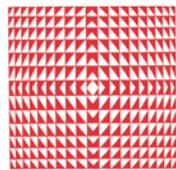


PREFERRED PRACTICE PATTERN®



Esotropia and Exotropia

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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 March 2012. The document was edited in response to the discussion and comments.

Preferred Practice Patterns Committee 2012

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The Esotropia and Exotropia PPP was then sent for review to additional internal and external groups and individuals in June 2012. All those returning comments were required to provide disclosure of relevant relationships with industry to have their comments considered. Members of the Pediatric Ophthalmology/Strabismus PPP Panel reviewed and discussed these comments and determined revisions to the document. The following organizations and individuals returned comments.

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Pediatric Ophthalmology/Strabismus Panel
Basic and Clinical Science Course Subcommittee
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FINANCIAL DISCLOSURES

In compliance with the Council of Medical Specialty Societies' Code for Interactions with Companies (available at www.cmss.org/codeforinteractions.aspx), relevant relationships with industry are listed. The Academy has Relationship with Industry Procedures to comply with the Code (available at <http://one.aao.org/CE/PracticeGuidelines/PPP.aspx>). A majority (87%) of the members of the Pediatric Ophthalmology/Strabismus Preferred Practice Pattern Panel 2011–2012 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 August 2012 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 <http://one.aao.org/CE/PracticeGuidelines/PPP.aspx>) 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 Policy, 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.
- ◆ Literature searches to update the PPP were undertaken in April 2011 in PubMed and the Cochrane Library and updated in March 2012. Complete details of the literature search are available at www.aao.org/ppp.



HIGHLIGHTED FINDINGS & RECOMMENDATIONS FOR CARE

Infantile esotropia and exotropia are associated with an increased risk of amblyopia. (*good evidence*)

Strabismus in children under 4 months of age sometimes resolves, particularly if the deviation is intermittent, or variable, or measures less than 40 prism diopters. (*good evidence*)

Repeat cycloplegic refraction is indicated when esotropia does not respond to the initial prescription of hyperopic refraction or when the esotropia recurs after surgery. (*strong recommendation, moderate evidence*)

Young children with intermittent exotropia and good fusional control can be followed without surgery. (*strong recommendation, moderate evidence*)

In patients with exotropia and a high accommodative convergence to accommodation ratio (AC/A), treatment with eyeglasses is generally preferred over surgery because of the risk of consecutive esotropia and diplopia after surgery. (*discretionary recommendation, moderate evidence*)

Esotropia that persists after exotropia surgery may place the patient at risk for amblyopia, diplopia, and loss of stereoacuity. (*moderate evidence*)

Children with untreated strabismus can have reduced binocular potential and impaired social interactions, and they can be subject to negative perceptions by others, which may affect their psychosocial quality of life. (*good evidence*)

SECTION I. ESOTROPIA



INTRODUCTION

DISEASE DEFINITION

Esotropia is a convergent misalignment of the visual axes. The scope of this section of the document is limited to the nonparalytic, nonrestrictive form of the disease 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.

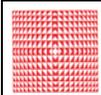
- ◆ Infantile esotropia
- ◆ Acquired esotropia
 - ◆ Accommodative esotropia
 - Accommodative refractive esotropia
 - Accommodative refractive esotropia with a high accommodative convergence to accommodation ratio (AC/A)
 - Accommodative non-refractive esotropia with a high AC/A ratio
 - ◆ Partially accommodative esotropia
 - ◆ Non-accommodative esotropia
- ◆ Other forms of esotropia

Infantile Esotropia

Infantile esotropia presents between the ages of 3 and 6 months.⁴ Intermittent esotropia during the first 3 months of life⁵⁻¹¹ is common and does not necessarily predict the development of constant strabismus. Children with infantile esotropia are at risk for amblyopia. Characteristics of infantile esotropia include the following:

- ◆ Onset before the age of 6 months without spontaneous resolution
- ◆ Nonaccommodative or partially accommodative etiology
- ◆ Constant angle of deviation that may increase with time¹²
- ◆ Frequent cross fixation with the fixing eye held in adduction
- ◆ Abnormal binocular visual function

Features that may not be present at the time of diagnosis include latent nystagmus, dissociated vertical deviation, oblique muscle dysfunction with A or V patterns, and optokinetic nystagmus asymmetry for nasal vs. temporal pursuit.



Infantile esotropia and exotropia are associated with an increased risk of amblyopia.
(*good evidence*)

**Esotropia and Exotropia PPP:
Section I. Esotropia**

Acquired Esotropia

Acquired forms of esotropia typically develop after age 6 months and may be accommodative, partially accommodative, or nonaccommodative in nature. Children with accommodative esotropia are at risk for amblyopia.

Accommodative Esotropia

Characteristics of accommodative esotropia include:

- ◆ An accommodative component that is usually associated with hyperopia
- ◆ Typical onset between the ages of 1 and 8 years with an average age of onset of approximately 2 years⁴; may appear in infancy^{4,13,14} or may reappear as a sequel to surgically corrected infantile esotropia^{15,16}
- ◆ May be precipitated by illness, fever, or minor trauma
- ◆ Binocular visual function that may be normal at the onset of deviation¹⁷

Etiology is usually related to excessive convergence in a child with bilateral hyperopia (usually more than 2.00 diopters [D]), correction of which eliminates the esotropia (accommodative refractive esotropia). Sometimes correction of the hyperopia results in normal alignment at distance fixation but a persistent esotropia at near (accommodative refractive esotropia with a high AC/A ratio). Less frequently, children have normal alignment at distance fixation with no significant hyperopia, but develop a constant or intermittent esotropia with near fixation (accommodative non-refractive esotropia with a high AC/A ratio).

