Esotropia and Exotropia
As a service to its members and the public, the American Academy of Ophthalmology has developed a series of guidelines called Preferred Practice Patterns that identify characteristics and components of quality eye care.

The Preferred Practice Pattern® guidelines are based on the best available scientific data as interpreted by panels of knowledgeable health professionals. In some instances, such as when results of carefully conducted clinical trials are available, the data are particularly persuasive and provide clear guidance. In other instances, the panels have to rely on their collective judgment and evaluation of available evidence.

Preferred Practice Patterns provide guidance for the pattern of practice, not for the care of a particular individual. While they should generally meet the needs of most patients, they cannot possibly best meet the needs of all patients. Adherence to these Preferred Practice Patterns will not ensure a successful outcome in every situation. These practice patterns should not be deemed inclusive of all proper methods of care or exclusive of other methods of care reasonably directed at obtaining the best results. It may be necessary to approach different patients’ needs in different ways. The physician must make the ultimate judgment about the propriety of the care of a particular patient in light of all of the circumstances presented by that patient. The American Academy of Ophthalmology is available to assist members in resolving ethical dilemmas that arise in the course of ophthalmic practice.

The Preferred Practice Pattern® guidelines are not medical standards to be adhered to in all individual situations. The Academy specifically disclaims any and all liability for injury or other damages of any kind, from negligence or otherwise, for any and all claims that may arise out of the use of any recommendations or other information contained herein.

References to certain drugs, instruments, and other products are made for illustrative purposes only and are not intended to constitute an endorsement of such. Such material may include information on applications that are not considered community standard, that reflect indications not included in approved FDA labeling, or that are approved for use only in restricted research settings. The FDA has stated that it is the responsibility of the physician to determine the FDA status of each drug or device he or she wishes to use, and to use them with appropriate patient consent in compliance with applicable law.

Innovation in medicine is essential to assure the future health of the American public, and the Academy encourages the development of new diagnostic and therapeutic methods that will improve eye care. It is essential to recognize that true medical excellence is achieved only when the patients’ needs are the foremost consideration.

All Preferred Practice Patterns are reviewed by their parent panel annually or earlier if developments warrant and updated accordingly. To ensure that all guidelines are current, each is valid for 5 years from the “approved by” date unless superseded by a revision.

Financial Disclosures:
This author has disclosed the following financial relationships occurring from January 2006 to August 2007:
# TABLE OF CONTENTS

**INTRODUCTION** ............................................................................................................................................. 2  
**SECTION I. ESOTROPIA**  
**ORIENTATION** ........................................................................................................................................... 3  
Entity ......................................................................................................................................................... 3  
Disease Definition ...................................................................................................................................... 3  
Infantile (Congenital Esotropia) .................................................................................................................... 3  
Accommodative Esotropia ............................................................................................................................. 4  
Differential Diagnoses of Esotropia ........................................................................................................... 4  
Patient Population ....................................................................................................................................... 4  
Activity ....................................................................................................................................................... 4  
Purpose ...................................................................................................................................................... 4  
Goals ......................................................................................................................................................... 4  
**BACKGROUND** ....................................................................................................................................... 5  
Epidemiology and Risk Factors .................................................................................................................. 5  
Natural History .......................................................................................................................................... 5  
Rationale for Treatment ............................................................................................................................. 5  
**PREVENTION AND EARLY DETECTION** .................................................................................................. 6  
**CARE PROCESS** ..................................................................................................................................... 7  
Diagnosis .................................................................................................................................................... 7  
History ..................................................................................................................................................... 7  
Examination ............................................................................................................................................. 7  
Assessment of Visual Acuity and Fixation Pattern .................................................................................. 7  
Ocular Alignment and Motility .................................................................................................................. 8  
Extraocular Muscle Function .................................................................................................................... 9  
Detection of Nystagmus ............................................................................................................................ 9  
Sensory Testing ....................................................................................................................................... 9  
Cycloplegic Retinoscopy/Refraction ......................................................................................................... 9  
Funduscopic Examination ......................................................................................................................... 10  
Additional Testing ................................................................................................................................... 10  
Management ............................................................................................................................................ 10  
Choice of Therapy .................................................................................................................................... 10  
Follow-up Evaluation ............................................................................................................................... 13  
Provider ................................................................................................................................................... 14  
Counseling and Referral .......................................................................................................................... 14  
**SECTION II. EXOTROPIA**  
**ORIENTATION** ...................................................................................................................................... 15  
Entity ....................................................................................................................................................... 15  
Disease Definition .................................................................................................................................... 15  
Patient Population .................................................................................................................................... 15  
Activity .................................................................................................................................................... 15  
Purpose ................................................................................................................................................... 15  
Goals ....................................................................................................................................................... 15  
**BACKGROUND** .................................................................................................................................... 16  
Epidemiology and Risk Factors ................................................................................................................ 16  
Natural History ....................................................................................................................................... 16  
Rationale for Treatment ........................................................................................................................... 16  
**PREVENTION AND EARLY DETECTION** ................................................................................................. 17  
**CARE PROCESS** ................................................................................................................................... 17  
Diagnosis .................................................................................................................................................. 17  
Management ........................................................................................................................................... 17  
Choice of Therapy .................................................................................................................................... 17  
Follow-up Evaluation ............................................................................................................................... 19  
Provider ................................................................................................................................................... 20  
Counseling and Referral .......................................................................................................................... 20  
**APPENDIX 1. SUMMARY OF MAJOR RECOMMENDATIONS FOR CARE** ............................................. 21  
**RELATED ACADEMY MATERIALS** .......................................................................................................... 26  
**REFERENCES** ......................................................................................................................................... 26
INTRODUCTION

The Preferred Practice Pattern® (PPP) guidelines have been written on the basis of three principles.

- Each Preferred Practice Pattern should be clinically relevant and specific enough to provide useful information to practitioners.
- Each recommendation that is made should be given an explicit rating that shows its importance to the care process.
- Each recommendation should also be given an explicit rating that shows the strength of evidence that supports the recommendation and reflects the best evidence available.

In the process of revising this document, a detailed literature search in Medline and the Cochrane Library for articles in the English language was conducted on the subject of esotropia and exotropia for the years 2001 to 2006. The results were reviewed by the Pediatric Ophthalmology/Strabismus Panel and used to prepare the recommendations, which they rated in two ways. The panel first rated each recommendation according to its importance to the care process. This “importance to the care process” rating represents care that the panel thought would improve the quality of the patient’s care in a meaningful way. The ratings of importance are divided into three levels.

- Level A, defined as most important
- Level B, defined as moderately important
- Level C, defined as relevant but not critical

The panel also rated each recommendation on the strength of evidence in the available literature to support the recommendation made. The “ratings of strength of evidence” also are divided into three levels.

- Level I includes evidence obtained from at least one properly conducted, well-designed randomized controlled trial. It could include meta-analyses of randomized controlled trials.
- Level II includes evidence obtained from the following:
  - Well-designed controlled trials without randomization
  - Well-designed cohort or case-control analytic studies, preferably from more than one center
  - Multiple-time series with or without the intervention
- Level III includes evidence obtained from one of the following:
  - Descriptive studies
  - Case reports
  - Reports of expert committees/organizations (e.g., PPP panel consensus with external peer review)

The evidence is that which supports the value of the recommendation as something that should be performed to improve the quality of care. The panel believes that it is important to make available the strength of the evidence underlying the recommendation. In this way, readers can appreciate the degree of importance the committee attached to each recommendation and they can understand what type of evidence supports the recommendation.

The ratings of importance and the ratings of strength of evidence are given in bracketed superscripts after each recommendation. For instance, “[A:II]” indicates a recommendation with high importance to clinical care [A], supported by sufficiently rigorous published evidence, though not by a randomized controlled trial [II].

This PPP is divided into two parts, with the management of esotropia described first followed by the management of exotropia. In both parts, the sections entitled “Orientation” and “Background” do not include recommendations; rather, they are designed to educate and provide summary background information and rationale for the recommendations that are presented in the Care Process section. A summary of the major recommendations for care for esotropia and exotropia is included in Appendix 1.
SECTION I. ESOTROPIA

ORIENTATION

ENTITY

Infantile and accommodative esotropias.

Esotropia, which includes entities with the following ICD-9 classifications:

- Nonaccommodative (378.00)
- Accommodative (378.35)
- Alternating (378.05)
- Alternating, with A pattern (378.06)
- Alternating, with V pattern (378.07)
- Alternating, with X or Y pattern (378.08)
- Monocular (378.01)
- Monocular, with A pattern (378.02)
- Monocular, with V pattern (378.03)
- Monocular, with X or Y pattern (378.04)
- Unspecified (378.00)

Esotropia, intermittent, which includes entities with the following ICD-9 classifications:

- Alternating (378.22)
- Monocular (378.21)

DISEASE DEFINITION

Infantile and accommodative esotropias are terms that define convergent misalignments of the visual axes. The scope of this document is limited to the nonparalytic, nonrestrictive form of the disease with an onset in childhood and with minimal or no limitation in range of motion of the eyes.

