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Esotropia and Exotropia Preferred Practice Pattern®

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PEDIATRIC OPHTHALMOLOGY/STRABISMUS PREFERRED PRACTICE PATTERN DEVELOPMENT PROCESS AND PARTICIPANTS

The **Pediatric Ophthalmology/Strabismus Preferred Practice Pattern Panel** members wrote the Esotropia and Exotropia Preferred Practice Pattern guidelines (PPP). The PPP Panel members discussed and reviewed successive drafts of the document, meeting in person twice and conducting other review by e-mail discussion, to develop a consensus over the final version of the document.

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The **Preferred Practice Patterns Committee** members reviewed and discussed the document during a meeting in June 2022. The document was edited in response to the discussion and comments.

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The Esotropia and Exotropia PPP was then sent for review to additional internal and external groups and individuals in July 2022. All those returning comments were required to provide disclosure of relevant relationships with industry to have their comments considered (indicated with an asterisk below). Members of the Pediatric Ophthalmology/Strabismus Preferred Practice Pattern Panel reviewed and discussed these comments and determined revisions to the document.

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In compliance with the Council of Medical Specialty Societies' Code for Interactions with Companies (available at <https://cmss.org/code-for-interactions-with-companies/>), relevant relationships with industry are listed. The Academy has Relationship with Industry Procedures to comply with the Code (available at www.aao.org/about-preferred-practice-patterns). A majority (100%) of the members of the Pediatric Ophthalmology/Strabismus Preferred Practice Pattern Panel 2021-2022 had no financial relationship to disclose.

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The disclosures of relevant relationships to industry of other reviewers of the document from January to October 2022 are available online at www.aao.org/ppp.

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OBJECTIVES OF PREFERRED PRACTICE PATTERN GUIDELINES

As a service to its members and the public, the American Academy of Ophthalmology has developed a series of Preferred Practice Pattern guidelines that **identify characteristics and components of quality eye care**. Appendix 1 describes the core criteria 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.

These documents 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 PPPs 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.

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 U.S. Food and Drug Administration (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 ensure 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 Pattern guidelines are reviewed by their parent panel annually or earlier if developments warrant and updated accordingly. To ensure that all PPPs are current, each is valid for 5 years from the approved by date unless superseded by a revision. Preferred Practice Pattern guidelines are funded by the Academy without commercial support. Authors and reviewers of PPPs are volunteers and do not receive any financial compensation for their contributions to the documents. The PPPs are externally reviewed by experts and stakeholders, including consumer representatives, before publication. The PPPs are developed in compliance with the Council of Medical Specialty Societies' Code for Interactions with Companies. The Academy has Relationship with Industry Procedures (available at www.aao.org/about-preferred-practice-patterns) to comply with the Code.

Appendix 2 contains the International Statistical Classification of Diseases and Related Health Problems (ICD) codes for the disease entities that this PPP covers. The intended users of the Esotropia and Exotropia PPP are ophthalmologists.

METHODS AND KEY TO RATINGS

Preferred Practice Pattern guidelines should be clinically relevant and specific enough to provide useful information to practitioners. Where evidence exists to support a recommendation for care, the recommendation should be given an explicit rating that shows the strength of evidence. To accomplish these aims, methods from the Scottish Intercollegiate Guideline Network¹ (SIGN) and the Grading of Recommendations Assessment, Development and Evaluation² (GRADE) group are used. GRADE is a systematic approach to grading the strength of the total body of evidence that is available to support recommendations on a specific clinical management issue. Organizations that have adopted GRADE include SIGN, the World Health Organization, the Agency for Healthcare Research and Quality, and the American College of Physicians.³

- ◆ All studies used to form a recommendation for care are graded for strength of evidence individually, and that grade is listed with the study citation.
- ◆ To rate individual studies, a scale based on SIGN¹ is used. The definitions and levels of evidence to rate individual studies are as follows:

I++	High-quality meta-analyses, systematic reviews of randomized controlled trials (RCTs), or RCTs with a very low risk of bias
I+	Well-conducted meta-analyses, systematic reviews of RCTs, or RCTs with a low risk of bias
I-	Meta-analyses, systematic reviews of RCTs, or RCTs with a high risk of bias
II++	High-quality systematic reviews of case-control or cohort studies High-quality case-control or cohort studies with a very low risk of confounding or bias and a high probability that the relationship is causal
II+	Well-conducted case-control or cohort studies with a low risk of confounding or bias and a moderate probability that the relationship is causal
II-	Case-control or cohort studies with a high risk of confounding or bias and a significant risk that the relationship is not causal
III	Nonanalytic studies (e.g., case reports, case series)

- ◆ Recommendations for care are formed based on the body of the evidence. The body of evidence quality ratings are defined by GRADE² as follows:

Good quality	Further research is very unlikely to change our confidence in the estimate of effect
Moderate quality	Further research is likely to have an important impact on our confidence in the estimate of effect and may change the estimate
Insufficient quality	Further research is very likely to have an important impact on our confidence in the estimate of effect and is likely to change the estimate Any estimate of effect is very uncertain

- ◆ Key recommendations for care are defined by GRADE² as follows:

Strong recommendation	Used when the desirable effects of an intervention clearly outweigh the undesirable effects or clearly do not
Discretionary recommendation	Used when the trade-offs are less certain—either because of low-quality evidence or because evidence suggests that desirable and undesirable effects are closely balanced

- ◆ The Highlighted Findings and Recommendations for Care section lists points determined by the PPP Panel to be of particular importance to vision and quality of life outcomes.
- ◆ All recommendations for care in this PPP were rated using the system described above. Ratings are embedded throughout the PPP main text in italics.
- ◆ Literature searches to update the PPP were undertaken in March 2021 and May 2022 in the PubMed database. Complete details of the literature searches are available on www.aao.org/ppp.

HIGHLIGHTED FINDINGS & RECOMMENDATIONS FOR CARE

Strabismus in children under 4 months of age sometimes resolves without treatment, particularly if the deviation is intermittent, variable, or measures less than 40 prism diopters.

Repeat cycloplegic refraction is indicated when esotropia does not respond to an initial prescription for hyperopia or when esotropia recurs after surgery.

Acquired esotropia should be evaluated and treated promptly.

Young children with intermittent exotropia and good fusional control can be followed without surgery because there is a low rate of deterioration to constant exotropia or reduced stereopsis.

Indications for surgery in intermittent exotropia include a progression to constant or nearly constant deviation, reduced stereopsis, and/or a negative effect on social interactions.

Unilateral recess-resect and bilateral lateral rectus recessions are both effective initial surgical procedures for the treatment of intermittent exotropia.

Convergence insufficiency occurs in children and adults, and symptoms with near viewing can often be improved using vergence exercises.

Simultaneous prism and cover testing measures the manifest angle of strabismus, and prism and alternate cover testing measures the total angle of misalignment. Both inform the ophthalmologist's decisions regarding management and surgical indications.

SECTION I. ESOTROPIA

INTRODUCTION

DISEASE DEFINITION

Strabismus describes any binocular misalignment. The most common types are esotropia (inward deviation) and exotropia (outward deviation). Esotropia is a convergent misalignment of the visual axes. The scope of this section is limited to the nonparalytic, nonrestrictive form of the disorder with 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 esotropia
- ◆ Acquired esotropia
 - ◆ Accommodative esotropia
 - Normal accommodative convergence/accommodation (AC/A) ratio
 - High AC/A ratio
 - ◆ Partially accommodative esotropia
 - ◆ Nonaccommodative esotropia
- ◆ Sensory esotropia
- ◆ Other (e.g., Duane syndrome, congenital fibrosis syndrome)

Infantile Esotropia

Infantile esotropia presents before the age 6 of months.⁴ Intermittent esotropia during the first 3 months of life⁵⁻¹¹ may occur and does not necessarily predict the development of constant strabismus. The presence of esotropia is confirmed by the cover-uncover test or Hirschberg light reflect test in younger children, and it is generally measured using the prism and alternate cover test or Krimsky test. Children with infantile esotropia are at risk for amblyopia, although the presence of cross-fixation may diminish this risk prior to surgical correction. Characteristics of infantile esotropia include the following:

- ◆ Onset before the age of 6 months without spontaneous resolution
- ◆ Nonaccommodative or partially accommodative etiology
- ◆ A constant angle of deviation that may increase with time¹²
- ◆ Frequent cross-fixation with the fixing eye in adduction

Additional features that may not be present at the time of diagnosis include latent or manifest latent nystagmus (see Detection of Nystagmus section under Examination), dissociated vertical deviation, oblique muscle dysfunction, A or V patterns, and optokinetic nystagmus asymmetry for nasal versus temporal pursuit.

Acquired Esotropia

Acquired forms of esotropia typically develop after age 6 months and may be accommodative, partially accommodative, or nonaccommodative. These children are at risk for amblyopia.¹³

Accommodative Esotropia

Characteristics of accommodative esotropia include the following:

- ◆ An accommodative component that is usually associated with hyperopia
- ◆ Typical onset between the ages of 1 and 4 years,¹⁴ with an average age of onset of approximately 2 years.⁴ However, it may appear in infancy,^{4, 15, 16} in older children,¹⁷ or develop after surgical correction of infantile esotropia^{18, 19}

- ◆ Development that may be precipitated by illness, fever, or minor trauma
- ◆ Binocular visual function that may be normal prior to the onset of deviation²⁰

The etiology is related to excessive convergence in a child who usually has bilateral hyperopia, and correcting it eliminates the esotropia (accommodative esotropia). Sometimes correction of the hyperopia results in normal alignment at distance fixation but a persistent esotropia at near (accommodative esotropia with a high AC/A ratio). Less frequently, children have normal alignment at distance fixation with no significant hyperopia, but they develop a constant or intermittent esotropia with near fixation (accommodative esotropia with a high AC/A ratio).