Partially Accommodative Esotropia

Children with acquired partially accommodative esotropia experience a partial improvement of their esotropia when they wear corrective lenses for their hyperopia, but they have a residual esotropia greater than 10 prism diopters at distance and near.

Nonaccommodative Esotropia

Children with nonaccommodative esotropia have an acquired esotropia that is approximately equal in size at distance and near fixation that does not improve with correction of refractive error with eyeglasses or the child has no significant refractive error.

Other Forms of Esotropia

A differential diagnosis of childhood esotropia includes cranial nerve VI palsy, esotropic Duane syndrome, sensory esotropia, restrictive esotropia, consecutive esotropia, and nystagmus blockage esotropia. Discussion of these entities is outside the scope of this PPP.

PATIENT POPULATION

Patients with childhood onset of esotropia.

CLINICAL OBJECTIVES

- ◆ Identify children at risk for esotropia
- ◆ Detect esotropia
- ◆ Detect and treat amblyopia that may cause, or be caused by, esotropia (see Amblyopia PPP¹⁸)
- ◆ Educate the patient and/or family/caregiver, as appropriate, of the diagnosis, treatment options, and care plan
- ◆ Inform the patient's other health providers of the diagnosis and treatment plan
- ◆ Treat the esotropia (align the visual axes) to promote and maintain binocular vision (fusion, stereopsis), prevent or facilitate treatment of amblyopia, and restore normal appearance
- ◆ Maximize quality of life by optimizing binocular alignment and visual acuity
- ◆ Monitor vision and binocular alignment, and modify therapy as appropriate



BACKGROUND

PREVALENCE AND RISK FACTORS

Strabismus describes any binocular misalignment. The most common types are esotropia and exotropia. Prevalence estimates of strabismus range from 1% to 6% in different populations.¹⁹⁻²⁷

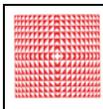
In the United States, esotropia and exotropia have similar prevalence rates, whereas in Ireland esotropia is reported five times more frequently than exotropia, and in Australia esotropia has been reported to be twice as frequent as exotropia.^{26,28} In Hong Kong and Japan, however, exotropia is more frequent than esotropia.^{22,29} Amblyopia can both cause and result from a manifest strabismus.^{4,30} Approximately 50% of children who have strabismus develop amblyopia.^{31,32}

Certain children are at higher risk for developing strabismus, including those with anisometropia and hyperopia, and they are at a greater risk of developing esotropia as hyperopia increases.^{28,33,34} Other at-risk groups include children who are neurodevelopmentally impaired³⁵⁻³⁸; were born prematurely^{39,40}; had low birth weight^{41,42}; had low Apgar scores⁴³; have craniofacial or chromosomal anomalies⁴³⁻⁴⁶; were exposed to alcohol in utero⁴⁷; or have a family history of strabismus.^{7,48-50}

The prevalence of esotropia increases with older age (e.g., higher prevalence at 48 to 72 months compared to 6 to 11 months), moderate anisometropia, and moderate amounts of hyperopia.^{27,28} In some families, a Mendelian inheritance pattern has been observed.⁵¹ The incidence of infantile esotropia is related to premature births and perinatal morbidity, genetic disorders, and detrimental prenatal environmental influences such as substance abuse and smoking.^{43,47,52-54} In the long term, reduction or prevention of those factors could result in a decrease in the incidence of infantile esotropia.

NATURAL HISTORY

Infantile esotropia, characterized as a large-angle, 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 measures less than 40 prism diopters, may have resolution of their esotropia by age 1 year.^{11,55,56} Because children with intermittent esotropia have normal alignment at least part of the time, the risk of abnormal binocularity is reduced.



Strabismus in children under 4 months of age sometimes resolves, particularly if the deviation is intermittent, or variable, or measures less than 40 prism diopters. (*good evidence*)

Acquired esotropia is more frequent than infantile esotropia⁵⁷ and usually presents between the ages of 1 year and 8 years.⁴ Onset as early as 2 months of age has been reported.^{4,14,39} Children with very early onset acquired esotropia are more likely to require extraocular muscle surgery despite correction of their refractive error with eyeglasses.³⁹ Accommodative forms of esotropia may begin as an intermittent deviation associated with fatigue, illness, or near viewing. Because younger children lose binocular vision rapidly, correcting the hyperopic refractive error as quickly as possible is advised.³⁰

RATIONALE FOR TREATMENT

The potential benefits of treatment for esotropia include promoting binocular vision and normal visual function in each eye.⁵⁸⁻⁶¹ If binocularity is achieved, the number of surgical procedures over a lifetime and overall cost to society may be reduced.^{62,63} Fusion and stereopsis are necessary for some careers and may be useful in others as well, such as athletic activities and activities of daily life.⁶⁴⁻⁶⁷ The appearance of crossed eyes may reduce employment opportunities.⁶⁷⁻⁶⁹ 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.^{64,66,67,70-73} In one study, children aged 5 years and older expressed a negative feeling about dolls that had been altered to be esotropic or exotropic.⁷⁴ In another study, elementary school teachers rated personal characteristics of children with esotropia and exotropia more negatively than orthotropic children.⁷¹ In a sample of children enrolled in the Multi-ethnic Pediatric Eye Disease Study, strabismus was associated with decreased general health-related quality of life in preschool children, based on the parents' proxy reporting.⁷⁵