Esotropia can be categorized in a variety of ways, usually based on age of onset or underlying causes.

Infantile (Congenital) Esotropia

Infantile esotropia usually presents between the age of 3 and 6 months.\(^1\) Intermittent esotropia during the first 3 months of life\(^2-8\) is common and does not necessarily predict the development of constant strabismus. For this PPP, features of infantile esotropia include the following:

- Onset before the age of 12 months without spontaneous resolution
- Nonaccommodative etiology
- Large constant angle of deviation
- Cross fixation
- Mild A and V pattern deviations

Features that may not be present at the time of diagnosis include:

- Latent nystagmus
- Dissociated vertical deviation
- Oblique muscle dysfunction
- Optokinetic nystagmus asymmetry
- Abnormal binocular vision
Accommodative Esotropia

For this PPP, features of accommodative esotropia include:

- An accommodative component usually associated with hyperopia.
- Esotropia not explained by sixth cranial nerve (CN VI) palsy, congenital anomalies of innervation (e.g., Duane syndrome), intraocular or orbital disease, or intracranial disease.

Most cases of accommodative esotropia are first noted between the ages of 1 and 8 years, with an average onset at 2.5 years. Children with accommodative esotropia are presumed to have had normal visual development before the onset of the strabismus. Parents may observe the onset of accommodative esotropia with illness, fever, or minor trauma. A substantial portion of these children may have subnormal binocular function at the onset of their deviation.

Accommodative esotropia is caused by excessive convergence resulting from bilateral hyperopia (usually more than 2.00 diopters [D]), an elevated accommodative convergence to accommodation (AC/A) ratio, or combinations thereof. Over 30% of children with +4.00 D or more of hyperopia develop esotropia by 3 years of age. Children with hyperopia who develop esotropia have higher AC/A ratios when compared with similarly matched hyperopic children without strabismus. The average amount of hyperopia in individuals with accommodative esotropia and high AC/A ratios is approximately +2.00 D, and in those with normal AC/A ratios it is approximately +4.50 D. Although most cases appear after 1 year of age, some appear in infancy and may reappear as a sequel to surgically corrected infantile esotropia. A progressive increase in the angle and frequency of deviation is common in these patients, and dysfunction of the oblique muscles may occur in up to 30% of cases.

Children with accommodative esotropia more frequently have amblyopia at presentation than children with infantile esotropia. It is not known whether developing amblyopia produces a further impediment to fusion or always requires prior fusion disruption.

Differential Diagnoses of Esotropia

Differential diagnoses of esotropia include CN VI palsy, Duane syndrome, acquired nonaccommodative esotropia, divergence insufficiency, and nystagmus blockage-type esotropia.

PATIENT POPULATION

Individuals with childhood-onset esotropia.

ACTIVITY

Evaluation, diagnosis, and management of infantile and accommodative esotropias.

PURPOSE

The purpose of treating infantile and accommodative esotropias is to restore normal ocular alignment, thereby promoting development and preservation of binocular vision.

GOALS

The goals of the patient care process are to do the following:

- PREDICT DISEASE
  - Identify children at risk for esotropia
- DIAGNOSE
  - Detect esotropia
  - Detect and treat amblyopia that may cause, or be caused by, esotropia
- INFORM
  - Educate the patient and family/caregiver of the diagnosis, treatment options, and care plan
  - Inform the patient’s other health providers of the diagnosis and treatment plan
TREAT

- Treat the esotropia (align the visual axes) in order to promote and maintain binocular vision (fusion, stereopsis), prevent or facilitate treatment of amblyopia, and restore normal appearance
- Limit the effects of amblyopia treatment on quality of life
- Lessen the effect of strabismus and amblyopia on employment and career choices

MONITOR

- Monitor vision and ocular alignment and modify therapy as appropriate

BACKGROUND

EPIDEMIOLOGY AND RISK FACTORS

Strabismus is any ocular misalignment. The most common types are esotropia (inwardly deviating eyes) and exotropia (outwardly deviating eyes). Prevalence estimates of strabismus range from 1% to 6% in different populations, with esotropia reported five times more frequently than exotropia in Ireland and twice as frequently in Australia. In Hong Kong and Japan, however, exotropia is more frequent than esotropia. Amblyopia can both cause and result from a manifest strabismus. Approximately 50% of children who have strabismus develop amblyopia on that basis.

Certain pediatric populations are at higher risk for developing strabismus, including children who are neurodevelopmentally impaired, were born prematurely or had low birth weight, had low Apgar scores, have craniofacial anomalies, high hyperopia, and a family history of strabismus. In some families, a Mendelian inheritance pattern has been observed. The incidence of infantile esotropia is related to premature births and perinatal morbidity, genetic disorders, and detrimental prenatal environmental influences such as substance abuse and smoking. Reduction or prevention of those factors can result in a decrease in the incidence of infantile esotropia.

NATURAL HISTORY

Up to one quarter of the esotropias appearing between 3 and 6 months of age will resolve over time and, thus, do not meet the definition of infantile esotropia used in this PPP. The magnitude of the angle of deviation may inversely correlate with the likelihood of resolution. Studies have found that if a constant esotropia of 40 prism diopters (PD) or greater was present at age 2 to 4 months, it was unlikely to resolve. Infantile esotropia rarely resolves spontaneously within the first year of life.

Accommodative esotropia is more frequent than infantile esotropia and usually presents between the ages of 1 year and 7 years. Onset as early as 4 months of age has been reported. It may begin as an intermittent deviation associated with fatigue, illness, or near viewing and progress to constant esotropia when untreated. Because younger children lose binocular vision rapidly, the chance of resolution with hyperopic correction alone is better if the duration of constant esotropia before treatment is shorter. In one study, 18% of patients with onset of accommodative esotropia before 1 year of age eventually underwent extraocular muscle surgery despite correction of refractive error with eyeglasses; only 4% of those with onset after age 2 years deteriorated despite correction of refractive error with eyeglasses.

RATIONALE FOR TREATMENT

The potential benefits of treatment for esotropia include promoting binocular vision and normal visual function in each eye. If binocularity is achieved, the number of surgical procedures over a lifetime and overall cost to society may be reduced. Fusion and stereopsis are necessary for some careers and may be useful in others as well as in athletic activities and activities of daily life. The appearance of crossed eyes may reduce employment opportunities. In addition, ocular alignment at any age enhances social interactions by normalizing appearance as well as eye
Normal ocular alignment is important for the development of a positive self-image and social eye contact.59,61,62,65,66

PREVENTION AND EARLY DETECTION

Prevent Blindness America has estimated that early identification and treatment of children with strabismus could have prevented amblyopia in approximately 5 million Americans.60 However, the benefit of early detection on improved visual outcomes has not been unequivocally demonstrated.69,70

Although the threshold of hyperopia that requires treatment has not been established, prescribing eyeglasses for at-risk children may reduce the risk of developing accommodative esotropia and/or amblyopia.71,72 In hyperopic patients, anisometropia is a risk factor for the development of accommodative esotropia.73

Table 1 provides guidelines for prescribing eyeglasses for young children.

| TABLE 1 | CONSENSUS GUIDELINES FOR PRESCRIBING EYEGLASSES FOR YOUNG CHILDREN[A,8]|  |
|----------|-----------------|---|---|---|
| Condition | Diopters | Age 0–1 year | Age 1–2 years | Age 2–3 years |
| Isometropia | (similar refractive error in both eyes) | | | |
| Myopia | ≥ -5.00 | ≥ -4.00 | ≥ -3.00 |
| Hyperopia (no manifest deviation)* | ≥ +6.00 | ≥ +5.00 | ≥ +4.50 |
| Hyperopia with esotropia† | ≥ +3.00 | ≥ +2.00 | ≥ +1.50 |
| Astigmatism | ≥ 3.00 | ≥ 2.50 | ≥ 2.00 |
| Anisometropia | | | | |
| Myopia | ≥ -2.50 | ≥ -2.50 | ≥ -2.00 |
| Hyperopia | ≥ +2.50 | ≥ +2.00 | ≥ +1.50 |
| Astigmatism | ≥ 2.50 | ≥ 2.00 | ≥ 2.00 |
| Additional Factors | | | | |
| History of previous amblyopia or strabismus surgery | | | | |
| Visual acuity | | | | |
| Acceptance of eyeglass wear | | | | |
| Possible accommodative esotropia/monofixation syndrome | | | | |
| Medical comorbidities | | | | |
| Developmental delay | | | | |

NOTE: These values were generated by consensus and are based solely on professional experience and clinical impressions, because there are no scientifically rigorous published data for guidance. The exact values are unknown and may differ among age groups; they are presented as general guidelines that must be tailored to the individual patient.