Partially Accommodative Esotropia

Children with acquired partially accommodative esotropia experience some improvement of their esotropia when they wear corrective lenses for hyperopia. Partially accommodative esotropia can present with normal or high AC/A.

Nonaccommodative Esotropia

Children with nonaccommodative esotropia have an acquired esotropia that is approximately equal in amount at distance and near fixation and have either no significant refractive error or no improvement in the angle of esotropia with correction of refractive error. If onset is acute and other neurological signs and symptoms are present, patients should be evaluated and treated promptly. Neuroimaging should be considered.

Adults may also develop an acute acquired comitant esotropia. This is particularly common in adults who are myopic.^{21, 22}

Sensory Esotropia

Sensory esotropia is associated with unilateral or bilateral vision loss on a structural basis.

Other

Consecutive esotropia occurs in some children with exotropia after surgery or, less commonly, spontaneously.

Other conditions that are associated with esotropia include Duane syndrome, congenital fibrosis syndrome, craniofacial abnormalities, and ocular myasthenia gravis.

Pseudoesotropia is not true strabismus but is caused either by certain facial features or a disparity between the visual and pupillary axes of the eyes (negative angle kappa).

PATIENT POPULATION

Patients with childhood onset of esotropia.

CLINICAL OBJECTIVES

- ◆ Identify children at risk for esotropia
- ◆ Detect esotropia
- ◆ Diagnose and treat amblyopia that may cause, or be caused by, esotropia (see Amblyopia PPP²³)
- ◆ Educate the patient and/or family caregiver, as appropriate
- ◆ Inform the patient's other health care providers of the diagnosis and treatment plan
- ◆ Treat the esotropia (align the visual axes) to promote and maintain binocular vision (fusion, stereopsis), prevent amblyopia or facilitate its treatment, and restore normal appearance
- ◆ Maximize quality of life by optimizing binocular alignment and visual acuity
- ◆ Monitor vision and binocular alignment, and modify treatment as appropriate

BACKGROUND

PREVALENCE AND RISK FACTORS

Prevalence estimates of strabismus range from 0.8% to 6.8% in different populations.²⁴⁻³⁹ In the United States, esotropia and exotropia have similar prevalence rates, whereas in Ireland esotropia has been reported five times more frequently than exotropia, and in Australia esotropia has been reported to be twice as frequent as exotropia.^{31, 34, 40} In Hong Kong, Singapore, Japan, and China, however, exotropia is more frequent than esotropia.^{27, 33, 36, 39, 41} Children with esotropia are at risk for amblyopia.^{42, 43}

Children are at higher risk to develop strabismus if anisometropia and/or hyperopia are present, and they are at a greater risk to develop esotropia as hyperopia increases.^{40, 44, 45} Other at-risk groups include children who are neurodevelopmentally impaired,⁴⁶⁻⁴⁹ are born prematurely,^{50, 51} have low birth weight,^{52, 53} have low Apgar scores,⁵⁴ have craniofacial or chromosomal anomalies,⁵⁴⁻⁵⁷ are exposed to smoking³⁶ or alcohol in utero,⁵⁸ or have a family history of strabismus.^{7, 59-61}

The prevalence of esotropia in children increases with age (e.g., higher prevalence at 4 to 6 years compared with 6 to 11 months), moderate anisometropia, and moderate amounts of hyperopia.^{32, 40} In some families, a Mendelian inheritance pattern has been observed.⁶² A large study of monozygotic and dizygotic twins found evidence of heritability for esodeviation, whereas no such association was found for exodeviation.⁶³ The incidence of strabismus is related to premature birth⁶⁴ and perinatal morbidity, genetic disorders, and detrimental prenatal environmental influences, such as substance abuse⁶⁵ and smoking.^{36, 54, 58, 66-68} One study looked at the effect of gestational age and birth weight in premature infants and found that a birth weight under 2000 grams conferred a large increase in risk of strabismus.⁶⁴

NATURAL HISTORY

Infantile esotropia, characterized as a constant esodeviation presenting before 6 months of age, is unlikely to resolve. However, some children in this age group who have esotropia that is intermittent or variable, or that measures less than 40 prism diopters, may have resolution of their esotropia by age 1 year.^{11, 69, 70} Because children with intermittent esotropia have normal alignment at least part of the time, the risk of abnormal binocularity is reduced.

Acquired esotropia is more frequent than infantile esotropia⁷¹ and usually presents between the ages of 1 and 4 years.⁴ Onset of accommodative esotropia as early as 2 months of age has been reported.^{4, 15, 16, 50} Children with very early onset accommodative esotropia are more likely to decompensate to partially accommodative esotropia and require extraocular muscle surgery despite correction of their refractive error with eyeglasses.^{50, 72} Accommodative forms of esotropia may begin as an intermittent deviation associated with fatigue, illness, or near viewing. Because younger children lose binocular vision rapidly, it is advisable to correct the hyperopic refractive error as quickly as possible.⁷³

RATIONALE FOR TREATMENT

The potential benefits of treatment for esotropia include promoting binocular vision and improving visual function in each eye.⁷⁴⁻⁷⁷ If binocularity is achieved, the number of surgical procedures over a lifetime and overall cost to society may be reduced.^{78, 79} Fusion and stereopsis are necessary for some careers and may be useful in sports such as baseball and activities such as needlepoint and watching 3D movies.⁸⁰⁻⁸³ The appearance of crossed eyes may reduce employment opportunities because of stigma and bias.⁸³⁻⁸⁵ In addition, binocular alignment is important for the development of a positive self-image and enhances social interactions by normalizing appearance as well as eye contact.^{80, 82, 83, 86-89} In one study, children aged 5 years and older expressed a negative feeling about dolls that had been altered to be esotropic or exotropic.⁹⁰ In another study, elementary school teachers rated personal characteristics of children with esotropia and exotropia more negatively than orthotropic children.⁸⁷ In a sample of children enrolled in the Multi-ethnic Pediatric Eye Disease Study, strabismus was associated with a decreased general health-related quality of life in preschool children, based on the parents' proxy reporting.⁹¹

CARE PROCESS

PATIENT OUTCOME CRITERIA

- ◆ Binocular motor alignment
- ◆ Binocular sensory status (fusion and stereopsis)
- ◆ Binocular and monocular visual acuity including binocular summation and inhibition^{92, 93}

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 strabismus should be considered and addressed promptly. Because binocular vision can degrade rapidly in young children, resulting in suppression and anomalous retinal correspondence, early diagnosis and treatment are essential.^{72, 73, 94}

The examination of a patient who has childhood-onset strabismus includes all elements of the comprehensive ophthalmic examination in addition to sensory, motor, refractive, and accommodative testing.^{95, 96}

History

Although a thorough history generally includes the following items, the details depend on the patient's particular problems and needs.

- ◆ Demographic data, including sex, date of birth, and identity of parent/caregiver
- ◆ Documentation of identity and relationship of historian
- ◆ 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, closing one eye, or other visual symptoms. Review of photographs and/or videos 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, gestational age, pertinent prenatal and perinatal history (e.g., alcohol, drug, and tobacco use during pregnancy), past hospitalizations and operations, and general health and development
- ◆ Pertinent review of systems, including history of head trauma and relevant systemic diseases
- ◆ Current and recently adjusted medications and allergies
- ◆ Family history, including eye conditions (strabismus, amblyopia, type of eyeglasses and history of wear, extraocular muscle surgery or other eye surgery, and genetic diseases)
- ◆ Social history (e.g., grade level in school, learning difficulties, behavior problems, or issues with social interactions)

Examination

When possible, the comprehensive strabismus examination should include the following elements:

- ◆ Visual acuity and verification of eyeglass prescription with a lensometer
- ◆ Binocular alignment at distance and near in primary gaze, up and down gaze, and horizontal gaze positions, if possible. If eyeglasses are worn, alignment testing should be performed with correction; alignment testing without correction may also be appropriate in some circumstances.
- ◆ Extraocular muscle function (ductions and versions, including incomitance such as found in some A and V patterns)
- ◆ General facial examination to note any pronounced dysmorphology or asymmetry
- ◆ Detection of latent or manifest nystagmus

- ◆ Assessment of head posture
- ◆ Sensory testing, including fusion and stereoacuity
- ◆ Cycloplegic retinoscopy/refraction
- ◆ Funduscopy examination
- ◆ Additional testing

Documenting the child's level of cooperation with the examination can be useful in interpreting the results and in making comparisons among examinations over time.