Children with untreated strabismus can have reduced binocular potential and impaired social interactions, and they can be subject to negative perceptions by others, which may affect their psychosocial quality of life. (*good evidence*)



CARE PROCESS

PATIENT OUTCOME CRITERIA

- ◆ Optimal binocular motor alignment
- ◆ Optimal binocular sensory status (fusion and stereopsis)
- ◆ Optimal visual acuity in each eye

DIAGNOSIS

The purpose of the comprehensive strabismus evaluation is to make the diagnosis, establish baseline status, and determine appropriate initial therapy. The possibility of restrictive, paralytic, or other neurologic causes (especially head trauma or increased intracranial pressure) for the strabismus should be considered. Because binocular vision can degrade rapidly in young children, resulting in suppression and anomalous retinal correspondence, early diagnosis and treatment are essential.^{50,76,77}

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.^{78,79}

History

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

- ◆ Demographic data, including gender, date of birth, and identity of parent/caregiver
- ◆ Documentation of identity and relationship of historian
- ◆ The identity of other pertinent health care providers
- ◆ The chief complaint and reason for the eye evaluation, including date of onset and frequency of the ocular misalignment; which eye is deviated and in what direction; and the presence or absence of diplopia, squinting, or other visual symptoms. Review of photographs of the patient may be helpful.
- ◆ Ocular history, including other eye problems, injuries, diseases, surgery, and treatments (including eyeglasses and/or amblyopia therapy)

- ◆ Systemic history, birth weight, gestational age, prenatal and perinatal history that may be pertinent (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 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 in school, learning difficulties, behavior problems, or issues with social interactions)

Examination

The comprehensive strabismus examination should include the following elements:

- ◆ Assessment of fixation pattern and visual acuity in each eye at distance and near
- ◆ Binocular alignment at distance and near in primary gaze and in up and down gaze positions, if possible
- ◆ Extraocular muscle function (ductions and versions, including incomitance such as found in some A and V patterns)
- ◆ Monocular and binocular optokinetic nystagmus testing for nasal-temporal pursuit asymmetry associated with infantile esotropia
- ◆ Detection of latent or manifest nystagmus
- ◆ 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.

Assessment of Fixation Pattern and Visual Acuity

Fixation

Visual acuity measurement of the infant and toddler involves a qualitative assessment of fixation and tracking (following) movements of the eyes. Fixation and following are assessed by drawing the child's attention to the examiner or caregiver's face (infants under 3 months) or to a hand-held light, toy, or other accommodative fixation target and then slowly moving the target. Fixation behavior can be recorded for each eye as "fixes and follows" or "central, steady, and maintained."

Fixation preference can be assessed by observing the vigor with which the child objects to occlusion of one eye relative to the other: children resist covering an eye when the fellow eye has limited vision.⁸⁰⁻⁸² Grading schemes can be used to describe fixation preference. For strabismic patients, fixation pattern is assessed binocularly by determining the length of time that the nonpreferred eye holds fixation. Fixation pattern can be graded by whether the nonpreferred eye will not hold fixation, holds momentarily, holds for a few seconds (or to or through a blink), or by observation of spontaneous alternation of fixation. For children with small-angle strabismus or no strabismus, the induced tropia test is typically done by holding a 10 to 20 prism diopter base-down prism over one eye and then the other eye and noting fixation behavior.⁸²⁻⁸⁴

Qualitative assessment of visual acuity should be replaced with a visual acuity test based on optotypes (letters, numbers, or symbols) as soon as the child can perform this task.

**Esotropia and Exotropia PPP:
Section I. Esotropia**

Visual Acuity

Recognition visual acuity testing, which involves identifying optotypes, including letters, numbers, or symbols, is preferred for assessment of visual acuity to detect amblyopia. The optotypes may be presented on a wall chart, computer screen, or hand-held card. Visual acuity is routinely tested at distance (10 to 20 feet or 3 to 6 meters) and at near (14 to 16 inches or 35 to 40 centimeters). Under ideal circumstances, visual acuity testing conditions should be standardized so that results obtained over a series of visits can be readily compared. High-contrast charts with black optotypes on a white background should be used for standard visual acuity testing.⁸⁵

A child's performance on a visual acuity test will be dependent on the choice of chart and the examiner's skills and rapport with the child. To reduce errors, the environment should be quiet. Younger children may benefit from a pretest on optotypes presented at near, either at the start of testing or in a separate session. Before monocular testing, the examiner should ensure that the child is able to perform the test reliably. Allowing children to match optotypes on the chart to those found on a hand-held card will enhance performance, especially in young, shy, or cognitively impaired children. Visual acuity testing of children with special needs can provide quantitative information about visual impairment and reduce concerns of parents/caregivers about the child's vision.⁸⁶ A shorter testing distance or flip chart can also facilitate testing in younger children.⁸⁷

Visual acuity testing should be performed monocularly and with refractive correction in place. Ideally the fellow eye is covered with an adhesive patch or tape. If such occlusion is not available or tolerated by the child, care must be taken to prevent the child from peeking and using the "covered" eye. Sometimes the child will not allow any monocular occlusion, in which case binocular visual acuity should be measured. Monocular visual acuity testing for patients with nystagmus requires special techniques such as blurring of the fellow eye with plus lenses or a translucent occluder rather than using opaque occlusion. Binocular visual acuity testing can also be performed for these patients to provide additional information about typical visual performance.