* May reduce the correction by up to 50% (but no more than 3.00 diopters) depending on the clinical situation.
† In higher hyperopes, reduction of the cycloplegic refraction may be necessary to achieve eyeglass acceptance.


DIAGNOSIS

The purpose of the comprehensive strabismus evaluation is to make the diagnosis, establish baseline status, and determine appropriate initial therapy. The possibility of restrictive, paralytic, or other neurologic causes (especially head trauma or increased intracranial pressure) for the strabismus should be considered. Because binocular vision can degrade rapidly in young children, resulting in suppression and anomalous retinal correspondence, early diagnosis and treatment are essential. 26,73,74

The examination of a patient who has childhood-onset strabismus includes all components of the comprehensive pediatric or adult ophthalmic evaluation in addition to the sensory, motor, refractive, and accommodative functions. 75,76

History

Although a thorough history generally includes the following items, the exact composition varies with the patient's particular problems and needs.

- Demographic data, including identification of parent/caregiver, and patient's gender and date of birth
- Documentation of identity and relationship of historian
- The identity of other pertinent health care providers
- The chief complaint and reason for the eye evaluation, including date of onset and frequency of the ocular misalignment; which eye is deviated and in what direction; and the presence or absence of diplopia, squinting, or other visual symptoms. Review of photographs of the patient may be helpful.
- Ocular history, including other eye problems, injuries, diseases, surgery, and treatments (including eyeglasses and/or amblyopia therapy)
- Systemic history; birth weight; prenatal and perinatal history that may be pertinent (e.g. alcohol, drug, and tobacco use during pregnancy); past hospitalizations and operations; general health and development
- Pertinent review of systems, including history of head trauma and relevant systemic diseases
- Family and social history including eye conditions (strabismus, amblyopia, type of eyeglasses and history of wear, extraocular muscle surgery or other eye surgery, and genetic diseases)
- Current medications and allergies

Examination

The comprehensive strabismus examination should include the following elements:

- Assessment of fixation pattern and visual acuity in each eye
- Ocular alignment and motility at distance and near
- Extraocular muscle function (ductions and versions including incomitance, such as A and V patterns)
- Detection of nystagmus
- Sensory testing
- Cycloplegic retinoscopy/refraction
- Funduscopic examination

Documentation of the child's level of cooperation with the examination can be useful in interpreting the results and in making comparisons between examinations.

Assessment of Visual Acuity and Fixation Pattern

The method of evaluating visual acuity will vary according to the age of the child and level of cooperation. Distance visual acuity should be determined monocularly whenever possible. To prevent peeking when a child's visual acuity is tested, the occluded eye should be completely...
Section I. Esotropia

covered. The child should not hold the occluder. An adhesive occluder patch is recommended to ensure assessment of monocular visual acuity. Monocular visual testing for patients with nystagmus may require special techniques such as blurring of the fellow eye; binocular testing also should be performed for these patients.

Under ideal circumstances, visual acuity testing conditions should be standardized in each examination room and at each visit so that the same viewing distance and lighting conditions are used. Some children are more amenable to testing at shorter distances. The testing distance, type of optotype, and whether the optotype is presented a line at a time or isolated, should be documented. Patients should be encouraged to learn optotype-equivalent tests at the earliest possible age.

Infant and Preverbal Child

Visual acuity measurement of the infant and preverbal child is limited to qualitative assessment of fixation, fixation preference, and tracking (following) movements of the eyes. These assessments are usually made by drawing the child’s attention to the examiner’s or family’s/caregiver’s face (infants under 3 months) or a toy either hand held or at 20 feet (6 meters). Because children resist covering the sound eye when the other has limited vision, the vigor with which the child objects to alternate occlusion of the eyes can be used to judge the relative quality of vision in each eye. Fixation behavior is recorded for each eye as “fix, follows, maintains” or the equivalent “central, steady, maintains.” An assessment of equality of vision also can be made by observing the child’s ability to maintain fixation when a prism is placed in the visual axis of each eye in turn. Several tests that have been described utilize different amounts of prism and different ways of introducing the prism. In the 10 PD base-up fixation test, the prism is introduced and the child’s fixation preference is observed. It is recorded as alternates or the preferred eye is the right/left and nonpreferred eye holds well, holds briefly, or shows no hold. With the 25 PD base-in test, there is equal vision if fixation alternates. This test does not distinguish equal vision from unequal vision in those children whose fixation does not alternate. Both induced tropia tests fail to differentiate amblyopia from fixation preference.

Verbal Preliterate Child

Quantitative visual acuity assessment in cooperative verbal children (at approximately age 3 years) involves recognition of symbols, tumbling E, or letters presented at a standardized distance, generally at 20 feet (6 meters). Linear targets or targets surrounded by crowding bars are preferred because these targets may help identify children with subtle amblyopia by detecting interocular differences in acuity. In such patients, testing with isolated figures may suggest symmetrical acuities or a false negative test. The crowding phenomenon is important in amblyopia and may result in inconsistencies in measured visual acuity because of decreased recognition of a target within others. The Vision in Preschoolers Study Group reported that a crowded line of Lea symbols was more accurate in detecting reduced visual acuity. Allen figures are not crowded and have been shown to be less accurate than other methods. Parents/caregivers can assist in the testing process by showing the child the figures or tumbling E game before the test. The tumbling E is used less frequently because it has a lower success rate in children when compared to picture charts.

Although it is not a measure of visual acuity, stereoacuity testing may detect fusional defects found in small-angle strabismus.

Literate Child

Snellen acuity is routinely tested at distance (about 20 feet or 6 meters) and, when appropriate, at near (about 13 inches or 0.33 meter). In some children, testing at 6 meters cannot be accomplished, but the child can be tested at 3 meters.

Ocular Alignment and Motility

Ocular alignment can be evaluated using a variety of clinical methods. When possible, a target that controls the patient's accommodation should be used for both distant and near fixation during assessment of the deviation regardless of the technique. The method of measuring the angle of
the deviation and the presence or absence of refractive correction should be documented. If the patient is uncooperative (especially a child under 3 years old or an individual with developmental delay), the angle may be estimated using the corneal light reflection test with or without prisms or by estimating the amount of eye movement to refixate on alternate-cover testing. The cover-uncover and alternate-cover tests using prisms are more accurate and used whenever feasible to quantify ocular misalignment in appropriate gaze and head positions. The simultaneous prism-and-cover test may provide additional useful information for patients with fusional vergences, where the ocular alignment under binocular viewing conditions is better than during alternate cover testing (e.g., monofixation syndrome).

**Extraocular Muscle Function**

Versions and ductions should be evaluated and any over- or underactions of extraocular muscles assessed and recorded. Inferior oblique muscle dysfunction, A or V patterns, or dissociated vertical or horizontal deviations may develop over time. The examiner should note any limitations in versions or ductions. The oculocephalic rotations maneuver (doll’s head) is particularly valuable in infants and young children and often reveals clinically normal ductions that cannot otherwise be documented, even with patch testing. Diseases associated with paresis/paralysis of the extraocular muscles are not included in the scope of this PPP.

**Detection of Nystagmus**

Nystagmus in the patient with esotropia may be manifest or latent. Manifest nystagmus is constantly present and may be horizontal, vertical, or torsional. It is typically symmetrical, although it may vary in magnitude, speed, and wave form, depending on the direction of gaze and other specific viewing conditions. Latent nystagmus (sometimes called occlusion nystagmus) is conjugate, predominantly horizontal, jerk oscillations of the eyes that are produced or exacerbated by monocular viewing. It is characterized by a slow drift away from the fixating eye, with rhythmic jerk redress movements to re-establish central fixation. The nystagmus is described as latent because it is typically perceptible or accentuated when one eye is occluded. Both manifest and latent nystagmus may coexist in the same patient.

**Sensory Testing**

The binocular sensory status should be assessed when feasible using Worth 4-dot testing and stereoaucity tests. Reliable data may be difficult to obtain in younger children. In the older strabismic (especially esotropic) patient, more detailed sensory testing is occasionally useful, especially if there is a history of diplopia. An orthoptic evaluation may be useful to further define the sensory status of the child.