For details on visual acuity, assessment of fixation pattern, and visual field testing, refer to the Pediatric Eye Evaluations PPP, Section II. Comprehensive Ophthalmic Examination.⁹⁵

Binocular Alignment and Motility

Binocular 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 alignment. The method of measuring the angle of esotropia and the presence or absence of refractive correction should be documented. If the patient is unable to participate in more sophisticated testing, the angle may be estimated using the corneal light-reflection test with or without prisms or by estimating the amount of eye movement required to refixate with alternate cover testing without prism. The prism and alternate cover test measures the total deviation and, as such, is used to quantify the amount of surgery required.⁹⁷ The simultaneous prism-and-cover test measures the manifest deviation and provides useful information for patients with fusional vergences, where the alignment under binocular viewing conditions is better than during alternate cover testing (e.g., monofixation syndrome). The simultaneous prism-and-cover test is used by many surgeons as a means of determining if strabismus surgery is indicated.

Extraocular Muscle Function

The examiner should evaluate versions (binocular motility) and ductions (monocular motility) and note any limitation, overaction, or incomitance (change in the angle of strabismus in different gaze positions). When versions are limited, full abduction on monocular duction testing can distinguish the child with infantile or accommodative esotropia from a child with paretic or restrictive esotropia, or esotropic Duane syndrome. Monocular occlusion and oculocephalic rotations (the "doll's-head maneuver" or vestibulo-ocular reflex) are particularly valuable in infants and young children and often reveal clinically normal ductions that may not otherwise be seen. Oblique muscle dysfunction, A or V patterns, and/or dissociated vertical or horizontal deviations should be documented. Diseases associated with paresis, paralysis, or restriction of the extraocular muscles are outside the scope of this PPP.

Detection of Nystagmus

Nystagmus in the patient with esotropia may be manifest, latent, or manifest-latent. Nystagmus is more common in patients with earlier-onset strabismus than in those with later-onset strabismus. Manifest nystagmus is present constantly and may be horizontal, vertical, and/or torsional. It is typically symmetrical, and it may vary in magnitude, speed, and wave form, depending on the direction of gaze and other specific viewing conditions. Latent nystagmus (also known as occlusion nystagmus) is conjugate, and it is characterized by horizontal jerk oscillations of the eyes that are seen under monocular viewing conditions. The nystagmus is described as latent because it is seen when one eye is occluded. It is the only form of nystagmus that reverses direction with a change in fixation. Latent nystagmus is characterized by a slow nasal drift of the fixating eye, followed by saccadic refixation. Manifest-latent (also known as fusion maldevelopment) nystagmus has an identical waveform as latent nystagmus but is evident under binocular viewing conditions, and its amplitude increases with monocular occlusion. Children with manifest-latent nystagmus often present with a head turn and hold the fixating eye in adduction. For this reason, documentation of preferred head posture is critical for surgical planning in patients with strabismus and/or nystagmus.

Although esotropia and nystagmus often coexist in infantile esotropia, it must be distinguished from nystagmus blockage syndrome in which children with infantile esotropia use excessive convergence to damp the amplitude of nystagmus. In these children, the magnitude of the esotropia seems to increase with prism neutralization of the deviation.

Sensory Testing

Sensory testing should be performed before motor testing since motor testing can disrupt ocular alignment. When feasible, the child's binocular sensory status should be assessed using Worth 4-Dot Testing and stereoacuity tests. Reliable data may be difficult to obtain in young children.⁹⁸ In the older strabismic (especially esotropic) patient, more detailed sensory testing is occasionally useful, especially if there is a history of diplopia. Sensory testing should be done before a patch or occluder dissociates the child's binocular status. An orthoptic evaluation, which could include Bagolini lenses, after-image testing, and synoptophore testing, may further define the sensorimotor status of the child.

Stereopsis occurs when the two slightly disparate images from the two eyes are cortically integrated. Many tests are available to determine stereopsis, including the Preschool Randot test, the Randot Test, the Random-Dot E Test, the TNO Test, the Frisby Test, the Stereo Fly Test, and the Lang Stereopsis Test.

The Worth 4-Dot Test assesses central fusion at distance and peripheral fusion at near. The patient wears the red-green eyeglasses and looks at a target with four lights (two green, one red, and one white). If the patient sees four lights, it indicates fusion. Two or three lights indicate monocular suppression, and five lights seen simultaneously indicate diplopia. Some patients with alternating monocular suppression may report five lights, though not all five are seen at once.

Cycloplegic Retinoscopy/Refraction

Determination of refractive error is important in the diagnosis and treatment of amblyopia and/or strabismus. Patients should undergo cycloplegic refraction by retinoscopy or autorefractometry with subjective refinement when necessary.^{99, 100} (*I+, Good, Strong*) Dynamic retinoscopy done prior to cycloplegia provides a rapid assessment of accommodation and may be helpful in evaluating a child with asthenopia who has high hyperopia or with accommodative insufficiency.^{101,102} Using this technique, the examiner evaluates the change in the retinoscopic reflex from a "with" motion toward neutrality when the patient shifts fixation from distance to a small target on the retinoscope.

Adequate cycloplegia is necessary for accurate retinoscopy in children because of their increased accommodative tone compared with adults. For details of cycloplegic eye drops, refer to the Pediatric Eye Evaluation PPP, Section II. Comprehensive Ophthalmic Examination.⁹⁵

Funduscopy Examination

Retinal or optic nerve abnormalities may be present in children with esotropia, in some cases producing sensory strabismus. Particular attention should be paid to the optic nerve for signs of swelling, pallor, or congenital anomalies. In addition, nasal or temporal displacement of the macula may give rise to pseudostrabismus (the appearance of strabismus when there is no shift by alternate cover testing in the presence of good fixation). 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 child with aligned eyes or mask the strabismus in a child with esotropia.¹⁰⁰ A negative angle kappa is seen less frequently and is usually associated with high myopia.

Additional Testing

Forced duction, force generation, and/or saccadic velocity testing 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 when the child is anesthetized. Detection of mechanical restriction may influence the surgical plan. Monocular and binocular optokinetic nystagmus testing for nasal-temporal pursuit asymmetry can be used to help confirm the diagnosis of infantile esotropia.

MANAGEMENT

Prevention

There is consensus that early detection and prompt management of strabismus and potential amblyogenic factors improve long-term visual and sensorimotor outcomes.

For children without esotropia, the threshold of hyperopia that requires treatment has not been established, but correction of hyperopia may reduce the risk of developing accommodative esotropia and/or amblyopia.¹⁰³⁻¹⁰⁶ (Refer to Table 3 in the Pediatric Eye Evaluations PPP, Section II. Comprehensive Ophthalmic Examination, for guidelines for correcting hyperopia in children.⁹⁵) For children with esotropia, the threshold for prescribing hyperopic eyeglasses is lower than for children without esotropia. For example, eyeglasses are generally prescribed for hyperopia of +1.00 D or more. For hyperopic patients, anisometropia is a risk factor for the development of accommodative esotropia.²⁰

Choice of Therapy

Treatment should be considered for all forms of esotropia, and binocular alignment should be established as soon as possible, especially in young children, to maximize binocular potential,^{75, 107} to prevent or facilitate treatment of amblyopia,^{43, 108} and to restore normal appearance. Significant refractive errors should be corrected. Amblyopia treatment is usually started before surgery because it may alter the angle of strabismus¹⁰⁹ and/or increase the likelihood of good postoperative binocularity.^{107, 110}

There is evidence that early surgical correction improves sensory outcomes for infantile esotropia, probably because the duration of constant esotropia is minimized.^{74, 75, 79, 111-113} However, there is no evidence that early surgery compared with later surgery improves motor outcomes.¹¹⁴ One study reported a higher success rate and a lower reoperation rate for patients with infantile esotropia treated with bilateral medial rectus recessions compared with unilateral strabismus surgery.¹¹⁵

Treatment for esotropia may include the following elements:

- ◆ Correction of refractive errors¹¹
- ◆ Bifocal eyeglasses¹¹⁶
- ◆ Prism therapy^{117, 118}
- ◆ Extraocular muscle surgery¹¹⁹
- ◆ Botulinum toxin injection^{120, 121} (*I+*, *Moderate*, *Discretionary*)
- ◆ Other pharmacologic agents

Treatment plans are formulated in consultation with the parent/caregiver and patient, if appropriate. The plans should be responsive to the preferences and expectations of the parent/caregiver and patient. The plans should account for the parent's/caregiver's and patient's perception of the existing alignment. If surgery is performed, it is important that the family/caregiver and ophthalmologist agree beforehand on the goals of treatment. For patients for whom the potential for binocularity is poor, surgery to restore normal appearance may be an appropriate treatment.

Correction of Refractive Errors

Correction of significant refractive errors should be the initial treatment for children with esotropia.^{46, 115} Eyeglasses are generally prescribed for +1.00 D or more of hyperopia in a child with new-onset esotropia. (Refer to Table 3 in the Pediatric Eye Evaluations PPP, Section II. Comprehensive Ophthalmic Examination, for guidelines for correcting refractive errors in children.⁹⁵) For patients with accommodative esotropia, realignment by cycloplegia-determined eyeglasses or contact lenses alone is successful in most cases.^{50, 122}

In general, a greater degree of hyperopia indicates a higher likelihood that the refractive error is an important etiologic factor of the esotropia.

The aim of treatment is to correct hyperopia sufficiently to restore alignment, and in most cases a prescription is given to correct the full refractive error as determined after cycloplegia. A manifest noncycloplegic refraction may be required to optimize visual acuity and binocular alignment in older children because correction of the full cycloplegic refractive error may blur their distance vision.

