intermittency, but it can result from associated anisometropia.

DIFFERENTIAL DIAGNOSIS

- *Infantile esotropia (p. 27):* Hyperopia does not eliminate or significantly reduce the esotropia.
- *Abducens nerve palsy (p. 100):* Esotropia will be nonconcomitant with an abduction deficit in the affected eye. Version testing would confirm an esotropia in primary position, which increases on gaze in the direction of the involved side. Neurological evaluation recommended.
- *Duane retraction syndrome (p. 77):* Abduction deficit is coupled with retraction of the globe and narrowing of the palpebral fissure on adduction of the affected eye.

WORK-UP

1. *History:* Age of onset? Laterality: which eye looks turned, or does the turn switch between the two? Frequency: do the eyes always looked crossed? Symptoms: has your child been rubbing or closing an eye recently? Has your child had a fever or ear infection recently? Developmental and neurological history? Family history of strabismus?
2. *Visual acuity:* Assess monocular acuities to check for amblyopia. The age of the child will determine the type of acuity measure to obtain. Options for the type of acuity testing depend upon age, maturity, and rapport with patient:
   - *Infants:* Preferential Looking method, such as Teller Acuity cards.
   - *Preschool-aged children:* Lea, HOTV, or Broken Wheel cards.
   - *School-aged children:* Snellen chart.
   - *For all ages:* A behavioral assessment of acuity can be obtained by observing the patient's monocular/binocular fixation pattern (p. 147).
3. *Assessment of deviation:* Measure magnitude at distance versus near, laterality, concomitancy, and frequency. Care should be taken to use an accommodative target to maximize chances of strabismus detection.
   - *Infants and toddlers:* Cover test, Hirschberg and Brüchner tests (p. 150), versions.
   - *Older children/Adults:* Cover Test, Hirschberg, modified Thorington (p. 153).
4. *Dry static retinoscopy:* Use of interactive distance targets may help stabilize patient fixation. Dry retinoscopy can often be

used as an indirect measure of plus acceptance in patients for whom a subjective refraction is not possible or reliable.

5. *Evaluating the effects of plus (+) at near:* AC/A in the young child can be indirectly evaluated by comparing the results of a cover test at near with those from a cover test with a +2.00 D lens OU. A reduction in the size of the strabismus through the plus lenses indicates an accommodative (or at least partially accommodative) component to the esotropia.
6. *Cycloplegic refraction (p. 179):* Mandatory in all cases in which strabismus, amblyopia, or significant refractive error is suspected. Recommended administration is as follows: 2 gtt of 1% cyclopentolate separated by 5 minutes. 1 gtt of 1% cyclopentolate can be used in infants under 1 year of age. Onset of maximum effect occurs approximately 30 minutes after instillation. The duration of action is approximately 6 to 12 hours.
7. *Ocular motility:* Check for restrictions or oblique overactions and/or nystagmus.
8. *Stereopsis:* Likely to be reduced or absent if strabismus is present.
9. *Evaluation of the anterior and posterior health of the eye to rule out a sensory cause for strabismus (p. 66):* A dilated fundus evaluation is mandatory for any young child with strabismus and/or amblyopia.

TREATMENT

Prescribe the appropriate amount of hyperopia needed to eliminate the strabismus. Correcting the strabismus, if possible, through optical means takes priority over any initial reduction in acuity secondary to the use of the glasses.

Determining the Appropriate Correction

For refractive Acc ET:

- Full cycloplegic hyperopic finding for constant wear is desirable. This correction should eliminate the esodeviation at distance and near. Once the prescription has been determined, a cover test should be performed to confirm the elimination of the strabismus.
- If full cycloplegic correction cannot be prescribed because it may be an obstacle to compliance, then maximum plus acceptance, as determined by a combination

of dry retinoscopy and the effect on distance acuity, should be substituted. If a distance esodeviation is present, care should be taken to ensure that it is eliminated with this distance prescription. Cover test at near will likely reveal residual esodeviation. If present, it needs to be corrected by an addition lens. Trial frame increasing powers of plus at near in combination with a cover test to determine the appropriate bifocal power (typically does not exceed +2.50 D). Once the prescription has been determined, a cover test should be performed to confirm the elimination of the strabismus at both distance and near.

For nonrefractive Acc ET:

- Full cycloplegic hyperopic finding for constant wear is desirable. This correction should eliminate the esodeviation at distance. Cover test at near will reveal residual esotropia which needs to be corrected by an addition lens. Trial frame increasing powers of plus at near in combination with cover test to determine the appropriate bifocal power (typically does not exceed +2.50 D). Once the prescription has been determined, a cover test should be performed to confirm the elimination of the strabismus at both distance and near.
- If full cycloplegic correction cannot be prescribed because it may be an obstacle to compliance, then maximum plus acceptance, as determined by a combination of dry retinoscopy and the effect on distance acuity, should be substituted. If a distance esodeviation is present, care should be taken to eliminate it with this distance prescription. The residual esodeviation present at near requires use of an addition lens as outlined above.

Additional Considerations

- If a spectacle prescription alone does not eliminate esotropia, consider the following:
  1. The residual esotropia is likely the result of a nonaccommodative component. Allow the patient to wear the prescription for several weeks. Reassess the strabismus and repeat cycloplegic refraction to investigate additional latent hyperopia/anisometropia.[6]

2. If cosmetically apparent strabismus persists and no additional refractive error is detected, surgical referral may be warranted.
3. If constant unilateral strabismus persists, amblyopia therapy should be initiated (p. 140).

- Parents should be educated that their child's eye turn may become more apparent when the child's glasses are removed. Parents should also be made aware that the glasses prescription may be subject to change.
- Full-time-wear, polycarbonate spectacles are mandatory in all cases. If a bifocal is indicated, a flat top 28 is recommended with the segment height set 3 mm below midpupil. If progressive bifocals are desired, it is advisable to increase the add by +0.50 D.
- Vision therapy and prism in combination with spectacle intervention are recommended by some to improve visual function.[5,7]

FOLLOW-UP

Follow cases every 2 months until strabismus has been controlled and becomes stable. Follow every 6 months for the next year and then continue yearly examinations. Carefully monitor patients for changes in refractive error, strabismus, and the development of amblyopia. Normal visual acuity, ocular alignment, and stereopsis should be attained shortly after appropriate treatment is initiated.

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3.3 Acute Acquired Comitant Esotropia

GENERAL INFORMATION

Acute acquired concomitant esotropia is a general term that describes the occurrence of an esotropia in a previously nonstrabismic patient. This large-angle insidious esotropia may be idiopathic, associated with serious neurological conditions, or result from prolonged occlusion of one eye.[1-4] Commonly occurring during early childhood,[1] the onset may coincide with febrile illness, emotional stress, or trauma.[2] With all types of acquired esotropia, it is imperative to differentiate benign from pathologic causes. Therefore, a full neurological evaluation is often prudent.

SYMPTOMS

- *Diplopia:* This is a common presenting symptom even in children as young as 4 years old.[1] Given the abrupt nature of the onset, patients find the diplopia to be quite disconcerting and may often be able to report the exact time of onset.
- Parents may observe eye closing, squinting, or eye covering while the child tries to alleviate the diplopia.

SIGNS

The following are characteristics of the esotropia:

- Moderate to large angle deviation (approximately 30 to 50 PD) is present.
- Magnitude is equal at both distance and near.
- *Concomitant:* Strabismus is of the same magnitude in all positions of gaze.
- "V" pattern estropia may be detected (larger deviation in downgaze and smaller deviation in upgaze).[1]

Associated signs of acute acquired concomitant esotropia include the following:

- *Refractive error:* Significant refractive error may or may not be present.
- AC/A ratio is normal.[4]
- Fusion is often possible when the angle of deviation is corrected with prism.

- *History of occlusion treatment* (e.g., for amblyopia, corneal abrasion): Occlusion may result in a breakdown of an already fragile binocular system.
- There is a previously documented esophoria.
- There is a history of neurological or intracranial disease.

DIFFERENTIAL DIAGNOSIS

An esotropia or esophoria that is manifest with a larger magnitude at distance than at near warrants a neurological consult due to the high probability of a neurologic and potentially serious cause.

- *Abducens nerve palsy (p. 100):* Esotropia will be nonconcomitant with an abduction deficit in the affected eye. Versions testing would confirm an esotropia in primary position, which increases on gaze in the direction of the involved side and may also be larger at distance. End-point nystagmus may be noted. Immediate neurological evaluation is recommended.
- *Decompensated phoria:* Onset tends to be gradual, and magnitude of strabismus is usually small or moderate.
- *Accommodative esotropia (p. 32):* A significant hyperopic refractive error that eliminates or significantly reduces the deviation would be revealed. Although typical onset occurs between 3 and 5 years of age, it can have a later onset. With acute onset esotropia, there is no observed accommodative component. Magnitude of deviation tends to be smaller in cases of accommodative esotropia.[3]
- *Divergence insufficiency (p. 53):* Esotropia or esophoria is greater at distance than at near. Distance diplopia that has progressively gotten worse is typically reported.
- *Divergence paralysis:* Esotropia is greater at distance than near, with normal versions.[5] Onset may be sudden with resulting diplopia. Neurological evaluation is imperative. This condition may be difficult to differentiate from bilateral CN VI paresis.

WORK-UP

1. *History:* Age/Time of onset? Laterality: which eye looks turned, or does the turn switch between the two? Frequency: do the eyes always look crossed? Is there a noticeable difference between near and distance? Diplopia (distance, near,

horizontal, vertical, or oblique)? Associated neurological signs or symptoms (headaches, vomiting, dizziness, lethargy)? Family history of strabismus? Associated neurological symptoms (poor balance/coordination, headache, vomiting, and nausea)? Trauma?

2. *Assessment of deviation:*
   - Measure magnitude at distance versus near, laterality, and frequency. This is accomplished with a cover test, Hirschberg test, and modified Thorington (p. 153).
   - *Concomitancy:* Measurement of the angle in nine positions of gaze is important to rule out a nonconcomitant deviation. This can be accomplished with prism cover test in all nine positions or modified Thorington.
3. *Ocular motility:* Carefully check for restrictions, overactions, and/or nystagmus to rule out lateral rectus paresis.
4. *Pupil testing:* Pupil abnormalities may indicate an underlying neurological cause.
5. *Cycloplegic refraction (p. 179):* Mandatory in all cases in which strabismus is suspected. Rule out latent hyperopia as a causative agent for the esotropia. Recommended administration: 2 gtt of 1% cyclopentolate separated by 5 minutes. 1 gtt 1% cyclopentolate can be used in infants under 1 year of age. Onset of maximum effect occurs approximately 30 minutes after instillation. The duration of action is approximately 6 to 12 hours.
6. *Evaluation of the anterior and posterior health of the eye to rule out a sensory cause for strabismus (p. 66) and any signs of neurological disease*: Carefully check optic nerve head for papilledema.

ADDITIONAL TESTING

Referral to a neuroophthalmologist is imperative in all cases of nonaccommodative sudden onset esotropia to rule out neurologic or intracranial disease, no matter whether associated neurological signs have been detected.

TREATMENT AND FOLLOW-UP

1. Determine and treat underlying cause if identified.
2. Spectacle intervention:
   - If hyperopia is present, assess the effect of the full cycloplegic on the magnitude of the deviation. A bifocal may be

indicated in some cases to minimize the esotropia at near. Although the spectacle intervention alone will not eliminate the esotropia, it may improve cosmesis by reducing the magnitude of the strabismus. A 2- to 4-week trial period with the spectacles and a follow-up progress evaluation will typically be sufficient to determine whether the glasses will be helpful.

- *Prism:* Although not typically successful because of the large magnitude of the deviation, a prism may prove useful in eliminating the diplopia in some cases. Determine the minimal amount of base-out prism that eliminates the symptoms and improves function by trial framing in conjunction with cover test. Fresnel prisms may be applied to a patient's glasses to assess the prescription and its relief of symptoms before grounding the prism into a pair of glasses. However, visual acuity may be degraded through the Fresnel prism, which may give the patient a false sense of what to expect with a more permanent prism prescription. Approximately five to six PD of base-out prism to each eye is the maximum amount that can be incorporated into a pair of glasses without sacrificing cosmesis and visual clarity. A prism adaptation test (p. 172) should be performed before prescribing to ensure that the patient does not adapt to the prism.

3. *Vision therapy:* Can be utilized to aid in the maintenance of fusion ranges after a prism correction has been prescribed. Referral to an optometrist specializing in this area is recommended.
4. *Surgery:* Given the large magnitude of the deviation and the minimal effect achieved through the use of spectacle intervention, surgery often becomes the only viable option. Studies of surgical interventions suggest a good prognosis for achieving alignment in cases of normal, high-grade stereopsis.[1,2] Surgery is often delayed for a period of 6 to 9 months to demonstrate stability. A referral to an experienced strabismic surgeon for consultation is warranted.
5. Co-manage the case with the surgeon:
   - Written communication between co-managing practitioners is suggested to minimize unnecessary and excessive testing. It is especially important to communicate in the report what the characteristics of the strabismus are and

whether significant refractive error was detected during your evaluation.
- *Monitor the patient for amblyopia:* Although rare in acute acquired esotropia, amblyopia may be a concern in young patients.
- Monitor the stability of the strabismus, particularly the magnitude of the deviation.
- Perform repeated assessment of the presence of neurological signs and symptoms, as well as of the internal and external health of the eye.

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3.4 Cyclic Esotropia

GENERAL INFORMATION

Cyclic esotropia, also referred to as circadian esotropia, is a rare form of esotropia that is based on a clock or circadian rhythm.[1-3] Patients exhibit a large-angle esotropia with sensory adaptations, followed by periods of normal binocularity in a cyclical pattern. The pattern tends to be observed in a 48-hour rhythm.[1-3]

SYMPTOMS

- No symptoms are usually reported because of the development of sensory adaptations; however, parents will typically have concerns about the appearance of the eyes.
- Eye rubbing or eye closure can be observed with the onset of this condition.

SIGNS

The following are characteristics of the esotropia:

- Large magnitude esotropia (approximately 50 PD), equal at distance and near, is followed by periods of normal binocularity.
- Esophoria may be expressed during the periods of normal binocular vision.
- *Frequency:* Cyclic esotropia tends to operate in 48-hour cycles, in which the child experiences 24 hours of esotropia followed by 24 hours of normal binocularity. After a period of time (months or years) the esotropia becomes constant.
- *Onset:* Mean age of onset is 3 to 4 years. It has been reported to occur secondarily to head trauma and surgery.[4,5]
- V pattern esotropia may be detected.[6]

Associated signs of cyclic esotropia include the following:

- *Refractive error:* Emmetropia to moderate hyperopia is present.
- *Sensory adaptations:* Suppression and anomalous retinal correspondence may develop during episodes of esotropia. When aligned, patients generally exhibit normal retinal correspondence (NRC), good stereopsis, and no strabismus.
- High AC/A may be detected.[6]

DIFFERENTIAL DIAGNOSIS

Any condition that can result in an intermittent strabismus should be considered as part of the differential diagnosis. Differentiation is typically based on the pattern of the strabismus and the presence or absence of associated signs. Among others, the following should be considered:

- *Divergence excess exotropia (p. 58):* This condition can develop at a young age. Young children with this condition will usually not exhibit strabismus during near testing and will show intermittent strabismus during distance testing. An occasional eye turn will likely be reported by the parent.
- *Accommodative esotropia (p. 32):* Mean age of onset is approximately 2 1/2 years of age, but early onset accommodative esotropia can be seen as early as 6 months of age. Esotropia will be larger at near than at distance, and moderate hyperopic refractive error will be detected. A hyperopic prescription will eliminate or appreciably reduce the amount of esotropia.

WORK-UP

Examining the patient over several consecutive days will give the eye care practitioner additional insight into the pattern of the strabismus.

1. *History:* Age of onset, frequency, associations with time of day or activities (distance versus near)? Developmental and neurological history? Recent trauma? Parents should be instructed to maintain a log of the periods of tropia to monitor the pattern of strabismus.
2. *Visual acuity:* Check for amblyopia, which due to periods of normal viewing is not likely to occur in cyclic esotropia:
   - *Infants:* Preferential Looking method, such as Teller Acuity cards.
   - *Preschool-aged children:* Lea symbols, HOTV, or Broken Wheel cards.
   - *School-aged children:* Snellen chart or Log MAR.
   - *For all ages:* A behavioral assessment of acuity can be obtained from the patient's response to forced occlusion, maintenance of fixation, and ductions (p. 147).
3. *Measurement of deviation:* Distance, near, and in all positions of gaze. Special care should be taken to break down fusion during cover test with prolonged occlusion to elicit an intermittent strabismus (perform alternating cover test for 15 seconds, and then cover one eye for 5 to 10 seconds).
4. *Ocular motility:* Check for restrictions, overactions, and/or nystagmus.
5. *Cycloplegic refraction (p. 179):* Mandatory to uncover any possible latent hyperopia. Recommended administration is as follows: 1 to 2 gtt of 1% cyclopentolate separated by 5 minutes. Onset of maximum effect comes approximately 30 minutes after instillation. The duration of action is approximately 6 to 12 hours.
6. Evaluation of the anterior and posterior health of the eye is mandatory in any young child with strabismus.

TREATMENT

- *Surgery:* This is the treatment of choice after all nonsurgical options have been exhausted and the deviation has become essentially constant. Referral to an experienced strabismic

surgeon is warranted. Prognosis for normal binocular vision is good. Recurrence of the cyclic esotropia has been reported in the literature.[7]

- Botulinum toxin type A has been utilized alternatively to surgery with less success.

Co-management

Written communication between co-managing practitioners is suggested to minimize unnecessary and excessive testing. It is especially important to communicate in the report what the characteristics of the strabismus are and whether significant refractive error was detected during your evaluation.

- *Monitor the patient for amblyopia:* Although it is rare in cyclic esotropia, amblyopia may be a concern if the deviation becomes constant or after surgery has been performed.
  1. *Repeated visual acuity testing:* The patient should be taught to respond to pediatric recognition visual acuity tests (LEA or HOTV) as soon as possible. A photocopy of the symbols for home instruction may facilitate this process.
  2. Assess the strabismus to check for changes in laterality (alternating versus bilateral and unilateral) and frequency (intermittent versus constant), and for the development of a strong fixation preference, which in turn may lead to amblyopia
- Monitor the stability of the strabismus for recurrence of the cyclic esotropia.[7]
- Repeated assessment of the integrity of the visual system (pupils, acuity, confrontation visual fields if possible, retina, and optic nerve) is appropriate.

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3.5 Microtropia

GENERAL INFORMATION

Also called *monofixation syndrome,*[1] this type of strabismus commonly goes undetected because of its lack of symptoms and normal cosmetic appearance. Clinically, one should suspect microtropia in all patients with otherwise unexplained minimal to moderate unilateral reduction in acuity and reduced stereopsis. Although microtropia may be present by 3 years of age, the lack of significant symptoms and an overt eye turn may result in a delay in diagnosis until patients seek their first complete eye examination. This condition can generally be classified according to three basic causes:

- *Primary microtropia:* No specific cause can be documented.
- *Treated large-angle esotropia:* Microtropia results after treatment (typically surgical) for a larger magnitude esotropia.
- *Anisometropia:* Microtropia detected in a patient with small to moderate amounts of anisometropia (usually hyperopia).[2-4]

SYMPTOMS

No symptoms are usually reported because of the development of sensory adaptations such as suppression and/or anomalous retinal correspondence. Patients may report reduced vision in one eye or the awareness of a "weak" eye. Cosmetically these patients do not appear to have a strabismus.

SIGNS[4–6]

- *Small-angle esotropia* (usually defined as less than 8 PD): This may not be apparent during cover test, but esophoria may be detected.
- *Amblyopia:* Patient exhibits reduced unilateral acuity in the range of 20/30 to 20/70 without the presence of an obvious amblyogenic factor.

- *Refractive error:* Small or moderate amounts of anisometropia may be present (typically hyperopic anisometropia).
- *Reduced or no stereo acuity:* Global stereopsis is typically absent or reduced; however, good local stereopsis has been reported.[7]
- Small central suppression occurs under binocular viewing conditions, with peripheral fusion still intact (see Work-up for further explanation).
- *Anomalous retinal correspondence:* Remapping of corresponding retinal points occurs to account for the lack of bifoveal fixation resulting from strabismus. This process results in subnormal fusion with a small central suppression.
- *Eccentric fixation:* Use of a nonfoveal point for fixation is evident under monocular conditions. Small degrees of relatively steady nasal eccentric fixation are commonly found in the involved eye, particularly in the presence of a small-angle esotropia (see Work-up for further explanation).

DIFFERENTIAL DIAGNOSIS

Microtropia is part of the differential diagnosis for all conditions manifesting unexplained unilateral reduction in acuity and central suppression. The challenge in diagnosing this condition is documenting the cause of the reduced vision. As always it is essential to perform a thorough dilated internal and external ocular health evaluation to rule out a pathologic cause of the reduced acuity. This condition differs from typical amblyopia (p. 140) in that the clinician may not be able to document directly the presence of an amblyogenic agent (amblyogenic strabismus, anisometropia, form deprivation). Instead, one must rely on testing that indirectly suggests the cause of the amblyopia.

WORK-UP

1. *History:* Previously treated esotropia and/or strabismus surgery? Family history of strabismus? If reduced acuity or a "better seeing eye" is reported by the patient, how long has this condition been present?
2. *Visual acuity:* This is often the main sign alerting the clinician that the patient has a problem. Perform monocular acuities to

check for amblyopia, which is usually manifest as a 1- to 3-line reduction in acuity.
   - *Preschool-aged children:* Lea symbols, HOTV, or Broken Wheel cards.
   - *School-aged children:* Snellen chart or Log MAR.
3. *Assessment of deviation:* The unilateral and alternating cover test will often reveal a difference in magnitude of the eso movement. For example, esophoria is often detected during the alternating cover test, but during the unilateral cover test there may be a very subtle movement suggestive of esotropia or no movement at all. Clinicians sometimes refer to the suggestive movement as a "flick." This flick is due to the small magnitude of the strabismus and the presence of eccentric fixation. Detection during the cover test depends on both the cooperativeness of the patient and the skill of the clinician.
4. *Assessment of eccentric fixation:* Eccentric fixation is the use of a nonfoveal point for viewing under monocular conditions. It is often a key part of the diagnosis of microtropia and clinically is best detected using Visuoscopy (p. 174). This procedure will often reveal a small, relatively steady nasal eccentric fixation in the eye with reduced acuity.
5. *Assessment of central suppression:* This may be performed by the use of several tests, and it is recommended that at least the following two tests be performed to ensure an accurate diagnosis:
   - *Four base-out test (p. 177):* Different responses are detected when this test is performed on the involved eye and on the other eye. Lack of movement in response to the prism suggests a small central suppression scotoma. This effect occurs because the target displaced by the prism remains within the suppression scotoma and the eye is therefore unaware it has moved (Fig. 3-3).
   - *Worth four-dot test:* Using the "walk away" technique will reveal a distance at which suppression is reported. Begin testing at 40 cm and slowly walk away from the patient, increasing testing distance up to 10 feet. The angular subtense of the target is largest at near and may overlap the suppression scotoma, resulting in a normal fusional response (patient reports four dots). Suppression is more commonly reported at the intermediate or remote distance of testing, where the angular subtense of the target is decreased and falls within the suppression scotoma.

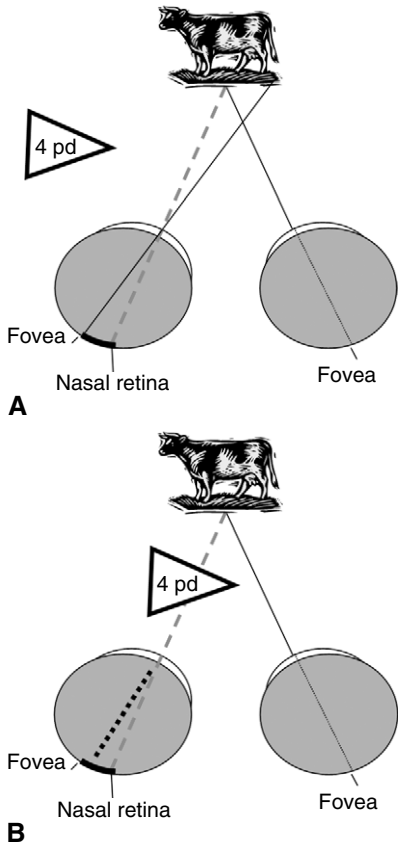

FIG. 3-3 4-PD base-out test in microtrope. A small central suppression resulting from a microtropia prevents the left eye from seeing that the image has moved as a result of interposing the prism. Although the image is shifted by the placement of the prism, it remains within the suppression.

6. *Stereopsis:* Performance is highly dependent on the type of stereopsis test used. Poor performance on small pixel stereo tests has been demonstrated in microtropia/monofixation syndrome.[8-10] Amblyopes have a difficult time passing the random dot E test when tested at 1.5 to 2 m, but they may perform normally on contour tasks such as Titmus circles.[10]

7. *Cycloplegic refraction (p. 179):* Mandatory in all cases in which strabismus, amblyopia, or an otherwise unexplained reduction in acuity is suspected. Especially in the case of microtropia, the clinician must take all measures to reveal the presence of anisometropia. Recommended administration is as follows: 2 gtt of 1% cyclopentolate separated by 5 minutes. 1 gtt of 1% cyclopentolate can be used in infants under 1 year of age. Onset of maximum effect comes approximately 30 minutes after instillation. The duration of action is approximately 6 to 12 hours.
8. Perform an evaluation of the anterior and dilated posterior health of the eye to rule out any pathologic cause behind the reduced acuity in one eye.

TREATMENT

In an adult

- With the absence of symptoms or cosmetic concerns, there is no need to treat the condition. In fact, the high likelihood of sensory adaptations (suppression, ARC) results in an increased risk of inducing intractable diplopia if typical amblyopia treatment (patching, antisuppression) is initiated.
- *If symptoms are present:* Microtropia may become unstable with age and the onset of presbyopia. In this case, prescribe correction for significant refractive error, use addition lenses, and consider referral to a vision therapy specialist to work on peripheral fusion techniques to help stabilize the condition.

In a young child (less than approximately 8 years of age)

Although symptoms are unlikely, the plasticity of the visual system at this age suggests that treatment aimed at remediation of the amblyopia is more likely to be successful with a reduced risk of inducing intractable diplopia. Treatment is similar to that of traditional amblyopia (p. 140), with the emphasis on correcting significant refractive error and initiating patching. Although additional measures may need to be taken in some cases with eccentric fixation, the use of traditional direct patching (patching the dominant eye) has been shown to be a viable first alternative.[11]

1. Correct significant refractive error.
2. Initiate direct patching (occlusion of the dominant eye).

3. Perform follow-up at 1-month intervals, monitoring for changes in acuity, eccentric fixation, and strabismus.
4. Continue patching as long as improvement is noted. If no improvement and no change in the eccentric fixation pattern occur, refer the patient to a specialist to initiate alternative techniques such as pleoptics, vision therapy, or indirect patching.

FOLLOW-UP

In adults

- If this is a first-time diagnosis, perform follow-up in 3 to 4 months to check for changes in acuity and to ensure that an accurate diagnosis has been made.
- If this is a long-standing condition, routine yearly follow-up is indicated.
- If the patient is symptomatic, monitor according to treatment.

In young children (less than approximately 8 years of age)

- If treatment has been initiated, monitor the patient according to the treatment outlined above.
- If no treatment has been initiated, follow up in 3 to 4 months to check for changes in acuity and to ensure that an accurate diagnosis has been made:
  1. Check for the development of amblyopia (p. 140). *Repeated visual acuity testing:* The patient should be taught to respond to pediatric recognition visual acuity tests (e.g., LEA or HOTV) as soon as possible. A photocopy of the symbols for home instruction may facilitate this process.
  2. Monitor the patient for stability of the strabismus or any increases in magnitude.
  3. Perform repeated assessment of the integrity of the visual system (pupils, acuity, confrontation visual fields if possible, retina, and optic nerve).

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3.6 Divergence Insufficiency

GENERAL INFORMATION

Divergence insufficiency (DI) refers to a clinical condition wherein the main characteristic is noted to be a concomitant esodeviation (phoria or tropia), the magnitude of which is greater at distance than at near. Often, the esodeviation is manifest as a phoria and with time decompensates into an intermittent strabismus. The onset of the strabismus tends to be later; therefore, intermittent diplopia is a common presenting symptom.[1-3] As with all types of acquired esotropia, care must be taken to differentiate functional from pathological conditions.[1-5]

SYMPTOMS

Patients typically report a long history of symptoms that may be exacerbated during distance viewing and night-driving and by fatigue.

- Patients may experience intermittent diplopia and generalized ocular discomfort, including headaches and strain, nausea, dizziness, blurred vision, motion sickness, and sensitivity to light.[1]
- Patients may have concerns about the appearance of their eyes.

SIGNS

Characteristics of esodeviation include the following:

- Magnitude greater at distance than near. Average magnitude is 16 PD at distance.[2]
- Esodeviation is concomitant.
- Usually alternating, but patients may show a fixation preference in one eye.

Associated signs of DI include the following:

- AC/A ratio is low.
- Negative fusional vergence is decreased.
- Significant refractive error may or may not be present.

DIFFERENTIAL DIAGNOSIS

An esotropia or esophoria with a larger magnitude at distance than at near warrants a neurological consult due to the high probability of a neurologic and potentially serious cause.