The choice and arrangement of optotypes on an eye chart can significantly affect the visual acuity score obtained.⁸⁸⁻⁹⁰ Optotypes should be clear, standardized, of similar characteristics, and should not reflect a cultural bias.⁸⁵ LEA Symbols (Good-Lite Co., Elgin, IL), a set of four symbol optotypes developed for use with young children, are useful because each optotype blurs similarly as the child is presented with smaller symbols, increasing the reliability that individual symbols will be identified.^{88,91} Another method for testing the young child uses a chart containing only the letters H, O, T, and V.^{88,92} Children who cannot name the symbols on the LEA Symbol chart or the letters on the HOTV chart may be able to match them using a hand-held card. Desirable optotypes for older children are LEA numbers and Sloan letters.⁹³ Snellen letters are less desirable because the individual letters are not of equal legibility and the spacing of the letters does not meet World Health Organization standards.^{85,94-96}

Several other symbol charts have serious limitations in testing visual acuity of young children. These include Allen figures,⁹⁷ the Lighthouse chart, and the Kindergarten Eye Chart.⁹⁸ In these charts, the optotypes are not standardized to blur equally and/or the optotypes are presented in a culturally biased or confusing fashion.⁹⁹ The Illiterate or Tumbling E chart is conceptually difficult for young children, leading to high untestability rates.⁹⁸ Appendix 3 lists the details of design of visual acuity testing charts. Some charts meet recommended criteria,⁸⁵ although many do not.



The choice and arrangement of optotypes (letters, numbers, symbols) on an eye chart can significantly affect the visual acuity score obtained. Preferred optotypes are standardized and validated. (*strong recommendation, good evidence*)

The arrangement of optotypes on the chart is important.⁹⁹ Optotypes should be presented in a full line of five whenever possible. Children should correctly identify the majority of optotypes on a line to “pass” the line. A similar number of optotypes on each line with equal spacing is preferred. In the setting of amblyopia, visual acuity testing with single optotypes is likely to overestimate acuity¹⁰⁰⁻¹⁰² because of the crowding phenomenon. In amblyopia, it is easier to discriminate an isolated optotype than one presented in a line of optotypes. Therefore, a more accurate assessment of monocular visual acuity is obtained in amblyopia with the presentation of a line of optotypes. Optotypes should not be covered or masked as the examiner points to each successive symbol in order to preserve the crowding effect of adjacent optotypes. If a single optotype must be used to facilitate visual acuity testing for some children, the optotype should be surrounded (crowded) by bars placed above, below, and on either side of the optotype to account for the crowding phenomenon and not overestimate visual acuity.¹⁰³⁻¹⁰⁵



Vision testing with single optotypes is likely to overestimate visual acuity in a patient with amblyopia. A more accurate assessment of monocular visual acuity is obtained with the presentation of a line of optotypes or a single optotype with crowding bars that surround (or crowd) the optotype being identified. (*strong recommendation, good evidence*)

The Teller Acuity Cards (Stereo Optical Co., Inc., Chicago, IL) are a test of forced preferential looking and can provide a general assessment of resolution visual acuity in young children and how the patient’s acuity compares with normative data, but this method of testing overestimates recognition visual acuity in children with amblyopia.^{106,107}

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 regardless of the technique. 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 to refixate with alternate-cover testing. The prism and alternate-cover test is more accurate and used whenever feasible to quantify binocular misalignment in appropriate gazes and head positions.¹⁰⁸ The simultaneous prism-and-cover test may provide additional useful information for patients with fusional vergences, where the binocular alignment under binocular viewing conditions is better than during alternate-cover testing (e.g., monofixation syndrome).

Extraocular Muscle Function

The examiner should evaluate versions (binocular motility) and ductions (monocular motility) and note any limitation or overaction. Full abduction on monocular duction testing can distinguish the child with infantile or accommodative esotropia from a child with parietic or restrictive esotropia, or esotropic Duane syndrome. Monocular occlusion and oculocephalic rotations (doll’s-head maneuver) are particularly valuable in infants and young children and often reveal clinically normal ductions that may not otherwise be documented. Inferior 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 not included in the scope of this PPP.

Detection of Nystagmus

Nystagmus in the patient with esotropia may be manifest, latent, or manifest-latent. It 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, although it may vary in magnitude, speed, and wave form, depending on the direction of gaze and other specific viewing conditions. Latent nystagmus (sometimes called occlusion nystagmus) is conjugate and is predominantly manifested as horizontal jerk oscillations of the eyes that are produced or exacerbated by monocular viewing. Latent nystagmus is characterized by a slow drift nasally of the fixating eye followed by saccadic refixation. The nystagmus is described as latent because it is typically perceptible or accentuated when one eye is occluded. Manifest-latent 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. Although esotropia and nystagmus often coexist in infantile esotropia, it must be distinguished from nystagmus blockage syndrome in which children with congenital 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

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 younger children.¹⁰⁹ 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 may further define the sensorimotor status of the child.

The Worth 4-dot test can assess peripheral or central fusion. Correct interpretation of the Worth 4-dot test is essential. The patient wears the red-green eyeglasses and looks at a target with four lights (two green, one red, and one white) in a darkened room. If the patient sees four lights it indicates peripheral 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. A small-distance Worth 4-dot target tests foveal fusion and suppression.

Stereopsis occurs when the two images of each eye are cortically integrated secondary to the slight disparity in the images. Many tests are available to determine stereopsis, including the Stereo Fly test, the Randot test, the Random-Dot E test, the TNO test, and the Lang stereopsis test.