Improved alignment after prescribing eyeglasses may take several weeks. If the esotropia persists, the cycloplegic refraction should be repeated before considering surgery because additional hyperopic refractive error may be uncovered. A repeat refraction should also be performed for those children initially well aligned in hyperopic eyeglasses who develop recurrent esotropia. Cycloplegia may be prescribed temporarily to facilitate compliance of eyeglass wear. Poor motor and sensory outcomes are likely if eyeglass compliance is poor.¹²³ In older children, gradual reduction of the hyperopic correction can be attempted if the deviation is controlled. The effect of such reductions in the hyperopic correction can be simulated in the office setting by placing minus lenses over the eyeglasses to ensure that binocular alignment is optimized while maintaining best-corrected visual acuity.¹²⁴

In general, eyeglasses to control an esodeviation are well tolerated by children, especially when there is visual improvement. Accurately fitting and properly adjusting the eyeglasses facilitates their acceptance. Head straps or flexible single-piece frames may be useful in infants and young children; cable temples and spring hinges are also helpful. Impact-resistant lenses provide greater safety; they are preferable for children, especially if they are amblyopic, and these lenses are often mandated by state law.

Bifocals

An esodeviation greater for near than for distant targets is found in some cases. Convergence excess is defined clinically as a near esodeviation 10 prism diopters D or greater than the distance deviation (high clinical AC/A ratio) with the use of full hyperopic correction. Bifocal treatment can be considered in patients with potential for sensory fusion who maintain essentially aligned eyes at distance but have a manifest esotropia at near while wearing their full hyperopic correction. If successful, bifocals may be necessary on a long-term basis to maintain binocular alignment for viewing near targets. An excellent initial response is associated with a lower likelihood that the bifocals can be withdrawn later without recurrence of the esotropia.¹¹⁶ Further, sensory outcomes may not be improved in patients treated with bifocals for high AC/A esotropia.^{125, 126}

Bifocals should be a flat-top (D-segment) type or an executive type, with the top of the bifocal bisecting the pupil in primary gaze in preschool children and a few millimeters lower in older children. The minimum strength of the bifocal can sometimes be estimated by office testing in trial frames or can be empirically prescribed +2.50 to +3.00 D and later reduced as tolerated. Reductions can be made later as part of a routine eyeglass change. Progressive bifocals offer cosmetic advantages and are often preferred by older children who have adapted well to standard bifocals. The transition zone should be placed several millimeters higher than the standard adult fitting.¹²⁷

Disadvantages of bifocals include a socially disfavored appearance and potential rejection of the glasses by the child. Some clinicians avoid bifocals because they believe that alignment at distance is sufficient to protect binocular vision.^{126, 128} 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^{129, 130} and possibly eliminate the need for bifocal wear without producing consecutive exotropia at distance.¹³¹⁻¹³⁴

Prism Therapy

Prisms are rarely useful in infantile esotropia, in part because the angle of deviation is usually too large to correct with prisms alone. In some patients with acquired esotropia less than 30 prism diopters who have diplopia, prism therapy may be beneficial in promoting

binocular vision.¹³⁵ Fresnel plastic prisms may also be used for preoperative prismatic adaptation to establish the full angle on which to base extraocular muscle surgery.¹¹⁸ The Prism Adaptation Study investigated the role of preoperative membrane (Fresnel) prisms to determine the maximum angle of the strabismus for surgical planning and to estimate fusional potential. Rates of surgical success, defined as a horizontal deviation of 8 prism diopters or less (measured with the simultaneous prism and cover test at distance fixation), were highest (90%) among those participants who responded to prisms (i.e., showed evidence of sensory fusion) and underwent extraocular muscle surgery for the adapted (larger) angle of esotropia.^{117, 118} However, because prism-adapted patients received greater amounts of surgery on average, it is possible that increasing surgical dosage for patients with potential for fusion without prism adaptation would have produced similar results. Fresnel prisms can cause blurred vision that some children find objectionable, which can cause poor compliance with eyeglasses. In addition, Fresnel prism use requires re-evaluation (additional office visits), and it may be unacceptable to children not otherwise wearing eyeglasses. For these reasons, prism adaptation is used selectively.

Amblyopia Treatment

Amblyopia treatment is typically initiated before surgical treatment of strabismus. The esotropia may increase or decrease with the treatment of amblyopia¹²⁶ (see Amblyopia PPP²³). 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.¹¹⁰

Extraocular Muscle Surgery

Children with esotropia should undergo surgical correction if eyeglasses and amblyopia management are insufficient to align the eyes,¹¹⁹ and strabismus surgery should be performed only when more conservative methods have failed or are unlikely to be of benefit. Surgery is rarely justified if the primary objective is to eliminate eyeglasses. Except for acquired symptomatic deviations, small-angle deviations of less than 12 prism diopters at distance or near are not usually considered for surgery.

Although some binocular vision and stereopsis can be restored after surgical alignment in many patients with infantile esotropia,^{76, 136} achieving high-grade stereopsis is rare.^{74, 75, 77} In contrast, the quality of stereopsis appears to be improved by prompt surgical realignment in patients with decompensated nonaccommodative or partially accommodative esotropia.^{75, 119, 137}

Most patients with infantile esotropia receive surgical intervention during childhood. One study reported that esotropia is most likely to recur during the first year after surgery, but the risk of developing a consecutive exotropia steadily increases for at least 10 years.¹³⁸ Achieving binocular alignment early in life (before age 2 years) to within 10 prism diopters of orthotropia increases the likelihood of achieving binocularity^{74, 75, 111-113} and may decrease the risk for the development of dissociated vertical deviation.¹³⁹

Whether or not there is surgical realignment of infantile esotropia, many affected children subsequently develop other motility problems, such as latent nystagmus, dissociated strabismus, and inferior oblique muscle overaction.^{140, 141} The presence of amblyopia¹¹⁰ or nystagmus¹⁴² is associated with an increased rate of requiring reoperation. In one study, infantile esotropia recurred postoperatively on an accommodative basis in 50% of patients and correlated with the magnitude of the hyperopia.⁷⁹

Extraocular muscle surgery is usually performed for the maximum distance angle of deviation when the individual is wearing full hyperopic correction; however, some surgeons use the maximum near deviation. For patients with a distance-near disparity (high AC/A ratio [i.e., 10 prism diopters or greater¹²⁹]), bilateral medial rectus recession usually reduces the AC/A ratio,¹⁴³ decreasing the need for bifocals by reducing the esodeviation at near fixation.¹³⁰ The higher the preoperative AC/A ratio, the greater the chance for postoperative normalization (i.e., restoring a more normal pattern of accommodative vergence)¹²⁹ and improving binocularity.¹³⁰ Prism adaptation for the near angle,¹⁴⁴ augmentation of the recession over amounts done with a normal AC/A ratio,¹⁴⁵ or posterior

fixation sutures¹³³ increase the likelihood of a satisfactory alignment and eventual weaning from bifocals.

The amount of surgery and the choice of surgical technique vary (e.g., methods of suture placement in the muscle and sclera, or measurement of recession or resection/plication). Although two-muscle surgery is most frequently performed, three- or four-horizontal-muscle surgery may be required for large-angle deviations.¹⁴⁶ Some clinicians believe that two-muscle surgery is the better initial choice even for large-angle deviations, regardless of magnitude, to reduce the risk of consecutive exotropia.¹⁴⁷ In patients with deviations more than 60 prism diopters, additional extraocular muscle surgery is more often required to achieve ocular alignment.¹⁴⁸

Adjustable sutures have been advocated as an adjunct to strabismus surgery to improve motor outcomes, especially for patients with restrictive disease or for those requiring reoperation. Its utility in children remains unproven.¹⁴⁹ Moreover, the adjustment is challenging to do in younger children who may not cooperate.

Results may be similar with different surgical procedures; one method may be chosen over another on the basis of preoperative diagnosis, angle of deviation at distance and near, 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 unilateral or ipsilateral surgery (single-muscle recession or recession with resection or plication) for patients with irreversible amblyopia or other causes of substantially reduced vision in one eye. 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.

Pharmacological Injection

Chemodenervation by injection of botulinum toxin into one or more extraocular muscles induces a temporary weakness by pharmacologically blocking the neuromuscular junction. Although the mechanism of long-term ocular realignment after chemodenervation in children is unknown, it likely results from contracture of the direct antagonist combined with motor and sensory adaptations that allow restoration of some degree of binocularity.¹⁵⁰⁻¹⁵² As with conventional extraocular muscle surgery, favorable prognostic indicators include good vision in each eye, absence of restricted eye movement, a small to moderate angle of esotropia, and the potential for binocular vision. Such treatment may be an alternative to conventional extraocular muscle surgery in selected patients,¹⁵³ but its value in managing infantile esotropia has not been definitively established.^{121, 154-158} (*I+*, *Moderate*, *Discretionary*) A disadvantage is that repeat injections are frequently needed. Risks of treatment include ptosis, vertical deviations, globe perforation, and tonic pupil.¹⁵⁹ Since treatment is administered by injection, treatment complications relate principally to trauma from the needle or toxin leakage. Careful injection technique and preinjection counseling are essential. Importantly, delayed binocular realignment may be disadvantageous in an infant with a rapidly developing visual system.