- *Divergence paralysis:* Esotropia greater at distance than near in the presence of neurological disease. Divergence paralysis has been associated with neurologic or intracranial diseases such as tumors, demyelinating disease, and head trauma. A referral for a full neurological evaluation is warranted.
- *Unilateral or bilateral abducens nerve palsy (p. 100):* Esotropia will be nonconcomitant with an abduction deficit in the affected eye. Versions testing would confirm an esotropia in primary position, which increases on gaze in the direction of the involved side. End-point nystagmus may be noted. An immediate neurological evaluation recommended.
- *Convergence excess (p. 7):* Esodeviation is concomitant; however, the magnitude and frequency are greater at near than at distance.
- *Basic esotropia or esophoria:* Esodeviation is concomitant; however, the magnitude and frequency are equal at distance and near.

WORK-UP

1. *History:* Age of onset? Laterality: which eye looks turned, or does the turn alternate? Frequency: do the eyes always look crossed? Is there a noticeable difference between near and distance? Associated neurological signs or symptoms? Duration of symptoms (sudden or gradual onset)?

2. *Assessment of deviation:* Direction, magnitude (distance versus near), laterality, conomitancy, and frequency. Measurement of the angle in 9 positions of gaze is important to rule out nonconcomitancy and can be accomplished by prism cover test or modified Thorington (p. 153).
3. *Ocular motility:* Perform a careful assessment of lateral rectus function, looking for any restrictions of abduction or overactions of the contralateral medial rectus.
4. *Pupil testing:* Pupil abnormalities may indicate an underlying neurological cause and are not present in divergence insufficiency.
5. *Stereopsis:* Likely to be normal at near and possibly reduced or absent at distance.
6. *Worth four-dot test:* This test will reveal fusion at near while the distance findings will result in suppression or in a diplopic response. Start by testing at 40 cm and slowly increase distance by walking away from the patient.
7. *Refraction:* Significant refractive error, especially hyperopia or significant anisometropia, should be corrected and the effect on the strabismus evaluated. Cycloplegic refraction (p. 179) is helpful in cases in which hyperopia is suspected.
8. Perform an evaluation of the anterior and posterior health of the eye to rule out a sensory cause for strabismus (p. 66) and any signs of neurological disease. Carefully assess the optic nerve head for papilledema. A dilated fundus evaluation is mandatory in any young child with strabismus.

ADDITIONAL TESTING

Referral to neuroophthalmologist is imperative in all cases of sudden onset esotropia to rule out neurologic or intracranial disease, whether or not associated neurological signs are detected.

TREATMENT AND FOLLOW-UP

- *Rule out serious underlying causes:* If one is identified, refer the patient for appropriate treatment.
- *Refractive correction:* Prescribe the most plus or least amount of minus correction the patient is able to accept: Full cycloplegic finding for at least distance wear is desirable. Trial frame to evaluate the effect on distance visual acuity OU and the

esodeviation. Because of the low AC/A ratio in these patients, this spectacle intervention alone may not eliminate the esotropia and thus may not eliminate the patient's symptoms.

- Prism has been shown to be the most efficacious treatment for DI.[1-3] There are many different philosophies about prism prescriptions. A systematic approach by trial framing, in conjunction with performing cover tests on the patient, aids in determining the minimal amount of base-out prism correction that should be prescribed to eliminate the patient's symptoms and improve vision function. Before grounding the prism into a pair of glasses, one may apply Fresnel prisms to the patient's glasses to assess the prescription and the level of relief it provides; however, visual acuity may be degraded by the Fresnel prism. Approximately 5 to 6 PD of base-out prism to each eye is the maximum amount prism that is cosmetically appealing and of good optical quality in a pair of glasses. Before a prescription is made, prism adaptation tests should be preformed to ensure that the patient does not adapt to the prism (p. 172).
  1. Perform prism adaptation test to determine whether the patient is a good candidate for a prism prescription. If no adaptation is noted, proceed to step 2.
  2. Ask the patient about his or her comfort level wearing the full amount of prism; recheck cover test through the prism prescription.
  3. Trial frame half the total amount of prism from step 2, ask the patient about comfort, and recheck the cover test through the prism.
  4. Depending on the patient's response, increase or decrease the prism appropriately until the least amount of prism that is subjectively appreciated by the patient has been realized.
- *Vision therapy:* If the above prescriptions with prism are unsuccessful or if a patient would like an alternative approach to treatment, vision therapy is a reasonable alternative and may improve visual function and eliminate symptoms in patients with DI. Treatment of DI with vision therapy is best accomplished under the supervision of a specialist in this area. The basic approach involves beginning training at near and gradually working up to distance.[1] The reader is encouraged to pursue additional sources for details concerning this subject.[6]

- *Surgery:* If all nonsurgical options are unsuccessful, if a patient wants another approach, or if a patient's strabismus has degraded to a large-magnitude constant strabismus, a referral to an experienced strabismus surgeon for consultation is warranted.

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4

Exodeviations

BARRY S. KRAN

4.1 Exotropia of the Divergence Excess Type

GENERAL INFORMATION

Divergence excess (DE) is an intermittent exotropia whose angle of deviation at distance is at least 10 to 15 prism diopters (PD) greater than at near, and whose frequency of turn can vary from 1% to 99% of the time.

Approximately 30% of strabismics are exotropic.[1] Most are identified in the first few years of life. About two thirds of exotropes are female.[2] One retrospective study of patients with an exodeviation (convergence insufficiency, divergence excess, basic exotropia) found that approximately 10% of the patients had DE.[3]

SYMPTOMS

- Photophobia is the most commonly reported symptom. The classic history includes the parental description of the patient's closing (squinting) one eye when outside.
- A common reason for referral is cosmesis.
- Asthenopic symptoms are rare unless a secondary binocular/accommodative issue is manifest at near.
- Diplopia is very rare. When reported, it is typically done so by a young child.

SIGNS

- A concomitant exotropia is greater at distance than at near (Fig. 4-1).

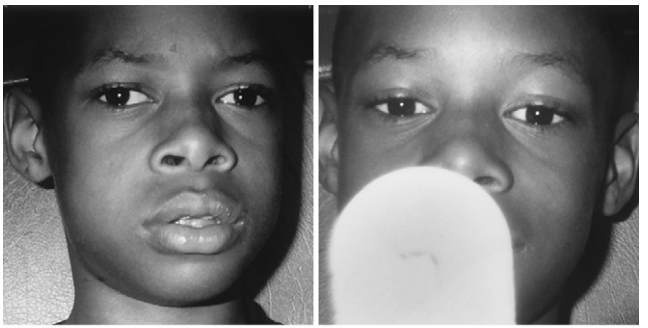

FIG. 4-1 Divergence excess type exotropia characterized by moderate angle right exotropia during distance fixation **(A)** but normal fusion during near fixation **(B).** (From Rutstein RP, Daum KM: Anomalies of binocular vision: diagnosis & management, St. Louis, 1998, Mosby.)

- Approximately 50% of patients may have a vertical component, which seems due in about two thirds of these cases to an overaction of the inferior oblique muscle, and in the remainder to the fact that the medial rectus is simply a stronger elevator when the eye is abducted.
- An exotropia of "V pattern" may be present.[2]
- When the eyes are aligned, patients generally exhibit normal retinal correspondence (NRC), good stereopsis, no strabismic amblyopia, and a good near point of convergence.
- When the deviation is present, these patients will exhibit either NRC with suppression or abnormal retinal correspondence (ARC).
- Stimulus-response AC/A ratios *may* be higher than average, and distance-near AC/A ratios are high.

DIFFERENTIAL DIAGNOSIS

- *Basic exotropia:* Concomitant strabismus with angles at distance and near equal or within 10 PD of each other. This is a descriptive term, not an indication of cause. Rule out sensory strabismus (p. 66) and pseudo DE. Treatment is tailored to the degree of symptoms and to cosmetic concerns (e.g., vision therapy, prism, surgery).

- *Pseudodivergence excess:* Ruled out by monocular occlusion for 45 to 60 minutes. If immediately after removing the patch the distance and near angles are within 10 to 15 PD of each other, then a case of true DE is ruled out.[1,2,4] This may affect treatment options.
- *Sensory strabismus (p. 66):* There is an ocular pathology or an amblyogenic factor as the causative agent.
- *Sudden-onset exotropia:* Trauma, tumor, infarction associated with nonconcomitancy and diplopia. Visual fields testing and referral to a specialist such as a neuroophthalmologist are warranted.

WORK-UP

1. Correct significant refractive error.
2. Perform binocular evaluation, including the following:
   - *Assessment of deviation:* Measure the magnitude distance versus near, laterality, concomitancy, and frequency:
     - *Infants and toddlers:* Cover test, Hirshberg and Brückner tests (p. 150), and versions
     - *Older children and adults:* Cover test, Hirshberg test, and modified Thorington (p. 153)
   - Vergences at both distance and near in phoropter or with prism bar (p. 156)
   - AC/A ratio
   - Near point of convergence (NPC)
   - Stereopsis
   - Accommodative amplitudes
3. *Cycloplegic refraction (p. 179):* Mandatory in all cases where strabismus, amblyopia, or significant refractive error is suspected. Recommended administration is as follows: 2 gtt of 1% cyclopentolate separated by 5 minutes. 1 gtt of 1% cyclopentolate can be used in infants under 1 year of age. Onset of maximal effect comes approximately 30 minutes after instillation. The duration of the action is approximately 6 to 12 hours.
4. Perform a thorough external and dilated internal ocular health assessment.

TREATMENT

The two most common approaches to the treatment of DE are vision therapy and surgery. Disagreement about the definition of a "cure" (cosmetic versus functional goal) has led to wide varia-

tions in reported success rates. That being said, it appears that vision therapy provides a higher level of functional success after a long treatment period (4 to 6 months), while surgery can provide "instant" cosmetic alignment.

Vision Therapy

A complete description of commonly used models of vision therapy is beyond the scope of this text. The following is a brief outline of one model. The reader is encouraged to consult additional sources for details concerning this subject.[1,4,5]

Flax/Brock Model[1,5]. Commence the patient's training at near (40 cm) with third-degree fusion targets, and over time increase the training distance, reducing the degree of the fusional target from third to second and ultimately to first. Standard monocular and binocular accommodative techniques, as well as combination techniques, are used. Postural awareness via monocular use of prisms horizontally and vertically is often employed as part of the early phase of therapy. Flax believed that a defective accommodative system contributes to the development of DE, and as such, plus at near to stabilize the binocular/accommodative systems is another ultimate goal.

Surgery

Although there is some debate in the literature, most physicians will not perform surgery until the patient is at least 4 to 5 years of age. Bilateral recession of the lateral rectus seems to be the preferred procedure regardless of the subtype. Initial, slight overcorrection is preferred. Postsurgical follow-up is required.

Botulinum Toxin A

One study suggests that botulinum toxin A is an appropriate treatment for children between 2- and 4-and-a-half years of age.[6] All subjects in this age range had the angle corrected to ±10 PD of orthophoria.

Minus Lenses[7-9]

The use of overcorrecting minus lenses to treat intermittent exotropia (not just DE) has been studied seriously since 1975.

This treatment is typically used in cases involving young patients not ready for vision therapy or surgery, or adults specifically concerned with cosmesis and unwilling to pursue other treatment options. Best results seem to be obtained with subjects whose angle of deviation is relatively small (less than 15 to 20 PD). One study found that such treatment did not cause additional myopia in comparison with a control population.[10]

Obviously the application of additional minus lenses will have a deleterious effect on distance vision and a profound effect at near on those who are prepresbyopic or presbyopic. Knowledge of the AC/A ratio in conjunction with the expectation that patients will not tolerate vision below 20/30 should guide the clinician through a determination of the "best" initial prescription. If the patient cannot tolerate the overminus, or if with the maximally tolerated overcorrecting minus the reduction in the deviation is not significant, then the patient is not a candidate for this technique. Patient education about the need to modify the prescription should be provided. Anticipate seeing the patient at least once more, approximately 1 month after dispensing. Once the patient is stable, follow up as needed. Ultimately the goal is slowly to remove the overcorrection while maintaining the alignment. This process is done on an individual basis over a variable amount of time, depending upon the patient's needs, age, and symptoms.

FOLLOW-UP

Follow-up will depend on the treatment (or lack thereof), as well as the history of the deviation. For example, a 2-year-old child first diagnosed with a concomitant intermittent exotropia only 15% of the time might be seen several times during the year, with parents instructed to note between visits which eye is deviated, by how much, and how often. Action can then be taken promptly if any signs appear that the condition is not a true intermittent exotropia, or if treatment becomes necessary. However, a 25-year-old patient with a long-standing DE who is not bothered functionally or cosmetically should be seen simply for regular care. Finally, a newly diagnosed case of intermittent exotropia needs to be worked up (and referred and/or followed closely) to determine whether the condition is merely a decompensated phoria (a care-

ful history is a necessary component), or whether an acquired condition, such as a tumor or neuromuscular disease, is present.

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4.2 Infantile/Congenital Exotropia

GENERAL INFORMATION

Infantile/congenital exotropia is an extremely rare entity.[1] Although the exact prevalence is debated, most agree that it is far less common than infantile esotropia.[2] Infantile exotropia has been classically defined as a large (30 to 80 PD) constant exotropia that develops during the first 6 to 12 months of life. The size of the angle may increase over time. Since many patients with infantile exotropia have alternating fixation, it is rare to find amblyopia. Unlike congenital esotropia, nystagmus is uncommon.[1] Recently, the notion that all infantile cases of XT are constant has been challenged, so one should follow closely patients

with intermittent XT for the first year or two of life, monitoring them for changes in frequency.[2]

SYMPTOMS

A parent or guardian may seek to address cosmetic concerns about the child. Symptoms reported on the part of the patient are obviously uncommon due to the age of diagnosis.

SIGNS

- A large constant (or infrequently intermittent) exotropia, usually associated with alternating fixation, is present.
- The patient may have coexisting ocular, craniofacial, or systemic abnormalities.

DIFFERENTIAL DIAGNOSIS

- *Sensory strabismus (p. 66):* There is an ocular pathology or an amblyogenic factor as the causative agent. Assessment of refractive error and a thorough internal and external ocular health assessment can usually rule out this condition.
- Some genetic conditions such as Down syndrome have an associated exotropia and/or craniofacial abnormalities that will result in a different natural history of the exotropia.
- *Divergence excess exotropia (p. 58):* This condition may develop at a young age. Young children with this condition will usually not show strabismus during near testing and will show intermittent strabismus during distance testing. An occasional eye turn will likely be reported by the parent.

WORK-UP

1. *History:* Onset of strabismus? Frequency of deviation? Recent changes in the strabismus? History of unilateral vision loss? Complications during the pregnancy, or during the perinatal or postnatal periods? Associated systemic neurological conditions?
2. *Measurement of visual acuity:* The age of the child will determine the type of acuity measure to obtain:

- *Infants:* Preferential Looking method, such as Teller Acuity cards.
- *Preschool-aged children:* Lea symbols, HOTV, or Broken Wheel cards.
- *School-aged children:* Snellen chart or Log MAR.
- *For all ages:* Behavioral assessment of acuity can be obtained by observing the patient's monocular/binocular fixation pattern (p. 147).

3. *Assessment of deviation:* Measure magnitude at distance versus near, laterality, concomitancy, and frequency:
   - *Infants and toddlers:* Cover test, Hirshberg and Bruchner tests (p. 150), and versions.
   - *Older children/Adults:* Cover test, Hirshberg test, and modified Thorington (p. 153).
4. *Cycloplegic refraction (p. 179):* Mandatory in all cases in which strabismus, amblyopia, or significant refractive error is suspected. Recommended administration is as follows: 2 gtt of 1% cyclopentolate separated by 5 minutes. 1 gtt of 1% cyclopentolate can be used in infants under 1 year of age. Onset of the maximal effect comes approximately 30 minutes after instillation. The duration of action is approximately 6 to 12 hours.
5. Slit-lamp evaluation of the anterior segment to rule out a sensory cause of the exotropia: Cataracts or a cloudy cornea.
6. *Dilated evaluation to rule out a sensory cause of the exotropia:* Vitreal opacity, retinal detachment, toxoplasmosis, or an anomalous optic nerve (hypoplasia).

ADDITIONAL TESTING

In spite of a negative work-up for another cause, prompt referral of a case of infantile XT to a pediatric ophthalmologist is prudent. Additional testing, including a comprehensive neurological work-up and electrophysiological testing, is often needed to confirm the diagnosis and to determine the cause.

TREATMENT AND FOLLOW-UP

- *Surgical intervention:* This is done typically later in life than in cases of infantile esotropia. After making a referral for a

confirmation of the diagnosis, you may comanage the patient with the surgeon.

- Co-management would include the following:
  1. Monitor the patient for the rare case of amblyopia (p. 140).
  2. *Repeated visual acuity testing:* The patient should be taught to respond to pediatric recognition visual acuity tests (LEA or HOTV) as soon as possible. A photocopy of the symbols for home instruction may facilitate this process.
  3. Monitor the patient for changes in refractive error (significant anisometropia).
  4. Perform repeated assessment of the integrity of the visual system (pupils, acuity, confrontation visual fields if possible, retina, and optic nerve).
  5. Assess the strabismus, watching for changes in frequency or the development of a strong fixation preference, which in turn may lead to amblyopia.
- Written communication between co-managing practitioners is suggested to minimize unnecessary and excessive testing.
- Finally, for patients who have a significant visual impairment or are multiply impaired, a referral for community-based functional visual assessment is warranted. Various vision professionals may perform this service, such as teachers for the visually impaired, orientation and mobility specialists, and certified low-vision specialists. The results of their evaluations, along with your input, could be critical in helping these patients maximize the use of their vision and/or employ other means for learning.

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4.3 Sensory Strabismus

GENERAL INFORMATION

Sensory strabismus is a rare condition generally thought to be caused by a monocular obstruction to vision resulting in a suffi-

cient loss of visual information such that a strabismus develops over time. However, bilateral (though asymmetric) conditions such as anisometropia and optic nerve anomalies may also result in a sensory strabismus.

The prevalence of sensory strabismus is high among patients with congenital vision loss, esotropia being at least as likely to develop as exotropia (if not more likely).[1,2] Beyond the age of 5, patients with an acquired vision loss are more likely to have an associated exotropia.[3]

SIGNS

- A large (30 to 60 PD) unilateral basic strabismus is present, one that may have an associated vertical deviation. The vertical deviation is more likely found in sensory esotropia.
- Visual acuity is significantly decreased in the affected eye.
- There is a possible history of a predisposing sensory-impeding event. A partial list includes anisometropia, cataracts, trauma, retinopathy of prematurity, staphyloma, foveal hypoplasia, persistent papillary membrane, vitreous hemorrhage, glaucoma, and retinal detachment.

SYMPTOMS

- In patients (old enough to express themselves) with an acquired sensory strabismus, changed or decreased vision may be a presenting symptom.
- Infrequently the patient reports pain with eye movement, headache, or photophobia.
- *Cosmetic complaints:* Parent/guardian or patient may complain of an eye turn or drifting eye. They may also report that under some circumstances "the eye looks funny."

DIFFERENTIAL DIAGNOSIS

The presence/absence of unilateral reduced acuity will play a major role in the differential diagnosis of sensory strabismus.

- *Infantile esotropia (p. 27) or infantile exotropia (p. 63):* A careful birth history, the absence of unilateral reduced acuity, and a thorough assessment of ocular health should differentiate these conditions from sensory strabismus.

- *Basic exotropia:* Constant exotropia with equal magnitude at distance and near with no associated predisposing sensory-impeding event.
- *Sudden-onset exotropia:* This condition may result from trauma (involving or not directly involving the orbit), tumor, or infarction. It is often associated with nonconcomitancy and diplopia. Aside from a thorough examination, visual fields testing and referral to a neuroophthalmologist is warranted. In sensory strabismus, the onset of the strabismus is not usually sudden.
- *Intermittent exotropia of the convergence-insufficiency type (p. 1):* Exotropia or phoria is larger at near than at distance, without unilateral reduction in acuity. Diplopia at near may be reported.
- *Intermittent exotropia of the divergence-insufficiency type (p. 53):* Exotropia or phoria is larger at distance than at near, without unilateral reduction in acuity.

WORK-UP

1. A history should include the following questions:
   - *General questions:*
     1. During previous eye exams, were problems noted, and if so, what treatments were prescribed?
     2. When was the onset and what has been the duration of the strabismus?
     3. What is the direction, and has there been any change in direction (e.g., in the case of a patient born with esotropia but now exhibiting exotropia)?
     4. If there is a history of strabismus surgery, one should ask the following: when and how many times was surgery performed, and what direction was the strabismus originally?
     5. If there is history of unilateral vision loss: why, how long, and what kind of previous and current care? Trauma? Systemic health? (It may be helpful to inquire about what other doctors the patient sees regularly.)
   - *Additional questions:*
     1. *Infants and preverbal children:* Were there birth complications? Head turns or tilts? Is the child's behavior consistent with unilateral vision loss (e.g., holding toys in front of one eye only)?

   2. *Verbal children and adults:* Is diplopia present? What are the patient's concerns (cosmesis, function, or both)? Are there associated neurological symptoms (poor balance or coordination) or signs (nystagmus, additional ocular motor deficits)?
2. *Measurement of visual acuity:* The age of the patient will determine the type of acuity measure to obtain:
   - *Infants:* Preferential Looking method, such as Teller Acuity cards.
   - *Preschool-aged children:* Lea symbols, HOTV, or Broken Wheel cards.
   - *School-aged children:* Snellen chart or Log MAR.
   - *For all ages:* A behavioral assessment of acuity can be made by observing the patient's monocular/binocular fixation pattern (p. 147).
3. *Assessment of deviation:* Magnitude at distance versus near, laterality, concomitancy, and frequency*:
   - *Infants and toddlers:* Cover test, Hirshberg and Brüchner tests (p. 150), versions testing.
   - *Older children/Adults:* Cover test, Hirschberg test, modified Thorington (p. 153).
4. *Cycloplegic refraction (p. 179)*: Mandatory in all cases in which strabismus, amblyopia, or a significant refractive error is suspected. Recommended administration is as follows: 2 gtt of 1% cyclopentolate separated by 5 minutes. 1 gtt 1% cyclopentolate can be used in infants under 1 year of age. Onset of maximal effect comes approximately 30 minutes after instillation. The duration of action is approximately 6 to 12 hours.
5. Slit-lamp evaluation of the anterior segment to rule out a sensory cause of the strabismus: Cataracts or a corneal opacity.
6. *Dilated evaluation to rule out a sensory cause of the strabismus:* Vitreal opacity, retinal detachment, toxoplasmosis, anomalous optic nerve (hypoplasia).

* If unilateral vision loss is present, care must be taken to utilize a fixation target that can be seen by the affected eye. If vision loss is severe, cover test (light as target), Brüchner test, and Hirschberg/Krimsky test may be used.

ADDITIONAL TESTING

Depending on the cause, consider referral to an appropriate specialist. If this is the first diagnosis, then referral is necessary. If it is a long-term and stable condition, monitoring may be sufficient.

TREATMENT

Even if good vision can be obtained in the affected eye, development or recovery of bifoveal fixation is rare. Avoidance of fusion or intractable diplopia is far more likely.[3] A lapse of 2 years or less between the sensory deprivation and the restoration of acuity is important for a good outcome.[4] However, there are case reports of good outcomes following even 15 years of deprivation.[5]

General Treatment

- Communication with the family and with those to whom you are referring the patient, as well as with the patient's primary care physician, is critical.
- Regardless of cause and treatment, a prescription of polycarbonate lenses, guidance concerning activities to avoid, and at the appropriate time, career counseling are all warranted.
- If patients have associated neurological conditions in addition to sensory strabismus, then referrals for community-based functional visual assessment may be warranted. Various vision professionals may perform this service, such as teachers for the visually impaired, orientation and mobility specialists, and certified low-vision specialists. The results of their evaluations, along with your input, could be critical in helping these patients maximize the use of their vision and/or employ other means for learning or for achieving maximal efficiency in the workplace.

Functional Cure

- *If the case is refractive in origin:* Referral to an optometrist competent in the care and management of strabismus and amblyopia should be considered. The doctor may consider full

optical correction (spectacles or contact lenses) as well as prismatic correction, which would be reduced as therapy proceeds.
- *If the case is due to a physical disruption of the sensory input:* Referral to an appropriate physician is recommended. The case may ultimately require several doctors working together in an attempt to achieve recovery of vision and binocular status. The list may include a cataract surgeon, a strabismus surgeon, and an optometrist to direct the postoperative vision therapy and/or low-vision care. In addition, neurologists may be involved.

Cosmetic Cure

- *Surgery:* The risk of diplopia from surgery in long-standing strabismic patients may be avoided because of the reduced unilateral acuity associated with sensory strabismus. Depending on the patient's level of cosmetic concern, the option of surgery should be presented and referral to a surgeon with experience in adult strabismus surgery considered.
- *Creative use of prism and spectacles:* The use of a cosmetic prism (prism prescribed with the base in the direction of the strabismus) in isolation or combined with overcorrecting minus lenses (refer to overcorrecting minus lens treatment on p. 61) can improve cosmesis by shifting the image of the eye presented to an external observer.

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4.4 Consecutive Exotropia

GENERAL INFORMATION

Consecutive exotropia is a condition in which a patient manifests an exotropia that had originally been an esotropia. The majority of cases are due to the surgical overcorrection of an esotropia, although this condition does occur in cases without surgical intervention.[1-5] The development of the consecutive exotropia may occur soon after the initial surgery or well into adulthood. Approximately 10% to 20% of early presented esotropes may, over time, progress to a consecutive exotropia.[4,5]

Why a significant percentage of moderate-angle esotropes (congenital or partially accommodative) go on to develop exotropia is not well understood at this time. Hyperopia greater than 4.50 diopters (D) is the strongest risk factor for the nonsurgical development of a consecutive exotropia.[1] Other potential factors include the level of fusion, the type of correspondence, the presence of amblyopia, and the level of accommodative function.[1,4,5,7,8] As a primary eye-care optometrist, you are less likely to see the recently postoperative esotrope who is now an exotrope, than you are to examine a patient whose status has changed months to years later. Thus, one must rule out other causes for the reported changes.

SIGNS

- A moderate to large, usually constant exotropia is present, one that may have an associated vertical deviation.
- The deviation *may* not be concomitant if the patient has had prior surgery. In either case, the review of previous records and/or photographs is critical to confirm the diagnosis, and to determine the time line of the change in direction and the magnitude of the deviation.
- There is a history of infantile or (partially) accommodative esotropia that may have been treated surgically and/or with spectacles.

SYMPTOMS

- A common reason for presentation is cosmesis.
- Patients may report an exotropia increasing (in frequency and size) over time.

- Diplopia is rare, but it may occur if the strabismic deviation changes in adulthood.

DIFFERENTIAL DIAGNOSIS

All types of exotropia can be included in the differential diagnosis. A history of esotropia, typically with surgical intervention, is the key finding in the proper diagnosis of consecutive exotropia. Even in light of a positive history of strabismus surgery, care should be taken to exclude any other possible coexisting problem (recent changes in vision, neurological conditions) that may be leading to the strabismus.

WORK-UP

1. *History:* Review old records and photos to document the course of development and any changes in the strabismus. Diplopia or sudden changes in acuity could indicate something recent (e.g., sensory XT, central lesion, or systemic problem). Appropriate referral may be warranted pending work-up.
2. *Visual acuity:* A sudden change in acuity is not associated with this condition, and care should be taken to rule out other, more ominous causes for the presenting signs and symptoms (some of which are mentioned throughout this section). The age of the patient will determine the type of acuity measure to obtain:
   - *Infants:* Preferential Looking method, such as Teller Acuity cards.
   - *Preschool-aged children:* Lea symbols, HOTV, or Broken Wheel cards.
   - *School-aged children:* Snellen chart or Log MAR.
   - *For all ages:* A behavioral assessment of acuity can be made by observing the patient's monocular/binocular fixation pattern (p. 147).
3. *Assessment of deviation:* Magnitude at distance versus near, laterality, concomitancy, and frequency (in comparison with previous records). If a significant negative change has occurred (e.g., the development of a large nonconcomitiancy), referral is indicated.
   - *Infants and toddlers:* Cover test, Hirshberg and Brüchner tests (p. 150), versions

- *Older children/Adults:* Cover test, Hirshberg test, modified Thorington (p. 153)

4. *Sensory fusion:* If the patient has a long-standing condition, it is likely that there have been multiple evaluations. Comparison of the patient's level of fusion during your work-up with that noted in the past is helpful in determining whether a significant change has occurred. One example of an easily employed test of sensory fusion is the Worth four dot test. If the patient reports suppression, then it is likely that a sudden change in the presentation of symptoms has not occurred. If the patient reports fusion without obvious alignment (extremely rare), then this would also imply a long-standing and well-adapted condition. However, if diplopia is reported, then this would indicate a recent and likely change in the status of the condition. The results of the Worth four dot test with compensatory prism in place would have important implications for treatment (surgical or vision therapy) once more ominous causes for the change in status had been ruled out.
5. *Slit-lamp evaluation of the anterior segment to rule out a sensory cause of the exotropia:* Cataracts or a cloudy cornea. It is also critical to determine whether the patient has had prior strabismus surgery. Often one can locate the point at which the surgeon approached the ocular muscles by noting changes (scarring and/or vascularization) in the bulbar conjunctiva or sclera. Be sure to assess each eye in extreme gaze left and right.
6. *Cycloplegic refraction (p. 179):* Mandatory in all cases in which strabismus, amblyopia, or significant refractive error is suspected. Recommended administration is as follows: 2 gtt of 1% cyclopentolate separated by 5 minutes. 1 gtt 1% cyclopentolate can be used in infants under 1 year of age. Onset of maximal effect comes approximately 30 minutes after instillation. The duration of action is approximately 6 to 12 hours.
7. *Dilated evaluation to rule out a sensory cause of the exotropia:* Vitreal opacity, retinal detachment, toxoplasmosis, anomalous optic nerve (hypoplasia). Papilledema should also be ruled out.