Cycloplegic Retinoscopy/Refraction

Determination of refractive errors is important in the diagnosis and treatment of amblyopia or strabismus. Patients should undergo cycloplegic refraction with retinoscopy and subjective refinement when possible.¹¹⁰ Before cycloplegia, dynamic retinoscopy provides a rapid assessment of accommodation and may be helpful in evaluating a child with asthenopia who has high hyperopia or the child with accommodative insufficiency.^{111,112}

Adequate cycloplegia is necessary for accurate retinoscopy in children due to their increased accommodative tone compared with adults. Cyclopentolate hydrochloride is useful because it produces rapid cycloplegia that approximates the effect of topical ophthalmic atropine 1% solution but with a shorter duration of action.¹¹³ Cyclopentolate 1% solution is typically used in term infants over 6 months old. The dose of cyclopentolate should be determined based on the child's weight, iris color, and dilation history. In eyes with heavily pigmented irides, repeating the cycloplegic eyedrops or using adjunctive agents such as phenylephrine hydrochloride 2.5% (has no cycloplegic effect) or tropicamide 0.5% or 1.0% may be necessary to achieve adequate dilation to facilitate retinoscopy. Tropicamide and phenylephrine may be used in combination to

produce adequate dilation size, but this combination may not be strong enough for adequate cycloplegia in children. A single eyedrop combination of cyclopentolate 0.2% and phenylephrine 1% is safe and effective for infants with dark irides.¹¹⁴ In rare cases, topical ophthalmic atropine sulphate 1% solution may be necessary to achieve maximal cycloplegia.¹¹³ The use of topical anesthetic prior to the cycloplegic reduces the stinging of subsequent eyedrops and promotes its penetration into the eye.¹¹⁵ Cycloplegic and dilating agents may be compounded in spray forms that provide similar dilation and cycloplegia with equal or greater patient satisfaction.¹¹⁶⁻¹¹⁸ Short-term side effects of cycloplegic and dilating agents may include hypersensitivity reactions, fever, dry mouth, rapid pulse, nausea, vomiting, flushing, and, rarely, behavioral changes.

Funduscopy Examination

Retinal or optic nerve abnormalities may lead to sensory strabismus. 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). 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.¹¹⁹ A negative angle kappa is seen less frequently and is usually associated with high myopia.

Additional Testing

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

MANAGEMENT

Prevention

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

Although the threshold of hyperopia in children without esotropia that requires treatment has not been established, correction of hyperopia may reduce the risk of developing accommodative esotropia and/or amblyopia. (See Table 1 for guidelines for correcting hyperopia in children.)¹²⁰⁻¹²² For children with esotropia, the threshold for prescribing hyperopic eyeglasses is lower than for those children without esotropia. For hyperopic patients, anisometropia is a risk factor for the development of accommodative esotropia.¹⁷

Choice of Therapy

All forms of esotropia should be considered for treatment. Binocular alignment should be established as soon as possible, especially in young children, to maximize binocularity,^{59,123} to prevent or facilitate treatment of amblyopia,^{32,124} and to restore normal appearance. Significant refractive errors should be corrected. Amblyopia treatment is usually started before surgery because this may alter the angle of strabismus¹²⁵ and/or increase the likelihood of good postoperative binocularity.^{123,126}

There is evidence that early surgical correction improves sensory outcomes for infantile esotropia, probably because the duration of constant esotropia is minimized.^{58,59,63,127-129} Given equal visual acuity in both eyes, there is no consensus among strabismus surgeons on the criteria for unilateral or bilateral surgery, nor is there good evidence to support unilateral versus bilateral surgery.¹³⁰

**Esotropia and Exotropia PPP:
Section I. Esotropia**

TABLE 1 GUIDELINES FOR REFRACTIVE CORRECTION IN INFANTS AND YOUNG CHILDREN

Condition	Refractive Errors (diopters)		
	Age <1 year	Age 1–2 years	Age 2–3 years
Isoametropia (similar refractive error in both eyes)			
Myopia	–5.00 or more	–4.00 or more	–3.00 or more
Hyperopia (no manifest deviation)	+6.00 or more	+5.00 or more	+4.50 or more
Hyperopia with esotropia	+2.50 or more	+2.00 or more	+1.50 or more
Astigmatism	3.00 or more	2.50 or more	2.00 or more
Anisometropia (without strabismus)*			
Myopia	–4.00 or more	–3.00 or more	–3.00 or more
Hyperopia	+2.50 or more	+2.00 or more	+1.50 or more
Astigmatism	2.50 or more	2.00 or more	2.00 or more

NOTE: These values were generated by consensus and are based solely on professional experience and clinical impressions because there are no scientifically rigorous published data for guidance. The exact values are unknown and may differ among age groups; they are presented as general guidelines that should be tailored to the individual child. Specific guidelines for older children are not provided because refractive correction is determined by the severity of the refractive error, visual acuity, and visual symptoms.

* Threshold for correction of anisometropia should be lower if the child has strabismus. The values represent the minimum difference in the magnitude of refractive error between eyes that would prompt refractive correction.