Bupivacaine can also be injected into extraocular muscles to strengthen muscles either in combination with chemodenervation of an antagonistic muscle with botulinum toxin or in combination with extraocular muscle surgery.^{160, 161}

Perioperative Care

Preoperative Management

Once a decision has been made to proceed with strabismus repair, preoperative counseling with the patient or parents/caregivers should include a realistic discussion of the goals of surgery, potential benefits of surgery, and risks of surgery and anesthesia. If the patient has any significant systemic risk factors for surgery, a pre-anesthesia evaluation with the primary care physician or specialty physician or anesthesiology service is essential.

Sometimes a tour of the surgical facility by the patient and family can relieve presurgical anxiety, especially for young children.

Postoperative Management

Pain management is usually limited to non-narcotic analgesics. Narcotics are avoided in children, if possible, because of the risk of nausea, vomiting, and dehydration. Antiemetics, such as ondansetron, may be used postoperatively to control nausea. Diet is advanced slowly in the first 24 hours following surgery. Many surgeons use a combination antibiotic-corticosteroid preparation for the first week after surgery, although its effectiveness in reducing the risk of postoperative infection is not proven. Parents should be advised of the risks and signs of postoperative complications, especially orbital cellulitis and slipped or lost muscle.

Follow-up Evaluation

Even when initial treatment results in good binocular alignment, follow-up is essential, because the child remains at high risk for developing amblyopia, losing binocular vision, and having a recurrence of strabismus. Children who are well aligned and do not have amblyopia may be followed every 6 to 12 months. As the child matures, the frequency of follow-up visits can be reduced.¹⁶² New or changing findings may indicate the need for more frequent follow-up examinations.

Children with esotropia should be assessed for hyperopia at least annually and more frequently if visual acuity decreases or the esotropia increases. Detection of uncorrected or undercorrected hyperopia is essential in the child with a recurrence of esotropia after successful initial treatment. Cyclopentolate 1% is effective in most patients for obtaining cycloplegia for refraction. In some patients, more hyperopia may be documented after 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 before concluding that the esotropia has a nonaccommodative component. Atropine 1% may be used to establish adequate cycloplegia when shorter-acting drugs are inadequate.¹⁶³

Recurrence of esotropia or consecutive exotropia that is not responsive to eyeglasses, patching, or medical treatment may indicate the need for repeat strabismus surgery if the magnitude of the strabismus is sufficient.

PROVIDER AND SETTING

Certain eye care services and procedures, including elements of the eye examination, may be delegated to appropriately trained and supervised auxiliary health care personnel under the ophthalmologist's supervision.¹⁶⁴ For cases in which the diagnosis or management is difficult, consultation with or referral to an ophthalmologist who specializes in the diagnosis and treatment of pediatric patients may be desirable. The operating ophthalmologist has the ultimate responsibility for the preoperative assessment and postoperative care of the patient, beginning with the determination of the need for surgery and ending with completion of the postoperative care contingent on medical stability of the patient. Postoperative care responsibilities may be ethically delegated to another nonoperating health care practitioner, whether as part of a co-management arrangement or as a transfer of care, under appropriate circumstances.¹⁶⁵⁻¹⁶⁷

COUNSELING AND REFERRAL

Childhood esotropia is a long-term problem that requires commitment from the patient and/or family/caregiver and the 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 include the family in a collaborative approach to therapy. Parents/caregivers of children who understand the diagnosis and rationale for treatment are more likely to adhere to treatment recommendations.^{168, 169}

SECTION II. EXOTROPIA

INTRODUCTION

DISEASE DEFINITION

Exotropia is a divergent misalignment of the visual axes. The scope of this section is limited to the following forms of exotropia:

- ◆ Infantile exotropia
- ◆ Intermittent exotropia
- ◆ Convergence insufficiency
- ◆ Sensory exotropia
- ◆ Other

Infantile Exotropia

Infantile exotropia appears before 6 months of age and is a constant exotropia that has many characteristics similar to infantile esotropia, including limited binocular potential, oblique muscle dysfunction, latent nystagmus, and dissociated vertical deviation. Neonates frequently have intermittent exotropia during the first 3 to 4 months of life; however, it rarely persists.⁷ Children with neurodevelopmental delay may have constant exotropia from infancy.¹⁷⁰ Children with infantile exotropia are at risk for amblyopia.

Intermittent Exotropia

Childhood-onset exotropia is typically intermittent and usually appears before 3 years of age, but it may be first detected later in childhood. The deviation often becomes manifest with fatigue, visual inattention, or illness when fusional compensatory mechanisms are compromised. The patient may close or cover one eye in bright light. Generally, the image from the deviated eye is suppressed and the patient does not report diplopia. Mild amblyopia occasionally occurs, but severe amblyopia is uncommon with intermittent exotropia.

Convergence Insufficiency

Older children and teenagers with convergence insufficiency typically have intermittent exotropia at near fixation, reduced fusional convergence amplitudes, and a remote near point of convergence. They often report asthenopic symptoms with near work.

Sensory Exotropia

Sensory exotropia is associated with unilateral or bilateral vision loss on a structural basis.

Other

Consecutive exotropia occurs in some children after surgery for esotropia.

Other conditions that are associated with exotropia include Duane syndrome, congenital fibrosis syndrome, craniofacial abnormalities, and ocular myasthenia gravis. Dissociated horizontal deviation is a divergent misalignment of the eyes and typically occurs in patients with a history of infantile esotropia.

Pseudoexotropia is not true strabismus but is caused by a disparity between the visual and anatomic axes of the eyes (positive angle kappa).

PATIENT POPULATION

Patients with childhood onset of exotropia.

CLINICAL OBJECTIVES

- ◆ Identify children at risk for exotropia
- ◆ Detect exotropia
- ◆ Detect and treat amblyopia that may be associated with exotropia (see Amblyopia PPP²³)
- ◆ Educate the patient, as appropriate, about the diagnosis, treatment options, and care plan
- ◆ Inform the patient's other health providers of the diagnosis and treatment plan
- ◆ When indicated, treat the exotropia (align the visual axes to promote and maintain binocular vision [fusion, stereopsis]), prevent amblyopia or facilitate its treatment, and restore normal appearance
- ◆ Maximize quality of life by optimizing binocular alignment and visual acuity¹⁷¹⁻¹⁷³
- ◆ Monitor vision and binocular alignment and modify treatment as appropriate

BACKGROUND

PREVALENCE AND RISK FACTORS

Exotropia occurs in approximately 1% of the population; intermittent exotropia is the most frequently reported type.^{31, 32, 174, 175} A large study of monozygotic and dizygotic twins found evidence of heritability for esodeviation, but no such association was found for exodeviation.⁶³ Exotropia has been associated with prematurity, perinatal morbidity, genetic disorders, detrimental prenatal environmental influences (e.g., maternal substance abuse and smoking), family history of strabismus, female sex, astigmatism, myopia, and anisometropia.^{31, 32, 40, 66, 171-173} One small retrospective population-based cohort study in the United States found that intermittent exotropia was twice as frequent in girls than in boys.¹⁷⁶ Clinic-based studies of children with infantile-onset exotropia found that half had associated ocular or systemic anomalies.^{60, 177}

Reduction or prevention of factors such as prematurity and maternal smoking during pregnancy, as well as diagnosis and treatment of myopia and myopic anisometropia, may reduce the incidence of exotropia.

NATURAL HISTORY

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 fixation, and symptoms. Some studies suggest that many patients who decline surgical correction appear to remain stable or spontaneously improve with observation alone,^{178, 179} but other studies report deterioration during long-term follow-up.¹⁸⁰ Von Noorden followed 51 patients ages 5 to 10 years with intermittent exotropia for an average of 3.5 years and found that an increase in angle size, decrease in fusional control, and/or development of suppression occurred in 75%.¹⁸¹ However, a more recent study of 109 patients followed for an average of 9 years found that there was no trend for worsening or improvement of the size or control of exodeviation angle.¹⁷⁹ In an observation arm of a large clinical trial, 28% of children 12 to 35 months old and 15% between 3 and 10 years old showed deterioration of intermittent exotropia when observed for 3 years.^{182, 183} A small proportion of children with intermittent exotropia eventually develop a constant deviation, which may cause binocular vision to deteriorate in some children.¹⁸⁴

The causes of exotropia are poorly understood. Proposed etiologies for exotropia include excess tonic divergence and mechanical or orbital factors.¹⁸⁵ Severe unilateral or bilateral vision loss may cause exotropia. Typically, unilateral poor vision in early childhood is more often associated with esotropia than with exotropia.