TREATMENT

- *Reduction of the hyperopic correction or the use of overcorrecting minus lenses:* This technique stimulates accommodation and can help

control the consecutive exotropia. It may be successful in younger children, but the acuity compromise often precludes this treatment in presbyopic adults. Trial framing with reduced plus correction and an examination of the effect on distance/near acuity, along with a cover test, can act as a guide for determining the correct prescription. This technique may be more successful in patients with a high AC/A ratio.

- *Vision therapy:* Once it has been demonstrated that only the angle of turn has increased and that one is in fact dealing with a case of consecutive XT, then one may carefully consider this option. A primary-care optometrist who does little VT should not consider performing therapy with this patient: if the therapy is approached in an incorrect manner, intractable diplopia may result. Most therapists, after a thorough evaluation, would consider using techniques to develop and improve peripheral fusion. As peripheral skills improve, careful attention to improvement of monocular skills and further reduction (or increased control) of the angle would be considered. The goal of most practitioners would most likely be a small angle strabismus that is not cosmetically noticeable, with excellent peripheral fusion and reasonable accommodative skills.
- Surgery
  1. *For cases presented soon after the initial surgery:* It is common for most surgeons to plan for a small overcorrection of the esotropia; however, an exotropia present 6 weeks after surgery may require additional surgery. The exotropia could be as the result of a slipped medial rectus muscle, but it is more likely due to an aggressive initial correction.[9] It is probable that the child who has undergone the surgery is still under a tight follow-up schedule with the surgeon, and patient (parent) education/support may be all that is required. Most surgeons will have made the parents aware of the risks and benefits of surgery, as well as the chances for and timing of a second surgery. It would be good practice to either call or send a note to the surgeon documenting your discussion with the parent and the need for maintaining follow-up.
  2. *For cases presented long after the initial surgery:* When providing a surgical consultation for a patient with a long-standing consecutive XT that is cosmetically noticeable, care should be taken by the surgeon to preoperatively assure the

improbability (one *cannot* assure impossibility) of either intractable diplopia or the inability to maintain alignment. A preferred version of a prism adaptation test may be used in this situation. Some might even have the patient use the proposed prism correction at home on a part-time basis to judge the patient's reaction and to determine the stability of the proposed correction.

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5

Mechanically Restrictive Strabismus

NICOLE QUINN

5.1 Duane Retraction Syndrome

GENERAL INFORMATION

Duane retraction syndrome (DRS) is a congenital ocular motility disorder, most likely caused by abnormal development of the abducens nerve and anomalous innervation of the lateral rectus muscle by the oculomotor nerve. It occurs more commonly in females and in the left eye. Approximately 20% of cases are bilateral and 10% are inherited.[1] Traditionally DRS is classified in three types (Box 5.1-1), with Type I occurring in the majority of cases. All three types show characteristic globe retraction and narrowing of the interpalpebral fissure in adduction.

BOX 5.1-1
Subtypes of DRS

Type I	Type II	Type III
Limited abduction	Limited adduction	Limited abduction
Minimal adduction limitation	Minimal abduction limitation	Limited adduction
Esotropia common	Exotropia common	Exotropia common

SYMPTOMS

- Asymptomatic
- Cosmetic concerns (eyes not moving together, anomalous head posture, strabismus)
- Diplopia

SIGNS

- There is a marked limitation in abduction (most common), in adduction, or in both (Box 5.1-1).
- Retraction of the globe (Fig. 5-1) and narrowing of the palpebral fissure of the affected eye during adduction characteristically occur; this process may be difficult to observe in infants; it may cause the appearance of enophthalmos or ptosis in the affected eye.
- A widening of the palpebral fissure of the affected eye occurs during abduction.
- Upshoots or downshoots of the affected eye occur in adducted position.
- The patient may have strabismus in primary gaze, most commonly esotropia of less than 30 diopters (D).
- If patients are strabismic in the primary gaze, there will be a compensatory head posture. (Esotropia will turn the head towards the affected eye; exotropia will turn the head away from the affected eye.)
- Stereopsis may be present, often decreased or only present with a compensatory head tilt.[2]
- Patients will exhibit a receded near point of convergence (NPC) and exotropia in the contralateral gaze (due to the adduction limitation).
- Associated ocular findings include crocodile tearing (tearing while chewing), epibulbar dermoids, nystagmus, coloboma, ptosis, anisocoria, iris heterochromia, optic nerve hypoplasia, cataracts, and Marcus-Gunn jaw-winking syndrome.[1,3-5]
- Systemic findings (Box 5.1-2) have been described in patients with DRS.[1,4,5]

DIFFERENTIAL DIAGNOSIS

- *Abducens nerve palsy (p. 100):* Esotropia with limited abduction, negative forced duction test, normal NPC, overaction of

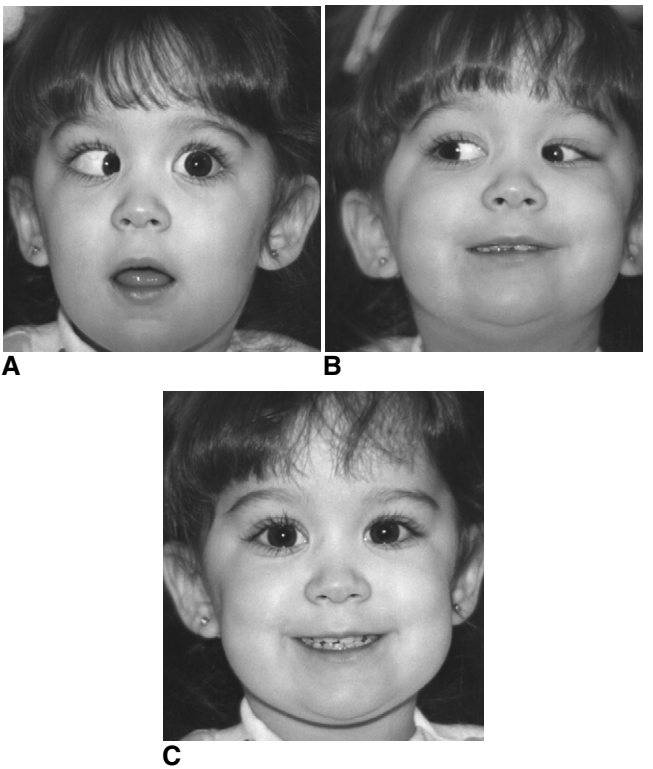

FIG. 5-1 Typical presentation of Duane's syndrome in a female patient affecting the left side. Note limited abduction of the left eye when patient looks to the left **(A),** retraction of the globe of the left eye in right gaze **(B),** and normal eye alignment in primary gaze **(C).** (From Moore BD: Eye care for infants and young children, Boston, 1997, Butterworth-Heinemann.)

contralateral medial rectus. No upshoots or downshoots are present.

- *Infantile esotropia (p. 27):* Large angle strabismus with no abduction deficit.
- *Möbius syndrome*[6]*:* Congenital sixth and seventh nerve palsies resulting in limitation of horizontal eye movements, especially abduction, and facial weakness. Möbius syndrome is usually bilateral. Esotropia and lagophthalmos are common. Vertical

BOX 5.1-2
Systemic Findings Associated With DRS

- Sensorineural deafness or hearing loss
- Skeletal abnormalities affecting feet, palate, rib, and vertebrae
- Cardiac abnormalities
- Goldenhar syndrome (vertebral abnormalities, epibulbar dermoids, facial hypoplasia, auricular malformations including preauricular skin tags)
- Klippel-Feil anomaly (fusion of cervical vertebrae with resultant torticollis)
- Wildervanck's syndrome (triad of DRS, deafness, and Klippel-Feil anomaly)
- Okihiro syndrome (DRS, congenital arm and hand abnormalities)

eye movements and Bell's phenomenon are typically preserved. Common associations include mental retardation, and tongue, craniofacial, and skeletal abnormalities. Möbius syndrome is easily confused with bilateral DRS. Patients may have crocodile tearing.[7] No globe retraction, upshoots, or downshoots are present.

- *Congenital oculomotor apraxia (p. 117):* Voluntary horizontal eye movements are impaired, involuntary eye movements intact; patient exhibits characteristic horizontal head thrusts.
- *Brown's syndrome (p. 82):* Motility restriction occurs during elevation and adduction; patients may have downshoots and widening of palpebral fissure in adduction.
- *Pseudo- or acquired DRS:* This condition becomes manifest later in life due to mechanical restriction, associated with an underlying cause such as orbital trauma, surgery or tumor, thyroid ophthalmopathy (p. 88), or orbital inflammatory disease.

WORK-UP

Although amblyopia is rare unless strabismus is present in the primary gaze, careful evaluation of acuity, refractive error, and stereopsis should be performed in all pediatric strabismus cases.

1. *History:* At what age did onset occur? Was it due to a viral illness or trauma (may suggest other cause)? Is diplopia present? Are there associated congenital defects (Box 5.1-2)? Hearing problems? A family history of DRS, strabismus, deafness, or associated syndromes (Box 5.1-2)?
2. *Externals:* Is head posture abnormal? Do photographs demonstrate a head tilt?

3. *Measurement of visual acuity:* The age of the child will determine the type of acuity measurement to obtain:
   - *Infants:* Preferential Looking method, such as Teller Acuity cards.
   - *Preschool-aged children:* Lea symbols, HOTV, or Broken Wheel cards.
   - *School-aged children:* Snellen Chart or Log MAR.
   - *For all ages:* A behavioral assessment of acuity can be obtained by observing the patient's monocular/binocular fixation pattern (p. 147).
4. *Ocular motility:* Measure versions and ductions, looking for limitations in horizontal movements, upshoots or downshoots in lateral gaze, and changes in globe position. Use Doll's head maneuver to elicit abduction in difficult patients (i.e., physically move the patient's head into the position that will require abduction of the affected eye in order to maintain fixation on a target, eliminating the need for a voluntary pursuit of the target).
5. *Cover test at distance and near:* Evaluate the patient in all gaze positions as well as with the head in a compensatory position.
6. *Binocularity:* Stereopsis may be reduced or present only with compensatory head tilt.

ADDITIONAL TESTING

1. *Forced duction testing:* Fibrotic medial rectus muscle is common in long-standing cases of DRS.
2. *Electrooculography (EOG):* Decreased saccadic velocity is present in DRS.
3. Perform hearing and ECG testing in cases in which other congenital defects are suspected.

TREATMENT

- If no strabismus is present in the primary gaze, or if the patient can maintain single binocular vision using a cosmetically acceptable head turn, routine follow-up and education regarding the condition are appropriate.
- Surgery may be indicated if there is significant strabismus or abnormal head posture present.

- If amblyopia is present, correct any significant refractive error and initiate patching therapy (p. 140).
- Refer the patient for a systemic evaluation to rule out associated features.

FOLLOW-UP

- If single binocular vision is present in the primary gaze, follow up the patient every year or prn.
- If amblyopia is present, see the patient every 4 to 8 weeks during patching therapy.
- If significant strabismus or a head tilt is present, refer the patient for a timely surgical evaluation.

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5.2 Brown's Syndrome

GENERAL INFORMATION

Brown's syndrome is a rare form of strabismus characterized by an inability to elevate the eye in adduction, due to abnormalities in the superior oblique (SO) muscle, tendon, or trochlea. It may be congenital or acquired, with several causes existing (Box 5.2-1).

Brown's syndrome is typically unilateral (bilateral in about 10% of cases[2]), with no predilection toward sex or laterality.[3] Affected patients rarely have an immediate family member with

BOX 5.2-1
Causes of Brown's syndrome

CONGENITAL

Inelastic tendon (most common[1])
Short tendon
Trochlear or tendon-trochlear complex abnormalities

ACQUIRED

Idiopathic
Tendon shortening after SO surgery
Damage/scarring after
- Scleral buckling surgery
- Glaucoma valve surgery
- Sinus surgery

Superior nasal orbital mass
Sinus infection
Blunt or direct trauma
Inflammation of trochlea associated with
- Rheumatoid arthritis
- Sjögren syndrome
- Systemic lupus erythematosus

Thyroid ophthalmopathy affecting SO

Brown's syndrome, but one third of patients with the congenital form may have a family history of strabismus or amblyopia.[1]

SYMPTOMS

- Cosmesis (eyes not moving together in the superior gaze).
- Asymptomatic.
- Diplopia in certain gaze positions.
- Pain or tenderness of the superior nasal orbit (may be present in acquired cases).

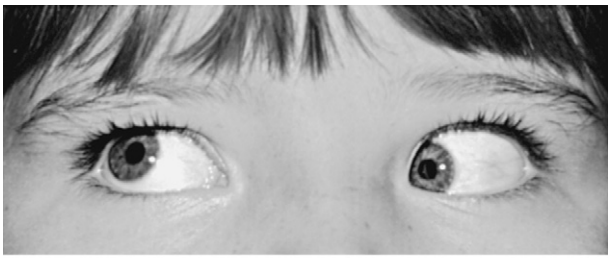
SIGNS

- *Congenital:* Restriction is usually constant, and the condition has been present since birth.
- *Acquired:* The patient may exhibit intermittent signs and symptoms, and the condition is more likely to resolve.

Due to the different causes, the diagnostic signs of Brown's syndrome are divided into features that are consistently present (primary) and features whose presence may vary among patients (variable).

Primary Features

- The patient is unable to elevate the eye during adduction (Fig. 5-2).



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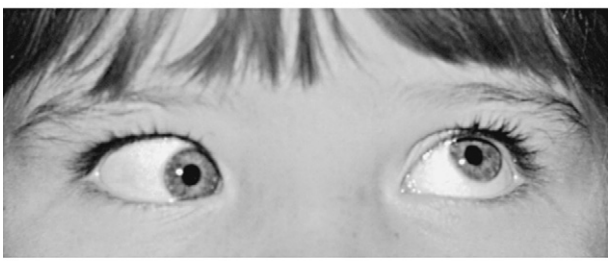

B

FIG. 5-2 Bilateral Brown syndrome demonstrated by inability to elevate either eye when adducted. (From Rosenbaum AL, Santiago AP: Clinical strabismus management, Philadelphia, 1999, WB Saunders.)

- During elevation in adduction, there is restricted forced duction of the affected eye.
- In abduction the patient experiences difficulty elevating that is significantly less than that experienced in adduction.[1]
- Minimal or no SO overaction is present.
- Small (less than 10 D) or no vertical deviation is present in the primary gaze. If vertical deviation is greater than 10 prism diopters (PD), one should suspect inferior oblique palsy, periocular scarring, or a superior nasal mass.[1]

Variable Features

- A "V" pattern (increased divergence in the upgaze) is present.
- The patient exhibits an anomalous head posture (chin up and face turned away from the affected eye).
- Contralateral inferior oblique overaction is present (in cases with an abduction deficit).

- Hypotropia is present in the primary position (being largest in cases of acquired traumatic Brown's syndrome).
- Downshoots occur during adduction.
- Widening of the palpebral fissure occurs during adduction.
- Fundus intorsion increases in the upgaze.[1]
- The patient may hear or feel "clicking" in the superior orbit during eye movements.

DIFFERENTIAL DIAGNOSIS

- *Inferior oblique palsy:* Associated SO overaction, A pattern, negative forced duction, results from Parks three-step test (p. 168) indicating inferior oblique palsy, and a larger vertical deviation in the primary gaze.
- *Primary superior oblique overaction:* Forced duction is negative; fundus intorsion is greater in the downgaze.
- *Duane retraction syndrome (p. 77):* There is typically a restriction in horizontal motility (most commonly abduction) accompanied by globe retraction. Patients with DRS may exhibit downshoots during adduction and a widening of the palpebral fissure during abduction.

The following conditions typically have an elevation deficit that is more severe in abduction than in adduction:

- *Double elevator palsy:* This condition may be present with ptosis or pseudoptosis.
- *Blowout fracture of the inferior orbital floor:* The patient has a history of trauma, enophthalmos, and infraorbital paresthesia. Radiographic studies can confirm this diagnosis.
- *Orbital floor adhesions:* The patient has a history of strabismus surgery (typically inferior oblique); depending on the location of the adhesion, the patient may exhibit an elevation deficit greater in adduction.
- *Congenital fibrosis syndrome:* This rare, familial condition is characterized by ptosis and chin elevation associated with severe limitation of eye movements (both during versions/ductions and forced duction). Eyes become fixed in inferior gaze. Typically bilateral, the condition can be asymmetric. Amblyopia, A pattern strabismus, and corneal exposure due to lagophthalmos and the lack of Bell's phenomenon are common. The condition may show a digitopalpebral sign (use a finger or hand to elevate the lid in order to remove obstruction of the visual axis).[4]

- *Thyroid ophthalmopathy* (p. 88): Patients with this condition exhibit proptosis, lid retraction, and lagophthalmos.

WORK-UP

Although amblyopia is rare unless strabismus is present in the primary gaze, careful evaluation of acuity, refractive error, and stereopsis should be performed in all pediatric strabismus cases.

1. *Case history:* When did the onset occur? Is there a history of orbital trauma, surgery, sinusitis, or inflammatory disease? Is there a family history of strabismus? Is diplopia present? Are the symptoms and signs intermittent or constant? Are the symptoms and signs improving? What is the effect on daily activities?
2. *Externals:* Is there a compensatory head turn or tilt (chin up and face turned away from the affected eye)?
3. *Measurement of visual acuity:* The age of the child will determine the type of acuity measure to obtain:
   - *Infants:* Preferential Looking method, such as Teller Acuity cards.
   - *Preschool-aged children:* Lea symbols, HOTV, or Broken Wheel cards.
   - *School-aged children:* Snellen chart or Log MAR.
   - *For all ages:* A behavioral assessment of acuity can be made by observing the patient's monocular/binocular fixation pattern (p. 147).
4. *Versions and ductions:* The affected eye shows reduced elevation that is most severe when the eye is adducted (the same degree of motility limitation on versions, ductions, and forced duction), and contralateral inferior oblique overaction may be present.
5. *Cover test:* Measure deviation in 9 positions of gaze; look for the presence and magnitude of strabismus in the primary gaze, and for the presence of a V pattern; mechanical restrictions found in Brown's syndrome may render this test ineffective in certain gaze positions.
6. *Parks three-step test (p. 168):* Rule out inferior oblique palsy.
7. *Objective fundus torsion:* Using direct or indirect ophthalmoscopy, assess the location of fovea relative to the optic disc in the primary gaze, upgaze, and downgaze. Normally the fovea is aligned with the lower third of the disc. If intorsion is

present, then the fovea will be located superior to the lower third of the disc. (In Brown's syndrome, fundus intorsion is greatest in the upgaze.)

ADDITIONAL TESTING

1. *Forced duction testing:* May be warranted if the above testing proves inconclusive (if there is a restriction when attempting elevation during adduction).
2. Using a stethoscope, listen for a "clicking" in the superior nasal orbit.
3. Lab tests, including rheumatoid factor and antinuclear antibody, are warranted when dealing with a case of acquired-onset Brown's syndrome with no history of surgery, trauma or underlying disease.[5]
4. Consider an orbital MRI or CT if the condition is of the acquired type, or if it is associated with orbital pain or inflammation.

TREATMENT

Treatment decisions are made based on the cause of the case of Brown's syndrome, with the goal of maximizing binocularity, preventing amblyopia, and relieving symptoms in affected patients. If present, amblyopia should be treated.

- Only education is required if strabismus is absent in the primary gaze and if the patient is asymptomatic. Recommend a head posture to prevent diplopia (chin up and the face turned away from the involved eye).
- Exercising the eye by repeatedly moving the eye into the restricted field has occasionally been shown to decrease signs and symptoms.[6-8]
- Surgery is appropriate if hypotropia is present in the primary gaze or if there is a cosmetically/functionally significant anomalous head posture. (Preservation of binocularity and prevention of amblyopia are the main concerns in young children). If possible, delay surgical intervention, because 6% to 7% of patients experience spontaneous resolution within 4 years of onset.[1] Acquired, nontraumatic cases are the most likely to resolve.
- Refer the case to a primary care doctor to rule out an underlying systemic cause in acquired cases with no known history of

surgery, trauma, or disease. Often cases with inflammatory causes can be treated with NSAIDS or with oral or injected steroids.

FOLLOW-UP

- *Cases not warranting treatment:* The appropriate follow-up time is 1 year or prn.
- *Acquired Brown's of unknown cause:* Provide immediate follow-up, until the cause has been determined.

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5.3 Thyroid-Related Ophthalmopathy

GENERAL INFORMATION

Thyroid-related (Graves') ophthalmopathy (TRO) is an autoimmune disease causing expansion of the orbital tissues (especially the extraocular muscles) and resulting in orbital congestion. The disease is most commonly associated with hyperthyroidism (especially Graves' disease), but it sometimes occurs in hypothyroidism or euthyroidism. Systemic and ocular abnormalities typically develop within 18 months of each other, with most patients developing hyperthyroidism first.

TRO is more common in females than in males (at a 6:1 ratio) and typically appears in patients between 40 and 60 years of age. Children are rarely and usually mildly affected. The active stage (inflammation of the lids, conjunctiva, and orbit) lasts 8 months to 3 years and is followed by an inactive phase in which signs may persist but typically do not worsen. About 60% of cases spontaneously regress after 5 years.[1]

SYMPTOMS

The symptoms of TRO include a foreign-body sensation, diplopia, photophobia, lacrimation, blurred vision, retroorbital discomfort (especially during eye movements), and cosmetic changes (eyelid swelling, proptosis).

SIGNS

Signs of TRO are typically bilateral but can be unilateral or asymmetric.

- *Proptosis:* TRO is the most common cause of bilateral and unilateral proptosis.
- *Eyelid retraction:* Involving the upper and lower lid.
- *Lid lag:* On the downgaze.
- Decreased blinking (stare) is evident.
- *Ocular motility restriction and strabismus:* The inferior and medial rectus muscles are most commonly affected. There may be coexisting horizontal and vertical deviations.
- *Eyelid and periorbital edema:* Worse in the morning.
- *Conjunctival chemosis and hyperemia:* Especially in the area of the rectus muscle insertions.
- Exposure-related corneal dryness is present.
- *Superior limbic keratoconjunctivitis (SLK)*: Thickening of the superior bulbar conjunctiva, punctate erosions of superior cornea, and increased mucus production are present.
- *Compressive optic neuropathy:* This condition affects approximately 6% and is more common in males and in patients over 50 years old.

DIFFERENTIAL DIAGNOSIS OF STRABISMUS IN TRO

Because TRO can affect several extraocular muscles alone or in combination, the differential is determined by the muscle or

muscles affected. If TRO is suspected, the following conditions must be considered:

- *Orbital pseudotumor or myositis:* This condition is more commonly unilateral. Computerized tomography (CT) shows thickened Tenon's capsule and tendon involvement; there is neither eyelid retraction nor limitation of elevation.
- *Orbital tumors:* A unilateral, palpable mass may be present, with proptosis in the direction opposite from the location of the tumor, which can be primary or metastatic.
- *Ocular myasthenia (p. 133):* Full versions and negative forced duction are present. Symptoms are intermittent and often variable. Patients with myasthenia are at greater risk for TRO.[3]

WORK-UP

1. *History:* When did the onset of symptoms and signs occur? How long have they persisted? Are symptoms stable or worsening? Does the patient use tobacco? (Smokers may be at increased risk.) Is the patient recently under personal stress? Is there a personal or familial history of thyroid dysfunction or autoimmune disease (myasthenia gravis)?
2. *Visual acuity:* This may be decreased as a result of optic neuropathy or corneal exposure.
3. *Lid lag evaluation:* Observe the position of the lid relative to the globe while the patient follows the target downwards. Jerky movements or a fine tremor of lightly closed lids may also be observed.
4. *Periorbital edema:* Measure the edema by placing a ruler in the upper eyelid fold and allowing the periorbital tissues to rest on it.
5. *Exophthalmometry:* Proptosis is present if the measurement is greater than 22 mm or if there is a difference of 2 mm between the eyes. In TRO, proptosis is straight out.
6. *Extraocular motility:* Normally the range of elevation and depression measures 5.5 mm. Most commonly elevation or abduction are affected.
7. *Cover test:* If strabismus is present, monitor it with a prism cover test.
8. *Corneal evaluation with fluorescein:* May reveal exposure keratitis or SLK.

9. *Intraocular pressure (IOP):* Increased IOP will be present on an attempted upgaze in some cases. TRO patients may be at high risk for ocular hypertension and open-angle glaucoma.[4]
10. *Optic nerve evaluation:* Check pupils, brightness comparison, color vision, baseline visual field. Rule out disc edema (present in 50% of cases).[1]

ADDITIONAL TESTING

1. *CT or magnetic resonance imaging (MRI):* Findings include enlarged rectus muscle bellies without tendon involvement. These tests can be used to rule out orbital tumors.
2. *Forced duction testing:* Restrictions may occur in all positions, but most commonly in the upgaze.
3. *Thyroid function screening:* Free T_4, TSH, and T_3.

TREATMENT

1. Refer the patient for diagnosis and treatment of the underlying thyroid disorder; systemic treatment may reduce lid retraction.
2. *Strabismus:*
   - Educate the patient regarding a compensatory head posture to reduce diplopia. Teach patients to point their noses in the direction of greatest diplopia.
   - *Prisms to relieve diplopia:* Use a ground prism if the deviation is less than 12 D and stable, a Fresnel prism for deviations up to 30 D and for unstable deviations. In nonconcomitant deviations, it is best to focus on establishing single vision in the primary gaze.
   - *Occlusion to eliminate diplopia:* Adhesive or tie-on patches, opaque contact lenses, or frosted lenses may be prescribed for the temporary relief of diplopia.
   - *Strabismus surgery:* Appropriate only in cases of inactive TRO with a stable angle of strabismus.
3. *Ocular irritation and exposure:*
   - The patient should use preservative-free artificial tears during the day and lubricating ointment at night.
   - Tape lids at night if nocturnal lagophthalmos is present.
   - Perform short-term tarsorrhaphy, if symptoms are severe.
   - *Lid surgery:* Only in cases involving chronic, severe exposure.

4. *Optic neuropathy:*
   - *Systemic corticosteroids and orbital radiation therapy*: Useful in active cases with soft tissue changes and optic nerve compression.
   - *Orbital decompression surgery:* Indicated in cases with severe proptosis or compressive optic neuropathy.
5. Tinted lenses for photophobia may be prescribed.
6. *Lid edema:*
   - Have the patient elevate the head of the bed.
   - Recommend medical treatment (diuretics).

FOLLOW-UP

Follow-up is based on the kind of signs and symptoms being treated and their severity.

- *Asymptomatic cases:* Refer the patient for systemic work-up. Monitor the case at intervals of 1 to 2 months during the active phase, and at intervals of 6 months to 1 year once the condition has become stable.
- *Diplopia treated with prism/occlusion:* Monitor the case at intervals of 6 to 8 weeks until the condition has become stable.
- *Exposure:*If the case is mild, monitor it at intervals of 1 to 2 months. If corneal ulceration is present, follow the case daily until it is resolved.
- *Optic neuropathy:* Refer the patient for immediate evaluation.

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7

Horizontal Gaze Disturbances

MELISSA L. RICE

7.1 Internuclear Ophthalmoplegia and One and a Half Syndrome

GENERAL INFORMATION

Internuclear ophthalmoplegia (INO) is caused by a lesion of the medial longitudinal fasciculus (MLF). It is characterized by adduction weakness of the eye on the side of the lesion.[1] Multiple causes for INO exist: multiple sclerosis (more common in young patients), ischemia (more common in elderly patients), and a brain stem tumor.[2] Bilateral lesions in the MLF result in bilateral INO and sometimes exotropia.[1]

One and a half syndrome is secondary to combined lesions of the abducens nucleus or pontine paramedian reticular formation (PPRF) and adjacent MLF on one side of the brain stem.[1] The name of this syndrome explains the clinical picture: one eye has no lateral movement and the other eye retains abduction only. Possible causes are similar to INO and include stroke, multiple sclerosis, pontine glioma, arteriovenous malformation, basilar artery aneurysm, posterior fossa tumor, and trauma.[2]

SYMPTOMS

If symptoms are present, the onset is typically painless. Patients may report diplopia, oscillopsia, or an inability to move their eye(s), but often they have no visual symptoms.

SIGNS

Internuclear Ophthalmoplegia

- Paralysis or paresis of adduction can be present in various amounts. Evaluate horizontal saccades; look for relative slowing of adducting movement.
- Nystagmus occurs during abduction of the eye contralateral to the lesion. For example, a patient with a right INO will have difficulty with the left gaze. The right eye will be unable to fully move to the left and the left eye will exhibit nystagmus during the attempt.
- *Adduction may be preserved during convergence.*
- Hypertropia on the side of the lesion may be present.
- Vertical saccades are normal.
- Bilateral INO often leads to additional findings: bilateral adduction paralysis/paresis, exotropia (coined WEBINO syndrome), gaze-evoked vertical nystagmus, impaired vertical pursuit, and decreased vertical vestibular responses.

One and a half syndrome

- INO and ipsilateral horizontal gaze palsy are present.
- The only movement preserved is abduction of the contralateral eye.
- Paralytic pontine exotropia is present in the primary gaze (exotropia of the eye opposite the side of the lesion).
- The patient may demonstrate abducting nystagmus on the side opposite the brain stem lesion.
- Vergence and vertical movements may be spared.