**Cycloplegic Retinoscopy/Refraction**

Determination of refractive errors is important in the diagnosis and treatment of amblyopia or strabismus. Patients should receive an accurate cycloplegic refraction either by retinoscopy or by subjective refraction. Prior to cycloplegia, dynamic retinoscopy provides a rapid assessment of accommodative function and may be helpful in evaluating a child with high hyperopia or possible accommodative insufficiency.

Cycloplegia is necessary for accurate refraction in children. Cyclopentolate is useful because it has a rapid onset and produces cycloplegia that approximates the effect of topical ophthalmic atropine but with a shorter duration of action. Cyclopentolate 1% is more frequently used; cyclopentolate 2% is also available. The strength of cyclopentolate should be determined based on the child’s weight, iris coloration, and dilation history. In eyes with heavily pigmented irides, adjunctive agents such as tropicamide and/or phenylephrine hydrochloride may be necessary to achieve adequate dilation. In rare cases, topical ophthalmic atropine may be necessary to achieve maximal cycloplegia. The use of topical anesthetic prior to the cycloplegic makes the cycloplegic sting less and promotes its penetration into the eye.
Funduscopic Examination

Retinal or optic nerve abnormalities may lead to strabismus. Fundus examination is preferably performed with the binocular indirect ophthalmoscope, at which time the relationship between the macula and optic nerve can be assessed. Temporal displacement of the macula (most often seen in patients with retinopathy of prematurity) may cause a positive angle kappa, with nasal displacement of the corneal light reflection. This can simulate exotropia in a straight-eyed child or mask the strabismus in a child with esotropia. A negative angle kappa is seen less frequently and is usually associated with high myopia.

Additional Testing

Forced duction and/or force generation tests may be useful if there is incomitance or other evidence of extraocular muscle restriction, or if paresis/paralysis is suspected. Generally, such testing in young children is not feasible as an office procedure. Many ophthalmologists perform forced duction testing routinely at the beginning of extraocular muscle surgery. Detection of mechanical restriction may alter the surgical approach.

MANAGEMENT

All forms of esotropia should be considered for treatment. Ocular alignment should be established as soon as possible, especially in young children, to maximize binocularity, prevent or facilitate treatment of amblyopia, and normalize appearance. In almost all cases, clinically important refractive errors should be corrected. Amblyopia treatment is usually started before surgery, because this may reduce the angle of strabismus or increase the likelihood of good postoperative binocularity.

Although there is no level I evidence that early surgical correction improves outcomes for infantile esotropia, uncontrolled studies support the belief of many clinicians that surgery by 6 months of age results in a better potential for binocularity than later surgery. The surgeon must balance the potential benefit of early surgery with the risk of producing consecutive exotropia. There is no consensus among strabismus surgeons on the criteria for unilateral or bilateral surgery, nor is there level I evidence to provide guidance as to which approach is superior.

Choice of Therapy

The following treatment modalities are used alone or in combination as required to achieve the therapeutic goal:

- Correction of refractive errors
- Bifocal lenses
- Prism therapy
- Amblyopia treatment
- Extraocular muscle surgery

Treatment plans are formulated in consultation with the patient and parent/caregiver. The plans should be responsive to their expectations and preferences, including the family’s/caregiver’s perception of the existing alignment, which may differ from the ophthalmologist’s, and what they hope to achieve with treatment. It is important that the family/caregiver and ophthalmologist agree on the goals of treatment before surgery is performed. A particularly challenging situation can arise when the surgical intervention might worsen the appearance of the child. In patients for whom the potential for binocularity is poor, surgery to normalize appearance is still appropriate treatment.

Correction of Refractive Errors

Corrective lenses should be the first treatment for refractive errors that can cause reduced visual acuity in one or both eyes or are likely to contribute to the esotropia. Such refractive errors include anisometropia, hyperopia, and, occasionally, high myopia. In general, eyeglasses are well tolerated by children, especially when there is visual improvement. Accurate fitting and maintaining proper adjustment facilitate acceptance. Straps may be useful in babies; cable temples...
Section I. Esotropia

and spring hinges are helpful in keeping eyeglasses on active young children. Polycarbonate lenses have greater safety and are preferable for children, especially if they are amblyopic. For patients with accommodative esotropia, realignment by eyeglasses or contact lenses alone is successful in most cases. In one study, only one of 26 (4%) of children with onset after 2 years of age required surgical intervention. In children with onset before 1 year of age, three of 17 (18%, P = NS) required surgery. In general, the greater degree of hyperopia indicates a higher likelihood that the refractive error is an important etiologic factor of the esotropia. Other factors that warrant initial treatment with eyeglasses include the presence of a variable angle of deviation; greater deviation at near; change of deviation after cycloplegia; and family history of hyperopia, amblyopia, accommodative esotropia, or eyeglasses in early childhood. It is usually worthwhile to prescribe eyeglasses for patients with esotropia who have greater than or equal to 1.50 D of hyperopia, to determine if there is an accommodative component, because children may develop esotropia as a result of relatively mild hyperopia or anisometropia. While often intolerant of eyeglasses, children with developmental delay should also be considered for such correction on a trial basis.

The aim of treatment is to correct hyperopia sufficiently to restore alignment, and, in most cases, the full refractive error should be prescribed. Undercorrection of the hyperopia sometimes improves compliance, especially in older children, and can provide acceptable ocular alignment. In such cases, prescribing less than the full cycloplegic refraction may restore clear vision and allow increasing hyperopic correction to be prescribed over time if required. A postcycloplegic refraction may be required to balance visual acuity and ocular alignment. In amblyopic patients, part-time occlusion of the dominant eye may improve alignment and sometimes obviates the need for surgery.

Maximum response to eyeglasses may take several months or longer. If misalignment persists, the full cycloplegic refraction should be prescribed before considering surgery. An increase of hyperopic correction should be prescribed if esotropia persists or recurs. Cycloplegia may be used temporarily to facilitate compliance in wearing eyeglasses. In preteens and teenagers, gradual reduction of the full cycloplegic hyperopic correction can be attempted if the deviation is controlled. Such reductions in the hyperopic correction are titrated against the angle of deviation.

Bifocals

An esodeviation greater for near than for distant targets is found in some cases. Convergence excess is defined clinically as an increased near esodeviation of 10 PD or greater compared with the distance deviation (high clinical AC/A ratio) with the use of full hyperopic correction. Bifocal treatment should be considered in patients with sensory fusion who maintain essentially straight eyes at distance but have a manifest esotropia at near (typically greater than 10 PD) while wearing their full hyperopic correction. If successful, bifocals may be necessary on a long-term basis to maintain ocular alignment for viewing near targets. Eliminating bifocals is feasible in approximately 60% of cases after an average of 5 years of use. However, excellent initial response is associated with a lower likelihood that the bifocals can be withdrawn later without recurrence of the esotropia.

For children 5 years of age and younger, the bifocal add should be prescribed as an executive or a flat-top type, with the top of the bifocal bisecting the pupil in primary gaze in younger children and a few millimeters lower (between the lower eyelid and lower pupillary border) in older children. While the minimum strength of the bifocal can sometimes be estimated by office testing in trial frames, it is usually more practical to prescribe a +2.50 to +3.00 D add for all patients requiring them. Later reductions can then be made as part of a routine eyeglasses change. Progressive bifocals offer some cosmetic advantages and can be used effectively in older children. However, the transition zone should generally be placed several millimeters higher than the standard adult fitting.

Disadvantages of bifocals include expense, appearance, and potential rejection by the child. A minority of clinicians avoid bifocals because they believe that alignment at distance is sufficient to protect binocular vision and that most children will outgrow their high AC/A ratio more easily wearing single-vision lenses. In some cases, strabismus surgery is appropriate in older children to reduce dependence on bifocals or to allow for transition to contact lenses. Surgical correction can
reduce the AC/A ratio\textsuperscript{107,108} and eliminate the need for bifocal wear without producing consecutive exotropia at distance.\textsuperscript{109-111}

Prism Therapy
Prisms are rarely useful in infantile esotropia in part because the angle of deviation is usually too large to correct. In selected subgroups of patients with accommodative esotropia, the use of press-on plastic prisms to promote binocular vision and establish the full angle for extraocular muscle surgery has been shown to be effective.\textsuperscript{103} In the late 1980s, the Prism Adaptation Study investigated the role of preoperative prisms in the surgical management of acquired esotropia. Prisms were used preoperatively to determine the maximum angle of the strabismus and to estimate fusion potential. Reported surgical success rates, defined as a horizontal deviation of 8 PD or less (measured with the simultaneous prism and cover test at distance fixation), were highest (90%) among those participants who responded to prisms and underwent extraocular muscle surgery for the adapted angle of esotropia.\textsuperscript{102,103} However, because prism-adapted patients received greater amounts of surgery on average, it is possible that increasing surgical dosage without prism adaptation would have produced similar results. In addition, press-on prisms cause visual symptoms that some children find objectionable, are costly, require re-evaluation (additional office visits), and may be unacceptable in children not otherwise wearing eyeglasses. For this reason, prism adaptation is used only selectively by most practitioners.