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

- ◆ Correction of refractive errors¹¹
- ◆ Bifocals¹³¹
- ◆ Prism therapy^{132,133}
- ◆ Amblyopia treatment¹²⁶
- ◆ Extraocular muscle surgery¹³⁴
- ◆ Botulinum toxin A injection¹³⁵
- ◆ Other methods

Treatment plans are formulated in consultation with the parent/caregiver and patient, if appropriate. The plans should be responsive to the expectations and preferences of the parent/caregiver and patient, including to their perception of the existing alignment, which may differ from the ophthalmologist's, and what they hope to achieve with treatment. It is important that the family/caregiver and ophthalmologist agree on the goals of treatment before surgery is performed. 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 is the initial treatment for children with esotropia (see Table 1). For patients with accommodative esotropia, realignment by cycloplegic-determined eyeglasses or contact lenses alone is successful in most cases.^{39,136} In general, a greater degree of hyperopia indicates a higher likelihood that the refractive error is an important etiologic factor of the esotropia. While children with developmental delay and strabismus may be less tolerant of eyeglasses, they may respond to correction of smaller amounts of ametropia. Additionally, children with a variable angle of esotropia or a greater deviation at near may respond to correction of even low hyperopia.

The aim of treatment is to correct hyperopia sufficiently to restore alignment, and in most cases a prescription to correct the full refractive error is written. Undercorrection of the hyperopia sometimes improves adherence, especially in older children. A manifest noncycloplegic refraction may be required to balance visual acuity and binocular alignment in older children.

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 used temporarily to facilitate adherence to wearing eyeglasses. 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 assessed in the office setting by placing minus lenses over the eyeglasses to ensure that binocular alignment is optimized.



Repeat cycloplegic refraction is indicated when esotropia does not respond to the initial prescription of hyperopic refraction or when the esotropia recurs after surgery.
(*strong recommendation, moderate evidence*)

In general, eyeglasses to control an esodeviation are well tolerated by children, especially when there is visual improvement. Accurate fitting and maintaining proper adjustment facilitate acceptance. Head straps or flexible single-piece frames may be useful in babies; cable temples and spring hinges are helpful in keeping eyeglasses on active young children. Polycarbonate lenses are more durable and thus provide greater safety; these are preferable for children, especially if they are amblyopic.

Bifocals

An esodeviation greater for near than for distant targets is found in some cases. Convergence excess is defined clinically as an increased near esodeviation of 10 prism diopters or greater compared with the distance deviation (high clinical AC/A ratio) with the use of full hyperopic correction. Bifocal treatment should be considered in patients with potential for sensory fusion who maintain essentially aligned eyes at distance but have a manifest esotropia at near (typically greater than 10 prism diopters) 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. Eliminating bifocals is feasible in approximately 60% of cases after an average of 5 years of use.¹³¹ An excellent initial response is associated with a lower likelihood that the bifocals can be withdrawn later without recurrence of the esotropia.¹³¹

For children 5 years of age and younger, the bifocal should be prescribed as an executive or a flat-top (D-segment) type, with the top of the bifocal bisecting the pupil in primary gaze in younger children and a few millimeters lower in older children. While the minimum strength of the bifocal can sometimes be estimated by office testing in trial frames, it is usually more practical to empirically prescribe a +2.50 to +3.00 D add for all patients requiring them. Reductions can be made later as part of a routine eyeglass change. Progressive bifocals offer some cosmetic advantages and are preferred in older children who have adapted well to standard bifocals. The transition zone should generally be placed several millimeters higher than the standard adult fitting.¹³⁷

Disadvantages of bifocals include expense, appearance, and potential rejection by the child. A minority of clinicians avoids bifocals because they believe that alignment at distance is sufficient to protect binocular vision. 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^{138,139} and eliminate the need for bifocal wear without producing consecutive exotropia at distance.¹⁴⁰⁻¹⁴²

Prism Therapy

Prisms are rarely useful in infantile esotropia, in part because the angle of deviation is usually too large to correct. In some patients with acquired esotropia, Press-On (3M™ Press-On™ Optics; 3M, Medical Specialties Division, St. Paul, MN) plastic prisms are used to promote binocular vision and establish the full angle on which to base extraocular muscle surgery.¹³³ The Prism Adaptation Study investigated the role of preoperative Press-On 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.^{132,133} 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. Press-On prisms cause visual symptoms that some children find objectionable (blurred vision with poor compliance with eyeglasses). In addition, using Press-On prisms requires re-evaluation (additional office visits) and 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 amblyopia (see Amblyopia PPP¹⁸).¹⁴³ 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.¹²⁶

Extraocular Muscle Surgery

Children with esotropia should undergo surgical correction if eyeglasses and amblyopia management are ineffective in aligning the eyes.¹³⁴ Strabismus surgery should be performed only when more conservative methods have failed or are unlikely to be of benefit. Surgery is rarely justified when the primary objective is to eliminate eyeglasses. Except for acquired symptomatic deviations in older children, 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 patients with infantile esotropia,^{60,144} achievement of high-grade stereopsis is rare.^{58,59,61} In contrast, the quality of stereopsis appears to be improved by prompt surgical realignment in patients with decompensated accommodative esotropia.^{59,134,145}

While most patients with infantile esotropia receive surgical intervention during childhood, it is unknown whether early treatment results in long-term motor alignment. However, achieving binocular alignment early in life (before age 2 years) to within 10 prism diopters of orthotropia increases the likelihood of achieving binocularity.^{58,59,127-129}

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.^{146,147} The presence of amblyopia¹²⁶ or nystagmus¹⁴⁸ is associated with an increased rate of requiring reoperation. In addition, esotropia may recur postoperatively on an accommodative basis in 50% of patients and correlates with the magnitude of the hyperopia.⁶³

Extraocular muscle surgery usually is performed for the distance angle of deviation when the individual is wearing full hyperopic correction; however, some surgeons use the maximum near deviation. For those individuals with a distance-near disparity (high AC/A ratio), bilateral medial rectus recession usually reduces the ratio.^{138,139} Prism adaptation for the near angle,¹⁴⁹ augmentation of the recession over amounts done with a normal AC/A ratio,¹⁵⁰ or posterior fixation sutures (Fadenoperation)¹⁴¹ increase the likelihood of a satisfactory alignment and eventual weaning from bifocals.