DIFFERENTIAL DIAGNOSIS

- *Myasthenia gravis (p. 133):* Any variation of ocular motor signs, including a possible variable adduction deficit, and fluctuating ptosis. Symptoms usually worsen with fatigue. Results of the Tensilon test may be positive.
- *Orbital disease (inflammatory pseudotumor, thyroid disease, tumor):* Proptosis, globe displacement. Pain may be present. Nystagmus is usually not present.
- *Surgical paresis of the medial rectus muscle:* Unilateral or bilateral paresis of adduction with nystagmus of the abducting eye has been reported after a weakening operation of the medial rectus

muscle. Forced duction testing confirms mechanical restriction of adduction caused by the artificial adhesion created between muscle and sclera by the sutures.[3]

- *Bilateral INO and associated sixth nerve palsy:* This may be confused with one and a half syndrome; however, adduction in one eye is spared. Suspect this disorder in cases with asymmetric horizontal gaze palsy in which one eye is more limited than the other.

WORK-UP

1. *History:* Time of onset? Duration of symptoms? Diplopia? Medical history? Trauma? Associated neurological symptoms?
2. *Neuroophthalmologic examination:* Visual acuity, color vision, cover testing, ocular motility (including pursuit and saccade testing, near point of convergence, pupil reactions, slit-lamp examination, and fundus evaluation).

ADDITIONAL TESTING

1. Patient should have a full medical work-up with appropriate blood work and lab tests (Tensilon test, sedimentation rate, CBC, syphilis serologic study, lumbar puncture, B12 level).
2. A neurological examination should be performed.
3. Magnetic resonance imaging with focus on the brain stem and midbrain will determine the location of the lesion.

TREATMENT

- Determine the cause and manage it with an appropriate specialist.
- If diplopia is present in the primary gaze, one can manage this by applying the correct power of Fresnel prism to the spectacles. If the deviation changes in the future, this mode of management can easily and inexpensively be adjusted. If the deviation remains consistent for several months, one can prescribe the prism to be ground in to the spectacle prescription. A prism adaptation test (p. 172) is the first step in determining the benefit of a prism prescription. If the patient is not a prism adapter, apply the least amount of prism power that eliminates diplopia. Furthermore, if the deviation is strictly horizontal, patients may find that by turning their heads, they can elimi-

nate diplopia. For patients with a right-side INO, turning the head to the left will reposition the eyes so that both eyes are positioned to the right but aimed straight ahead. Using the same degree of head turn would be critical to prevent diplopia. This may be difficult for some patients.

FOLLOW-UP

- The case should be followed as the medical doctor has directed.
- Prism management should be monitored if the patient's symptoms change.

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7.2 Congenital Ocular Motor Apraxia

GENERAL INFORMATION

Congenital ocular motor apraxia (COMA) becomes manifest between the ages of 4 and 8 months.[1] This condition is more common in males than in females and improves over time. Pathogenesis of COMA is unknown; however, the consensus is that the disorder is the result of a central "miswiring."[2]

SYMPTOMS

Presenting symptoms include the following: visual inattentiveness, unusual eye and/or head movements, and perceived blindness by the pediatrician and/or parents.[1]

SIGNS

- *Thrusting horizontal head movements:* The child turns his or her head rapidly in the desired direction of gaze, overshooting the target. The eye movement follows the head movement, and

when foveal fixation occurs, the head is slowly moved back to midline with vestibulo-ocular reflex (VOR) keeping the eyes on the target.[3] Head movements typically develop by 6 months of age; however, they may be absent or delayed in children with poor head control.

- Repetitive blinking occurs when the child attempts to change fixation.
- Abnormal saccades are present, including the following:
  1. The child displays an inability to execute voluntary horizontal saccades (this can be asymmetrical), with intact involuntary saccadic ability.
  2. Vertical saccades and pursuits are unaffected.
  3. *Abnormal optokinetic nystagmus (OKN) response:* Rotate an OKN drum at approximately 2 seconds per rotation. The patient's eyes should follow the direction of rotation slowly with a quick corrective saccadic back, picking up fixation again. This pattern will repeat as long as the drum is rotated. In a patient with COMA one will observe a large slow phase but no corrective saccade.
- Horizontal pursuits are usually normal.
- VOR is intact.
- Patient may have strabismus.

DIFFERENTIAL DIAGNOSIS

- *Blindness:* The patient exhibits an inability to fixate on or follow targets after 3 or 4 months of age secondary to an ocular pathology, delayed maturation of the visual system, or high refractive error.
- *Acquired ocular motor apraxia (AOMA) (p. 120):* The patient has difficulty initiating horizontal saccades; most cases also exhibit defects in the vertical direction; other neurological signs are noted. AOMA is usually secondary to bilateral frontal or frontoparietal lesions.
- *Nystagmus:* Usually a bilateral, involuntary, rhythmic movement in a side-to-side or up-and-down pattern. It is typically faster in one direction of the movement. Vision loss may occur, as in the case of congenital motor nystagmus, or the nystagmus may be a consequence of vision loss, as in sensory nystagmus. This disorder appears in its manifestation distinctly different from the abnormal eye movements associated with COMA.

- *Spasm nutans:* This condition is characterized by a triad of nystagmus, head nodding, and anomalous head positions (e.g., torticollis). Its onset occurs during the first year of life and spontaneously remits within 2 to 8 years. Saccades remain intact in patients with spasm nutans.

WORK-UP

1. *History:* Age of onset? Family history? Other developmental delays?
2. Perform a complete eye examination, including evaluation of saccadic movements, pursuits, alignment, cycloplegic retinoscopy (p. 179), and an ocular health evaluation. VOR testing can be done by rotating the infant at arm's length or spinning the child in a chair and observing eye movements. OKN testing can be done with an OKN drum.
2. A complete systemic and developmental evaluation by pediatrician and a pediatric neurologist is recommended.
3. Neuroimaging may be indicated.

ADDITIONAL TESTING

Results of electroretinogram (ERG) and visual evoked potential (VEP) testing are normal.

TREATMENT

The natural history of patients with COMA is such that the condition improves over time but may not completely resolve. COMA does not affect visual acuity; however, the ability to fixate on objects is impaired. Families should be reassured and be referred to their pediatrician for management. In addition, children with COMA frequently have developmental delays (speech, motor)[1]: they are usually children of normal intellect, but they struggle with reading.

FOLLOW-UP

Perform a full eye exam as indicated. A multidisciplinary team, including a neuroophthalmologist, a neurologist, and a pediatrician, should follow patients with COMA.

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7.3 Acquired Ocular Motor Apraxia (Balint's Syndrome)

GENERAL INFORMATION

Acquired ocular motor apraxia (AOMA) can become manifest at any age and is caused by acute, bilateral, frontal, or frontoparietal lesions. The following diseases can be associated with AOMA: Huntington's disease, multiple sclerosis, and Wilson's disease.

Balint's syndrome is secondary to bilateral posterior parietal lesions. In addition to AOMA, it is associated with inaccurate arm pointing (optic ataxia) and disturbance of visual attention (simultanagnosia).[1]

SYMPTOMS

Patients may note an impaired ability to read and simultanagnosia (inability to perceive more than one object at a time). Frequently they will exhibit associated visual field defects or dementia.

SIGNS

- Voluntary control saccades and/or smooth pursuits are absent, with preservation of some reflexive movements. (Patients may use head movements to assist in initiating gaze shift.)
- Random eye movements and vestibulo-ocular reflex are intact.
- Vertical saccades may be involved. (If AOMA is limited to the vertical plane, then it is secondary to bilateral lesions at the mesencephalic-diencephalic junction.[2])

- Optic ataxia (inaccurate arm pointing) is associated with Balint's syndrome.
- Simultanagnosia is associated with Balint's syndrome.

DIFFERENTIAL DIAGNOSIS

- *Congenital ocular motor apraxia (COMA) (p. 117):* This condition becomes manifest between 4 and 8 months of age, with thrusting horizontal head movements, repetitive blinking with changes in fixation, and an inability to initiate voluntary saccades. Vertical saccades and pursuits are unaffected, and the vestibulo-ocular reflex (VOR) is intact. Patients with COMA may have strabismus.

WORK-UP

1. *History:* Time of onset? Medical history? Surgery? Trauma?
2. *Ocular motility/Saccades/Pursuit evaluation:* Inability to initiate saccades on command. Using an eye-head movement with a blink helps shift the gaze. Smooth pursuit may be impaired.
3. VOR testing is normal.
4. *Ocular health:* Patients can have optic nerve pallor depending on the cause; otherwise, ocular health is normal.

ADDITIONAL TESTING

1. A neurological examination should be performed.
2. Magnetic resonance imaging will determine the location of the lesion.

TREATMENT

The patient should be treated as indicated if it is secondary to a treatable lesion or condition. Ocular findings may improve over time.

FOLLOW-UP

Monitor the condition with the neurologist and neuroophthalmologist.

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8

Vertical Gaze Disturbances

MELISSA L. RICE

8.1 Vertical Gaze Palsy

GENERAL INFORMATION

Vertical gaze palsy more commonly affects upgaze or both upgaze and downgaze. The least common vertical palsy is isolated downgaze palsy.[1] Nuclei and tracts in the midbrain and thalamus control vertical eye movements, and damage to these areas can result in vertical gaze palsy.[2]

SYMPTOMS

- *Upgaze palsy:* Inability to open eyes, ptosis, difficulty looking upwards. A spouse or parent may report inward deviation.[3,4] Cases may be asymptomatic visually if the ptosis is not too severe.
- *Downgaze palsy:* Increased difficulty reading or performing other near tasks due to the inability to look downwards. Patients may experience problems using stairs or doing other visually guided tasks that rely on downgaze.

SIGNS

Upgaze palsy

- The patient is unable to rotate the eyes upward.
- Ptosis occurs bilaterally or unilaterally.
- Vestibulo-ocular reflex (VOR) is preserved.

Downgaze palsy

- The patient is unable to rotate the eyes downward.
- *Apparent ptosis in downgaze:* The lids move down, but eye movements are limited.
- *Magnetic resonance imaging:* A lesion appears in the midbrain, thalamus, rostral interstitial nucleus of the medial longitudinal fasciculus, posterior commissure, interstitial nucleus of Cajal, or Darkshevich's nucleus.

DIFFERENTIAL DIAGNOSIS

- *Orbital floor fracture:* There is a history of blunt trauma that may restrict upgaze and downgaze movement; enophthalmos, ecchymosis, and edema are present. Results of forced duction testing are positive secondary to entrapment of the inferior rectus.
- *Progressive supranuclear palsy (p. 127):* Found in elderly patients, this disorder is associated with falling, mental slowing, and difficulty swallowing/speaking. Vertical saccades will be slow, then small amplitude, followed by loss of vertical gaze.
- *Dorsal midbrain syndrome (p. 130):* Patients exhibit a limitation of upgaze, middilated pupils with light-near dissociation, retraction of lids, convergence spasm, convergence-retraction nystagmus, downgaze paresis, anisocoria, ptosis, and absence of Bell's phenomenon.

WORK-UP

1. *History:* Time of onset? Duration of symptoms? Trauma? Associated neurological symptoms?
2. *Neuroophthalmologic examination:* Visual acuity, color vision, cover testing, ocular motility (including pursuit and saccadic evaluation), pupil reactions, slit-lamp examination, and fundus evaluation should be performed.

ADDITIONAL TESTING

1. A neurological examination should be performed.
2. Magnetic resonance imaging (MRI) will determine the location of the lesion.

TREATMENT

1. Determine the cause and manage the case with a neurologist.
2. Often the palsy persists. For downgaze palsy, two pairs of glasses are recommended: one for distance and one for near. A base-down yoked prism should be trial framed to shift gaze upward, increasing the patient's comfort and decreasing symptoms during the act of reading. Start with the patient's reading prescription plus 6 prism diopters (PD) in each eye and increase the level of correction until improvement in visual comfort is noted. Remember to educate patients fully about the importance of wearing the prism prescription exclusively for reading because of the accompanying distortion and blur. In cases of upgaze palsy, a ptosis crutch could be considered if the lid position is interfering with visual function. Typically visual function is not as affected because the upgaze is used with much less frequency. Furthermore, educate patients to tip their chin up or head back when looking in upgaze.

FOLLOW-UP

- The case should be followed by a medical doctor to ensure the appropriate systemic treatment.
- Follow the case to evaluate the effectiveness of the yoked prism after 4 to 6 weeks of use and then as needed.

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8.2 Monocular Elevation Paresis

GENERAL INFORMATION

Monocular elevation paresis is a rare acquired disorder of sudden onset resulting from an interruption stretching from the vertical gaze center in the pretectum to the ocular motor complex.[1,2] This condition may be the first manifestation of a third-nerve palsy (p. 105) in patients with neurofibromatosis type II.[3]

SYMPTOMS

Patients may note diplopia on upgaze, or they may exhibit no symptoms. They may report having to hold their chin up to avoid diplopia.

SIGNS

- Monocular loss of upgaze occurs, resulting in diplopia only in upgaze.
- Abnormalities of lid position or movement may be present.
- In the early stages of the disorder, Bell's phenomenon is usually normal; however, it may become reduced in relation to the degree of the paresis.

DIFFERENTIAL DIAGNOSIS

- *Thyroid ophthalmopathy (p. 88):* This disorder commonly involves restriction of the inferior rectus (limited elevation); therefore, a vertical deviation is measured. On examination the following may be noted: lid retraction, lid lag, exophthalmos, and orbital congestion. Results of forced duction testing are positive.
- *Myasthenia gravis (p. 133):* There is a history of ptosis, along with diplopia that worsens with fatigue and has a tendency to improve. Results of additional testing by edrophonium chloride (Tensilon test) are positive.
- *Orbital floor fracture:* There is a history of blunt trauma. Enophthalmos is present. Results of forced duction testing are positive.
- *Skew deviation:* There is measured vertical deviation in the primary position.

WORK-UP

1. *History:* Diplopia? Time of onset? Is there a medical history of thyroid problems or myasthenia gravis? Trauma? Associated neurological symptoms (poor balance/coordination)?
2. *Versions:* Limitation of elevation is noted on adduction, abduction, and primary positions.
3. Cover testing reveals no vertical misalignment in the primary position or downgaze.
4. *Pupil reactions:* These can be normal; however, depending on the site of the lesion, the pupils may be involved.

ADDITIONAL TESTING

Negative forced duction testing, radiological testing, and edrophonium chloride (Tensilon) tests should be performed.

TREATMENT

Since the deviation is in the upgaze, cases are usually asymptomatic.

FOLLOW-UP

Patients should be managed by physicians familiar with the underlying disease. Perform follow-up eyecare as needed.

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8.3 Progressive Supranuclear Palsy (Steele-Richardson-Olszewski Syndrome)

GENERAL INFORMATION

Progressive supranuclear palsy (PSP) is a degenerative condition that affects persons later in life. This disease often produces a

disturbance of vertical eye movements early in its course, but this disturbance has been noted to happen late in the course of the disease or not at all.[1,2] The disease is characterized by abnormal eye movements, axial rigidity, a tendency to fall backwards, difficulties with swallowing and speech, and mental slowness.[3,4] PSP occurs with equal frequency in patients of either sex and leads to death in 6 to 10 years.

SYMPTOMS

Patients' chief complaint may be that they are having a difficult time reading, that they cannot see the food on their plate, or that they are having a difficult time walking down the stairs.[5]

SIGNS

- An initial impairment of vertical saccades occurs, followed by upgaze and horizontal limitation.
- Smooth pursuits are relatively well preserved; however, these may be lost over time.
- Oculocephalic movements are normal; however, neck rigidity may make doll's head maneuvers difficult to perform.
- Blink rate is markedly decreased.
- Axial dystonia is greater than appendicular dystonia.
- Eyelid apraxia is present.
- In the later stages of the disease, patients may develop ocular misalignment and diplopia.

DIFFERENTIAL DIAGNOSIS

- *Parkinson's disease (PD):* Upgaze is initially affected, then hypometric saccades in all directions of gaze develop. Impaired downward gaze and slow vertical saccades are more characteristic of PSP. Eye movements in patients with PSP are not improved by dopaminergic drugs as are eye movements in cases of PD.
- *Whipple disease:* This is a rare multisystem disorder characterized by weight loss, abdominal pain, gastrointestinal bleeding, diarrhea, and steatorrhea. It may involve or be confined to the nervous system. The ocular motility of patients with Whipple disease can mimic PSP: initially vertical saccades are involved,

but with time all eye movements may be lost. A highly characteristic finding is pendular vergence oscillations and concurrent contraction of the masticatory muscles. Whipple disease can be diagnosed using polymerase chain reaction (PCR) analysis and treated with antibiotics.

- *Basal ganglia disorders:* A variety of disorders can mimic PSP, including diffuse Lewy body disease, idiopathic striopallidodentate calcification, autosomal dominant Parkinsonism and dementia with pallidopontonigral degeneration, and multiple system atrophy.
- *Dorsal midbrain syndrome (p. 130):* Patients exhibit bilateral paresis of upgaze, middilated pupils with light-near dissociation, convergence retraction nystagmus, and retraction of lids. This syndrome is caused by tumors, strokes, multiple sclerosis, congenital aqueductal stenosis, neurosyphilis, and trauma
- *Upgaze palsy (p. 123):* Patients are unable to elevate the eyes; vertical optokinetic responses are absent; patients may have downward and inward deviation. This palsy is usually secondary to hypertensive thalamic hemorrhages or to infarction in the midbrain.

WORK-UP

1. *History:* Time of onset? Diplopia? Medical history? Associated neurological symptoms (poor balance/coordination)?
2. *Versions/Saccades:* Patients may exhibit a limitation in the upgaze. Slowing of vertical saccades is noted initially. Pursuits may be normal but may worsen with time. Convergence movement may be impaired.
3. Cover testing reveals no misalignment in the early stages of the disease, but misalignment may develop as the disease progresses.
4. *Ocular health:* Patients may have eyelid apraxia; otherwise, ocular health is normal.

ADDITIONAL TESTING

Eye movement recordings, evoked potentials, MRI, magnetic resonance spectroscopy, or positron emission tomography (PET) scans may be helpful in supporting the diagnosis.

TREATMENT

No treatment is known. Manage the diplopia if it is symptomatic. In addition, because patients with PSP cannot look down, they may benefit from separate reading and distance spectacles.

FOLLOW-UP

- Patients with PSP should be managed by a neurologist.
- Follow the case if symptomatic for diplopia to determine the effectiveness of prism use.

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8.4 Dorsal Midbrain Syndrome (Parinaud's Syndrome)

GENERAL INFORMATION

Lesions of the posterior commissure produce bilateral paresis of the upgaze.[1] This disorder is known by a variety of names, including dorsal midbrain syndrome, Parinaud's syndrome, Koeber-Salus-Elschnig syndrome, and Sylvian aqueduct syndrome.[2]

This condition is found in younger men with pineal tumors, in women with multiple sclerosis, and in older men who have had a stroke.[3] Other causes include congenital aqueductal stenosis, neurosyphilis, trauma, and infiltrating tumors in the region of the aqueduct and superior colliculus.

SYMPTOMS

Patient may report difficulty looking upward.

SIGNS

- *Hallmark sign:* Limitation of upgaze. As the condition progresses, attempts to look upward are replaced by a convergence movement and retraction into the orbit.
- Pupils are middilated (4 to 5 mm in diameter) with light-near dissociation.
- *Collier sign:* Retraction of lids occurs in the primary position.
- *Additional findings:* Convergence spasm, convergence-retraction nystagmus, downgaze paresis, anisocoria, ptosis, and absence of Bell's phenomenon may be present.

DIFFERENTIAL DIAGNOSIS

- *Monocular elevation paresis (p. 126):* Monocular loss of upgaze occurs, with diplopia in upgaze only, and abnormalities in lid position and/or movement.
- *Upgaze palsy (p. 123):* Patients are unable to elevate their eyes; vertical optokinetic responses are absent; downward and inward deviation may be present. This palsy is usually secondary to hypertensive thalamic hemorrhages or to infarction in the midbrain.
- *Parkinson's disease:* Upgaze is affected initially, then hypometric saccades in all directions of gaze develop. Eye movements in cases of PD improve in response to dopaminergic drugs.
- *Progressive supranuclear palsy (p. 127):* This disorder is found in elderly patients and is associated with falling, mental slowing, and difficulty swallowing and/or speaking. Vertical saccades will be slow, then small amplitude, followed by loss of vertical gaze. Eye movements is cases of PSP are not improved by dopaminergic drugs.

WORK-UP

1. *History:* Time of onset? Duration of symptoms? Diplopia? Trauma? Associated neurological symptoms?
2. *Versions:* A bilateral limitation of upgaze is observed; convergence movement with retraction may be observed with OKN

or vertical saccades. Downgaze limitation may be noted as well. Check saccades for a decreased velocity of vertical upward saccades (downward saccades may be involved as well). Pursuits may or may not be abnormal.

3. *Pupil testing:* Middilated pupils may be noted, with light pupillary reaction to light being less than near pupillary response. Possible anisocoria.

TREATMENT/FOLLOW-UP

Systemic disease or involvement should be managed by a physician.

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9

Ocular Myasthenia Gravis

NICOLE QUINN

GENERAL INFORMATION

Ocular myasthenia gravis (MG) is an autoimmune disease that causes a decrease in available acetylcholine receptors at the neuromuscular junctions, resulting in progressive muscular fatigue and weakness.

Ocular symptoms are the presenting complaint in a majority of cases, with the levator palpebrae, extraocular muscles, and orbicularis oculi most commonly affected.[1] Systemic symptoms (Box 9-1) are often present at diagnosis or develop within 3 years. Most commonly onset occurs between the ages of 20 and 49, with women affected more often and earlier than men. Children are occasionally affected.[2]

SYMPTOMS

Patients classically describe their symptoms as variable, becoming worse at the end of the day or after physical exertion.

BOX 9-1
Symptoms of Generalized MG

- Limb, neck, or facial muscle weakness
- Change in voice quality (slurred or nasal)
- Difficulty chewing
- General fatigue and weight loss
- Difficulty swallowing or breathing (warrants immediate referral)

- Ptosis
- Diplopia (may report both horizontal and vertical diplopia)
- Blurry vision at distance or near

SIGNS

Lid

- *Ptosis:* May start unilaterally but becomes bilateral. Asymmetry is common. Ptosis may shift from eye to eye. It is most pronounced at the end of the day or after strain has been placed on muscles. Enhancement of ptosis is present (ptosis is worsened if contralateral eyelid is manually elevated).
- *Lid retraction:* May be present after periods of upwards gaze, in unilateral ptosis, as an attempt to elevate affected eyelid, or as Cogan's sign (Work-up). (Fig. 9-1.)
- Lagophthalmos is present with associated corneal exposure.
- Ectropion of the lower eyelid is present.

Ocular Motility

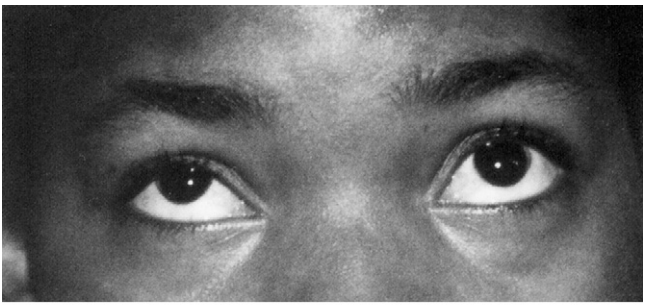
- *Limitations in ocular motility:* May affect a single muscle (most commonly medial recti, inferior recti, or superior oblique[1]) or a combination of muscles. The limitation may be unilateral or bilateral. Forced duction is negative.
- *Strabismus:* The pattern depends on the muscles affected. The strabismus is often intermittent, nonconcomitant, and variable in magnitude.

Binocular Vision/Accommodation and Pupil

- *Accommodative dysfunction:* This is present as an accommodative insufficiency, spasm, or rapidly fatiguing accommodative performance; the condition is not improved by vision training.[3]
- Magnitude of phoria and vergence fatigue increase throughout the day.[3]
- There is no clinically observable effect on pupillary response.

DIFFERENTIAL DIAGNOSIS

Because MG can mimic underaction of an individual muscle or a combination of extraocular muscles, the differential is deter-



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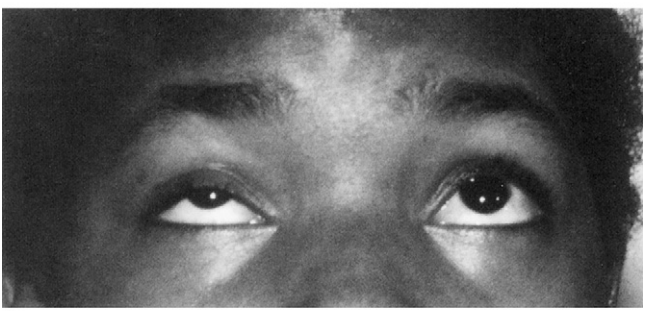

B

FIG. 9-1 Patient with ocular myasthenia demonstrating minimal ptosis of right upper lid **(A)** that increases after sustained gaze **(B).** (From Good WV, Hoyt CS: Strabismus management, St. Louis, 1995, Butterworth-Heinemann.)

mined by the muscle/s affected. Conditions presenting with ptosis should also be considered.

- *Binocular vision or accommodative dysfunctions:* These typically improve with vision therapy and are not associated with ptosis or other lid findings (Signs); there are no restrictions of ocular motility; strabismus, if present, is concomitant.
- *Thyroid-related ophthalmopathy (TRO) (p. 88):* Lid retraction, proptosis, and local inflammation are common; results of forced duction testing are positive; there is no associated ptosis. MG patients have a high risk for TRO.
- *Chronic progressive external ophthalmoplegia:* Slowly progressive ptosis and limitation in eye movements begin in young

adulthood. The condition, which may be inherited, is painless, sometimes asymmetric, with constant symptoms. Diplopia is rare, pupils are normal. One may raise the eyebrows and tilt the head back in order to see under lids. This condition is often associated with cardiac, neurological, and retinal pigment epithelial abnormalities. Results of Tensilon test are negative; muscle histology shows ragged red fibers.[4]

- *Double elevator palsy:* Unilateral restriction of upgaze is present with ipsilateral ptosis.
- *Internuclear ophthalmoplegia/One and a half syndrome (p. 114):* Adduction of the affected eye is limited; contralateral eye shows horizontal nystagmus in abduction[5]; ptosis is absent. Symptoms are not worsened by fatigue.
- *Möbius syndrome:* Abduction limitation with associated facial weakness. Symptoms are typically bilateral and constant.
- *Ocular motor nerve palsy (p. 105):* Variable presentations, depending on the aspect of nerve affected. This condition may have pupil involvement. There is no orbicularis weakness or association with fatigue.
- *Superior oblique palsy (p. 93):* Limited depression in the affected eye is greatest when the eye is adducted. Vertical deviation is nonconcomitant. Three-step test *(p. 168)* identifies superior oblique.
- *Congenital fibrosis of extraocular muscles:* Ptosis and chin elevation are associated with severe limitation of eye movements. Symptoms are typically constant.

WORK-UP

1. *History:* Onset? Are symptoms worse at end of day or after exertion? Variability in presence and severity of symptoms? Recent emotional stress or infection (may trigger onset of symptoms)? Medication? (D-Penicillamine and aminoglycosides produce pseudo-MG and worsen symptoms, respectively.) Other new systemic problems (Box 9-1)? Family or personal history of thyroid disease or other autoimmune disease?
2. *Externals:* Is there a head tilt to compensate for ptosis or strabismus?
3. *Ptosis:* Measure palpebral aperture and distance from upper lid to pupil margin; document variations throughout the exam; reassess after having the patient maintain upgaze for

1 minute or having the patient repeatedly open and close eyes (both should increase the amount of ptosis).

4. *Cogan's lid twitch sign:* Instruct the patient to look down for 10 to 20 seconds and then to look straight ahead; upper eyelid will overshoot then slowly return to resting position (may also twitch several times before becoming stable).
5. *Orbicularis weakness:* Instruct the patient to squeeze eyelids tightly closed. Weakness is present if the examiner can open the lids using fingers.
6. *Versions and ductions:* Restrictions of any muscle may be present; observe the patient's ability to hold fixation in extreme gaze (may be unable to maintain extreme gaze).
7. *Cover test/Maddox rod (pp. 152-153):* Assess deviation in all 9 positions; look for subtle, intermittent, and nonconcomitant deviations; measurements vary with time of day and level of fatigue.
8. *Pupils:* clinically normal in cases of MG.
9. *Vergence/Accommodative testing:*
   - *Accommodation:* Amplitude and accuracy may appear normal on initial testing. Facility shows fatigue with repeated effort.
   - *Vergence:* Phoria and vergence findings may progressively worsen as the day progresses.
10. Perform slit-lamp assessment of lagophthalmos and corneal exposure.
11. *Sleep/Rest test:* Evaluate the amount of ptosis and ocular motility restriction before and immediately after 30 minutes of rest with eyes closed: ptosis and/or ocular motility will temporarily improve in cases of MG.
12. *Ice test:* Evaluate ptosis of one eye (second eye used as control) before and after 2 minute application of ice to the affected eyelid: a transient 2-mm improvement in ptosis occurs in cases of MG 90% of the time; it may not affect ocular motility.[6]

ADDITIONAL TESTING

1. *Tensilon (edrophonium chloride) testing:* IV injection results in improvement in ptosis and ophthalmoplegia for approximately 2 minutes; the examiner must objectively quantify changes, because false negatives may occur.
2. *Prostigmin (neostigmine bromide) test:* Used if Tensilon test is negative and in children.[1]

3. An antiacetylcholine receptor antibody assay should be performed. (This antibody is not always present in individuals with only ocular symptoms.)
4. Magnetic resonance imaging or computed tomography should be performed to rule out the presence of an underlying intracranial lesion.
5. *Repetitive nerve stimulation and single fiber electromyography:* Amplitude of action potential generated by muscle decreases over time.
6. *Chest x-ray or computed tomography:* Rule out hyperplasia or tumor of the thymus gland.
7. *Thyroid function studies:* T_4, T_3, TSH.