RATIONALE FOR TREATMENT

The potential benefits of treatment for exotropia include promoting binocular vision and normal visual function in each eye, as well as relieving asthenopia and eye fatigue.^{186, 187} Normal binocular alignment promotes a positive self-image.⁸⁸ The appearance of misaligned eyes impairs self-image and social interactions and reduces employment opportunities via bias.^{83-87, 89, 90} In one study, children

aged 5 years and older expressed a negative feeling about dolls that had been altered to be esotropic or exotropic.⁹⁰ In another study, elementary school teachers rated personal characteristics of children with esotropia and exotropia more negatively than orthotropic children.⁸⁷ In a sample of children enrolled in the Multi-ethnic Pediatric Eye Disease Study, strabismus was associated with a decreased general health-related quality of life in preschool children, based on the parents' proxy reporting.⁹¹ After strabismus surgery, adults have reported improved confidence, self-esteem, and interpersonal interactions.⁸⁴ There is evidence that the severity of exotropia negatively affects a child and/or their parent's quality of life,^{188, 189} whereas surgical intervention may have a positive impact on a child's quality of life.^{190-192, 200} Studies have shown improvement in life quality after surgery for intermittent exotropia.^{191, 193}

CARE PROCESS

PATIENT OUTCOME CRITERIA

- ◆ Binocular motor alignment
- ◆ Binocular sensory status (fusion and stereopsis)
- ◆ Visual acuity in each eye
- ◆ Patient-reported quality of life

DIAGNOSIS

The purpose of the initial comprehensive strabismus evaluation is to confirm the diagnosis, establish baseline status, inform the patient and/or family/caregiver, and determine therapy. Secondary causes for the strabismus should be considered, including restrictive and paralytic deviations caused by head trauma or increased intracranial pressure.

The examination of a patient who has childhood-onset strabismus includes all elements of the comprehensive ophthalmic examination in addition to sensory, motor, refractive, and accommodative testing.^{95, 96} The esotropia section of this document contains details of the comprehensive strabismus evaluation, and examination elements specific to exotropia are discussed in this section on exotropia.

History

The medical history should include an estimate of the proportion of waking time that the eyes appear to be misaligned, whether there is an ability to control the deviation, when the deviation occurs (e.g., when tired, ill, daydreaming, or viewing distant objects), and whether the frequency of the deviation is changing. In addition, it is helpful to ascertain whether one or both eyes are observed to deviate.

Examination

Sensory tests (e.g., stereopsis) should be done before visual acuity and alignment measurements, which may dissociate the eyes by monocular occlusion and cause reduced stereoacuity measurement or interfere with assessment of control of the exodeviation.

The examination includes an assessment of the fusional control of the exodeviation at both distance and near fixation. The deviation is recorded as constant exotropia (XT), intermittent exotropia (X(T)), or exophoria (X). Fusional control can vary substantially from visit to visit or even within the same visit. Various scales have been developed to characterize control of exodeviations.¹⁹⁴⁻¹⁹⁶ Indicators of progression include worsening control, reduction in stereoacuity, and/or development of suppression. Some practitioners augment near stereoacuity tests with an assessment of distance stereoacuity, which may detect reduced distance fusional control.^{197, 198}

Binocular 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 when measuring alignment. The method of measuring the angle of exotropia and the presence or absence of refractive correction should be documented. If the patient is unable to

participate in more sophisticated testing, the angle may be estimated using the corneal light-reflection test with or without prisms or by estimating the amount of eye movement required to refixate with alternate cover testing without prism. The prism and alternate cover test measures the total deviation and, as such, is used to quantify the amount of surgery, if required.⁹⁷ The simultaneous prism-and-cover test measures the manifest deviation and provides useful information for patients with fusional vergence, where the alignment under binocular viewing conditions is better than during alternate cover testing.

MANAGEMENT

All forms of exotropia should be monitored, and some require treatment. Young children with intermittent exotropia and good fusional control should be followed without surgery.¹⁹⁹ Deviations that are present most or all of the time often require treatment. However, the optimal therapy for exotropia, the long-term benefit of early surgical correction, and the relative merits of bilateral versus unilateral surgery are not well established.²⁰⁰ Amblyopia is uncommon in patients with intermittent exotropia, but, if present, should be treated.

Choice of Therapy

Current treatment practices include the following:

- ◆ Correction of refractive errors
- ◆ Stimulating accommodative convergence (overcorrection of myopia or undercorrection of hyperopia)
- ◆ Patching (antisuppression) therapy
- ◆ Treating amblyopia
- ◆ Prism therapy
- ◆ Convergence exercises
- ◆ Extraocular muscle surgery
- ◆ Botulinum toxin injection¹²¹ (*I+*, *Moderate*, *Discretionary*)

Correction of Refractive Errors

In the setting of an exodeviation, corrective lenses should be prescribed for any clinically significant refractive error that causes reduced vision in one or both eyes.¹⁹⁹ Improved retinal-image clarity often improves the control of the exotropia.²⁰¹ Such refractive errors include myopia, high hyperopia, astigmatism, and significant anisometropia. In one study, myopia was found in more than 90% of exotropic patients by 20 years of age.²⁰² 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 of the exodeviation. If hyperopic correction is necessary, the amount prescribed is the least amount needed to promote good vision while still promoting accommodative convergence to control the exodeviation. Such correction may be the full cycloplegic refraction,²⁰¹ but it is often less than the full amount.

Stimulating Accommodative Convergence

If fusional control of intermittent exotropia is suboptimal despite providing image clarity with refractive correction, it may be improved in many cases by increasing myopic correction in myopes, reducing hyperopic correction in hyperopes, or prescribing myopic correction in ametropes. Some patients, in particular older patients and adults, may not tolerate this therapy because of visual discomfort or decreased visual acuity. Some studies suggest that overcorrecting minus-lens therapy stimulates accommodation without increasing myopia.^{203, 204} However, in a recent study of children age 3 to 10 with intermittent exotropia, this form of treatment appears to be effective, but the result is often not maintained and is associated with a myopic shift.²⁰⁵

Patching Therapy

In some cases, part-time patching (e.g., 2 to 6 hours daily) may improve fusional control^{206, 207} and/or reduce the angle of strabismus, particularly in the 3- to 10-year age group. It may

be done on the preferred eye or, in the absence of a fixation preference, prescribed to alternate between eyes.

Part-time patching, either unilateral or alternating, has been utilized as a treatment for intermittent exotropia. Deterioration of exotropia is uncommon. Two randomized clinical trials have determined that with or without patching, deterioration is uncommon, and part-time patching may slightly lower the probability of deterioration.^{208, 209}

Treating Amblyopia

In children with exotropia, treatment for amblyopia²³ may improve fusional control, decrease the angle of the exodeviation, and thereby improve the postoperative success rate in those requiring strabismus surgery. Because amblyopia is uncommon in intermittent exotropia,¹⁷⁴ the presence of reduced visual acuity without an obvious etiology (e.g., anisometropia or ocular structural abnormality) should alert the ophthalmologist to consider additional diagnoses, such as subtle optic nerve or retinal abnormalities.

Prism Therapy

Patients with intermittent exotropia do not typically have diplopia, so prisms are not generally prescribed. However, some patients with intermittent exotropia have convergence insufficiency. In these cases, base-out prism can be used during convergence exercises (see the following subsection). In cases of symptomatic convergence insufficiency exotropia that is refractory to exercises, base-in prism can be included in eyeglasses to improve comfort while reading, although one study found this treatment was no better than placebo in children.²¹⁰

Convergence Exercises for Convergence Insufficiency

Orthoptic therapy may improve fusional control in children or adults with convergence insufficiency and with small- to moderate-angle exodeviations (i.e., 20 prism diopters or less), with the goal of strengthening fusional convergence amplitudes.^{211, 212} Children and adults with the convergence insufficiency type of exotropia (exotropia greater at near) and asthenopic symptoms with near viewing (typically reading) may be good candidates for orthoptic therapy. Near point of convergence exercises on an accommodative target are useful if the near point of convergence is distant. Convergence exercises with a base-out prism may be beneficial once the near point of convergence improves. Treatment is tapered as symptoms improve, and it may need to be resumed if symptoms recur. Other treatments include computer-based convergence exercises and in-office orthoptics.²¹³⁻²¹⁶

Extraocular Muscle Surgery

Surgical intervention is considered if the exodeviation is constant, if it occurs so frequently or is so large as to be unacceptable to the child or parent/caregiver, or if symptoms are not relieved by corrective lenses and nonsurgical treatment. Observing the control and size of the deviation under daily-life conditions is essential when making the decision to perform extraocular muscle surgery. Other preoperative considerations include age, refractive error, and the accommodative convergence/accommodation (AC/A) ratio. A change in refractive correction may increase or decrease the measured deviation and influence surgical planning. Measurements of exotropia with best optical correction should be repeated using accommodative targets at near, distance, and if possible, at remote distance (e.g., while a patient looks down a hallway or out of a window). Thirty minutes of monocular occlusion (patch test) may help to elicit the full deviation.

If the distance angle exceeds the near angle by at least 10 prism diopters, -2.00 D lenses are placed over the usual refractive correction. If there is a significant decrease in the distance angle, a high AC/A ratio is diagnosed. In these patients, a nonsurgical approach may be warranted because there is a risk of consecutive esotropia with diplopia or asthenopia at near fixation.²¹⁷

The timing of surgery for exotropia depends on the child's neurodevelopmental status and the frequency of the deviation. For constant infantile-onset exotropia, early surgery is indicated to improve sensory outcomes, although normal binocular function is rarely

achieved. When the deviation is intermittent, many ophthalmologists defer surgery in young children with fusion to avoid complications associated with postoperative esotropia. These complications include suppression, amblyopia, and loss of binocular vision, particularly stereoacuity. However, excellent stereoacuity can be found in many patients who have undergone early surgery.^{218, 219} Some studies suggest that earlier surgery (patients 3 to 5 years of age) may actually have superior surgical outcomes compared with older patients.²²⁰⁻²²² Many surgeons elect to wait until the deviation is very frequent or there are significant psychosocial implications.