Amblyopia Treatment
The control of an esotropic deviation may occasionally be improved following the treatment of amblyopia (see Amblyopia PPP\textsuperscript{112}). Treatment of amblyopia is not a cure for strabismus. Surgical treatment of esotropia in the presence of moderate to severe amblyopia has a lower success rate than in the presence of mild or no amblyopia.\textsuperscript{96}

Extraocular Muscle Surgery
Children or adults with esotropia should undergo surgical correction if eyeglasses and amblyopia management are ineffective in straightening the eyes.\textsuperscript{104} Strabismus surgery should be performed only when more conservative methods have failed or are unlikely to be of benefit. Surgery is rarely justified when the primary objective is to eliminate the eyeglasses.\textsuperscript{[A:III]} Except for acquired symptomatic deviations in older children or adults, small-angle deviations of less than 12 PD at distance or near are not usually considered for surgery.\textsuperscript{[A:III]}

Although some binocular vision and stereopsis can be restored after surgical alignment in infantile esotropia,\textsuperscript{55,113} achievement of high-grade stereopsis is rare.\textsuperscript{53,54,56} In contrast, the quality of stereopsis appears to be improved by prompt surgical realignment in decompensated accommodative esotropia.\textsuperscript{54,104,114} Because variability in the angle of deviation may predict resolution, documentation of a stable angle of deviation is generally warranted before surgical intervention.\textsuperscript{2,49 [A:III]}

While most patients with infantile esotropia receive surgical intervention during childhood, it is unknown whether early treatment results in improved outcomes with respect to long-term motor alignment. However, achieving ocular alignment early in life (before age 2 years) to within 10 PD of orthotropia increases the likelihood of achieving binocularity.\textsuperscript{53,54,98-100}

Whether or not there is surgical realignment of infantile esotropia, many affected children later develop other motility problems, such as occlusion nystagmus, dissociated strabismus, or inferior oblique muscle overaction.\textsuperscript{17,115} The presence of amblyopia\textsuperscript{96} or nystagmus\textsuperscript{116} is associated with an increased rate of requiring reoperation. In addition, esotropia may recur on an accommodative basis in 50% of patients and correlates with the magnitude of the hyperopia.\textsuperscript{58}

Extraocular muscle surgery usually is performed for the distance angle of deviation when the individual is wearing full hyperopic correction; however, some surgeons use the maximum near deviation. For those individuals with a distance-near disparity (high AC/A ratio), bilateral medial rectus recession usually reduces the ratio.\textsuperscript{107,108} Prism adaptation for the near angle,\textsuperscript{117} augmentation of the recession over amounts done with a normal AC/A ratio,\textsuperscript{118} or posterior fixation suture (Fadenoperation),\textsuperscript{110} increases the likelihood of a satisfactory alignment and eventual weaning from bifocals.
The amount of surgery and the choice of surgical technique may vary (e.g., methods of suture placement in the muscle and sclera, or measurement of recession or resection). Although two-muscle surgery is most frequently performed, sometimes three- or four-horizontal-muscle surgery may be required for large-angle deviations. Some clinicians believe that two-muscle surgery is the better option for all deviations, regardless of magnitude, to reduce the risk of consecutive exotropia.

Results may be similar with different procedures; one method may be chosen over another on the basis of preoperative diagnosis, angle of deviation, technical ease, anatomical exposure, the need for an assistant, presence of scar tissue, and other factors such as physician preference and experience. Bilateral medial rectus muscle recessions are commonly performed as the initial surgical procedure. Most surgeons prefer uniocular surgery (single-muscle recession or recession/resection) for patients with irreversible amblyopia or substantially reduced vision in one eye on an anatomic basis. Operating on both eyes may be preferable in specific clinical circumstances, such as V pattern esotropia with inferior oblique-muscle overaction or null-point nystagmus with compensatory face turn. Detailed discussion of the surgical indications and management of complex deviations is beyond the scope of this publication.

Other Methods

Eye exercises are not indicated in patients with esotropia. Training in diplopia recognition (antisuppression training) and vergence amplitudes is ineffective in the treatment of most esotropic patients and may occasionally produce permanent diplopia, especially in patients with monofixation syndrome.

Chemodenervation by injection of botulinum toxin type A in one or more extraocular muscles induces a temporary weakness by pharmacologic blockade of the neuromuscular junction. While the mechanism of long-term ocular realignment is unknown, it is likely that the primary effect comes from contracture of the direct antagonist rather than long-term paresis of the injected muscle. As with conventional extraocular muscle surgery, favorable prognostic indicators include good vision in each eye, absence of restricted eye movement, and potential for binocular vision. Such treatment may be an alternative to conventional extraocular muscle surgery in selected patients, but its value in managing infantile esotropia has not been established. Disadvantages include the frequent need for repeat injection; iatrogenic ptosis, which may increase the risk for amblyopia; and need for general anesthesia. Importantly, delayed ocular realignment may be disadvantageous in an infant with a rapidly developing visual system.

Miotic agents such as echothiophate are cholinesterase inhibitors, which reduce accommodative effort and convergence by stimulating ciliary muscle contraction and reducing pupillary size. Although sometimes effective, this method is less desirable than using corrective lenses because of a risk of adverse systemic side effects such as diarrhea, asthma, and/or increased salivation and perspiration as well as increased risk associated with the administration of certain agents used in general anesthesia. The potential for ocular side effects, including cataract, retinal detachments, and iris cysts, exists. While traditionally used as topical 0.06% drops, the drug is only commercially available in the stronger 0.125% preparation.

Follow-up Evaluation

Even when initial treatment results in good ocular alignment, follow-up is essential, since the child remains at high risk for developing amblyopia, losing binocular vision, and having a recurrence of strabismus. Until visual maturity is reached, periodic evaluations are necessary (see Table 2).

During the teenage years, and if the examination has been stable, follow-up evaluations are appropriate every 1 to 2 years thereafter. New or changing findings may indicate the need for more frequent follow-up examinations.

In children with esotropia, hyperopia should be regularly assessed, typically every 1 to 2 years, depending on age and clinical circumstances. More frequent cycloplegic examinations are indicated in cases with changes in acuity, amblyopia, or unstable alignment. Cyclopentolate 1% is effective and convenient for office use. In some patients, more hyperopia may be documented with regular eyeglass wear. If the esotropia appears to be accommodative in etiology but is not controlled with the current eyeglasses, repeat cycloplegic refraction should be performed.
prior to concluding a nonaccommodative cause.\textsuperscript{[A:III]} Atropine sulfate 1\% may be used to establish adequate cycloplegia when shorter acting drugs are inadequate.\textsuperscript{91}

### TABLE 2 FOLLOW-UP EYE EXAMINATION GUIDELINES FOR CHILDREN WITH ESOTROPIA\textsuperscript{[A:III]}

<table>
<thead>
<tr>
<th>Age</th>
<th>Interval</th>
</tr>
</thead>
<tbody>
<tr>
<td>0 - 1 year</td>
<td>3 - 6 months</td>
</tr>
<tr>
<td>1 - 5 years</td>
<td>6 - 12 months</td>
</tr>
<tr>
<td>5 years</td>
<td>12 - 24 months</td>
</tr>
</tbody>
</table>

*NOTE: More frequent visits may be necessary if amblyopia is present (see Amblyopia PPP\textsuperscript{112}) or if there is a recent deterioration of alignment.*

---

### PROVIDER

Certain diagnostic procedures may be delegated to appropriately trained and supervised auxiliary personnel. The interpretation of results, diagnosis, and management of disease, including surgical correction and follow-up, require the high degree of medical and surgical training, clinical judgment, and experience of the ophthalmologist.\textsuperscript{[A:III]} Consultation with or referral to a pediatric ophthalmologist or general ophthalmologist with expertise in the diagnosis and treatment of strabismus may be desirable for cases in which the diagnosis, etiology, or management plan is unclear, or the esotropia appears unresponsive to treatment.

Diagnostic evaluation and treatment by an orthoptist can be a useful adjunct to the ophthalmologic care of patients with esotropia. The orthoptist's initial and subsequent assessments of visual function as well as monitoring of amblyopia and its treatment may supplement the ophthalmologist's management. The orthoptist's participation in the patient's care process may include assessing binocular function (suppression, diplopia, or fusion); determining the accommodative component and evaluating the usefulness of bifocals; using prisms diagnostically and/or therapeutically pre- and postoperatively; and monitoring amblyopia.