TREATMENT

- Refer patient immediately to neurologist/neuroophthalmologist to confirm diagnosis and for evaluation of systemic features. Medical treatments include anticholinesterase drugs (pyridostigmine [Mestinon]), immunosuppressants, thymectomy, and plasmapheresis.
- *Ptosis:* If severe, tape lid or use ptosis crutch. Surgery should be performed only when the condition has been stable for 3 to 4 years.
- *Diplopia:* Fresnel prisms or occlusion may be used to eliminate variable diplopia (adhesive or tie-on patches, opaque contact lenses, or frosted lenses may be prescribed); a ground-in prism may be used if the deviation is stable; strabismus surgery should be performed only if the deviation has been stable for over 12 months.
- *Exposure keratitis due to lagophthalmos:* Preservative-free artificial tears during the day and lubricating ointment at night; if the condition is severe, lid taping.

FOLLOW-UP

- *Diplopia treated with prism or occlusion:* Follow up every 8 to 12 weeks.
- *Exposure:* If the condition is mild, follow up every 1 to 2 months; if corneal ulceration has occurred, follow up daily until resolved.

- If the patient experiences difficulty breathing or swallowing, immediate medical attention is needed.

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10

Amblyopia

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GENERAL INFORMATION

Amblyopia can be defined as reduced vision not reparable solely by optical correction, and not caused by ocular anomalies or pathologies of a developmental or acquired nature. Amblyopia is a diagnosis of exclusion *and inclusion*. In addition to ruling out all pathological causes for the decreased vision, one must find evidence of a specific amblyogenic cause (form deprivation, strabismus, refractive error). The earlier the amblyogenic event occurs and the longer it is present prior to treatment, the greater the likelihood that the amblyopia will develop.

Onset occurs invariably prior to the sixth year of life and most often becomes manifest as a unilateral condition, but it can be bilateral. The bilateral form is typically milder and easier to treat.

TYPES OF AMBLYOPIA

- *Deprivation amblyopia:* This type is caused by congenital or very early acquired degradation of visual stimuli (most commonly congenital cataracts, but sometimes complete ptosis or corneal opacification) in one or both eyes resulting in disruption of normal visual development. Deprivation amblyopia is considered the most severe and rarest form of amblyopia.
- *Strabismic amblyopia:* This type is caused by constant unilateral strabismus. It results more commonly from esotropia than from exotropia because of the likely intermittent nature of exotropia early in life; due to the great likelihood of sensory

adaptations, strabismic amblyopia is considered more difficult to treat than refractive amblyopia.

- *Refractive amblyopia:* Caused by significant and prolonged optical blur in one or both eyes (Table 10.1-1), this type of amblyopia often develops later than other types of amblyopia, thus allowing for at least some period of relatively normal visual development. Consequently, the depth of amblyopia is shallower and more amenable to treatment at a later age.

SYMPTOMS

Virtually no symptoms are present in young children. Older patients may report or parents may observe behavior suggestive of visual difficulties (squinting, eye rubbing, eye closure, head turning, holding objects in front of one eye only). A history of ocular surgery or of eye turning or patching may be present.

SIGNS

- *Decreased vision that does not respond to optical correction:* The magnitude of the vision loss depends on the category, severity, and duration of the amblyogenic insult. Typically, acuity falls within the range of 20/40 to 20/100, but it may be worse in the strabismic and form deprivation varieties. Amblyopia rarely if ever leaves an eye with only light perception.
- *Congenital or very early acquired media opacity, constant unilateral strabismus, or significant refractive error:* In amblyopia these symptoms must be present before the age of 6.
- Single-letter acuity may be better than whole-chart acuity (also known as crowding phenomenon).
- *Ambiguous endpoint during visual acuity testing:* The patient may get several letters wrong over a wide range of acuity levels.
- Stereopsis is diminished or absent.

TABLE 10.1-1.
Guidelines for Potentially Amblyogenic Refractive Error[6]

Ametropia	Anisometropia	Isoametropia
Hyperopia	Greater than 1.00 PD	Greater than 5.00 PD
Myopia	Greater than 3.00 PD	Greater than 8.00 PD
Astigmatism	Greater than 1.50 PD	Greater than 2.50 PD

ADDITIONAL SIGNS

- Contrast sensitivity is decreased, especially at high spatial frequencies.
- Amplitude of accommodation in the amblyopic eye may be reduced.
- Some reports[1-2] suggest that an afferent pupillary defect or overall depression of pupillary response may be present in the amblyopic eye. If so, it is typically a subtle defect (less than 0.6 log units). Large defects should raise suspicions about a neurological cause for the vision loss.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis for amblyopia includes any ocular condition that may result in decreased acuity.

The presence in a child under the age of 6 months of a media opacity due to a cataract, a corneal or vitreal opacity, or ptosis strongly suggests deprivation amblyopia. A unilateral constant strabismus suggests strabismic amblyopia. Significant refractive error (typically hyperopia, or against the rule or oblique astigmatism) suggests refractive amblyopia. A thorough internal and external health evaluation and correction of any significant refractive error is essential before the final diagnosis can be made. Thereafter, the vision should not continue to decrease. Any subsequent decrease in vision is an indication that amblyopia is not the sole cause of the reduced vision and that the case necessitates a further work-up.

WORK-UP

The major elements in the work-up of suspected amblyopia include history, visual acuity, binocularity, ocular motility, refraction, and assessment of ocular health. The age at examination makes modifications of standard examination techniques often necessary.

1. *History:* If strabismus is present, when was the onset and how long has it persisted? Has there been previous treatment? If the previous treatment was for amblyopia, at what age did the patient receive it, was it appropriate, and was there compliance? History of visual deprivation (cataracts, ptosis)? Surgery? Family history of lazy eye or strabismus?

2. *Measurement of visual acuity:* Amblyopes perform better when isolated letters are used instead of a full chart. The use of isolated letters only may lead to an overestimation of acuity and decrease the chances of detecting amblyopia. Many pediatric acuity charts use "crowded letters" to improve the likelihood of detecting decreased vision secondary to amblyopia. A large difference between a patient's whole-chart and isolated-letter acuity is a good prognostic indicator that the amblyopia will improve during treatment, with the isolated-letter acuity being the end goal of treatment.
   - *Infants:* Preferential Looking method, such as Teller acuity cards. Note that Teller acuity cards are a resolution task and may overestimate visual acuity in strabismic amblyopes.
   - *Preschool-aged children:* Lea symbols, HOTV, or Broken Wheel cards.
   - *School-aged children:* Snellen chart or Log MAR.
   - *For all ages:* A behavioral assessment of acuity can be obtained by observing the patient's monocular/binocular fixation pattern (p. 147).
3. *Assessment of deviation:* Compare magnitude at distance versus near; measure laterality, concomitancy, and frequency:
   - Infants and toddlers: *Cover test, Hirshberg and Brüchner tests (p. 150), versions.*
   - *Older children and adults:* Cover test, Hirshberg, modified Thorington (p. 153).
   - If unilateral vision loss is present, care must be taken to use a fixation target that can be seen by the affected eye. If the patient's vision loss is severe, a cover (light as target), Brüchner, Hirshberg, or Krimsky test (p. 154) may be used.
4. *Cycloplegic refraction (p. 179):* The recommended administration is 1 to 2 gtt of 1% cyclopentolate hydrochloride separated by 5 minutes. 1 gtt of 1% cyclopentolate can be used in infants under 1 year of age. Onset of maximum effect occurs approximately 30 minutes after instillation. The duration of action is approximately 6 to 12 hours.
5. Assess stereopsis:
   - Stereo smile for infants.
   - Preschool random-dot stereogram or random-dot E test for preschool children.

6. *Fixation status:* Visuscopy (p. 174) is an easy test for the presence of eccentric fixation, which may explain decreased vision and lead to a more accurate measurement of strabismus.
7. A complete external and internal ocular health examination, including dilated fundus evaluation, is mandatory.

TREATMENT AND FOLLOW-UP

Comprehensive care of the child with amblyopia requires communication with pediatricians, educators, and other professionals associated with the child and the family. The child with deprivation amblyopia should be expeditiously referred to a pediatric ophthalmologist. In all cases of amblyopia, the earlier the treatment is initiated, the greater the likelihood of success, but advanced age alone is not a reason to withhold treatment.[3-4] Special considerations (e.g., diplopia) are required when treating adults with strabismic amblyopia, and referral to a specialist is recommended.

Treatment of amblyopia consists of several components.

1. *Proper refractive correction:* Eliminating optical blur and providing an optimal environment for amblyopia therapy are essential. It is useful to give the patient a short period of time (4 to 6 weeks) with proper optical correction alone before the initiation of occlusion or other amblyopia therapy. Polycarbonate lenses are mandatory.
   - *Anisometropia:* The full amount must be corrected. It is acceptable to decrease the overall amount of the hyperopia to increase compliance, but the anisometropic difference between the two eyes must always be maintained: for example, if the results of cycloplegic refraction are OD +5.00 OS +1.00, prescribe OD +4.00 OS plano.
   - *Astigmatism:* The full amount needs to be corrected. A partial astigmatic correction will lead to a residual blur on the retina and will be an obstacle to successful treatment.
   - *Hyperopia and esotropia:* Prescribe the appropriate amount of hyperopia needed to eliminate the strabismus if there is an accommodative component to the esotropia (p. 32). Correction of the strabismus through optical means, if possible, takes precedence over considerations of any initial reduction in distance acuity secondary to the glasses.

2. *Occlusion:* Occlusion of the dominant eye to force use of the amblyopic eye has long been the primary means of treating amblyopia. Part-time patching (4 to 6 hours per day) in conjunction with visual motor activities (mazes, video games, coloring, etc.) is typically sufficient treatment for the majority of amblyopia cases.
   - *Adhesive patches:* Several brands exist, including Coverlet, Opticlude, Ortopad, and Nextcare. These patches may cause skin irritation in some children. Use of warm compresses before removing the patch may reduce irritation. Certain brands such as Ortopad and Nextcare may be less likely to cause irritation and are preferred for part-time patching.
   - *Pharmacological penalization:* Works well in patients that are noncompliant with adhesive patches. Recent studies have shown that use of 1 gtt of 1% atropine ophthalmic solution instilled daily is as effective as traditional patching with minimal side effects.[5] Penalization is most effective in the presence of hyperopia in the nonamblyopic eye to provide sufficient blur to force the child to use the amblyopic eye at near. Based on near point blur, patients are likely to switch fixation if the acuity in the amblyopic eye is at least 20/100.

VISIT SEQUENCE AND FOLLOW-UP

1. *Initial visit:* Correct significant refractive error; perform follow-up in 4 to 6 weeks.
2. *Second visit:* Ensure compliance with prescription wear; assess visual acuity, stereopsis, overrefraction, and ocular health. If a difference in acuity exists between the two eyes, then begin patching.
   - *Adhesive patch:* Instruct patients to wear the patch 4 to 6 hours a day and/or after school. Educate them about the importance of remaining in a safe environment while wearing the patch. Perform follow-up in 1 month.

OR

   - *Atropine:* Patients should begin by instilling 1 gtt of 1% atropine ophthalmic solution each morning in the good eye. Hat and sunglasses should be worn outdoors. Have patients return in 1 week to ensure that a fixation switch has

occurred. This can be done by measuring the visual acuity of both eyes at distance and near. If the vision is at least two lines better in the amblyopic eye for either distance, then it is likely that the patient is using that eye for certain tasks. Perform follow-up in 1 month.

3. *Subsequent visits:* Ensure compliance with the patching; assess visual acuity, stereopsis, overrefraction, and ocular health. Monitor patients monthly until acuity shows improvement. Once improvement has been noted, monitor patients every 2 to 3 months. If acuity stabilizes for 2 to 3 visits, stop occlusion and follow-up 3 months to monitor for regression.
4. Monitor patients under 6 years of age carefully for the occurrence of reverse amblyopia. This is the development of amblyopia in the dominant eye secondary to patching. The younger the patient, the greater is the risk of occurrence, and the more frequent should be the follow-up visits.

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11

Diagnostic Techniques

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11.1 Monocular/Binocular Fixation Pattern (Fix, Follow, and Maintain)

The Monocular/Binocular Fixation Pattern procedure is considered a default method of indirectly measuring acuity or testing for the presence of amblyopia in infants, toddlers, and difficult-to-test

children. Most commonly used in cases of strabismic amblyopia, this procedure has been the subject of debate in the literature.[1-4] It is important to note that an observable fixation preference may occur in the presence of strabismus with or without amblyopia.[1]

PROCEDURE

- The examiner is seated across from patient in a fully lit room.
- An appropriate and high-interest accommodative target is used, such as small toy or sticker.

Monocular Fixation Pattern (Fix and Follow)

1. Occlude the patient's eye with a hand or thumb, while moving the toy (2 cm per second) into cardinal positions of gaze.
2. Observe the ability of the patient to fixate and accurately follow the object as it moves.
3. Repeat the procedure while covering the other eye.

Binocular Fixation Pattern (Maintain)

Standard fixation preference testing (SFPT). The following procedure is performed on patients with strabismus greater than 10 prism diopters (PD):

1. Hold the toy approximately 40 cm in front of the patient, with both of the patient's eyes open.
2. Wait and watch for the patient spontaneously to switch fixation between the two eyes.
3. If spontaneous alternation does not occur, occlude the preferred eye with a hand or thumb to force the nonpreferred eye to fixate on the target.
4. Remove the occlusion and note whether the nonpreferred eye is able to maintain fusion.

10-PD Fixation test[2]. The following procedure is performed on nonstrabismic patients, or on patients with strabismus of less than 10 PD:

1. The procedure is identical to that of SFPT, except the examiner holds a 10-PD loose prism in front of one eye either base-up or base-down, creating diplopia.
2. Note whether the patient alternates fixation between the two eyes.
3. If a fixation preference is noted, switch prism to the other eye.

INTEPRETATION

Monocular Fixation Pattern

- *Normal response:* The patient exhibits an equal ability to fix and accurately follow the object into all positions with no preference during forced occlusion. (The patient does not object when one eye is occluded.)
- Nystagmus, differences in quality of movement, or objection to covering one eye are considered abnormal findings.
- If the patient objects to the occlusion of only one eye, repeat the procedure to ensure that the objection is not a result of fatigue during testing. A negative response to occlusion of one eye only may indicate poor vision in the eye not occluded.
- A patient who objects to the occlusion of both eyes is considered untestable.

Binocular Fixation Pattern (SFPT and 10-PD Test)

- *Normal response:* The patient alternates fixation between the two eyes or has a very slight fixation preference for the dominant eye.
- Anything greater than a slight fixation preference is an abnormal response and may signify unequal acuity or the presence of amblyopia. Fixation preferences can be graded as follows[1-3]:
  (0) The nonpreferred eye immediately loses fixation when the preferred eye is uncovered.
  (1) The nonpreferred eye holds fixation for 1 to 2 seconds but loses fixation before the blink.
  (2) The nonpreferred eye holds fixation up to the blink.
  (3) The nonpreferred eye is able to hold fixation through the blink.
  (4) The patient freely alternates fixation, with no fixation preference demonstrated.

RECORD

- *FFM OD = OS:* The patient demonstrates an equal ability to fixate and follow monocularly with no preference to forced occlusion: Grade 4 fixation preference binocular.
- If an abnormality is detected, then describe the observation (e.g., patient fixes and follows with OD only, objects to occlusion of OD, Grade 1 fixation preference binocular).

REFERENCES

1. Kipf R: Binocular fixation pattern, Arch Ophthalmol 94:401-405, 1976.
2. Wright KW and others: Reliability of fixation preference testing in diagnosing amblyopia, Arch Ophthalmol 104:549-53, 1986.
3. Sener EC, et al: The reliability of grading the fixation preference test for the assessment of interocular visual acuity differences in patients with strabismus, J AAPOS 6:191-194, 2002.
4. Atilla H, et al: Poor correlation between "fix-follow-maintain" monocular/binocular fixation pattern evaluation and presence of functional amblyopia, Bin Vis Strab Qrtly 16:85-90, 2001.

11.2 Brückner Test

The Brückner test is easy to administer on patients of all ages and provides information concerning the presence of strabismus, refractive error (anisometropia), media opacities, and pupil size. Although it is not necessarily the most sensitive test for determining these conditions, its ease of administration and versatility make it a useful clinical tool.[1]

PROCEDURE

1. Seat the patient in a dimmed room.
2. Sit approximately 80 to 100 cm from the patient.
3. Shine the direct ophthalmoscope toward the patient's eyes to illuminate both eyes simultaneously.
4. Look through the ophthalmoscope and adjust the lenses for the working distance and your own refractive error. (The fundus reflex should be in focus.)
5. Observe the following:
   - Relative color and brightness of the fundus reflexes in comparison with one another.
   - Relative pupil size with the ophthalmoscope rheostat first dimmed and then set to the maximum brightness.
   - Corneal reflexes (Hirschberg test).

INTERPRETATION

A brighter reflex in one eye compared to the other may indicate one of the following:

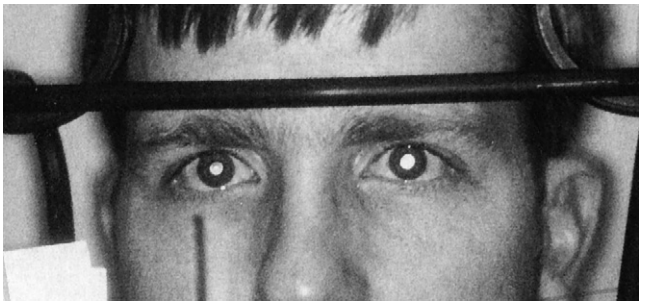


FIG. 11-1 Brighter fundus reflex in the left eye signifies a small angle left esotropia during the Brückner test. (From Griffin JR, Grisham JD: Binocular anomalies: diagnosis & vision therapy, 4 ed, 2002, Butterworth-Heinemann.)

- Strabismus in the brighter eye (Fig. 11-1)
- High refractive error in the dimmer eye
- Media opacity in the dimmer eye

CONSIDERATIONS

- It is important to consider the above three possibilities when a difference in brightness is noted. Refractive error can be accounted for by performing retinoscopy; media opacities have a very characteristic appearance and can be confirmed with a slit-lamp exam; strabismus can be confirmed with a cover test.
- Additional insight into the presence of strabismus can be obtained by observation of the corneal reflexes created by the ophthalmoscope light during this test. The results of this observation are interpreted in a manner identical to that for interpreting the Hirschberg test.
- Relative pupil size in both dim and bright light can be assessed simultaneously by observation of the fundus reflexes during adjustment of the rheostat.

REFERENCE

1. Griffin JR, Cotter SA: The Bruckner test: evaluation of clinical usefulness, Am J Optom Physiol Optics 63:957, 1986.

11.3 Maddox Rod With Prism

The Maddox rod can be used to assess the magnitude, direction, and concomitancy of either a phoria or a tropia, but it cannot differentiate between the two. The rod is used in combination with a penlight so that the eyes are dissociated, with one eye viewing the light and the other a red line. Crossed or uncrossed diplopia will be created and can be measured through the application of a prism bar.

PROCEDURE

1. Hold the Maddox rod over the right eye with rods oriented horizontally to test the horizontal deviation, vertically to test the vertical deviation.
2. Hold the light source (penlight) at 40 cm for near testing.
3. The patient will see both a light and a red line. Ask the patient to look (with both eyes open) at the light and to make a judgment about the location of the line in relation to the light.

INTERPRETATION OF HORIZONTAL MEASUREMENT

- *The patient reports line going through the light:* orthophoria.
- *The patient reports line to the left of the light (crossed diplopia):* Exodeviation. Using a prism bar, add base-in prism to OD until the line is centered with respect to the light.
- *The patient reports line to the right of the light (uncrossed diplopia):* Esodeviation. Using a prism bar, add base-out prism to OD until the line is centered with respect to the light.
- This procedure can be repeated in all positions of gaze to measure concomitancy. A change of 5 PD or more in any gaze is considered significant.

INTERPRETATION OF VERTICAL MEASUREMENT

- *The patient reports line going through the light:* Orthophoria.
- *Patient reports line above the light:* Right hypodeviation (or left hyperdeviation). Add base-up prism to the right eye until the patient sees the line go through the light.
- *The patient reports the line below the light:* Right hyperdeviation (or left hypodeviation). Add base-down prism to the right eye until the patient sees the line go through the light.

- This procedure can be repeated in all positions of gaze and with head tilts to assess concomitancy and to isolate a potentially paretic muscle. A change of 5 PD or more in any gaze is considered significant.

CONSIDERATIONS

The line produced from the Maddox rod may fluctuate slightly, so it is best for patients to wait a couple of seconds for the line to stabilize. Urge patients to concentrate on looking at the light with their left eye and then to make a judgment about the placement of the line.

11.4 Maddox Rod With Scale (Modified Thorington)

The Maddox rod can be used to assess the magnitude, direction, and concomitancy of either a phoria or a tropia, but the rod cannot differentiate between the two. The Maddox rod used in combination with a penlight dissociates the two eyes such that one eye is viewing the light and the other a red line. Crossed or uncrossed diplopia will be created and can be measured through the application of a modified scale or Thorington card. This test is simple to perform and has been shown to be more reliable than other methods of phoria measurement.[1] It is also readily adaptable to measuring concomitancy and the AC/A ratio.

PROCEDURE AND INTERPRETATION

1. Hold the Maddox rod over the right eye, with rods oriented horizontally to test the horizontal deviation, vertically to test the vertical deviation.
2. Hold the Thorington card or scale at 40 cm while shining a light through the center hole.
3. The patient will see both a light and a red line. Ask the patient to look (with both eyes open) at the light and to make a judgment about the location of the line in relation to the light.
4. Simply ask the patient to read the point on the scale at which the line falls.

5. The horizontal measure may be repeated through +1.00 diopter (D) lenses OU to assess the AC/A ratio.
6. Concomitancy of either the horizontal or the vertical phoria can be assessed by simply moving the card into different positions of gaze. A change of 5 PD or more in any gaze is considered significant.

CONSIDERATIONS

If the line is shifting, have the patient wait several seconds before responding. It is sometimes helpful to instruct patients to concentrate on looking at the light with their left eye and then to observe where the line falls on the scale. Ensure that they are keeping letters/numbers clear on the card at all times.

REFERENCE

1. Rainey BB and others: Inter-examiner repeatability of heterophoria test, Optom Vis Sci 75:719-726, 1998.

11.5 Hirschberg/Krimsky Test

First described in the 1940's,[1] the Hirschberg/Krimsky test is a practical but relatively gross method of measuring strabismus. Prism is used to "neutralize" the appearance of the corneal reflex during Hirschberg testing. The use of a light as a target makes it especially useful when a patient has a significant reduction in monocular visual acuity. The test is most useful in large angle and sensory strabismus.

PROCEDURE

Hirschberg Test

1. Sit across from the patient at eye level and shine a focal light source towards the patient's eyes. (The testing distance is greater than 50 cm.)
2. Hold the penlight below the preferred eye and close the other eye.
3. Direct the penlight toward the bridge of the patient's nose and instruct the patient to look at the light.

4. Occlude the patient's left eye and observe corneal reflex. The reflex may appear in the following positions:
   - Nasal to pupil center (positive angle lambda)
   - Directly in the center of the pupil (zero angle lambda)
   - Temporal to center of the pupil (negative angle lambda)
5. Repeat while occluding the patient's right eye.
6. Repeat while both of the patient's eyes are open and compare corneal reflexes to one another.

Krimsky Test

7. If a difference between corneal reflexes is detected, select a prism from the loose prism set or prism bar, using the approximated magnitude from the Hirschberg test.
8. Interpose the prism before the *fixating* eye. The prism will cause versional eye movement, shifting the fixating and the nonfixating eyes:
   - Base-out to neutralize esotropia
   - Base-in to neutralize exotropia
9. Increase or decrease the prism until the corneal reflex of the nonfixating eye matches the appearance of the fixating eye (without prism).

INTERPRETATION

1. The normal appearance of the corneal reflex is slightly nasal to pupil center.
2. 1 mm of displacement = 22 PD.
3. Under monocular conditions, if the corneal reflex of one eye differs from that of the other eye, this may indicate eccentric fixation:
   - *Corneal reflex displaced nasally:* Temporal eccentric fixation
   - *Corneal reflex displaced temporally:* Nasal eccentric fixation
   - *Corneal reflex displaced superior:* Inferior eccentric fixation
   - *Corneal reflex displaced inferior:* Superior eccentric fixation
4. Under binocular conditions if the corneal reflexes of the two eyes differ, strabismus is indicated.
   - *Corneal reflex displaced nasally:* Exotropia
   - *Corneal reflex displaced temporally:* Esotropia
   - *Corneal reflex displaced superior:* Hypotropia
   - *Corneal reflex displaced inferior:* Hypertropia

CONSIDERATIONS

Infants may have an increased angle kappa, which may mimic an exotropia if it is not accounted for.

REFERENCE

1. Krimsky E: The management of binocular imbalances, Philadelphia, 1948, Lea and Febiger.

11.6 Step Vergence Testing

Performed in free space, this test may be easier to use with children or nonresponsive adults by incorporating components both objective (examiner watches eye movements) and subjective (patient reports diplopia).

PROCEDURE

1. Instruct the patient to fixate on the appropriate target.
   - *For distance vergence ranges:* Target should be an isolated letter 2 lines above the best corrected acuity at 20 feet.
   - *For near vergence ranges:* Target should be an isolated letter 2 lines above best corrected acuity at 40 cm. For young children, a small (2 to 3 cm), detailed sticker can be substituted to increase attention.
2. Hold the prism bar over the right eye.
   - *Positive fusional vergences:* Hold the prism bar with the apex of the prism pointing toward the nose.
   - *Negative fusional vergences:* Hold the prism bar with the apex of the prism pointing toward patient's right ear.
3. Instruct the patient as follows:
   - "Look at the target at all times."
   - "I am going to put different lenses in front of your eye and you need to tell me if the target becomes blurry or double, or if it moves in one direction."
4. Starting with the 1 PD increment, slowly increase the amount of prism (approximately 1 step per second).
5. Increase the prism until the patient reports that the target is blurry. Make a mental note of this number.
6. Continue to increase the prism until the patient reports that the target is double and stays double. Make a mental note of this number.

7. Increase the prism an additional step and say, "Now tell me when the target becomes single again."
8. Decrease the prism until the patient recovers single vision. Make a mental note of this number.

INTERPRETATION

- Record the blur/break/recovery point (e.g., 12/16/10). If a blur point is not reported, substitute an "x" for the blur point value.
- Normative values at near are as follows:
  - *Children (ages 7 to 12):* BI x/12/7 BO x/23/16
  - *Adults:* BI x/13/10 BO x/19/14.

  For a more complete listing of normative values refer to other sources.[1-3]
- Compare the compensating vergence to the phoria finding (i.e., positive fusional vergence to exophoria, negative fusional vergence to esophoria) using Sheard's criterion. (Compensating vergence should be at least twice the phoria.)

CONSIDERATIONS

- If both positive and negative fusional vergences are to be measured, it is best to measure the negative ranges before the positive.
- The 25 PD increment can be briefly interposed before the patient's right eye to illustrate diplopia before testing begins.
- It is common for patients not to report blur during this test.
- In addition to having the patient report points of blur, break, and recovery, monitor eye movements when each PD step is made. The right eye and left eye should shift in the direction of the prism apex, followed by the left eye's making a subsequent fusional movement back toward the base. The "break point" can be recognized easily by the lack of fusional movement on the part of the left eye. Additionally, if patients are experiencing diplopia, they may make a quick saccadic movement between the true image and the diplopic image. This fact is especially important when one is testing children, who may give unreliable responses.

REFERENCES

1. Wesson MD: Normalization of prism bar vergences, Am J Optom Physiol Opt 59:628-633, 1982.

2. Scheiman M and others: A normative study of step vergence in elementary schoolchildren, J Am Optom Assoc 60:276-280, 1989.
3. Scheiman M, Wick B: Clinical management of binocular vision, Philadelphia, 2002, Lippincott Williams and Wilkins.

11.7 Vergence Facility

Vergence facility is a valuable but often overlooked diagnostic tool. It is the common and perhaps the only clinical measure of sustained and dynamic vergence ability, and without it the diagnosis of certain types of vergence dysfunctions cannot be made. Rather than assessing the range of convergence and divergence, it investigates the patient's ability to respond rapidly to alternating convergence and divergence demands and to sustain that response over an extended period of time.[1] Until recently the procedure and normative values were not well defined. However, a series of studies conducted by Gall, et al., have shed new light on this important clinical tool, offering guidance in its use and evaluation.[2,3]

PROCEDURE

1. The patient is seated comfortably while viewing an isolated vertical row of 20/30 letters held at 40 cm.
2. Instruct the patient that you will be introducing different lenses before his or her eyes and that the goal is to make the target "single and clear."
3. Instruct the patient to inform you as soon as the target becomes "single and clear" either by tapping on the table or by using a hand signal.
4. It may be helpful to demonstrate to the patient the following:
   - *Diplopia:* Introduce a large magnitude prism before the eye.
   - *Suppression:* Introduce a large magnitude prism before the eye in various orientations while one of the patient's eyes is occluded. Draw attention to the way in which the image shifts in space.
   - The difference between blur and the distortions induced by looking through a prism, demonstrated by alternating between a high plus lens and the 12-base-out prism.
5. Begin by using the 12-base-out prism and flip to the 3-base-in prism as soon as the patient indicates "clear and single" vision.