Surgery consists of bilateral-lateral rectus-muscle recessions or unilateral-lateral rectus-muscle recession and medial rectus-muscle strengthening. Some surgeons prefer bilateral surgery when the distance deviation exceeds the near deviation and unilateral surgery when the near deviation is greater than the distance deviation. When poor vision is present in one eye, unilateral surgery on that eye typically is preferred. Bilateral surgery is preferable when there is an A or V pattern with or without significant oblique overaction. Upshift of both lateral rectus muscles improves a V pattern and downshift improves an A pattern. In the setting of exotropia, small vertical deviations typically do not require vertical-muscle surgery. A single muscle may be operated for a small-angle exotropia.

Although some surgeons prefer symmetric surgery (e.g., bilateral-lateral rectus-muscle recession), with recession amounts based on the distance deviation, excellent results are also obtained from unilateral two-muscle surgery (lateral rectus-muscle recession and medial rectus resection).^{223, 224} In a recent, large clinical trial, 197 children between 3 years old up to 11 years with basic-type intermittent exotropia were randomized to bilateral-lateral rectus recession or unilateral recess-resect. At 6 months and 3 years after surgery there was no significant difference between the two groups in the proportion with suboptimal outcome. Surgeons elected to reoperate by 3 years in nine participants (10%) in the bilateral-lateral rectus recession group and in four participants (5%) in the unilateral recess-resect group.²²⁵ Similar outcomes from the two procedures is supported by recent meta-analyses.^{226, 227} (#227: *I+*, *Good*, *Strong*) (#228: *I+*, *Good*, *Strong*)

Esotropia that occurs immediately following surgery often causes diplopia. Some studies have reported that this overcorrection is usually temporary and may increase the likelihood of satisfactory long-term binocular alignment,^{228, 229} but another study reported a variable and unpredictable outcome following early overcorrection.²³⁰ The duration of follow-up likely influences motor outcomes.^{230, 231} When a consecutive esotropia persists for several weeks, placement of temporary membrane prisms, that are slowly reduced in power, can be helpful. When unsuccessful, additional surgery is often required for the consecutive esotropia.

Although approximately 80% of patients have good alignment 6 months postoperatively after bilateral-lateral rectus-muscle recession,²³² long-term results are less favorable and recurrence is common over time.^{228, 233} Outcomes may be improved with a combination of surgical and nonsurgical (orthoptic/occlusion) therapy during management of a child with exotropia.²³⁴ Use of an adjustable suture technique (in older children and adults) has not been shown to improve outcomes in uncomplicated intermittent exotropia.^{149, 235}

Botulinum Toxin Injection

Chemodenervation by injection of botulinum toxin into one or more extraocular muscles has been used as initial, secondary, and adjunctive treatment for exotropia. In a randomized study (n = 30; 20 with exotropia) of adjustable suture muscle surgery or chemodenervation by injection of botulinum toxin for adults with horizontal, nonaccommodative ocular misalignment, botulinum toxin treatment was less successful (29% vs. 77%) than surgery.²³⁶ There is insufficient evidence to make treatment recommendations for botulinum toxin treatment for exotropia.¹²¹ (*I+*, *Moderate*, *Discretionary*)

Perioperative Care

Refer to the subsection Perioperative Care in Section I. Esotropia of this PPP.

Follow-up Evaluation

Children with exotropia require follow-up evaluations to monitor the magnitude and frequency of the deviation, visual acuity, and binocularity. Young children with constant or poorly controlled exotropia or with postoperative consecutive esotropia are at risk for developing amblyopia, and they should be followed more frequently. Postoperative consecutive esotropia may also precipitate loss of stereoacuity. Prescribing base-out prism in eyeglasses is occasionally useful to alleviate diplopia associated with postoperative esotropia. There is a moderate risk of recurrent exotropia. The frequency of 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. Children with good fusional control of intermittent exotropia and without amblyopia are typically examined every 6 to 12 months. By age 7 to 10 years, the frequency of ophthalmological examinations may be reduced.

Follow-up evaluation includes frequency of any deviation, adherence to treatment plan (if any), ocular motility assessment, and an update of refractive correction, if needed.

PROVIDER AND SETTING

Certain eye care services and procedures, including elements of the eye examination, may be delegated to appropriately trained and supervised auxiliary health care personnel under the ophthalmologist's supervision.¹⁶⁴ For cases in which the diagnosis or management is difficult, consultation with or referral to an ophthalmologist who specializes in the diagnosis and treatment of pediatric patients may be desirable. The operating ophthalmologist has the ultimate responsibility for the preoperative assessment and postoperative care of the patient, beginning with the determination of the need for surgery and ending with completion of the postoperative care contingent on medical stability of the patient. Postoperative care responsibilities may be ethically delegated to another nonoperating health care practitioner, whether as part of a co-management arrangement or as a transfer of care, under appropriate circumstances.¹⁶⁵⁻¹⁶⁷

COUNSELING AND REFERRAL

Childhood exotropia is a long-term problem that requires commitment from the patient and/or family/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 include 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.^{168, 169}

SOCIOECONOMIC CONSIDERATIONS FOR STRABISMUS

There is consensus that timely and appropriate eye care can significantly improve children's quality of life and can reduce the burden of eye disease. Timely treatment of strabismus relies on early diagnosis.⁹⁵ Therefore, early and regular vision screenings are important to detect this and other conditions.

Many children 3 to 6 years of age or younger in the United States have never undergone a vision screening.^{237, 238} Children in low-income families, in uninsured families, and in racial and ethnic minority groups may fare worse.²³⁷⁻²⁴⁰ Studies indicate that Latino and African American children and children living below the federal poverty level are more likely to be uninsured and receive fewer and less intensive services relative to white children.^{239, 241-243} There is evidence that these race/ethnicity disparities are reflected in eye care services as well as in other health services.²⁴¹ It is still unclear whether these disparities in eye care services are due to underdiagnosis and undertreatment of certain conditions in minority children, a lower prevalence of treatable eye conditions in certain populations, racial/ethnic differences in access to care or in preferences for treatment, or a combination of these factors.²³⁹

Barriers to eye care extend beyond inadequate screening and diagnosis. Few screening programs ensure access to eye examinations and treatment for children who fail screening. In one large study, only about half of children who fail vision screening are seen by eye care providers in follow-up.²⁴⁴ Barriers to care may

include inadequate information, lack of access to care, and/or financial or insurance coverage difficulties.^{245, 246} Children with diagnosed eye conditions require greater use of medical services than children without such conditions, and their families incur higher out-of-pocket expenditures.²⁴¹ In keeping with other measures of disparities in the provision of health services, non-Hispanic whites and families of higher socioeconomic status may be more likely to obtain follow-up eye care.²⁴⁵

Children with untreated strabismus begin to suffer socially before the age of 6 years,⁹⁰ experience negative perceptions by their teachers,⁸⁷ and, in general, have a reduced psychosocial quality of life.^{247, 248} In addition, later employment prospects can be affected by strabismus.²⁴⁹ In one small (n = 140) time trade-off utility study, the majority of adults with strabismus would trade a portion of their life expectancy in return for being rid of their strabismus.²⁵⁰ Treatment studies indicate that appropriate management of strabismus can improve both functional and psychosocial outcomes, even into adulthood.²⁵¹⁻²⁵³

State legislatures have attempted to close the gap in children's eye care by mandating some form of vision screening for children.²⁵⁴ Legislative efforts have focused primarily on early detection of vision problems in young children. Leaders in these efforts have stressed the importance of funding mechanisms to support such programs, specifically advocating reimbursement of vision screening in the primary care setting as a pathway to success.²⁵⁴

Optimal eye and vision care for children involves an organized program of vision screening in the primary care and community settings. It also includes referral for comprehensive ophthalmic examinations when indicated and provision of refractive aids as needed. There remains a pressing need for studies to assess the impact of these interventions over time and across diverse populations.²⁵⁵

APPENDIX 1. QUALITY OF OPHTHALMIC CARE CORE CRITERIA

*Providing quality care
is the physician's foremost ethical obligation, and is
the basis of public trust in physicians.
AMA Board of Trustees, 1986*

Quality ophthalmic care is provided in a manner and with the skill that is consistent with the best interests of the patient. The discussion that follows characterizes the core elements of such care.

The ophthalmologist is first and foremost a physician. As such, the ophthalmologist demonstrates compassion and concern for the individual, and utilizes the science and art of medicine to help alleviate patient fear and suffering. The ophthalmologist strives to develop and maintain clinical skills at the highest feasible level, consistent with the needs of patients, through training and continuing education. The ophthalmologist evaluates those skills and medical knowledge in relation to the needs of the patient and responds accordingly. The ophthalmologist also ensures that needy patients receive necessary care directly or through referral to appropriate persons and facilities that will provide such care, and he or she supports activities that promote health and prevent disease and disability.

The ophthalmologist recognizes that disease places patients in a disadvantaged, dependent state. The ophthalmologist respects the dignity and integrity of his or her patients and does not exploit their vulnerability.

Quality ophthalmic care has the following optimal attributes, among others.