The amount of surgery and the choice of surgical technique may vary (e.g., methods of suture placement in the muscle and sclera, or measurement of recession or resection). Although two-muscle surgery is most frequently performed, sometimes three- or four-horizontal-muscle surgery may be required for large-angle deviations.¹⁵¹ Some clinicians believe that two-muscle surgery is the better option for all deviations, regardless of magnitude, to reduce the risk of consecutive exotropia.¹⁵²

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 to be proven.¹⁵³

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

Botulinum Toxin A Injection

Chemodenervation by injection of botulinum toxin type A into one or more extraocular muscles induces a temporary weakness by pharmacologic blockade of the neuromuscular junction. While the mechanism of long-term ocular realignment 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. 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,¹⁵⁴ but its value in managing infantile esotropia has not been definitively established.^{135,155-159} Disadvantages include the frequent need for repeat injection(s), especially with larger preoperative angles; iatrogenic ptosis, which may increase the risk for amblyopia; and the need for general anesthesia. Importantly, delayed binocular realignment may be disadvantageous in an infant with a rapidly developing visual system.

Other Methods

There is no role for most types of vision training for children with esotropia.¹⁶⁰ Training in diplopia recognition (antisuppression training) and strengthening vergence amplitudes is ineffective in the treatment of most esotropic patients and may occasionally produce permanent diplopia, especially in patients with monofixation syndrome.

Cholinesterase inhibitors, such as echothiophate iodide, reduce accommodative effort and convergence by stimulating ciliary muscle contraction (pupillary size is also reduced). Although sometimes effective, long-term use of this method is less desirable than using corrective lenses because of a risk of adverse systemic side effects such as diarrhea, asthma, and/or increased salivation and perspiration as well as increased risk associated with the administration of certain agents (e.g., succinylcholine chloride) used in general anesthesia.¹⁶¹ Potential ocular side effects include cataract, retinal detachment, and iris cysts, which may encroach on the visual axis.¹⁶²⁻¹⁶⁴ Some ophthalmologists prescribe phenylephrine 2.5% eyedrops twice daily to be used concurrently with the cholinesterase inhibitor to reduce the risk of iris cyst formation. Echothiophate iodide may be difficult to obtain in the United States.

**Esotropia and Exotropia PPP:
Section I. Esotropia**

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 4 to 6 months. As the child matures, the frequency of follow-up visits can be reduced.¹⁶⁵ New or changing findings may indicate the need for more frequent follow-up examinations.

In children with esotropia, hyperopia should be assessed at least annually and more frequently if visual acuity decreases or the esotropia increases. Detection of uncorrected 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.¹¹³

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

Diagnostic evaluation and treatment by an orthoptist can be a useful adjunct to the ophthalmologic care of patients with esotropia.

COUNSELING AND REFERRAL

Childhood esotropia is a long-term problem that requires commitment from the patient and/or family/caregiver, as appropriate, 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 pediatric patients who understand the diagnosis and rationale for treatment are more likely to adhere to treatment recommendations.^{166,167}

SECTION II. EXOTROPIA



INTRODUCTION

DISEASE DEFINITION

Exotropia is a divergent misalignment of the visual axes. Exotropia can be classified as follows:

- ◆ Infantile exotropia
- ◆ Intermittent exotropia
- ◆ Convergence insufficiency
- ◆ 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, inferior oblique overaction, and dissociated vertical deviation. Neonates frequently have intermittent exotropia within the first 3 to 4 months of life; however, it rarely persists.⁷ Children with neurodevelopmental delay may have constant exotropia from infancy.



Infantile esotropia and exotropia are associated with an increased risk of amblyopia.
(*good evidence*)

Intermittent Exotropia

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

Convergence Insufficiency

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

Other

Sensory exotropia is associated with unilateral or bilateral vision loss on a structural basis. Consecutive exotropia occurs in some children after surgery for esotropia. Sensory and consecutive exotropias are not within the scope of this document.

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 caused by positive angle kappa, which is a disparity between the visual and anatomic axes of the eyes.

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 PPP¹⁸)
- ◆ Educate the patient and/or family/caregiver, as appropriate, of the diagnosis, treatment options, and care plan
- ◆ Inform the patient's other health providers of the diagnosis and treatment plan
- ◆ Treat the exotropia (align the visual axes to promote and maintain binocular vision (fusion, stereopsis), prevent or facilitate treatment of amblyopia, and restore normal appearance
- ◆ Maximize quality of life by optimizing binocular alignment and visual acuity
- ◆ Monitor vision and binocular alignment and modify therapy as appropriate



BACKGROUND

PREVALENCE AND RISK FACTORS

Exotropia occurs in approximately 1% of the population; intermittent exotropia is the most frequently reported type.^{26,27,168,169} Exotropia has been associated with prematurity, perinatal morbidity, genetic disorders, detrimental prenatal environmental influences such as maternal substance abuse and smoking, family history of strabismus, female sex, astigmatism, and aniso-astigmatism.^{26,27 28,52} One small retrospective population-based cohort study in the United States found that intermittent exotropia was twice as frequent in girls than in boys.¹⁷⁰ Clinic-based studies of children with infantile-onset (congenital) exotropia found that half had associated ocular or systemic anomalies.^{49,171}