6. Continue the process for 60 seconds, counting the number of cycles that the patient is able to complete (cpm). A "clear and single" response through both the 12-base-out and the 3-base-in prism is considered one cycle.
7. Although this test can be repeated at distance, its main clinical application may be better served at near.[2]

INTERPRETATION

The normative value suggested by Gall, et al., is 15 cpm.[2]

REFERENCES

1. Daum KM: Vergence infaciltiy. In Eskeridge JB, Amos JF, Bartlett JD, editors: Clinical procedures in optometry, Philadelphia, 1991, Lippincott.
2. Gall R, Wick B, Bedell H: Vergence facility: establishing clinical utility, Optom Vis Sci 75:731-742, 1998.
3. Gall R, Wick B, Bedell H: Vergence facility and target type, Optom Vis Sci 75:727-730, 1998.

11.8 Monocular Estimation Method

The Monocular Estimation Method (MEM) is a reliable and simple way of assessing the accuracy of the accommodative system. It is an objective test that can be performed on patients of all ages.

PROCEDURE

1. The patient is seated out of the phoropter, wearing appropriate spectacle correction.
2. An age-appropriate MEM card should be positioned on the retinoscope. If an MEM card is not available, 20/30- to 20/40-sized letters or figures of a similar size may be substituted.
3. The room illumination should be bright enough so that the patient can clearly see the intended target.
4. Hold retinoscope and card at a distance of 40 cm from the patient.
5. Ask the patient to fixate on the letters on the card while you sweep the horizontal meridian of the eye. (Significant astigmatism should not be present if the appropriate near prescription is being worn.)

6. Estimate the dioptral value of the reflex and briefly interpose loose lenses to neutralize the reflex.
7. Repeat for the other eye.

INTERPRETATION

- The magnitude of the lens required to neutralize the reflex is the lag or lead of accommodation. Plus lenses indicate a lag of accommodation and minus lenses indicate a lead.
- The normal response is 0.33 +/–0.34 PD.[1-2]

CONSIDERATIONS

- Interpose lenses briefly during neutralization. If left in place too long, lenses may alter the accommodative stimulus and hence your measurement.
- Patients should be reminded to keep the target clear during testing. Increased accommodation may result if patients are asked to read the words, instead of their simply fixating on one word and keeping it clear.

REFERENCES

1. Rouse MW, Hutter RF, Shiftlett R: A normative study of the accommodative lag in elementary school children, Am J Optom Physiol Opt 61:693-7, 1984.
2. Tassinari JT: Monocular Estimate Method Retinoscopy: central tendency measures and relationship to refractive status and heterophoria, Optom Vis Sci 79:708-714, 2002.

11.9 Accommodative Facility

Accommodative facility testing evaluates the ability of the accommodative system to respond to a changing stimulus over time. Performed both binocularly and monocularly, this test is helpful in differentiating between primary accommodative problems and those secondary to a vergence dysfunction. Current research has suggested changes in the traditional way of employing this test: namely, the application of different normative values, lens powers, and test distances based on accommodative amplitude.[1-4] These changes have been recommended for testing patients over the age of 30.[5]

PROCEDURE (FOR PATIENTS UNDER THE AGE OF 30)

1. The patient is seated comfortably, wearing distance correction or a near correction (if warranted) while viewing an appropriate target.
   - *Adults:* The target should be a line of letters one or two levels above the patient's best corrected acuity.
   - *Children:* A modified target may be used in children with whom you do not want to rely solely on the subjective response of "clear." An accommodative rock card or similar target one or two levels above the patient's best corrected acuity is recommended. The patient is instructed to read the letters/words instead of simply responding "clear."
2. Begin testing binocularly with +2/–2 D flip lenses. It is important to check for suppression when performing this test OU,[6] especially with patients who have a large phoria or intermittent tropia. This may be done with a Polaroid bar reader and Polaroid glasses. The bar reader is simply placed over the target that you are using for testing, and the patient wears the glasses.
3. Instruct the patient that you are going to introduce different lenses in front of his or her eyes, and that the goal is to keep the print "clear and single."
   - Explain to the patient that "blurry" means no longer being able to discern the letters on the target.
   - By momentarily occluding either eye, demonstrate to the patient how the target may appear with one eye suppressed.
4. Holds the flip lenses in front of the patient, starting with the plus lenses. Ask the patient to indicate when the print is clear and single by saying "clear."
5. Switch between the plus and minus lenses as soon as the patient says "clear."
6. Repeat for one minute counting the cycles completed. One cycle is a "clear" response through both the plus and minus lenses.
7. Testing may be stopped if the patient is unable to clear either the plus or minus.

Monocular Testing (If Needed)

1. The procedure here is identical to that in the binocular portion, except that one eye is occluded and monitoring for suppression is not necessary.

2. Repeat the procedure with the left eye occluded and then with the right eye occluded.

PROCEDURE (FOR PATIENTS OVER THE AGE OF 30)

The procedure is identical to that described for patients under the age of the 30, except that lens selection and testing distance are determined according to Table 11.9-1.

TABLE 11.9-1
Amplitude Scaled Facility[3,4]

Amplitude of accommodation		Test set-up	
Diopters	Distance (cm)	Test distance (cm)	Flip lens power
22.5	4.5	10.0	±3.25
20.0	5.0	11.0	±3.0
18.25	5.5	12.0	±2.75
16.75	6.0	13.5	±2.5
15.5	6.5	14.5	±2.25
14.25	7.0	15.5	±2.5
13.25	7.5	16.5	±2.0
12.5	8.0	18.0	±2.0
11.75	8.5	19.0	±1.75
11.0	9.0	20.0	±1.75
10.5	9.5	21.0	±1.5
10.0	10.0	22.0	±1.5
9.5	10.5	23.5	±1.5
9.0	11.0	24.5	±1.5
8.75	11.5	25.5	±1.25
8.25	12.0	26.5	±1.25
8.0	12.5	28.0	±1.25
7.75	13.0	29.0	±1.25
7.5	13.5	30.0	±1.0
7.25	14.0	31.0	±1.0
7.0	14.5	32.0	±1.0
6.75	15.0	33.5	±1.0
6.5	15.5	34.0	±1.0
6.25	16.0	35.5	±1.0
6.0	16.5	37.0	±1.0
5.75	17.5	38.5	±1.0
5.5	18.0	40.5	±0.75
5.25	19.0	42.5	±0.75
5.0	20.0	44.5	±0.75
4.75	21.0	47.0	±0.75
4.5	22.0	49.5	±0.75

Scoring criteria: patients scoring less than 10 cycles per minute are likely to be symptomatic.

INTERPRETATION

- In many instances patients will be unable to clear plus, minus, or both, in which case "fails + or –" should be recorded.
- The examiner should note whether patients have more difficulty with plus or minus even if they are able to clear both and record their observations.
- Record the cycles per minute (cpm) and compare this number to the normative values. For a complete listing of the normative values for patients between the ages of 6 and 30, the reader is encouraged to consult other sources.[3,5] The following is a general guideline to be used when evaluating the number of cycles that should at the least be achieved.
  - *Patients under 13 years of age:* 8 cpm monocular, 6 cpm binocular.
  - *Patients over 13 years of age:* 10 cpm monocular or binocular.

CONSIDERATIONS

1. Although the matter is debated, it is the author's opinion that this test should be performed under binocular conditions first:
   - If the patient does well binocularly, monocular findings may be of little value.
   - If the patient has trouble binocularly, then monocular testing should most certainly be conducted.
2. *Target selection:* To obtain more reliable results, different targets may be used for children and adults[1:]
   - *Adults:* Printed material one or two lines above the patient's best correct acuity is recommended.
   - *Children:* Use an accommodative rock card one or two lines above the patient's best corrected acuity. The patient is asked to read off each word or letter instead of simply responding "clear."
3. The use of normative values is important, but equally important is monitoring the patient's behavior during testing for relative difficulty or inability to clear either the plus or minus lenses.

REFERENCES

1. Scheiman M and others: Normative study of accommodative facility in elementary school children, Am J Optom Physiol Opt 65:127-134, 1988.

2. Wick B and others: Clinical testing of accommodative facility: part I. A critical appraisal of the literature, Optometry 73:11-23, 2002.
3. Yothers TL, Wick B, Morse SE: Clinical testing of accommodative facility: part II. Development of an amplitude-scaled test, Optometry 73:91-102, 2002.
4. Wick B, Gall R, Yothers T: Clinical testing of accommodative facility: part III. Masked assessment of the relation between visual symptoms and binocular test results in school children and adults, Optometry 73:173-81, 2002.
5. Scheiman M, Wick B: Clinical management of binocular vision, ed 2, Philadelphia, 2002, Lippincott Williams and Williams.
6. Burge S: Suppression during binocular accommodative rock, Optometric Monthly 79:867-72, 1979.

11.10 NSUCO Ocular Motor Test (The Maples)

The NSUCO ocular motor test is a standardized observational method for assessing pursuits and saccades. Requiring no special equipment and easily administered, this test has been shown to be both repeatable and reliable.[1]

PROCEDURE

1. The patient is instructed to *stand* with feet shoulder-width apart and arms at his or her side.
2. The examiner is positioned directly across from the patient at eye level.
3. Two fixation targets are needed for testing. Targets should be of different colors and approximately 2 to 4 cm in size. For purpose of this explanation, one target will be red and the other white.
4. The patient is *not* given any instructions concerning head movements during this test.

Saccades

1. Targets are held 40 cm from the patient and separated by 20 cm.
2. The patient is instructed simply to do the following:
   - "Look at the white target when I say white and look at the red target when I say red."
   - "Do not look at the target until I call the color."

3. Calling out alternating target colors, have the patient complete 5 round trips for a total of 10 saccades.
4. Observe the accuracy, ability, head movements, and body movements of the patient (Table 11.10-1).

Pursuits

1. One target is held 40 cm from the patient.
2. The patient is instructed to watch the target as it is moved.
3. Move the target in two clockwise circles (20 cm diameter), two counterclockwise circles (20 cm diameter), and one sweep across the midline.
4. Observe the accuracy, ability, head movements, and body movements of the patient (Table 11.10-2).

INTERPRETATION

- The patient's performance is assessed for both pursuits and saccades according to four categories: accuracy, ability, head movements, and body movements.
- Record results according to the grading scale (Table 11.10-1 and 11.10-2).

TABLE 11.10-1
Saccade Testing: NSUCO Scoring Criteria

Scale	Ability	Accuracy	Head and body movement
1	Completes less than 2 round trips	Large under- or overshooting noted one or more times	Large movement of head/body at any time
2	Completes 2 round trips	Moderate under- or overshooting noted one or more times	Moderate movement of head/body at any time
3	Completes 3 round trips	Constant slight over- or undershooting noted more than 50% of the time	Consistent slight movement of head/body greater than 50% of the time
4	Completes 4 round trips	Intermittent slight over- or under-shooting noted less than 50% of the time	Intermittent slight movement of head/body less than 50% of time
5	Completes 5 round trips	No over or undershooting	No movement of head/body

TABLE 11.10-2
Pursuit Testing: NSUCO Scoring Criteria

Scale	Ability	Accuracy	Head and Body Movement
1	Cannot complete one half rotation in either direction	Refixations more than 10 times	Large movement of head/body at any time
2	Completes one half in either direction	Refixations 5 to 10 times	Moderate movement of head/body at any time
3	Completes one rotation, but not two rotations	Refixations 3 or 4 times	Consistent slight movement of head/body greater than 50% of time
4	Completes two rotations in one direction, but less than 2 in the other direction	Refixations two times or less	Intermittent slight movement of head/body less than 50% of the time
5	Completes two rotations in either direction	No refixations	No movement of head/body

- Results can be compared to normative values,[2] or repeated with the same patient to check for changes.

CONSIDERATIONS

- Note that the patient is asked to stand during this test and that no instructions concerning head or body movements are given.
- Eyedrop bottles (e.g., Tropicamide and Phenylephrine) are readily available, have two differently colored caps, and can be used as fixation targets for this test.

REFERENCES

1. Maples WC, Ficklin TW: Interrater and test-retest reliability of pursuits and saccades, J Am Optom Assoc 59:549-552, 1988.
2. Maples WC: NSUCO ocular motor test, Santa Ana, CA, 1995, Optometric Extension Program.

11.11 Developmental Eye Movement Test

The Developmental Eye Movement test (DEM) is the only visual-verbal test of ocular motor skills that takes into account the automaticity of the verbal response.[1] The purpose is to assess the eye movements associated with the task of reading. Although questions of reliability have surfaced,[2] the DEM remains a clinical tool that is easy to administer and interpret, and one that is well suited for a primary care clinical setting. A complete set of instructions, normative data, and examiners and test booklets are available through the Bernell Corporation.* The DEM is designed to be administered and normative data exists for children 6 to 12 years of age.

PROCEDURE[3]

A complete explanation of the test procedure is provided in the DEM Examiners Booklet. The following is a brief outline of the testing procedure intended to familiarize the reader with the test.

1. The patient is tested individually in a location without distraction.
2. Begin by opening the test booklet and having the patient read the pretest. The purpose of the pretest is to ensure that the patient is familiar with numbers. The pretest is not timed and is usually administered only to younger children.
3. Turn the booklet to Test A (vertical test) and instruct the patient to read the numbers down the two columns as fast as possible, using only his or her eyes. Time the patient's performance and follow along on a scoring sheet, noting any errors (omissions, additions, substitutions, and transpositions).
4. Turn the booklet to Test B (vertical test) and repeat step 3.
5. Turn the booklet to Test C (horizontal test) and instruct the patient to read carefully but as fast as possible the numbers across the rows. Time the patient's performance and follow along on the score sheet, noting any errors (omissions, additions, substitutions, and transpositions).

* The DEM may be obtained from the Bernell Corporation, 4016 North Home Street, Mishawaka, IN 46545, 800-348-2225.

INTERPRETATION

1. Scoring is done as follows:
   - Take into account the number and type of errors to obtain a score of adjusted time.
   - Scores for Test A and B are added to obtain a Vertical score.
   - The score for Test C is the Horizontal score.
   - A ratio equal to the Horizontal score/Vertical score is calculated.
   - All scores are compared to the age-adjusted normative values.
2. Scores are interpreted as follows:
   - *Normal performance:* Normal Horizontal, Vertical, and Ratio scores
   - *Ocular motor dysfunction:* Elevated Horizontal, normal Vertical, elevated Ratio
   - *Automaticity problem:* Elevated Horizontal, elevated Vertical, normal Ratio
   - *Ocular motor and automaticity problem:* Elevated Horizontal, elevated Vertical, elevated Ratio

REFERENCES

1. Garzia RP and others: A new visual-verbal saccade test: the developmental eye movement test (DEM), J Am Optom Assoc 61:124-135, 1990.
2. Rouse MW, Nestor EM, Parot CH: A re-evaluation of the reliability of the developmental eye movement test, Optom Vis Sci 61(Suppl):90, 1991.
3. Richman JE, Garzia RP: Developmental Eye Movement Test: examiners booklet, The Bernell Corporation.

11.12 Parks Three-Step Test

First described in 1958,[1] the Parks three-step test is a procedure designed to determine which of the eight cyclovertical muscles is paretic when a nonconcomitant vertical deviation exists. The three steps are as follows:

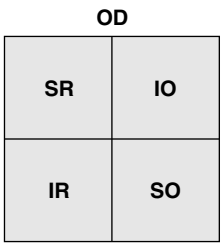
1. Determine whether the right eye is hyper or hypo in primary position.
2. Determine whether the vertical deviation increases in the right or left gaze.

3. Determine whether the vertical deviation increases on head tilt to the left or right (Bielschowsky head tilt test).

PROCEDURE AND INTERPRETATION

Record the results of the three steps on the form below. Upon completion there will be the following notation:

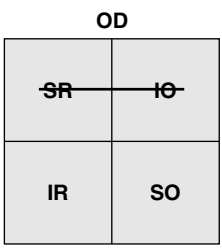
- A muscle crossed out three times, indicating a paretic muscle in the OD, or
- A muscle that has not been crossed out at all, indicating a paretic muscle OS



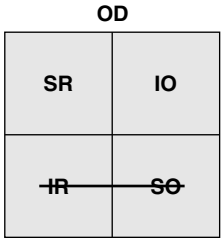
The diagram is drawn from the examiner's point of view looking at the patient. For the sake of simplicity, only the right eye is considered. This means that the examiner must convert any deviation in reference to the right eye. For example, a left hypo equals right hyper, and left hyper equals right hypo.

Step 1

Is the right eye hyper or hypo?



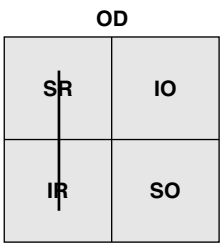
If right hypo, cross out elevators



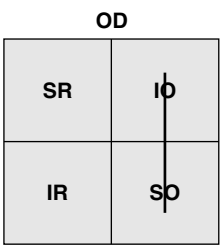
If right hyper, cross out depressors

Step 2

Does the deviation increase when the patient looks to the right or left?



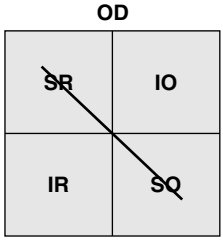
If deviation increases in right gaze



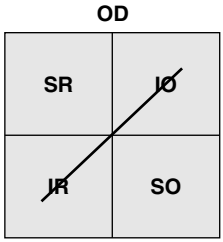
If deviation increases in left gaze

Step 3

Does the deviation increase when the patient tilts the head to the right or left?



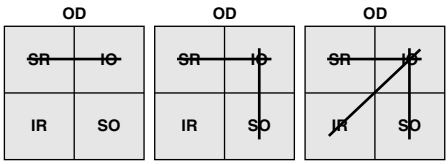
If deviation increases in right tilt



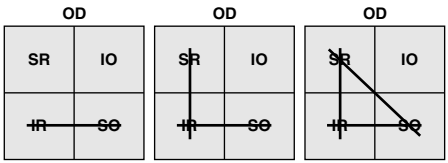
If deviation increases in left tilt

Examples

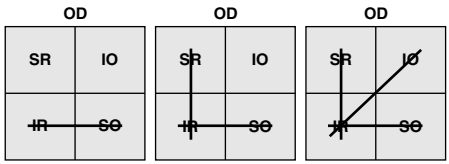
1. R hypo, L Gaze, L Tilt = RIO



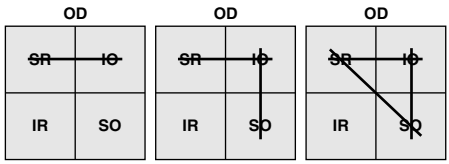
2. R hyper, R Gaze, R Tilt = LIO



3. L hypo (R hyper), R Gaze, L Tilt = RIR



4. L hyper (R hypo), L Gaze, R Tilt = LIR



CONSIDERATIONS

- Measurement of the deviation for the three steps can be accomplished by use of the cover test, or a Maddox rod with prism or scale (pp. 152-153).
- The three-step test may not work in long-standing noncomitant deviations in which spread of concomitancy has occurred.

REFERENCE

1. Parks M: Isolated cyclovertical muscle palsy, Arch Ophthalmol 60:1027, 1958.

11.13 Prism Adaptation Test

Although most widely used and studied as a presurgical tool,[1-3] the prism adaptation test (PAT) has nonsurgical applications as well.[4-5] As it is described here, its main use is in the evaluation of a patient before prescribing prism glasses. The version below is abbreviated for clinical purposes. It is a quick and easy way to assess a patient's ability to adapt to prism, providing a valuable insight into whether or not the patient will be a good candidate for a prism prescription. The logic behind the test is that if adaptation to a prism is demonstrated in a short period of time, the

patient is likely to adapt to the prism glasses and receive only transient benefit from wear.

PROCEDURE

1. Using the Maddox rod with prism or scale (pp. 152-153), measure the phoria/tropia.
2. Neutralize the phoria/tropia using the appropriate magnitude and direction of prism.
3. Remeasure the deviation to ensure that it has been completely neutralized by the prism. (The patient should have an orthophoria while wearing the prism.) If not, make the appropriate changes.
4. Place the prism in a trial frame and instruct the patient to wait *15 minutes* while wearing the prism. (Inform the patient that the prism may cause some discomfort during this period.)
5. While the patient is wearing the neutralizing prism, reassess phoria/tropia with a Maddox rod with prism or scale.

INTERPRETATION

- If the phoria/tropia remains neutralized (measured orthophoria), the patient is *not* a prism adapter.
- If the phoria/tropia shows residual deviation, interpret the results as follows:
  - If the deviation with the prism is less than the amount of the initially measured phoria/tropia, the patient is a partial prism adapter.
  - If the deviation with the prism is equal to the amount of the initially measured phoria/tropia, the patient is a complete prism adapter.

CONSIDERATIONS

- Generally patients who do not adapt to the prism are more likely to have prolonged relief of symptoms while wearing the prism prescription.
- Patients who completely or partially adapt may only experience transient relief with the prism prescription, and a vision therapy program may be a better option.

REFERENCES

1. Aust W, Welge-Lussen L: Preoperative and postoperative changes in the angle of squint following long-term, preoperative prismatic compensation. In Fells P, editor: The first congress of the International Strabismological Association, St Louis, 1971, Mosby.
2. Prism Adaptation Study Research Group: Efficacy of prism adaptation in the surgical management of acquired esotropia, Arch Ophthalmol 108:1248-1256, 1990.
3. Cotter S: Clinical uses of prism: a spectrum of applications, St. Louis, 1995, Mosby.
4. Rutstein RP, Eskeridge JB: Clinical evaluation of vertical fixation disparity. III. Adaptation to vertical prism, Am J Optom Physiol Opt 62(9):585-590.
5. Schor CM: The relationship between fusional vergence eye movements and fixation disparity, Vision Res 19:1359-1367, 1979.

11.14 Visuoscopy

Easily performed using an ophthalmoscope with a calibrated grid target, visuoscopy is used to detect and assess eccentric fixation (Fig. 11-2). Eccentric fixation is the use of a nonfoveal point to fixate objects centrally under monocular conditions.[1] Commonly found in strabismic amblyopia, not only can this condition for decreased acuity,[2,3] but it is also a prognostic indicator and a key finding in microtropia (p. 47).[4]

PROCEDURE

Perform this test on the dominant eye first.

1. Instruct the patient to occlude the nondominant eye and to look into the distance.
2. Focus on the retina using the ophthalmoscope with grid.
3. Instruct the patient to look directly into the light and at the center of the grid. Note the location of the foveal reflex in relation to the center of the grid.
4. Observe for approximately 15 to 20 seconds.
5. The dominant eye acts as a control. When you observe the foveal reflex in the dominant eye, it should be in center of the grid but may show some instability.
6. Instruct the patient to occlude the dominant eye and repeat the procedure with the nondominant eye.

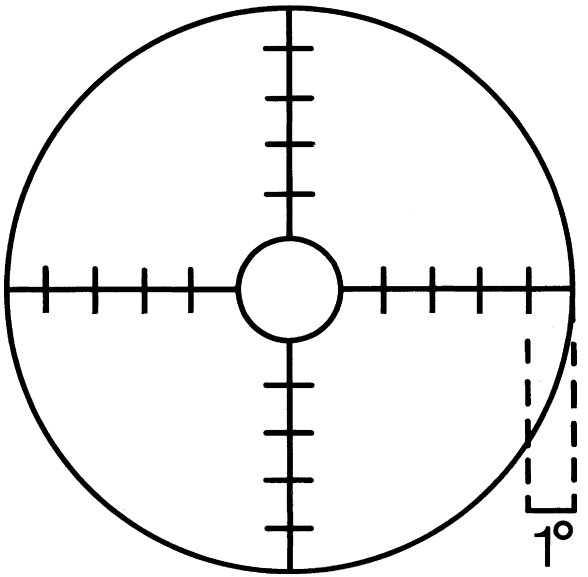


FIG. 11-2 Visuoscopy target found on Welch Allyn ophthalmoscope with marks separated by 1 degree. (From Ciuffreda KJ, Levi DM, Selenow A: Amblyopia: basic & clinical aspects, Boston, 1991, Butterworth-Heinemann.)

INTERPRETATION

First, determine whether the foveal reflex is in the center of the grid:

- If foveal reflex is in the center of the grid, there is central fixation.
- If foveal reflex is not in the center of the grid, there is eccentric fixation.

Central or Eccentric Fixation

Determine stability (steady or unsteady): Unsteady fixation is signified by a foveal reflex that continually shifts its position. Most central fixators and almost all eccentric fixators exhibit some degree of unsteadiness, so it is important to compare the dominant eye to the nondominant eye and note any differences.

Eccentric Fixation

Determine the location (part of the retina being used to fixate centrally on the grid): Note where the center of the grid lies in relation to the foveal reflex (Fig. 11-3).

- Grid center is *temporal* to the foveal reflex: Temporal eccentric fixation.
- Grid center is *nasal* to the foveal reflex: Nasal eccentric fixation.
- Grid center is *superior* to the foveal reflex: Superior eccentric fixation.
- Grid center is *inferior* to the foveal reflex: Inferior eccentric fixation.
- Grid center is *superior and nasal* to the foveal reflex: Superior/nasal eccentric fixation.

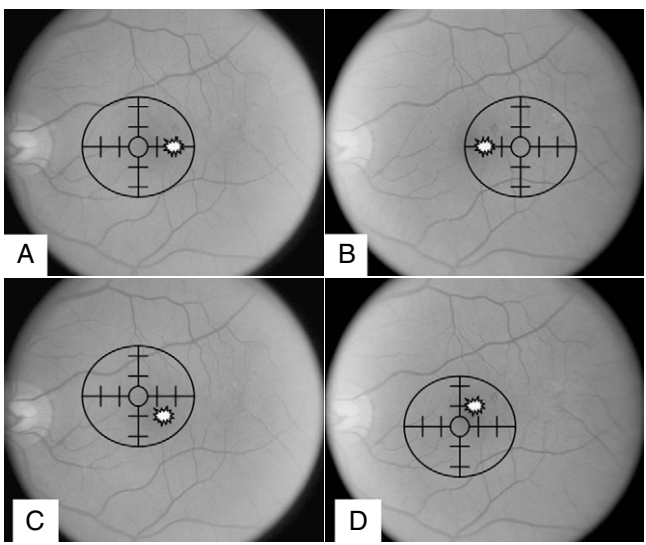


FIG. 11-3 Visuoscopy. Simulation of a visuoscopy grid being cast upon the retina of left eye. The direction of the eccentric fixation (EF) can most easily be determined by comparing the location of the grid to the foveal reflex. **A,** Grid is located nasal to foveal reflex, indicating nasal EF. **B,** Grid is located temporal to foveal reflex, indicating temporal EF. **C,** Grid is located superior and nasal to foveal reflex, indicating superior nasal EF. **D,** Grid is located inferior and nasal to foveal reflex, indicating inferior nasal EF.

Determine the magnitude in PD (distance of the eccentric point from the fovea): Note where on the grid the foveal reflex is falling. For Welch Allyn, each concentric circle on the grid denotes 1 PD of eccentric fixation. Consult the manuals of other ophthalmoscopes for calibration.

CONSIDERATIONS

- If testing a young child, first shine the grid pattern on the palm of your hand and have the child point to the center to determine whether the child understands the test concept.
- It is often helpful to lower the rheostat and use a green filter on the ophthalmoscope to reduce glare and increase your ability to see the foveal reflex.
- Larger magnitude eccentric fixation is typically more unsteady and results in larger reductions of acuity.
- The test cannot be performed in patients without discernible foveal reflex.

REFERENCES

1. Cline D, Hofstetter HW, Griffin JR: Dictionary of visual science, Radnor, Penn, 1980, Chilton.
2. Schapero M: Amblyopia, Philadelphia, 1971, Chilton.
3. Flom MC, Kirschen DG, Bedell HE: Acuity in eccentrically fixation amblyopes (letter), Am J Optom Physiol Opt 57:191-194, 1980.
4. Helveston EM, von Noorden GK: Microtropia: a newly defined entity, Arch Ophthalmol 78:272-81, 1967.

11.15 Four Base-Out Test

Image displacement, obtained by using a weak base-out prism and observing the resulting binocular (version) and monocular (fusional) eye movements, is a quick and sensitive screening procedure to determine whether or not bifoveal fusion or suppression of one fovea is present.[1] Required cooperation on the part of the patient is limited to attention and the ability to hold fixation during the test. Thus, this is a very easy test to administer to patients of all ages. Central suppression is most commonly observed in monofixation syndrome or microtropia, but it may also occur due to a pathological cause.[2]

PROCEDURE

1. The patient wears optimal refractive correction and is directed to fixate on a distance (isolated letter one line above visual acuity in the poorer-seeing eye, or point light source).
2. The patient is instructed to keep both eyes open and to fixate on the intended target.
3. Use a 4Δ base-out prism, quickly placing it over the patient's right eye and then removing it. Repeat several times, observing the movement of both the right and left eyes in response to the prism.
3. Repeat for the left eye.
4. Remember to keep reminding the patient to fixate on the intended target throughout testing.

INTERPRETATION

- The following constitutes a normal response (explained as though the prism were placed over the right eye):
  1. The right eye will adduct in response to image displacement because of the prism.
  2. The left eye will concurrently make a versional (abduct) movement equal in size.
  3. The left eye will then make a fusional (adduct) movement back to its original position.
  4. Record as "–" 4 BO OD.
- The following constitutes an abnormal response (explained as though the prism were placed over the right eye):
  1. The right eye makes no movement in response to the prism.
  2. The left eye does not move when the prism is placed before the right eye.
  3. Record as "+" 4 BO OD. This indicates a small suppression of the fovea in the right eye.

CONSIDERATIONS

- Insert and remove the prism multiple times before each eye to increase your chance of correct interpretation.
- To be complete, the test must be performed on both eyes.
- If patients have low positive fusional vergences (e.g., convergence insufficiency), they may be unable to fuse the prismatic

demand; hence the fusional movement of the eye without the prism may be absent.