- ◆ The essence of quality care is a meaningful partnership relationship between patient and physician. The ophthalmologist strives to communicate effectively with his or her patients, listening carefully to their needs and concerns. In turn, the ophthalmologist educates his or her patients about the nature and prognosis of their condition and about proper and appropriate therapeutic modalities. This is to ensure their meaningful participation (appropriate to their unique physical, intellectual, and emotional state) in decisions affecting their management and care, to improve their motivation and compliance with the agreed plan of treatment, and to help alleviate their fears and concerns.
- ◆ The ophthalmologist uses his or her best judgment in choosing and timing appropriate diagnostic and therapeutic modalities as well as the frequency of evaluation and follow-up, with due regard to the urgency and nature of the patient's condition and unique needs and desires.
- ◆ The ophthalmologist carries out only those procedures for which he or she is adequately trained, experienced, and competent, or, when necessary, is assisted by someone who is, depending on the urgency of the problem and availability and accessibility of alternative providers.
- ◆ Patients are assured access to, and continuity of, needed and appropriate ophthalmic care, which can be described as follows.
 - ◆ The ophthalmologist treats patients with due regard to timeliness, appropriateness, and his or her own ability to provide such care.
 - ◆ The operating ophthalmologist makes adequate provision for appropriate pre- and postoperative patient care.
 - ◆ When the ophthalmologist is unavailable for his or her patient, he or she provides appropriate alternative ophthalmic care, with adequate mechanisms for informing patients of the existence of such care and procedures for obtaining it.
 - ◆ The ophthalmologist refers patients to other ophthalmologists and eye care providers based on the timeliness and appropriateness of such referral, the patient's needs, the competence and qualifications of the person to whom the referral is made, and access and availability.
 - ◆ The ophthalmologist seeks appropriate consultation with due regard to the nature of the ocular or other medical or surgical problem. Consultants are suggested for their skill, competence, and accessibility. They receive as complete and accurate an accounting of the problem as necessary to provide efficient and effective advice or intervention, and in turn they respond in an adequate and timely manner.

- ◆ The ophthalmologist maintains complete and accurate medical records.
- ◆ On appropriate request, the ophthalmologist provides a full and accurate rendering of the patient's records in his or her possession.
- ◆ The ophthalmologist reviews the results of consultations and laboratory tests in a timely and effective manner and takes appropriate actions.
- ◆ The ophthalmologist and those who assist in providing care identify themselves and their profession.
- ◆ For patients whose conditions fail to respond to treatment and for whom further treatment is unavailable, the ophthalmologist provides proper professional support, counseling, rehabilitative and social services, and referral as appropriate and accessible.
- ◆ Prior to therapeutic or invasive diagnostic procedures, the ophthalmologist becomes appropriately conversant with the patient's condition by collecting pertinent historical information and performing relevant preoperative examinations. Additionally, he or she enables the patient to reach a fully informed decision by providing an accurate and truthful explanation of the diagnosis; the nature, purpose, risks, benefits, and probability of success of the proposed treatment and of alternative treatment; and the risks and benefits of no treatment.
- ◆ The ophthalmologist adopts new technology (e.g., drugs, devices, surgical techniques) in judicious fashion, appropriate to the cost and potential benefit relative to existing alternatives and to its demonstrated safety and efficacy.
- ◆ The ophthalmologist enhances the quality of care he or she provides by periodically reviewing and assessing his or her personal performance in relation to established standards, and by revising or altering his or her practices and techniques appropriately.
- ◆ The ophthalmologist improves ophthalmic care by communicating to colleagues, through appropriate professional channels, knowledge gained through clinical research and practice. This includes alerting colleagues of instances of unusual or unexpected rates of complications and problems related to new drugs, devices, or procedures.
- ◆ The ophthalmologist provides care in suitably staffed and equipped facilities adequate to deal with potential ocular and systemic complications requiring immediate attention.
- ◆ The ophthalmologist also provides ophthalmic care in a manner that is cost-effective without unacceptably compromising accepted standards of quality.

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APPENDIX 2. INTERNATIONAL STATISTICAL CLASSIFICATION OF DISEASES AND RELATED HEALTH PROBLEMS (ICD) CODES

Esotropia, which includes entities with the following ICD-10 classifications:

	ICD-10 CM
Nonaccommodative (unspecified)	H50.00
Accommodative	H50.43
Alternating	H50.05
Alternating with A pattern	H50.06
Alternating with V pattern	H50.07
Alternating with X or Y pattern (with other noncomitancies)	H50.08
Monocular	H50.01–
Monocular with A pattern	H50.02–
Monocular with V pattern	H50.03–
Monocular with X or Y pattern (with other noncomitancies)	H50.04–
Intermittent, alternating	H50.32
Intermittent, monocular	H50.31–
Unspecified	H50.00

CM = Clinical Modification used in the United States; (–) = 1, right eye; 2, left eye.

Additional Information:

- For bilateral sites, the final character of the codes indicates laterality. Esotropia and exotropia do not have bilateral codes. Therefore, if the condition is bilateral, assign separate codes for both the left and right side.
- When the diagnosis code specifies laterality, regardless of which digit it is found in (i.e., 4th digit, 5th digit, or 6th digit), most often you will find:
 - Right is 1
 - Left is 2

Exotropia, which includes entities with the following ICD-10 classifications:

ICD-10 CM	
Alternating	H50.15
Alternating with A pattern	H50.16
Alternating with specified noncomitancy not elsewhere classifiable (includes alphabetical patterns)	H50.18
Alternating with V pattern	H50.17
Monocular	H50.11–
Monocular with A pattern	H50.12–
Intermittent unspecified	H50.30
Alternating, intermittent	H50.34
Monocular, intermittent	H50.33–
Unspecified	H50.10

CM = Clinical Modification used in the United States; (–) = 1, right eye; 2, left eye

Additional Information for ICD-10 Codes:

- For bilateral sites, the final character of the codes indicates laterality. Esotropia and exotropia do not have bilateral codes. Therefore, if the condition is bilateral, assign separate codes for both the left and right side.
- When the diagnosis code specifies laterality, regardless of which digit it is found in (i.e., 4th digit, 5th digit, or 6th digit), most often you will find:
 - Right is 1
 - Left is 2

APPENDIX 3. LITERATURE SEARCHES FOR THIS PPP

Literature searches of the PubMed database were conducted on March 2021; the search strategies were as follows. Specific limited update searches were conducted after May 2022. The searches had added filters for randomized controlled trials and systematic reviews and date limiters to capture literature published since 2017. The panel analyzed 566 studies of which 29 were included in the PPP.

Literature searches of the PubMed and Cochrane databases were conducted in May 2022; the search strategies were as follows.

Esotropia: "esotropia"[MeSH Terms] OR esotropia[tiab]

Exotropia: "exotropia"[MeSH Terms] OR exotropia[tiab]

Prevalence – Esotropia: ("esotropia/epidemiology"[MeSH Terms] OR "esotropia/ethnology"[MeSH Terms] OR "esotropia"[MeSH Terms]) AND ("prevalence"[MeSH Terms])

Prevalence – Exotropia: (exotropia/epidemiology[mh] OR exotropia/ethnology[mh] OR exotropia[mh]) AND ("prevalence"[MeSH Terms])

Risk Factors - Esotropia: ("esotropia/epidemiology"[MeSH Terms] OR "esotropia/ethnology"[MeSH Terms] OR "esotropia"[MeSH Terms]) AND ("risk factors"[tiab] OR "risk factors"[mh])

Risk Factors - Exotropia: (exotropia/epidemiology[mh] OR exotropia/ethnology[mh] OR exotropia[mh]) AND ("risk factors"[tiab] OR "risk factors"[mh])

Socioeconomic Esotropia: ("esotropia"[MeSH Terms] OR esotropia[tiab]) AND (socioeconomic factors[mh] OR "socioeconomic factors"[tiab])

Socioeconomic Exotropia: ("exotropia"[MeSH Terms] OR exotropia[tiab]) AND (socioeconomic factors[mh] OR "socioeconomic factors"[tiab])

RELATED ACADEMY MATERIALS

Basic and Clinical Science Course

- Pediatric Ophthalmology and Strabismus (Section 6, 2022-2023)

Clinical Statements – Free download available at <http://one.aao.org/guidelines-browse?filter=clinicalstatement>.

- Amblyopia Is a Medical Condition (2017)
- Adult Strabismus Surgery (2017)

Focal Points

- Adult Strabismus (2016)
- Childhood Vision Screening (2018)

Ophthalmic Technology Assessment - Published in *Ophthalmology*, which is distributed free to Academy members; links to abstracts and full text available at www.aao.org/ota.

- Adjustable Sutures in the Treatment of Strabismus (2022)
- Binocular Treatment of Amblyopia (2019)

- Botulinum Toxin Injection for the Treatment of Strabismus (2018)
- Effectiveness of Laser Refractive Surgery to Address Anisometropic Amblyogenic Refractive Error in Children (2022)

Patient Education Downloadable Handout

- Amblyopia (2022)
- Amblyopia Patching (2022)
- Pseudostrabismus (2022)
- Strabismus Children (2022)

Patient Education Video

- Strabismus Surgery for Children (Pediatrics Patient Education Video Collection)
- Treating Amblyopia (Pediatrics Patient Education Video Collection)

Preferred Practice Pattern® Guidelines - Free download available at www.aao.org/ppp.

- Adult Strabismus (2019)
- Comprehensive Adult Medical Eye Evaluation (2020)
- Esotropia and Exotropia (2022)
- Pediatric Eye Evaluations (2022)

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