### COUNSELING AND REFERRAL

Childhood esotropia is a long-term problem that requires commitment from the parent/caregiver and ophthalmologist to achieve the best possible outcome.

The ophthalmologist should discuss the findings of the evaluation with the patient, when appropriate, as well as with the parent/caregiver. The ophthalmologist should explain the disorder and recruit the family in a collaborative approach to therapy.\textsuperscript{[A:III]} Parents/caregivers of pediatric patients who understand the diagnosis and rationale for treatment are more likely to adhere to treatment recommendations.\textsuperscript{124,135}
SECTION II. EXOTROPIA

ORIENTATION

ENTITY
Exotropia in children, which includes entities with the following ICD-9 classifications:

- Alternation (378.15)
- Alternation with A pattern (378.16)
- Alternation with specified noncomitancy not elsewhere classifiable (378.18) (includes alphabetical patterns)
- Alternation with V pattern (378.17)
- Monocular (378.11)
- Monocular with A pattern (378.12)
- Monocular with specified noncomitancy not elsewhere classifiable (378.14) (includes alphabetical patterns)
- Monocular with V pattern (378.13)
- Intermittent unspecified (378.20)
- Alternating, intermittent (378.24)
- Monocular, intermittent (378.23)
- Unspecified (378.10)

DISEASE DEFINITION
Exotropia is a divergent misalignment of the visual axes. The scope of this document is limited to the nonparalytic, nonrestrictive form of the disease with an onset in childhood and with minimal or no limitation in range of motion of the extraocular muscles. Exotropia associated with unilateral vision loss on a structural basis is not within the scope of this document.

Exotropia is usually characterized by the frequency of the deviation. The onset of exotropia often is in early childhood, and the condition is commonly intermittent. Sensitivity to light with closure of the deviating eye (squinting) is frequent. Congenital and profound unilateral visual impairment may also result in infantile exotropia.

PATIENT POPULATION
Individuals with childhood-onset exotropia.

ACTIVITY
Evaluation, diagnosis, and management of exotropia.

PURPOSE
The purpose of managing exotropia is to restore normal ocular alignment.

GOALS
The goals of the patient care process are to do the following:

- PREDICT DISEASE
  - Identify children at risk for exotropia
- DIAGNOSE
  - Detect exotropia
  - Detect and treat amblyopia that may cause, or be caused by, exotropia
Section II. Exotropia

◆ INFORM
  - Educate the patient and family/caregiver of the diagnosis, treatment options, and care plan
  - Inform the patient’s other health providers of the diagnosis and treatment plan

◆ TREAT
  - Treat the exotropia (align the visual axes) in order to promote and maintain binocular vision (fusion, stereopsis), prevent or facilitate treatment of amblyopia, and restore normal appearance
  - Limit the effects of amblyopia treatment on quality of life
  - Lessen the effect of strabismus and amblyopia on employment and career choices

◆ MONITOR
  - Monitor vision and ocular alignment and modify therapy as appropriate

BACKGROUND

EPIDEMIOLOGY AND RISK FACTORS

Exotropia is the most common type of exodeviation and occurs in approximately 1% of the population; intermittent exotropia is the most frequently reported type. In Hong Kong and Japan, exotropia is more frequent than esotropia. In Ireland and Australia, however, esotropia is the more frequent type of strabismus reported. One population-based study found that intermittent exotropia was twice as frequent in girls than boys. A study of children with infantile-onset (congenital) exotropia found that half had associated ocular or systemic anomalies. Certain pediatric populations are at higher risk for developing strabismus, including children who have a neurodevelopmental impairment; were premature or had low birth weight; have a history of low Apgar scores; have craniofacial anomalies, high hyperopia, or a family history of strabismus.

NATURAL HISTORY

Exotropia is typically intermittent and its onset is usually before 3 years of age, but it may be detected for the first time much later in childhood. The deviation often becomes manifest at times of fatigue, visual inattention, or illness when fusional compensatory mechanisms are compromised. The patient may close one eye in bright light. Generally, the image from the deviated eye will be suppressed and the patient will be asymptomatic; if suppression does not occur the patient will experience intermittent diplopia. Often only one eye will be suppressed and deviate spontaneously. Mild amblyopia occasionally occurs, but clinically important amblyopia is uncommon in the context of an intermittent deviation.

Although classifications derived from presumed etiologic bases have been used, exotropia is usually described clinically on the basis of frequency of the deviation, laterality, magnitude at distance and at near, and symptoms. Although some ophthalmologists believe that the frequency of the deviation increases with time, the natural history has not been unequivocally established. Although some reports suggest that many patients who decline surgical correction appear to remain stable or spontaneously improve with observation alone, others show deterioration during long-term follow-up. However, if the deviation becomes constant, binocular vision can deteriorate or become lost. It is unknown whether loss of binocular vision or deterioration of ocular alignment is the primary event in causing progression towards constancy. The causes of exotropia are poorly understood. Proposed etiologies include excess tonic divergence and mechanical or innervational orbital factors. Severe unilateral or bilateral vision loss of any etiology may cause exotropia, but is not the topic of this PPP.

RATIONALE FOR TREATMENT

The potential benefits of treatment for exotropia include promoting binocular vision and normal visual function in each eye. The importance of normal ocular alignment on the development of a
positive self-image should not be underestimated; the appearance of misaligned eyes impairs self-image and social interactions and may reduce employment opportunities.52,56,144

PREVENTION AND EARLY DETECTION

There is an increased incidence of exotropia in children with a history of premature birth, perinatal morbidity, genetic disorders, and detrimental prenatal environmental influences such as maternal substance abuse and smoking.45 Reduction or prevention of those factors as well as diagnosis and treatment of myopia and myopic anisometropia can result in a decreased incidence of exotropia.

CARE PROCESS

DIAGNOSIS

The purpose of the initial comprehensive strabismus evaluation is to make the diagnosis, establish baseline status, inform the family/caregiver, and determine therapy. The possibility of restrictive, paralytic, or other neurologic causes (especially head trauma or increased intracranial pressure) for the strabismus should be considered.

The examination of a patient who has childhood-onset strabismus includes all components of the comprehensive pediatric or adult ophthalmic evaluation in addition to the sensory, motor, refractive, and accommodative functions.75,76 The history, examination, and additional testing are the same as for a patient with esotropia (see pp. 7-10).

MANAGEMENT

All forms of exotropia should be considered for treatment.143 In most cases, ocular alignment should be re-established as soon as possible, especially in young children, if the deviation is manifest a large percentage of the time.143 However, the optimal modes of therapy for exotropia, the long-term benefit of early surgical correction, and the superiority of bilateral versus unilateral surgery are not well established.125 Amblyopia is uncommon in patients with intermittent exotropia, but it should be treated if present.143

Choice of Therapy

The following treatment modalities may be used alone or in combination as required to achieve the therapeutic goal:

- Correcting refractive errors143
- Overcorrecting minus lenses143
- Patching (antisuppression therapy)143
- Amblyopia treatment143
- Prism therapy143
- Convergence exercises for convergence insufficiency143
- Extraocular muscle surgery143

Correcting Refractive Errors

Corrective lenses should be prescribed for any clinically significant refractive error that causes reduced vision in one or both eyes.143 Improved retinal image clarity often improves the control of the exotropia.146 Such refractive errors include all degrees of myopia, hyperopia greater than 3.0 D, astigmatism, and anisometropia. Correcting even mild amounts of myopia may be beneficial. Correction of mild to moderate amounts of hyperopia is not generally recommended for patients with intermittent exotropia, because reducing accommodative convergence can worsen the control or size.
Section II. Exotropia

of the exodeviation. However, patients with greater amounts of hyperopia and/or hyperopic anisometropia (greater than 1.5 D) may have improved fusional control of the exotropia after optical correction. Typically, the hyperopic lens correction prescribed is less than the full cycloplegic refraction to preserve some accommodative convergence.

Overcorrecting Minus Lenses
Over-minus eyeglass correction (up to 2.0 D over the patient’s own refractive error) may help to stimulate accommodative convergence and improve the fusional control of the exotropia. This therapy may not be well tolerated visually by some young patients. Although overcorrecting minus-lens therapy stimulates excessive accommodation, studies suggest that this does not increase myopia. It is most useful in low-grade myopes or other patients already wearing eyeglasses.