REFERENCES

1. Irvine SR: A simple test for binocular fixation: clinical application useful in the appraisal of ocular dominance, amblyopia ex anopsia, minimal strabismus and malingering, Am J Ophthamol 27:740, 1944.
2. Carlson NB and others: Clinical procedures for ocular examination, ed 2, New York, 1996, McGraw-Hill.

11.16 Cycloplegic Refraction

The use of cycloplegic drugs to block the response of the ciliary muscle is an essential part of obtaining an accurate measurement of refractive error in children and many adults. A cycloplegic refraction should be performed on all children as part of a baseline finding, and should be repeated regularly if a patient has strabismus or amblyopia.

CHOICE OF CYCLOPLEGIC AGENT

A full discussion of the advantages and disadvantages of all the available cycloplegic agents is beyond the scope of this book. The reader is urged to consult other sources for a more comprehensive discussion of this topic.[1] It is the author's opinion that the best clinical method for ensuring an accurate cycloplegic refraction is the use of 2 gtt of 1% cyclopentolate separated by 3 to 5 minutes. Maximum cycloplegia occurs 30 to 60 minutes after instillation of the second drop, leaving an average of 1.25 D of residual accommodation.[2] However, it is important to note that these numbers may vary depending on the age of the patient and the color of the iris.[3]

PROCEDURE

1. Check the external health of the eye, including angle estimation and, if possible, intraocular pressure. Angle estimation testing may be performed by the penlight shadow test on young children unwilling to sit behind a slit lamp. It is often not possible to obtain an accurate measurement of the pressure.

2. Instill 2 gtt of 1% cyclopentolate separated by 3 to 5 minutes with punctual occlusion. 1 gtt of 1% or 0.5% cyclopentolate can be used in infants under 1 year of age. Although it is preferable to instill 1 drop of a topical anesthetic before the cyclopentolate, it is often not possible in young children.
3. After approximately 30 minutes check each eye to ensure cycloplegia. It is important to note that pupil dilation does not necessarily follow the same course as cycloplegia and should not be used as an indicator.[3] The following are two ways to ensure cycloplegia depending on age.
   - *Patients able to recognize letters:* Perform accommodative amplitude test to measure residual accommodation.[4]
   - *Infants or uncooperative patients:* Perform retinoscopy looking for any fluctuations in the reflex as the patient changes fixation from distance to near.
4. Perform retinoscopy, using your normal working distance. Loose lenses or a lens rack is recommended over a phoropter.
5. Given the artificial state of accommodation, a subjective refraction is typically of little clinical value–unless the patient had subnormal best-corrected acuity under dry conditions and the information will be helpful in formulating a diagnosis.

INTERPRETATION

Interpret the findings as you would normal retinoscopy findings.

CONSIDERATIONS

- Common side effects include stinging, blur, light sensitivity and less commonly diffuse ocular redness and facial flush.[5] Although more serious side effects, including CNS disturbances and psychotic episodes, are relatively rare, they have been reported in several cases,[6,7] and the clinician should be aware of them. The effects typically subside within 4 to 6 hours with no permanent consequences.[1]
- Adequate dilation may not be achieved, especially in patients with dark irides. Use of an additional drop of phenylephrine hydrochloride is often warranted to ensure adequate dilation.

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2. Priestly BS, Medine MM: A new mydriatic and cycloplegic drug. Compound 75 G.T, Am J Ophthalmol 34:572-575, 1951.
3. Manny RE and others: 1% Cyclopentolate hydrochloride: another look at the time course of cycloplegia using an objective measure of the accommodative response, Optom Vis Sci 70:651-665, 1993.
4. Carlson NB and others: Clinical procedures for ocular examination, ed 2, New York, 1996, McGraw-Hill.
5. Jones LW, Hodes DT: Possible allergic reactions to Cyclopentolate hydrochloride: case reports and literature review of uses and adverse reactions, Ophthalmic Physiol Opt 11:16-21, 1991.
6. Binkhorst RD and others: Psychotic reaction induced by Cyclopentolate, Am J Ophthalmol 56:1243-1245, 1963.
7. Shihab ZM: Psychotic reaction in an adult after topical Cyclopentolate, Ophthalmologica 181:228-230, 1980.

11.17 Delayed Subjective Refraction

A noncycloplegic method of maximizing the relaxation of accommodation, delayed subjective refraction is used as part of a subjective refraction. This method is not to be used as a substitute for cycloplegia but is still helpful in cases in which latent hyperopia or an accommodative spasm is suspected.

PROCEDURE

1. Begin with the routine protocol for subjective refraction with binocular balance.
2. Once the endpoint with best-corrected visual acuity OD, OS, and OU has been obtained, insert the near point rod and target used for negative relative accommodation (NRA).
3. Perform normal NRA while the patient is looking through your final prescription as determined by the subjective refraction. Direct the patient's attention to the *near target* and slowly increase the lenses by +0.25-D steps OU.
4. Once the patient reports sustained blur, remove near point rod and direct the patient's attention to the 20/400 "E" on the *distance chart*.

5. If the 20/400 "E" is reported by the patient to be "blurry," reduce by –0.25-D steps OU until the "E" is reported to be "clear".
6. Once the 20/400 "E" is reported to be "clear," slowly reduce the power of the lenses by –0.25-D steps OU while gradually revealing increasingly small acuity levels.
7. The endpoint is achieved when the patient is able comfortably to read the 20/20 line OU.

INTERPRETATION

If the endpoint of this procedure results in more plus or less minus than was determined by the subjective refraction, that fact is an indication that the patient was overaccommodating.[1,2]

CONSIDERATIONS

- When reducing the lens power during this procedure, take your time and allow the patient's accommodation to relax.
- During this procedure it is important to ensure that the patient is not viewing a "blurred" image for a prolonged period of time. Excessive "blur" may result in overaccommodation and defeat the purpose of the procedure. Present multiple acuity levels simultaneously so that the patient is always being presented with a line that is "clear." For example, if the patient is able to read the 20/60 line, present the 20/60, 20/50, and 20/40 lines while decreasing the power of the lenses.

REFERENCES

1. Grosvenor T: How to keep your patient from accommodating, Optom Weekly June:44-46, 1976.
2. Carlson NB and others: Clinical procedures for ocular examination, ed 2, New York, 1996, McGraw-Hill.

11.18 Double Maddox Rod

The double Maddox rod test is a subjective test for torsion, one particularly useful when differentiating a late-onset problem from an early-onset problem that has decompensated over time.[1] In cases involving the latter, patients will typically not report torsion. This test may yield results different from those obtained by

objective tests of torsion, such as direct visualization of the vertical relationship of the macula in comparison with the optic disc.[2] The double Maddox rod test should be performed on all patients with a vertical deviation, head tilt, or complaints of "tilting" images. As is the case for vertical deviations in general, paresis of the superior oblique is the most common cause of torsion.

PROCEDURE (FIG. 11-4)

Testing is performed in both primary gaze and downgaze at 40 cm.

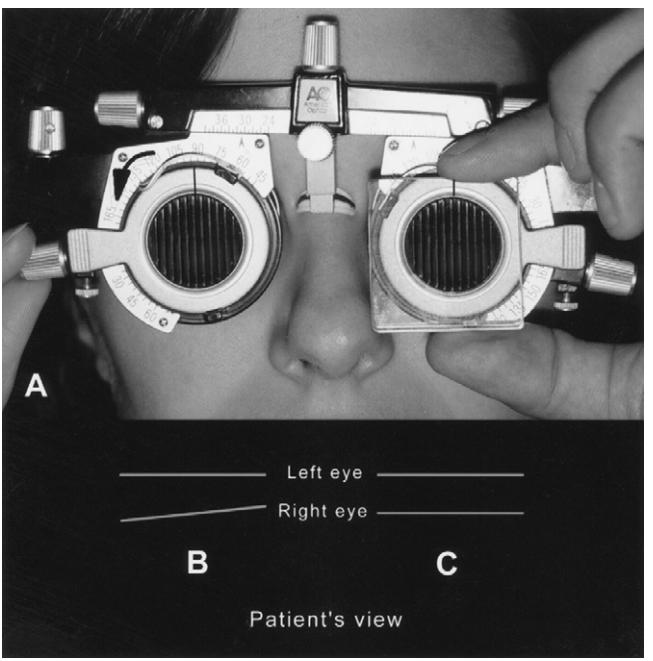

FIG. 11-4 **A,** Typical set-up for double Maddox rod test. Vertical prism may be placed before the right or left eye. Top line will be seen by the eye with the prism in front of it. **B,** Patient initially reports bottom line tilted, indicating excyclotorsion. **C,** Rotating Maddox rod in front of the right eye with image tilted until lines become parallel indicates magnitude of excyclotorsion. (From Rosenbaum AL, Santiago AP: Clinical strabismus management, 1999, Philadelphia, WB Saunders.)

1. The patient is seated comfortably in the exam chair, wearing the trial frame with the following:
   - Optimal refractive correction
   - A red Maddox rod oriented at 90 degrees (striations vertical) placed before *each* eye
   - A 6 base-down prism placed before the right eye
2. Hold a focal light source at 40 cm and direct the patient to fixate on the light.
3. The patient will see two red lines (roughly horizontally oriented), with the line on top being seen by the right eye and the line on the bottom being seen by the left eye.
4. Tell the patient that he or she will see two lines, and ask whether the lines are parallel to the floor or whether one appears tilted.
5. If the patient reports that one of the lines is tilted, rotate the Maddox rod placed before that eye until the two lines appear parallel.
6. Repeat the measurement three times and record the average.[3]

INTERPRETATION

- If the patient reports two lines parallel to each other and to the floor: *no* torsion.
- If the patient reports that one line is tilted in comparison with the other: torsion in that eye.
  - The magnitude of the Maddox rod rotation needed to achieve parallel is the degree of torsion.
- If a tilted image is viewed by the right eye, consider the following:
  - Clockwise rotation of the Maddox rod indicates intorsion.
  - Counterclockwise rotation of the Maddox rod indicates extorsion.
- If a tilted image is viewed by the left eye, consider the following:
  - Clockwise rotation of the Maddox rod indicates extorsion.
  - Counterclockwise rotation of the Maddox rod indicates intorsion.

CONSIDERATIONS

- In the event that the paretic eye has greater acuity than the nonparetic eye, the patient may fixate with the paretic eye

during testing and therefore report torsion in the nonparetic eye.[1,4]

- The use of one white Maddox rod and one red Maddox rod is another commonly employed method but may result in torsion being reported in the eye viewing through the red Maddox rod.[5]

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12

Primary Care Diagnosis and Vision Therapy for Non-Strabismic Binocular Vision Disorders

ERIK M. WEISSBERG

Three Order Binocular Work-up for Near Point Non-Strabismic Binocular Vision Disorders

The following work-up is broken down into three "orders."

- *First order:* Brief and typically sufficient to diagnose most near point nonstrabismic binocular vision disorders, this phase should be included as part of the routine testing performed by a primary care optometrist and will be described in the greatest detail in the following section.
- *Second order:* This phase represents additional testing for cases in which one is unable to obtain a clear diagnosis from the first-order work-up. This phase may need to be performed in a follow-up examination.
- *Third order:* More specialized testing, this phase may require referral.

Before testing begins ensure that the following two points have been considered:

1. Take a historical account of the symptoms.
   - *Are symptoms consistent or inconsistent with a functional binocular vision problem?*

Although symptoms may differ slightly for each binocular vision problem, there is often overlap. Common symptoms include but are not limited to the following: eyestrain and headaches with

near work; words running together; loss of place, skipping lines, or rereading lines; avoidance of near tasks; and fatigue after short periods of near work.[1-2] Onset is typically gradual. It is worth noting that young children may not report, or be unaware of, symptoms consistent with a binocular vision problem. It is important to probe school performance, avoidance of reading, inattention, or any other behaviors that may indicate a potential binocular vision problem.

- *Are the symptoms linked to eye use?*

Symptoms are typically associated with time of day (worse toward the end of the day), or they follow a pattern (eyes feel better on weekend, hurt after school, hurt after reading). If no pattern exists, it is unlikely that the patient has a functional binocular vision problem.

- *Have potential medical or neurological disorders been ruled out?*

Additional questions pertaining to any recent illnesses, changes in medications, changes in appetite or sleeping patterns, dizziness, trauma, tingling in extremities, severe headaches, and fainting should uncover any inconsistencies or suspicious signs that may indicate a nonfunctional etiology. A minimum data base consisting of visual acuity, pupils, color vision, ductions/versions, cover test, confrontational fields, and a thorough internal and external ocular health evaluation is strongly recommended for any patient with complaints of the binocular vision type. Additionally, any absence or reduction in stereopsis should raise suspicion that a strabismus, microtropia, or problem other than a nonstrabismic binocular vision disorder is present.

2. Consider significant uncorrected refractive error as a possible cause.
   - Specifically anisometropia, hyperopia, and/or astigmatism may be involved.
   - If detected, consider 6- to 8-week trial of the spectacles and a follow-up progress evaluation to look at the effect on binocular function and symptoms.
   - It is possible that a binocular problem and refractive error may coexist.

FIRST *ORDER WORK-UP* (FIG. 12-1)

1. Begin with Cover Test

- *Exophoria greater than 6 prism diopters (PD):* Suspect convergence insufficiency (p. 1); the patient may have an associated accommodative dysfunction.

- *Any esophoria:* Suspect convergence excess (p. 7), accommodative excess (p. 20), or latent hyperopia.
- *Fluctuations:* Suspect an unstable binocular system or latent hyperopia. Poor attention or an inappropriate cover test target may lead to this finding as well.
- *Normal findings:* Does not rule out a vergence/accommodative problem; proceed to the next step.

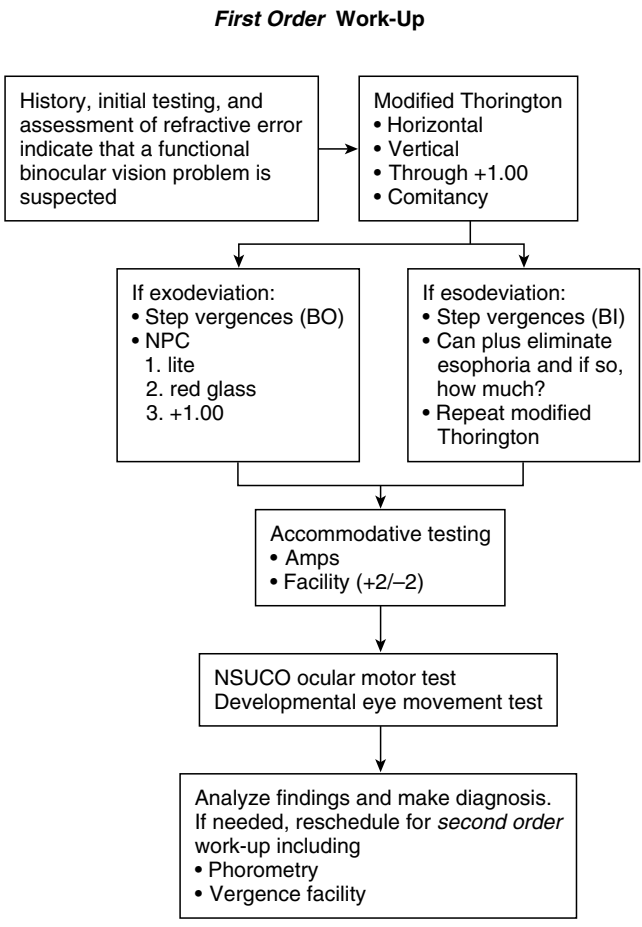

FIG. 12-1 First order work-up.

2. Modified Thorington (p. 153)

- Measure lateral phoria.
- Repeat lateral phoria through +1.00 lenses OU to determine AC/A.
- Measure vertical phoria.
- If lateral or vertical deviation is found, probe concomitancy by moving the target into nine cardinal positions of gaze.
  1. If deviation is nonconcomitant, an additional work-up may be required; consider a referral.
  2. If concomitant vertical deviation exists, it is worth probing the effect of a vertical prism (p. 99).

3. Step Vergences (p. 156)

If exophoria has been found in Steps 1 or 2:

- Perform positive fusional vergences and suspect convergence insufficiency.
- Use Sheard's criterion to compare phoria to compensating vergence. (Compensating vergence should be twice the phoria measurement). This method has been shown to predict symptoms in exophoric patients.[3,4]
- Perform near point convergence (NPC).
  1. *Light as target:* A break point greater than 6 cm is found in symptomatic patients who have a convergence insufficiency.[1-2]
  2. *Red glass over one eye and light as target:* This method is used as a provocative test and may result in immediate diplopia or degradation of NPC.
  3. *Light as target through +1.00 lenses OU:* This method tests for a convergence insufficiency with accommodative insufficiency (p. 20). If NPC improves, the patient is likely to benefit from plus lenses for near work. If NPC degrades further or shows no change, the patient more likely requires orthoptic intervention.

If esophoria has been found in Steps 1 or 2:

- Perform negative fusional vergence range and suspect convergence excess.
- Use Sheard's criterion to compare phoria with compensating vergence; this method, however, is not as helpful in cases of esophoria. Rather, the magnitude of esophoria may correlate

more closely with symptomatic patients.[3-5] Thus the management strategy is aimed at eliminating or reducing the size of the esophoria.

- Repeat modified Thorington through varying powers of plus to see how much is needed to eliminate or to make a significant reduction in the esophoria. Use as a guide for prescribing plus for near the least amount of plus that eliminates or greatly reduces the esophoria.

If no significant phoria (greater than 6 XP, or any degree of esophoria) has been found in Steps 1 or 2:

- Perform positive and negative fusional ranges to rule out vergence instability or general skills problem (p. 11).
- Repeat NPC with light 10 times, looking for a large recession. Normal recession will be less than 1 cm, but patients suffering from convergence insufficiency will recede by approximately 4 cm.[6]

4. Amplitude of Accommodation (Push up Method)

- Hofstetter's minimum (15 -0.25(age)) will give you expected results based on the patient's age.
- If hyperopia has been detected previously, it must be considered at this time. Low amplitude would suggest either an accommodative insufficiency or latent hyperopia. Asymmetric amplitude of accommodation should make one suspect uncorrected anisometropia.

5. Facility with +2/–2 Flippers (Flexibility of Accommodation) (p. 160)

- *Unable or slow to clear plus OU:* Convergence insufficiency or accommodative excess is likely. Repeat monocular testing. If the patient has no trouble with plus or minus, convergence insufficiency is likely. If the patient fails plus on monocular testing, accommodative excess is likely.
- *Unable or slow to clear minus OU:* Convergence excess or accommodative insufficiency is likely. Repeat monocular testing. If the patient has no trouble with plus or minus, convergence excess is likely. If the patient fails minus on monocular testing, accommodative insufficiency is likely.

- *Unable or slow to clear plus and minus OU:* Vergence or accommodative infacility is likely. Repeat monocular testing. If the patient has no trouble with plus or minus, vergence infacility is likely. If the patient fails either or both monocularly, accommodative infacility is likely.
- In addition to comparing results to the norms, pay attention to differences in patient responses to plus or minus and watch for fatigue toward the end of testing.

6. NSUCO Ocular Motor Test (the Maples) (p. 164)

The NSUCO ocular motor test is a simple standardized observational test that requires virtually no extra equipment and minimal time. Assessing a patient's ocular motility is essential when performing a binocular vision work-up.

7. Developmental Eye Movement Test (p. 167)

The Developmental Eye Movement test (DEM) is a standardized visual-verbal test specifically designed to differentiate functional ocular motor problems from visual-verbal processing difficulties.

Summary and Conclusion

At this point, the most common functional binocular vision problems have been examined. Approaching testing in this manner, you are seeking the most prevalent problems first. This knowledge will direct you in the appropriate referral, patient education, and case management. If the diagnosis remains unclear, then proceed to the *second order* binocular work-up or consider making a referral.

SECOND *ORDER WORK-UP*

Care should be taken to schedule this appointment coincident with the time of day at which symptoms are reported by the patient. Many of the tests in the second-order work-up employ the use of the phoropter. The artificial testing environment may stress the visual system and increase the chance of detecting a problem.

1. Abnormal or Inconsistent Findings

Repeat any abnormal or inconsistent findings noted during the first-order work-up and confirm the type and pattern of symptoms experienced by the patient.

2. Near Lateral Phoria (in Phoropter)

- Measure lateral phoria.
  1. Repeat lateral phoria measurement through +1.00 OU to determine AC/A.
  2. Exophoria of more than 6 PD and low AC/A are consistent with convergence insufficiency.
  3. Esophoria and high AC/A are consistent with convergence excess.
- Measure vertical phoria, if detected.
  1. Repeat cover test and modified Thorington for repeatability of measurement.
  2. If vertical phoria is consistently present, test for nonconcomitancy.
  3. If nonconcomitant, consider a referral.
  4. If vertical phoria is concomitant and consistently present, it is worth probing the effect of a vertical prism.

3. Vergence Testing (in Phoropter)

- *Measure positive fusional vergences:* Reduced positive fusional vergences or poor recovery indicate convergence insufficiency.
- *Measure negative fusional vergences:* Reduced negative fusional vergences or poor recovery indicate convergence excess.

4. Negative Relative Accommodation/Positive Relative Accommodation (NRA/PRA)

- Reduced NRA may indicate accommodative excess or convergence insufficiency.
- Elevated NRA may indicate uncorrected hyperopia and is a sign that the patient may benefit from plus correction for near tasks.

- Reduced PRA may indicate accommodative insufficiency or convergence excess.
- Elevated PRA may indicate accommodative excess.[6]

5. Fusion Cross Cylinder (FCC)

- A larger than normal lag (greater than +1.00) suggests accommodative insufficiency.
- A lead of accommodation (less than plano) suggests an accommodative excess.
- A variable response may indicate an unstable accommodative system or uncorrected refractive error.

6. Vergence Facility 3BI/12BO (p. 158)

This test may prove valuable in cases in which no clear diagnosis exists.

Summary and Conclusion

If the diagnosis remains unclear at this time, then consider referral for a *third-order* work-up. More specialized testing such as fixation disparity or aniseikonia work-up may be needed. If fluctuating, inconsistent, and nonrepeatable results predominated, one must consider the reliability of the patient and/or the possible presence of an underlying neurological condition (e.g., myasthenia gravis).

SUPPORTING CASE ANALYSIS FOR FIRST ORDER WORK-UP

CASE ANALYSIS 1

History

A 10-year-old boy reports at his first eye exam a history of "uncomfortable eyes" and sleepiness associated with reading. He began noticing the problem about two years ago, but it seems to go away during the summer. He admits to reading very little during the summer months. He feels that it is getting worse, and the

teacher has noticed that he rubs his eyes constantly during class. Homework at night is often difficult. He denies any diplopia but does report that the print appears blurry sometimes and that words seem to run together. He has no problem with distance vision. He denies headaches, but his mother chimes in to report that he does complain of headaches on occasion, usually at night after school. Further history to probe nonfunctional causes is negative.

Signs and symptoms consistent with a functional binocular vision anomaly have been documented and associated with eye use.

Initial testing
DVA 20/20 and NVA 20/25 OD, OS, OU
PERRLA D + C –MG
EOM: SAFE, but you do notice a change in facial expression during testing. The patient appears bothered.
Color Vision: WNL
Cover test: 8 exophoria at near and ortho at distance
Stereo: Randot 30 seconds
FCF: Full
Retinoscopy and Subjective: OD +0.25 sph, OS +0.50 sph

Although the magnitude of the exophoria lies just outside of normal limits, the most likely diagnosis at this time is convergence insufficiency.

Efficiency Exam

Modified Thorington Horizontal: 10 exophoria (concomitant)
Vertical: Orthophoria
+1.00: 12 exophoria, AC/A 2/1
Step vergences: BO at near ×/10/6
NPC lite: 8 cm to 10 cm OS out diplopia
Red glass: Diplopia
+1.00: 4 cm/6 cm
Amps: 8 D OD, OS; Hofstetter's minimum is 15-.25(age) = 12
Facility +2/–2: The patient is unable to clear minus OU, OD, OS.
MEM: +0.75 sph OD, OS

The failure to meet Sheard's criterion confirms the convergence insufficiency, a finding that is further supported by the receded near point of convergence. It is not uncommon to elicit

immediate diplopia when retesting the NPC through red glass in patients with a symptomatic convergence insufficiency.

The accommodative profile of this patient points to an associated accommodative insufficiency. This diagnosis is based on the facility and amplitude of accommodation findings. The improvement noted in NPC through +1.00 lenses is a favorable prognostic indicator for the management of this case with a reading prescription. The power of the spectacles should be determined through trial framing, and the amount of plus acceptance based on MEM. Vision therapy remains an option for this patient, but a trial period with plus lenses for near is a good alternative management strategy.

CASE ANALYSIS 2

History

14-year-old female reports a chief complaint of difficulty seeing the board in school. She notes that the blur is worse toward the end of the day but also admits that she sits in the back of the room for her last three classes. Upon further questioning, she states that although she is doing well in school, she doesn't really enjoy reading. Her mother elaborates: it has always been a struggle to get her to sit down and read, and when she does, she holds the reading material progressively closer to her face. Further history to probe nonfunctional causes is negative.

At this point, although there are no clear signs of a near point binocular problem, the clinician should be suspicious because of the patient's avoidance of reading. Furthermore, the distance blur may signify an accommodative problem, the development of myopia, or uncorrected astigmatism.

Initial testing

DVA 20/20 and NVA 20/20 OD, OS, OU
PERRLA D + C –MG
EOM: SAFE
Color Vision: WNL
Cover test: 8 exophoria at near and ortho at distance
Stereo: Randot 30 seconds
FCF: Full

Retinoscopy and Subjective: OD: –0.25 –0.50 × 085; OS: pl –0.75 × 095

Efficiency Exam
Modified Thorington Horizontal: 10 exophoria (concomitant)
Vertical: Orthophoria
+1.00: 14 exophoria, AC/A 4/1
Step vergences: BO at near 8/14/6
NPC lite: 8 cm to 10cm OS out diplopia
Red glass: Diplopia
+1.00: No improvement; the patient reports blur.
Amps: 14 D OD, OS; Hofstetter's minimum is 15-0.25(age) = 12
Facility: The patient is unable to clear plus, OD, OS, OU.
MEM: Plano to +0.25 (fluctuating) sph OD, OS

For reasons similar to those expressed in Case #1, the most likely diagnosis at this point, despite the normal AC/A, is a convergence insufficiency. Closer analysis of the findings indicates that this patient's accommodative system is responding in a manner very different from that of the patient in Case #1. This convergence insufficiency has an accommodative excess associated with it. The initial report of transient distance blur and the NPC through +1.00 are the first indicators of this association. The positive fusional vergence is low, but the normal AC/A allows this patient to receive support from accommodative convergence. However, during sustained tasks the extra accommodation turns into overaccommodation and can cause strain and distance blur after reading. Plus spectacles for near would have a detrimental effect on this patient, and vision therapy is the treatment of choice.

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General Vision Therapy Program for the Primary Care Optometrist

GENERAL INFORMATION

Vision therapy is a series of exercises designed to improve visual efficiency. The following is a description of a general program with the primary care optometrist in mind. It should be used as a starting point and may evolve as the clinician gains experience in this area. It is designed to treat the major types of near point nonstrabismic binocular vision and accommodative disorders. Although all patients will start with the same general program, it is flexible enough to reflect the different pace and needs of each individual patient and diagnosis.

GETTING STARTED

1. This program concentrates on developing the three major visual skills required for visual efficiency:
   - Vergence
   - Accommodation
   - Ocular motility
2. There are several training techniques available for each visual skill. We have chosen those techniques that are easily administered and require minimal capital investment.
   - Vergence: Brock String, Loose Prisms, Tranaglyph
   - Accommodation: Flippers, Hart Chart
   - Ocular motor: Saccadic Workbook, Michigan Tracking, Circling E's

   Equipment may be obtained through the Bernell Corporation: 4016 North Home Street, Mishawaka, IN 46545, 800-348-2225.
3. Training for each visual skill will involve multiple stages, requiring different techniques or the modification of an existing technique (Table 12.2-1). Each technique will have a specifically stated endpoint. (See the following sections for

TABLE 12.2-1
Sequence Of Training Techniques For The Three Major Visual Skills

Order	Vergence	Accommodation	Ocular motor*
1	Brock String	Monocular Letter Chart Rock	Monocular Saccadic Workbook
2	Loose Prisms	Monocular Lens Rock	Monocular Michigan Tracking
3	Smooth Tranaglyph	Binocular Letter Chart Rock	Monocular Circling E's
4	Jump Tranaglyph	Binocular Lens Rock	Binocular Saccadic Workbook
5	Jump Tranaglyph with Binocular Lens Rock		Binocular Michigan Tracking
6	Free Space Circles		Binocular Circling E's
7	Free Space Circles with Binocular Lens Rock		

*Additional ocular motor skills to incorporate can be simple visual motor tasks such as tracing pictures, coloring, and video games. Furthermore, the Letter Chart Rock (see above) can be modified to have the patient read off every other letter or the first and last letters only to train saccades. These techniques should initially be performed under monocular conditions, and once the eyes equalize in their ability the patient can move on to binocular conditions.

step-by-step instructions for each technique.) Once patients are able to reach the endpoint, they will proceed to the next stage. In this way, a general program can be designed that will allow people to start in the same place but to move quickly through the skills that do not give them difficulty.

4. Start by choosing the first technique for each visual skill outlined in Table 12.2-1 (e.g., Brock String, Monocular Letter Chart Rock, Monocular Saccadic Workbook). Patients should be trained in all three areas due to the often inseparable interaction between the three major visual skills. An in-office assessment should be performed to evaluate a patient's performance on this task.
   - If the patient is *unable* to reach the endpoint of this task, then this technique should be assigned for home therapy and progress reassessed at the next visit.

- If the patient *is able* to reach the endpoint of the task, then immediately assess performance on the subsequent task for that visual skill. Continue until you reach a technique the endpoint of which the patient cannot easily achieve. This is the task that should be assigned for home therapy.

5. Home therapy should consist of no more than three tasks that the patient should perform for a total of 20 to 30 minutes per day, 5 days per week. This means that each home task is to be performed for 5 to 7 minutes. A follow-up appointment should be scheduled every 1 to 2 weeks to assess the patient's progress and to change techniques accordingly. Depending on compliance and the severity of the problem, a complete vision therapy program should take approximately 2 to 3 months. If a patient is complying with the program but is not improving or is getting worse, then reconsider your original diagnosis and consider a consult.

ANATOMY OF AN ORTHOPTIC APPOINTMENT

A scheduled visit during an orthoptic program consists of five major elements and usually takes from 45 minutes to 1 hour.

1. History
   - *Compliance:* Determine whether or not the patient has been keeping up with the home therapy. This will provide insight into the expected progress and motivation of the patient.
   - *Subjective improvement:* Changes or alleviation of symptoms are one of the major outcome measures relied upon to determine success of a vision therapy program. A symptom survey can be used at each exam to document any changes. Recapping the original symptoms to remind the patient what the original complaint was may be of value in certain cases.
   - *Relative difficulty of techniques:* Information concerning which techniques have been perceived to be easy or difficult gives valuable insight into the patient's progress and may be used to modify the program.
2. *Repeat key diagnostic findings:* Changes in key diagnostic findings are the second major outcome measure used to evaluate the success of an orthoptic program and should be repeated at the beginning of each session. For example, if a receded NPC and low positive fusional vergences were the key

diagnostic tests for a case of convergence insufficiency, then those two tests should be repeated to evaluate progress.

3. *The patient demonstrates the home therapy techniques:* This demonstration will provide insight into compliance and progress. If a patient cannot demonstrate how to do the techniques, then he or she obviously has not been doing them. Additionally, this is the time to correct any mistakes in technique protocol and to document and monitor any changes in a patient's ability. Such changes are the third outcome measure used to evaluate success of an orthoptic program.
4. *Investigate changes in the program:* Working with the patient on the currently assigned techniques and trying the next phase in the technique will enable you to determine whether endpoints have been reached and subsequently what changes if any need to be made in the program. This is the time to determine what is to be assigned for the upcoming week(s) of home therapy.
5. *Wrap-up:* Repeat the instructions for the home techniques, educate the patient about the importance of compliance, ensure motivation, summarize any improvement noted in the three outcome measures (symptoms, changes in diagnostic findings, changes in therapy techniques), and outline goals for the next session.

SUMMARY

This program is based on a pyramidal approach to vision therapy. Starting with a broad base, patients will move quickly through those skills in which they are stronger and will have greater difficulty with the tasks for which their skills are weaker. Again, this is a general program to get the primary care practitioner started in the implementation of a vision therapy program. As the clinician gains experience, this approach can be modified with additional training procedures. If a patient is not responding or if symptoms seem to be getting worse, consider a referral to an optometrist specializing in this area to confirm your diagnosis or to implement a more elaborate vision therapy program.

Vision Therapy Techniques

Vergence Techniques

BROCK STRING

Purpose

1. To develop in the patient the "feeling" of converging and diverging
2. To develop the ability to voluntarily converge
3. To normalize the near point of convergence

Procedure

1. Approximately 1 m of string strung with two beads is used for this task.
2. The patient is instructed to hold the string taut and against the bridge of the nose, while tying the other end to a fixed object such as a door knob.
3. Set one bead (red bead) at the far end of the string and the other bead (green bead) at 40 cm.
4. Ask the patient to look at the closer bead and to describe what is seen. Because of physiological diplopia the patient should report one green bead and two red beads. In addition, two strings should be perceived crossing at the green bead,

with one string extending from the right eye and the other appearing to extend from the left eye (Fig. 12-2).

5. Ask the patient to fixate on the far bead (red), and he or she should now report one red bead with the strings crossing at the red bead. Two green beads will be seen.
6. Explain the following observations to the patient by saying, "This is a procedure that is designed to teach you how to

FIG. 12-2 A, Patient's view of Brock string during fixation on the near bead.

improve your ability to cross your eyes. The technique is designed to provide you with feedback about what your eyes are doing at all times. The way the visual system works is such that wherever your eyes are pointing, you perceive single vision. All other objects in front of or behind the object you are looking at will be seen as double. Look at the green bead and you will see one green bead, two red beads behind it and a string that crosses right at the green bead and forms the letter *X* or *Y*. The strings should look as though they are extensions of your right and left eyes. Where you perceive the two strings cross is actually the point at which your eyes

FIG. 12-2 *cont'd* **B,** Patient's view of Brock string during fixation on the far bead. (From Press LJ: Applied concepts in vision therapy, St. Louis, 1997, Mosby.)

are aimed. Thus, if you are trying to look at the green bead but the strings appear to cross farther away than the bead, this is an indication that you are looking too far away. Use this information to correct your eye position and to look closer."

7. If the patient experiences difficulty accomplishing any of the goals listed above, use the following suggestions:
   - Suggest that the patient try to get the "feeling" of looking close and of crossing his or her eyes.
   - Have the patient touch the bead that he or she is trying to fuse. This kinesthetic feedback is sometimes enough to help the patient achieve single vision.
   - Use binocular minus lenses to stimulate accommodative convergence.
8. Once the patient is able to fuse the near and far beads, fixation should be maintained on the near bead for 5 seconds and then switched to the far bead and maintained for 5 seconds. (This is considered a cycle.)
9. Have the patient perform 10 cycles, move the near bead 5 cm closer, and repeat.
10. Continue moving the near bead closer until the patient can successfully converge to a distance of 5 cm from the nose.

Alternative Skill to Be Practiced: "Bug on a String"

1. Using the same set-up, remove all beads from the string.
2. The patient begins by fixating on the end of the string and slowly converging along the string until he or she reaches a point approximately 5 cm from his or her nose.
3. Instruct the patient to diverge his or her eyes along the string until he or she reaches the far point of the string.
4. Repeat until the patient is able to perform this task with ease for 2 to 3 minutes.
5. To facilitate explanation of this task, you may instruct the patient to move his or her eyes along the string as though following a "bug" slowly crawling up and down the length of the string.

Endpoint

1. The patient can successfully converge on a bead placed 5 cm from the nose and can perform 10 cycles comfortably.

2. The patient should be able to perform the "bug on a string" task for 2 to 3 minutes with ease.
3. The patient should appreciate the different kinds of feeling and effort associated with converging and diverging.

LOOSE PRISMS

Purpose

1. To increase the patient's ability to converge and diverge.
2. To develop the "feeling" of converging and diverging.

Procedure

The patient should be positioned in a room with multiple 20/40-size targets at varied distances. Targets do not have to be letters (e.g., light switch, paper clip on desk, sticker on wall). Care should be taken to have one target approximately 3 m away, one target approximately 40 cm away, and one at an intermediate distance.

Convergence training

1. The patient is instructed to begin by holding the 4 *base-out* loose prism in the right hand.
2. While fixating on the distance target, the patient should interpose the prism before the right eye and try to the make the double image back into one. (Diplopia should be noted for no longer than 3 to 5 seconds before fusion.)
3. As soon as the image is perceived as single, the patient should remove the prism before the right eye and again make the double image back into one.
4. Repeat 10 times steps 2 and 3.
5. Repeat steps 2 through 4 for both the intermediate and near distances.
6. If successful, repeat entire the sequence but increase the prism by multiples of 4.
7. If the patient is unable to successfully complete the process for a larger prism, reduce its strength by 2 PD and repeat.
8. Note the following:
   - Diplopia may not be noticed with smaller amounts of prism.
   - The patient should be encouraged to try to "cross" his or her eyes during this technique.

- The patient should be encouraged not only to concentrate on a centrally fixated target, but also to take notice of peripheral objects.
- The procedure is performed with both eyes open, and it is not necessary to switch the prism to the left eye.

Divergence training

1. The patient is instructed to begin by holding 4 *base-in* loose prism in the right hand.
2. While fixating on the distance target, the patient should interpose the prism before the right eye and try to the make the double image back into one. (Diplopia should be noted for no longer than 3 to 5 seconds before fusion.)
3. As soon as the image is perceived as single, the patient should remove the prism before the right eye and again make the double image back into one.
4. Repeat 10 times steps 2 and 3.
5. Repeat steps 2 through 4 for both the intermediate and near distances.
6. If successful, repeat the entire sequence but increase the prism by multiples of 4.
7. If the patient is unable to successfully complete the process for a larger prism, reduce its strength by 2 PD and repeat.
8. Note the following:
   - Diplopia may not be noticed with smaller amounts of prism.
   - The patient should be encouraged to try to "relax" the eyes during this technique.
   - The patient should be encouraged not only to concentrate on the centrally fixated target, but also to take notice of peripheral objects.
   - The procedure is performed with both eyes open, and it is not necessary to switch the prism to the left eye.

Endpoint

1. The patient can successfully fuse a 20 base-out loose prism at distance, intermediate, and near.
2. The patient can successfully fuse a 12 base-in loose prism at intermediate and near, and an 8 base-in loose prism at distance.

3. The patient can appreciate the different kinds of feeling and effort associated with converging and diverging.

SMOOTH TRANAGLYPH (SMOOTH VERGENCE TRAINING)

Purpose

The purpose of the Smooth Tranaglyph is to increase positive and negative fusional vergence amplitudes.

Procedure

1. The patient is seated comfortably and wears red/green glasses while holding the "Clown" or "Bunny" tranaglyph set to zero prism demand. The "red" sheet should be placed on top of the "green."
2. Ask the patient to describe what he or she sees. The patient should be able to do the following:
   - Describe the picture and indicate that parts of the picture appear to be floating closer than other parts.
   - See the boxes with an *R* aligned over an *L*.
3. Determine whether the patient is able to appreciate blur:
   - Slowly increase the convergence demand until the patient reports that the picture is blurry.
   - Decrease the convergence demand until the patient regains clear vision.
4. Determine whether the patient is able to appreciate diplopia.
   - Slowly increase the convergence demand until the patient loses fusion.
   - Decrease the convergence demand until the patient regains single, clear vision.
5. Tell the patient to concentrate on the picture instead of the *R* and *L*.
6. Slowly move the red sheet to the right to create a small amount of convergence demand, and ask the patient to try to keep the picture clear and single, and to describe what he or she is seeing. The patient should notice that the target becomes smaller and moves closer.
7. Slowly move the red sheet to the left to create a small amount of divergence demand, and ask the patient to try to keep the picture clear and single, and to describe what he or she is

seeing. The patient should notice that the target becomes larger and moves farther away. This is SILO (small/in, large/out).

8. Explain to the patient that these are all feedback cues (blur, diplopia, SILO, and float) and will be used throughout this technique.

Convergence

1. Set the targets at zero prism demand.
2. Have the patient slowly move the red sheet to the right until he or she notices blur, diplopia, or suppression.
3. When blur, diplopia, or suppression has been noted, instruct the patient to close his or her eyes and then to reopen them to see whether single, clear vision is retained.
   - If yes, continue to move the red sheet to the right until blur, diplopia, or suppression is reported and cannot be overcome, and then continue with step 4.
   - If no, continue with step 4.
4. Slowly move the red sheet to the left until single and clear vision is once again attained.
5. Repeat, starting from step 2.
6. If the patient is having difficulty, do the following:
   - Instruct the patient to focus on a point in front of the tranaglyph. The patient may hold a pen or a finger 5 to 10 cm in front of the tranaglyph to facilitate this point.
   - Instruct the patient to concentrate on all the pictures instead of just focusing on the central picture.
   - Remind the patient to "cross" his or her eyes during this technique.

Divergence

1. The same procedure is followed as that described for convergence, except the red sheet is moved to the left.
2. If a patient is having difficulty, do the following:
   - Instruct the patient to focus on a point behind the tranaglyph. The patient may hold a pen or a finger (although he or she won't be able to see it) 5 to 10 cm behind the tranaglyph to facilitate this point.
   - Instruct the patient to concentrate on all the pictures instead of just focusing on the central picture.
   - Remind them the patient to "relax" his or her eyes during this technique.

Endpoint

1. 25 PD base-out
2. 12 PD base-in
3. In addition to achieving the above endpoint, the patient should strive for minimization of the gap between the blur/break point and the recovery of single, clear vision.

JUMP TRANAGLYPH (JUMP CONVERGENCE TRAINING)

Purpose

The purpose of the Jump Tranaglyph is to increase positive fusional vergence amplitudes.

Procedure

1. The patient is seated comfortably and wears red/green glasses while holding the "Clown" or "Bunny" tranaglyph set to zero prism demand. The "red" sheet should be placed on top of the "green."
2. Ask the patient to describe what he or she sees. The patient should be able to do the following:
   - Describe the picture and indicate that parts of the picture appear to be floating closer than other parts.
   - See the boxes with an *R* aligned over an *L*.
3. Determine whether the patient is able to appreciate blur:
   - Slowly increase the convergence demand until the patient reports that the picture is blurry.
   - Decrease the convergence demand until the patient regains clear vision.
4. Determine whether the patient is able to appreciate diplopia:
   - Slowly increase the convergence demand until the patient loses fusion.
   - Decrease the convergence demand until the patient regains single, clear vision.
5. Tell the patient to concentrate on the picture instead of the *R* and *L*.
6. Slowly move the red sheet to the right to create a small amount of convergence demand, and ask the patient to try to keep the picture clear and single and to describe what he or

she is seeing. The patient should notice that the target becomes smaller and moves closer.
7. Slowly move the red sheet to the left to create a small amount of divergence demand, and ask the patient to try to keep the picture clear and single and to describe what he or she is seeing. The patient should notice that the target becomes larger and moves farther away. This is SILO (small/in, large/out).
8. Explain to the patient that these are all feedback cues (blur, diplopia, SILO and float) and will be used throughout this technique.

Training Jump Convergence

1. Have the patient move the red sheet of the tranaglyph to the 4 base-out position and maintain single and clear vision.
2. Instruct the patient to switch fixation between distance (3 m) and the tranaglyph held at 40 cm, doing this 10 times. Each time the patient should be able to maintain single and clear vision when looking at the tranaglyph.
   - If the patient is successful, increase the tranaglyph in 4-PD steps, repeating step 2 for each new setting.
   - If the patient is unable to maintain single and clear vision, decrease the tranaglyph setting by 2-PD steps and repeat step 2 for the new setting.
3. If the patient is having difficulty, do the following:
   - Instruct the patient to focus on a point in front of the tranaglyph. The patient may hold a pen or a finger 5 to 10 cm in front of the tranaglyph to facilitate this point.
   - Instruct the patient to concentrate on all the pictures instead of just focusing on the central picture.

Endpoint

The endpoint for jump convergence training is 25 PD base-out.

JUMP TRANAGLYPH WITH BINOCULAR LENS ROCK

Purpose

The purpose of the Jump Tranaglyph with Binocular Lens Rock is to increase positive fusional vergence amplitudes though variable accommodative demands.

Procedure

1. The patient is seated comfortably and wears red/green glasses while holding the "Clown" or "Bunny" tranaglyph set to zero prism demand. The "red" sheet should be placed on top of the "green."
2. Ask the patient to describe what he or she sees. The patient should be able to do the following:
   - Describe the picture and indicate that parts of the picture appear to be floating closer than other parts.
   - See the boxes with an *R* aligned over an *L*.
3. Tell the patient to concentrate on the picture instead of the *R* and *L*.

Training Jump Convergence

1. Have the patient move the red sheet of the tranaglyph to the 4 base-out position and maintain single and clear vision.
2. Instruct the patient to maintain single and clear vision while alternately viewing through +1.00 and –1.00 lenses.
3. The patient should flip between both lens powers 10 times. Each time the patient should be able to maintain single and clear vision when looking at the tranaglyph.
   - If the patient is successful, increase the tranaglyph in 4-PD steps, repeating step 2 for each new setting.
   - If the patient is unable to maintain single and clear vision, decrease the tranaglyph setting by 2-PD steps and repeat step 2 on the new setting.

Endpoint

The endpoint for jump convergence training with binocular accommodative rock is 25 PD base-out through +1.00/–1.00 lenses.

FREE SPACE CIRCLES

Purpose

The purpose of Free Space Circles is to increase convergence and divergence ability.

Procedure

Convergence training

1. The patient holds the *opaque* eccentric circle card 40 cm from the nose.
2. Instruct the patient to fixate on a pencil tip that is placed *in front* of the card, slightly below and between the two circles (red and green) that are closest together.
3. Instruct the patient to continue fixating on the pencil tip while slowly moving it *closer* to the nose.
4. The patient should notice the red and green circles getting blurry and double. (Remind the patient to fixate on the pencil tip and to notice just the circles.)
5. The patient should notice that one of the green circles and one of the red circles merge, creating a third circle and possibly blurry circle.
6. The patient should be encouraged to ignore all of the other circles and to attempt to clear the newly created third circle by moving the pencil tip slightly forward or backward.
7. The newly created third circle (red and green mixed) should appear to float off of the page.
8. The patient should do the following:
   - Maintain fixation on the circle for approximately 10 seconds.
   - Maintain fixation on the circle without the aid of the pencil tip.
   - Maintain fixation on the circle after momentarily looking into the distance.
9. After accomplishing the goals in step 8, the patient should move up to the next set of circles and repeat the procedure until the circles on the top of the card can be fused.
10. If the patient has difficulty, suggest the following:
    - "Continue to use the pencil."
    - "Try to get the 'feeling' of looking close or crossing your eyes."
    - "Try moving the card slightly forward or backward."

Divergence training

1. The patient holds the *clear* eccentric circle card 40 cm from the nose.
2. Instruct the patient to fixate on a pencil tip that is placed *behind*, slightly below and between the two circles (red and green) that are closest together.

3. Instruct the patient to continue fixating on the pencil tip while slowly moving it *farther* from the nose.
4. The patient should notice the red and green circles getting blurry and double. (Remind the patient to fixate on the pencil tip and to notice only the circles.)
5. The patient should notice that one of the green circles and one of the red circles merge, creating a third circle and possibly blurry circle.
6. The patient should be encouraged to ignore all of the other circles and to attempt to clear the newly created third circle by moving the pencil tip slightly forward or backward.
7. The patient should do the following:
   - Maintain fixation on the circle for approximately 10 seconds.
   - Maintain fixation on the circle without the aid of the pencil tip.
   - Maintain fixation on the circle after momentarily looking into the distance.
8. After accomplishing the goals in step 7, the patient should move up to the next set of circles and repeat the procedure until the circles on the top of the card can be fused.
9. If the patient has difficulty, suggest the following:
   - "Continue to use the pencil."
   - "Try to get the 'feeling' of looking far or relaxing your eyes."
   - "Try moving the card slightly forward or backward."

Endpoint

The patient should be able to do the following:

- Maintain fixation on the circle for approximately 10 seconds.
- Maintain fixation on the circle without the aid of the pencil tip.
- Maintain fixation on the circle after momentarily looking into the distance.

FREE SPACE CIRCLES WITH BINOCULAR LENS ROCK

Purpose

The purpose of this training method, Free Space Circles with Binocular Lens Rock, is to increase convergence and divergence ability while stimulating and relaxing accommodation.

Procedure

Convergence training

1. The patient holds the *opaque* eccentric circle card 40 cm from the nose.
2. Instruct the patient to fixate and fuse circles (as described in the "free space fusion" section for convergence training), starting with the circles that are closest together.
3. The patient should continue to maintain fixation on the circle while alternately viewing through +1.00 and –1.00 flip lenses for 10 "flips."
4. The patient moves to the next set of circles and repeats these steps.

Divergence Training

1. The patient holds the *clear* eccentric circle card 40 cm from the nose.
2. Instruct the patient to fixate and fuse circles (as described in "free space fusion" section for divergence training), starting with the circles that are closest together.
3. The patient should continue to maintain fixation on the circle while alternately viewing through +1.00 and –1.00 flip lenses for 10 "flips."
4. The patient moves to the next set of circles and repeats these steps.

Endpoint

The patient should be able to maintain fixation on the circle through 10 "flips" between +1.00 and –1.00 lenses

Accommodative Techniques

MONOCULAR LETTER CHART ROCK

Purpose

- Restore normal monocular accommodative amplitude
- Restore normal monocular accommodative facility

Procedure

1. Place a chart of 20/40 distance letters on a wall (10 lines with 5 letters per line).
2. The patient occludes the left eye and stands 40 cm from the distance chart.
3. The patient slowly backs away from the chart until the letters have just become blurry or the patient has reached a distance of 3 m. This distance is how far the patient should stand from the chart during this technique.
4. Ask the patient to hold a small chart of 20/40 near letters (10 lines with 5 letters per line) at 40 cm and to call off the letters on the top line while slowly moving the chart closer.
5. When the patient can no longer keep it clear, he or she moves the near chart 2.5 cm farther away and then shifts focus to the first line of the distance chart. The patient repeats the far-to-near change for each letter on the first line of both charts (reading off one letter on the distance chart and then one letter on the near chart.)
6. After completing line one, the patient moves the near chart to 40 cm and calls off letters on the second line as the chart is slowly moved closer. Repeats step 5.
7. The patient repeats steps 5 and 6 until all 10 lines are complete.
8. If the patient experiences difficulty at any level, suggest that he or she try to get the "feeling" of looking close and of crossing his or her eyes with the small chart, or with the large chart have the patient try to get the feeling of relaxing or staring.
9. Have the patient repeat the procedure with the right eye occluded.

Endpoint

- The patient successfully clears the near chart held at a distance equal to age-appropriate amplitude, clears the distant chart at 3 m, and shifts and maintains focus while switching back and forth between the two distances.
- The patient should be able to appreciate the different feelings and efforts associated with clearing the print while viewing the far and near charts.

MONOCULAR LENS ROCK

Purpose

- Normalize accommodative amplitude for both stimulation and relaxation
- Normalize accommodative facility

Procedure

1. Occlude the patient's left eye.
2. Age-appropriate reading material is held at 40 cm.
3. The patient is asked to hold two lenses, one in the right hand and one in the left hand. (A flipper can be used as well.)
4. Start with +0.50 D and +1.00 D.
5. The patient is asked to clear the print while alternating between the two lenses.
6. The goal is to achieve 20 cycles (clearing each lens) in 1 minute.
7. If the patient experiences difficulty, do the following:
   - Suggest that the patient try to get the "feeling" of looking close with the minus lenses, or of relaxing or staring with the plus lenses.
   - Decrease the demand by moving the card away until the print is clear and then moving it back to 40 cm for the minus lenses, or move the card closer until the print clears and then move it back to 40 cm for the plus lenses.
8. Have the patient repeat the procedure with the right eye occluded.
9. If the patient is able to achieve the endpoint for one lens design, move on to the next.

Lens Designs

- +0.50/+1.00
- +0.50/+2.00
- +0.50/−0.50
- +1.00/−1.00
- +1.00/−2.00
- +2.00/−3.00
- +2.00/−4.00
- +2.00/−5.00

Endpoint

The endpoint for all lens designs will be first clearing the lenses successfully and then clearing 20 cycles per minute.

BINOCULAR LETTER CHART ROCK

Purpose

- Restore normal binocular accommodative amplitude
- Restore normal binocular accommodative facility

Procedure

1. Place a chart of 20/40 distance letters on a wall (10 lines with 5 letters per line).
2. The patient stands 40 cm from the distance chart.
3. The patient slowly backs away from the chart until the letters have just become blurry or the patient has reached a distance of 3 m. This distance is how far the patient should stand from the distance chart during this technique.
4. Ask the patient to hold a small chart of 20/40 near letters (10 lines with 5 letters per line) at 40 cm and to call off the letters on the top line while slowly moving the chart closer.
5. When the patient can no longer keep it clear, he or she moves the near chart 2.5 cm farther away and then shifts focus to the first line of the distance chart. The patient repeats the far-to-near change for each letter on the first line of both charts (reads off one letter on the distance chart and then one letter on the near chart).
6. After completing line one, the patient moves the near chart to 40 cm and calls off letters on the second line as the chart is slowly moved closer. Repeat step 5.
7. The patient repeats steps 5 and 6 until all 10 lines are complete.
8. If the patient experiences difficulty at any level, suggest that he or she try to get the "feeling" of looking close and of crossing his or her eyes with the small chart, or with the large chart have the patient try to get the feeling of relaxing or staring.

Endpoint

- The patient successfully clears the near chart held at a distance equal to age-appropriate amplitude, clears the distant chart at

3 m, and shifts and maintains focus while switching back and forth between the two distances.
- The patient should be able to appreciate the different feelings and efforts associated with clearing the print while viewing the far and near charts.

BINOCULAR LENS ROCK

Purpose

The purpose of the Binocular Lens Rock is to normalize accommodative amplitude and facility under binocular conditions.

Procedure

1. Age-appropriate reading material is held at 40 cm.
2. The appropriate lens design should be incorporated into a "flipper."
3. Start with +0.50 D OU and +1.00 D OU.
4. The patient is asked to clear the print while alternating between the two lens powers.
5. The goal is to achieve 20 cycles (clearing each lens) in 1 minute.
6. If the patient experiences difficulty, do the following:
   - Suggest that patient try to get the "feeling" of looking close with the minus lenses, or of relaxing or staring with the plus lenses.
   - Decrease the demand with the minus lenses by moving the card away until the print is clear and then moving it back to 40 cm, or with the plus lenses by moving the card closer until the print clears and then moving it back to 40 cm.
   - Decrease the demand by decreasing the power of the lenses for either plus or minus.
7. If patient is able to achieve the endpoint for one lens design, move on to the next design.
8. If suppression is suspected (e.g., the patient has intermittent strabismus), a bar reader and Polaroid glasses should be incorporated into the procedure design.

Lens Designs

- +0.50/+1.00
- +0.50/+2.00

- +0.50/–0.50
- +1.00/–1.00
- +1.00/–2.00
- +2.00/–3.00
- +2.00/–4.00
- +2.00/–5.00

Endpoint

The endpoint for all lens designs will be first clearing the lenses successfully and then clearing 20 cycles per minute.

Ocular Motor Techniques

SACCADIC WORKBOOK (MONOCULAR AND BINOCULAR)

Purpose

The Saccadic Workbook improves the accuracy of saccadic eye movements.

Procedure

1. Begin by having the patient occlude the left eye with a tie-on patch.
2. Instruct the patient to sit at a desk with Workbook Level I open to the first page.
3. Instruct the patient to read from left to right, starting with the first section of numbers and calling out each underlined number. Do not time the patient's performance.
4. Repeat for 2 to 3 minutes and then repeat with the right eye patched.
5. Initial stages of training concentrate on successful completion of the task; later stages concentrate on increased efficiency, and the patient's performance time is monitored.
6. The pages of the workbook, as well as the different levels (I, II, and III), get progressively more difficult due to decreased spacing between symbols and the use of multiple symbols.

7. This exercise may also be performed binocularly once the patient has demonstrated equal efficiency with both the right and left eyes.
8. If difficulty is encountered, do the following:
   - Encourage the patient to use kinesthetic feedback by using a finger to help the eyes track.
   - Have the patient use a ruler or typoscope to isolate each line while reading along in the section.

Endpoint

- Although the endpoint may vary slightly for each patient, generally, patients should be able to complete a section (one block of numbers) in approximately 10 to 15 seconds, monocularly and binocularly.
- The ability of both eyes should be equal under monocular conditions.

ANN ARBOR (MICHIGAN) TRACKING (MONOCULAR AND BINOCULAR)

Purpose

The purpose of the Ann Arbor Tracking method is to improve accuracy and ability of eyes to track using a combination of saccade- and pursuit-type eye movements.

Procedure

1. Begin by having the patient occlude the left eye with a tie-on patch.
2. Instruct the patient to sit at a desk with the Michigan Tracking book opened to the first page.
3. Instruct the patient to read from left to right, starting with the first paragraph of letters and circling each successive letter of the alphabet. For example, tell the patient, "Look for the first *A* and circle it, then find the first *B* and circle it, and so on."
4. The patient should be able to find all letters of the alphabet represented, *A* through *Z*. If the patient has skipped a letter or missed its first appearance in order, then he or she will not be able to finish the entire alphabet and should go back to find the mistake.

5. Repeat for 2 to 3 minutes and then repeat with the right eye patched.
6. Initial stages of training concentrate on successful completion of the task; later stages concentrate on increased efficiency, and the patient's performance time is monitored.
7. The pages of the book get progressively more difficult by decreasing the spacing between symbols and decreasing the overall size of the symbols.
8. This exercise can also be performed binocularly once the patient has demonstrated equal efficiency with both the right and left eyes.

Alternately, a similar training technique can be performed with an identical procedure, but instead the patient finds all of the *e*'s or dots the *o*'s in a newspaper or magazine. The disadvantage of this technique is that the examiner gives up some control of the font size, quality, and spacing. Because of the close spacing and small font of newspaper print, this technique is typically considered to be more difficult than that involving the Michigan Tracking Book.

If difficulty is encountered, do the following:

1. Encourage the patient to use kinesthetic feedback by using a finger or pen to help the eyes track.
2. Have the patient use a ruler or typoscope to isolate each line while reading along in the section.

Endpoint

- Although the endpoint may vary slightly for each patient, generally patients should be able to complete a section (one paragraph of letters) in approximately 60 seconds with no errors.
- The ability of both eyes should be equal under monocular conditions.

Index

Page numbers followed by f indicate figures; t, tables; b, boxes.

B

E

F

N

O

P

R

S

T

W