Patching (Antisuppression Therapy)
The exotropic patient usually suppresses the input from the temporal retina of the deviating eye. This will occur even in patients who alternate the eye that is used to fixate. Patching therapy may be beneficial as an antisuppression technique in children with intermittent exotropia. Part-time patching of the dominant eye appears to have sensory and motor benefits in some cases. These benefits include a reduction in the size of the scotoma as measured on haploscopic devices, improved fusional control, or a reduction in the angle of strabismus. Part-time patching may be done on the preferred eye or patching can be alternated in the absence of pre-existing fixation preference. It is most useful as a temporizing measure prior to surgery in infants or young children who are borderline surgical candidates. However, interruption of binocular vision can be counterproductive in patients with precarious control.

Amblyopia Treatment
Patching of the preferred eye to treat exotropic patients who show evidence of amblyopia may improve fusional control, decrease the angle of the exodeviation, or improve the postoperative success rate in patients who require strabismus surgery. Because amblyopia is uncommon in intermittent exotropia, the presence of reduced visual acuity without an obvious etiology (e.g., anisometropia or structural ocular abnormality) should alert the ophthalmologist to consider additional diagnoses.

Prism Therapy
Base-in Fresnel prisms can be used to neutralize the angle of the exodeviation and to maintain fusional control. However, prolonged use of prisms is impractical for most patients and may cause an undesirable reduction in fusional convergence amplitudes. The use of base-in Fresnel prisms to neutralize the exodeviation preoperatively may be helpful in identifying this group of patients who are at risk for a surgical overcorrection. In addition, prism therapy is occasionally useful to alleviate diplopia associated with transient postoperative esotropia.

Convergence Exercises for Convergence Insufficiency
Orthoptic therapy may improve fusional control in patients with intermittent exotropia. Patients with small- to moderate-angle exotropia (i.e., 20 PD or less) may respond to fusional convergence training to build fusional convergence amplitudes. Antisuppression therapy and diplopia awareness are used to stimulate the fusional vergence system. Patients with the convergence insufficiency type of exotropia (exotropia greater at near) and symptoms at near viewing (typically reading) may be good candidates for orthoptic therapy. Near point of convergence exercises on an accommodative target are recommended if the near point of convergence is distant. Convergence exercises with a prism can be beneficial once the near point of convergence improves. To perform convergence exercises, the patient holds a base-out prism of designated power over one eye while viewing an accommodative target at near.
Section II. Exotropia

Extraocular Muscle Surgery

Surgical correction may be considered if the deviation occurs so frequently or is so large as to be unacceptable to the patient or parent/caregiver, or if symptomatology is not relieved by corrective lenses and patching. The observations of the control and size of the deviation under daily life conditions are essential in making the decision to perform extraocular muscle surgery.

In addition to the angle of strabismus, preoperative considerations include refractive error, clinical AC/A ratio, and the age of the patient. An increase in the hyperopic corrective lenses may increase or decrease the measured deviation and influence surgical planning. Measurements of exotropia with new optical correction should be repeated using accommodative targets at near, distance, and if possible, at remote distance.\[A:III\] In patients with evidence of clinically elevated AC/A ratio, the degree of abnormality may be quantified using –2.00 lenses at distance or +2.00 to +3.00 lenses at near. In addition to assessment of ductions and versions, determination of the deviation in lateral gaze positions may be useful, because patients with a high AC/A ratio or lateral-gaze incomitance may be at risk for surgical overcorrection.

Some ophthalmologists believe that surgical correction should be deferred until after the age of 2 years to avoid complications associated with postoperative esotropia. These complications include suppression, amblyopia, and loss of binocular vision, particularly stereoaucity. However, excellent stereoaucity can be found in patients who have undergone early surgery\[155,156\] and it is not known whether surgery before age 2 years affects the prognosis for normal sensory outcome. Alignment before age 7 years, before 5 years of strabismus duration, or while the deviation is intermittent appears to increase the likelihood and quality of stereopsis.\[157\]

Although most surgeons prefer symmetric surgery (e.g., bilateral lateral rectus muscle recession) based on the distance deviation, patients with normal or low AC/A ratios may benefit from unilateral surgery (lateral rectus muscle recession and medial rectus resection).\[158,159\] Esotropia that typically occurs immediately following bilateral lateral rectus recession causes diplopia but increases the likelihood of satisfactory long-term ocular alignment.\[160,161\] While approximately 80% of patients are well aligned 6 months postoperatively after bilateral lateral rectus muscle recession,\[162\] long-term results are less favorable and recurrence is common over time.\[160,163\] Outcomes may be improved with a combination of surgical and nonsurgical therapy.\[164\] Use of adjustable suture techniques (older children and adults) has not been shown to improve outcomes in uncomplicated intermittent exotropia.\[165,166\]

Follow-up Evaluation

Children with exotropia require follow-up evaluations to monitor the magnitude and frequency of the deviation, visual acuity, and binocularity (see Table 3). Young children with constant or poorly controlled exotropia are at risk for developing amblyopia. Postoperative esotropia is associated with amblyopia and may precipitate loss of stereoaucity. The frequency of the follow-up evaluations is based on the age of the child, the ability to obtain an accurate visual acuity, and the control of the deviation. Yearly examinations are appropriate until visual maturity is reached, but they may be reduced in frequency thereafter if the strabismus has been stable.\[A:III\]

Follow-up evaluation includes interval history, tolerance to treatment (if any), and routine examination and testing of ocular motility.

<table>
<thead>
<tr>
<th>Age</th>
<th>Interval</th>
</tr>
</thead>
<tbody>
<tr>
<td>0 - 1 year</td>
<td>3 - 6 months</td>
</tr>
<tr>
<td>1 - 5 years</td>
<td>6 - 12 months</td>
</tr>
<tr>
<td>5 years</td>
<td>12 - 24 months</td>
</tr>
</tbody>
</table>

**Note:** More frequent visits may be necessary if patching therapy is being administered, or if there is a recent deterioration of alignment.
PROVIDER

Certain diagnostic procedures may be delegated to appropriately trained and supervised auxiliary personnel. The interpretation of results, diagnosis, and management of disease, including surgical correction and follow-up, require the high degree of medical and surgical training, clinical judgment, and experience of the ophthalmologist. Consultation with or referral to a pediatric ophthalmologist or general ophthalmologist with expertise in the diagnosis and treatment of strabismus may be desirable for cases in which the diagnosis, etiology, or management plan is unclear, or the exotropia appears unresponsive to treatment.

Diagnostic evaluation and treatment by an orthoptist can be a useful adjunct to the ophthalmologic care of patients with exotropia. The orthoptist's initial and subsequent assessments of visual function as well as monitoring of amblyopia and its treatment may supplement the ophthalmologist's management. The orthoptist's participation in the patient's care process may include assessing binocular function (suppression, diplopia, or fusion); determining the accommodative component and evaluating the usefulness of bifocals; using prisms diagnostically and/or therapeutically pre- and postoperatively; and monitoring amblyopia.

COUNSELING AND REFERRAL

The ophthalmologist should discuss the findings of the evaluation with the patient, if appropriate, as well as the parent/caregiver. The ophthalmologist should explain the disorder and recruit the family in a collaborative approach to therapy. Parents/caregivers of pediatric patients who understand the diagnosis and rationale for treatment are more likely to adhere to treatment recommendations.
SECTION I. ESOTROPIA

DIAGNOSIS

The purpose of the comprehensive strabismus evaluation is to make the diagnosis, establish baseline status, and determine appropriate initial therapy. The possibility of restrictive, paralytic, or other neurologic causes (especially head trauma or increased intracranial pressure) for the strabismus should be considered. Because binocular vision can degrade rapidly in young children, resulting in suppression and anomalous retinal correspondence, early diagnosis and treatment are essential.\textsuperscript{1,3}

The examination of a patient who has childhood-onset strabismus includes all components of the comprehensive pediatric or adult ophthalmic evaluation in addition to the sensory, motor, refractive, and accommodative functions.\textsuperscript{4,5}

History

Although a thorough history generally includes the following items, the exact composition varies with the patient's particular problems and needs.

- Demographic data,\textsuperscript{[A:III]} including identification of parent/caregiver, and patient's gender and date of birth
- Documentation of identity and relationship of historian\textsuperscript{[B:III]}
- The identity of other pertinent health care providers\textsuperscript{[A:III]}
- The chief complaint and reason for the eye evaluation,\textsuperscript{[A:III]} including date of onset and frequency of the ocular misalignment; which eye is deviated and in what direction; and the presence or absence of diplopia, squinting, or other visual symptoms. Review of photographs of the patient may be helpful.
- Ocular history,\textsuperscript{[A:III]} including other eye problems, injuries, diseases, surgery, and treatments (including eyeglasses and/or amblyopia therapy)
- Systemic history; birth weight; prenatal and perinatal history that may be pertinent (e.g. alcohol, drug, and tobacco use during pregnancy); past hospitalizations and operations; general health and development\textsuperscript{[A:III]}
- Pertinent review of systems,\textsuperscript{[B:III]} including history of head trauma and relevant systemic diseases
- Family and social history,\textsuperscript{[A:III]} including eye conditions (strabismus, amblyopia, type of eyeglasses and history of wear, extraocular muscle surgery or other eye surgery, and genetic diseases)
- Current medications and allergies\textsuperscript{[A:III]}

Examination

The comprehensive strabismus examination should include the following elements:

- Assessment of fixation pattern and visual acuity in each eye\textsuperscript{[A:III]}
- Ocular alignment and motility at distance and near\textsuperscript{[A:III]}
- Extraocular muscle function (ductions and versions including incomitance, such as A and V patterns)\textsuperscript{[A:III]}
- Detection of nystagmus\textsuperscript{[A:III]}
- Sensory testing\textsuperscript{[A:III]}
- Cycloplegic retinoscopy/refraction\textsuperscript{[A:III]}
- Funduscopic examination\textsuperscript{[A:III]}

Documentation of the child’s level of cooperation with the examination can be useful in interpreting the results and in making comparisons between examinations.
MANAGEMENT

All forms of esotropia should be considered for treatment.\[^{A:III}\] Ocular alignment should be established as soon as possible, especially in young children, to maximize binocularity,\[^{6,7}\] prevent or facilitate treatment of amblyopia,\[^{8,9}\] and normalize appearance.\[^{A:III}\] In almost all cases, clinically important refractive errors should be corrected.\[^{A:III}\] Amblyopia treatment is usually started before surgery, because this may reduce the angle of strabismus\[^{10}\] or increase the likelihood of good postoperative binocularity.\[^{7,11}\] [\^A:III\]

Choice of Therapy

The following treatment modalities are used alone or in combination as required to achieve the therapeutic goal:

- Correction of refractive errors\[^{12}\] [\^A:I\]
- Bifocals\[^{13}\] [\^A:II\]
- Prism therapy\[^{14,15}\] [\^A:II\]
- Amblyopia treatment\[^{11}\] [\^A:III\]
- Extraocular muscle surgery\[^{16}\] [\^A:III\]

Treatment plans are formulated in consultation with the patient and parent/caregiver. The plans should be responsive to their expectations and preferences,\[^{A:III}\] including the family’s/caregiver’s perception of the existing alignment, which may differ from the ophthalmologist’s, and what they hope to achieve with treatment. It is important that the family/caregiver and ophthalmologist agree on the goals of treatment before surgery is performed.

Follow-up Evaluation

Even when initial treatment results in good ocular alignment, follow-up is essential, since the child remains at high risk for developing amblyopia, losing binocular vision, and having a recurrence of strabismus. Until visual maturity is reached, periodic evaluations are necessary.\[^{17}\] [\^A:II\] During the teenage years, and if the examination has been stable, follow-up evaluations are appropriate every 1 to 2 years thereafter.\[^{18}\] [\^A:I\] New or changing findings may indicate the need for more frequent follow-up examinations.

COUNSELING AND REFERRAL

Childhood esotropia is a long-term problem that requires commitment from the parent/caregiver and ophthalmologist to achieve the best possible outcome.

The ophthalmologist should discuss the findings of the evaluation with the patient, when appropriate, as well as with the parent/caregiver. The ophthalmologist should explain the disorder and recruit the family in a collaborative approach to therapy.\[^{A:III}\] Parents/caregivers of pediatric patients who understand the diagnosis and rationale for treatment are more likely to adhere to treatment recommendations.\[^{19,20}\]
SECTION II. EXOTROPIA

DIAGNOSIS
The purpose of the initial comprehensive strabismus evaluation is to make the diagnosis, establish baseline status, inform the family/caregiver, and determine therapy. The possibility of restrictive, paralytic, or other neurologic causes (especially head trauma or increased intracranial pressure) for the strabismus should be considered.

The examination of a patient who has childhood-onset strabismus includes all components of the comprehensive pediatric or adult ophthalmic evaluation in addition to the sensory, motor, refractive, and accommodative functions.4,5

History
Although a thorough history generally includes the following items, the exact composition varies with the patient's particular problems and needs.

- Demographic data, including identification of parent/caregiver, and patient's gender and date of birth
- Documentation of identity and relationship of historian
- The identity of other pertinent health care providers
- The chief complaint and reason for the eye evaluation, including date of onset and frequency of the ocular misalignment; which eye is deviated and in what direction; and the presence or absence of diplopia, squinting, or other visual symptoms. Review of photographs of the patient may be helpful.
- Ocular history, including other eye problems, injuries, diseases, surgery, and treatments (including eyeglasses and/or amblyopia therapy)
- Systemic history; birth weight; prenatal and perinatal history that may be pertinent (e.g. alcohol, drug, and tobacco use during pregnancy); past hospitalizations and operations; general health and development
- Pertinent review of systems, including history of head trauma and relevant systemic diseases
- Family and social history, including eye conditions (strabismus, amblyopia, type of eyeglasses and history of wear, extraocular muscle surgery or other eye surgery, and genetic diseases)
- Current medications and allergies

Examination
The comprehensive strabismus examination should include the following elements:

- Assessment of fixation pattern and visual acuity in each eye
- Ocular alignment and motility at distance and near
- Extraocular muscle function (ductions and versions including incomitance, such as A and V patterns)
- Detection of nystagmus
- Sensory testing
- Cycloplegic retinoscopy/refraction
- Funduscopic examination

 Documentation of the child’s level of cooperation with the examination can be useful in interpreting the results and in making comparisons between examinations.

MANAGEMENT
All forms of exotropia should be considered for treatment. In most cases, ocular alignment should be re-established as soon as possible, especially in young children, if the deviation is manifest a large percentage of the time. However, the optimal modes of therapy for exotropia, the long-term benefit of early surgical correction, and the superiority of bilateral versus unilateral surgery are not well established. Amblyopia is uncommon in patients with intermittent exotropia, but it should be treated if present.
Appendix I. Summary of Major Recommendations for Care

Choice of Therapy

The following treatment modalities may be used alone or in combination as required to achieve the therapeutic goal:

- Correcting refractive errors\textsuperscript{[A:III]}
- Overcorrecting minus lenses\textsuperscript{[A:III]}
- Patching (antisuppression therapy)\textsuperscript{[A:III]}
- Amblyopia treatment\textsuperscript{[A:III]}
- Prism therapy\textsuperscript{[A:III]}
- Convergence exercises for convergence insufficiency\textsuperscript{[A:III]}
- Extraocular muscle surgery\textsuperscript{[A:III]}

Follow-up Evaluation

Children with exotropia require follow-up evaluations to monitor the magnitude and frequency of the deviation, visual acuity, and binocularity. The frequency of the follow-up evaluations is based on the age of the child, the ability to obtain an accurate visual acuity, and the control of the deviation. Yearly examinations are appropriate until visual maturity is reached, but they may be reduced in frequency thereafter if the strabismus has been stable.\textsuperscript{[A:III]}

Follow-up evaluation includes interval history, tolerance to treatment (if any), and routine examination and testing of ocular motility.

COUNSELING AND REFERRAL

The ophthalmologist should discuss the findings of the evaluation with the patient, if appropriate, as well as the parent/caregiver. The ophthalmologist should explain the disorder and recruit the family in a collaborative approach to therapy.\textsuperscript{[A:III]} Parents/caregivers of pediatric patients who understand the diagnosis and rationale for treatment are more likely to adhere to treatment recommendations.\textsuperscript{[9,20]}

REFERENCES

Appendix I. Summary of Major Recommendations for Care


RELATED ACADEMY MATERIALS

**Basic and Clinical Science Course**
- Pediatric Ophthalmology and Strabismus (Section 6, 2007-2008)

**LEO Clinical Update Course on CD-ROM**
- Pediatric Ophthalmology and Strabismus (2003)

**Patient Education**
- Eye Safety for Children brochure (2005)
- Personal-Eyes Printable™ Patient Handouts on CD-ROM (some handouts available in Spanish) (2007)
- Pseudostrabismus brochure (2006)
- Strabismus brochure (2005)

To order any of these materials, please call the Academy’s Customer Service number, 866.561.8558 (U.S. only) or 415.561.8540 or visit [http://www.aao.org/store](http://www.aao.org/store).

REFERENCES


156. Saunders RA, Trivedi RH. Sensory results after lateral rectus muscle recession for intermittent exotropia operated prior to two years of age. J AAPOS. In press.