In the long-term, reduction or prevention of factors such as prematurity and maternal smoking during pregnancy as well as diagnosis and treatment of myopia and myopic anisometropia (see Table 1) 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, and symptoms. Some studies suggest that many patients who decline surgical correction appear to remain stable or spontaneously improve with observation alone,^{172,173} but others report deterioration during long-term follow-up.¹⁷⁴ 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%.¹⁷⁵ 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.¹⁷³ Another study of 371 children with intermittent exotropia who were followed for 2 years found that only 0.5% decompensated to a constant exotropia.¹⁷⁶ If the deviation becomes constant, binocular vision can deteriorate.¹⁷⁷ The causes of exotropia and of its natural history are poorly understood. Proposed etiologies for exotropia include excess tonic divergence and mechanical or innervational orbital factors.¹⁷⁸ Severe unilateral or bilateral vision loss may cause exotropia. Typically, unilateral poor vision in early childhood is associated with esotropia rather than exotropia.

RATIONALE FOR TREATMENT

The potential benefits of treatment for exotropia include promoting binocular vision and normal visual function in each eye. Normal binocular alignment promotes a positive self-image.⁷² The appearance of misaligned eyes impairs self-image and social interactions and reduces employment opportunities.^{67-71,73,74} In one study, children aged 5 years and older expressed a negative feeling about dolls that had been altered to be esotropic or exotropic.⁷⁴ In another study, elementary school teachers rated personal characteristics of children with esotropia and exotropia more negatively than orthotropic children.⁷¹ In a sample of children enrolled in the Multi-ethnic Pediatric Eye Disease Study, strabismus was associated with decreased general health-related quality of life in preschool children, based on the parents' proxy reporting.⁷⁵ After strabismus surgery, adults have reported improved confidence, self-esteem, and interpersonal interactions.⁶⁸ Similar studies have not been done to assess the effect of strabismus surgery on these traits in children.



Children with untreated strabismus can have reduced binocular potential and impaired social interactions, and they can be subject to negative perceptions by others, which may affect their psychosocial quality of life. (*good evidence*)



CARE PROCESS

PATIENT OUTCOME CRITERIA

- ◆ Optimal binocular motor alignment
- ◆ Optimal binocular sensory status (fusion and stereopsis)
- ◆ Optimal visual acuity in each eye

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 etiologies 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.^{78,79} Specific issues for exotropia are discussed in this section; the Esotropia section of this document contains additional details of the comprehensive strabismus evaluation.

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, and when the deviation occurs (e.g., when tired, ill, daydreaming, or viewing distant objects). In addition, it is helpful to include any observation of alternation or of only one eye drifting out.

Examination

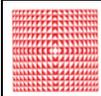
Sensory tests (e.g., stereopsis) should be done before visual acuity and alignment measurements, which may dissociate the eyes by monocular occlusion and cause reduced stereoacuity measurement or interfere with assessment of control of the exodeviation.

Esotropia and Exotropia PPP:
Section II. Exotropia

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 or XP). Fusional control can vary substantially from visit to visit or even within the same visit. Various scales have been developed to further characterize control of exodeviations.^{179,180} 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 fusional control at a distance with an intermittent exotropia.^{181,182}

MANAGEMENT

All forms of exotropia should be monitored and some will require treatment. Young children with intermittent exotropia and good fusional control can be followed without surgery.¹⁷⁶ Deviations that are present most or all of the time require treatment. However, the optimal modes of therapy for exotropia, the long-term benefit of early surgical correction, and the relative merits of bilateral vs. unilateral surgery are not well established.¹⁸³ Amblyopia is uncommon in patients with intermittent exotropia, but, if present, it should be treated.



Young children with intermittent exotropia and good fusional control can be followed without surgery. (*strong recommendation, moderate evidence*)

Choice of Therapy

Current treatment practices are listed below. Some of these treatments are under evaluation in randomized trials.

- ◆ Correction of refractive errors
- ◆ Stimulating accommodative convergence (overcorrection of myopia or undercorrection of hyperopia)
- ◆ Patching (antisuppression) therapy
- ◆ Amblyopia treatment
- ◆ Prism therapy
- ◆ Convergence exercises for convergence insufficiency exotropia
- ◆ Extraocular muscle surgery
- ◆ Botulinum toxin A injection¹³⁵

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. Improved retinal-image clarity often improves the control of the exotropia.¹⁸⁴ 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.¹⁸⁵ 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 should be the least amount needed to promote good vision and stimulate accommodative convergence to control the exodeviation; such correction can be the full cycloplegic refraction,¹⁸⁴ 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 by increasing myopic correction in myopes, reducing hyperopic correction in hyperopes, or prescribing myopic correction in ametropes. Some patients may not tolerate this therapy because of visual discomfort or decreased visual acuity. Studies suggest that overcorrecting minus-lens therapy stimulates accommodation without increasing myopia.^{186,187} It is most useful in patients with low-grade myopia and in those already wearing eyeglasses.

Patching Therapy

Patching therapy in children with intermittent exotropia may improve control of the exotropia even in the absence of amblyopia. In some cases, part-time patching (e.g., 2 to 6 hours daily) may improve fusional control^{188,189} and/or reduce the angle of strabismus. Part-time patching may be done on the preferred eye. In the absence of a fixation preference, patching is prescribed to alternate between eyes.

Amblyopia Treatment

In children with exotropia, treatment for amblyopia¹⁸ may improve fusional control, decrease the angle of the exodeviation, and/or improve the postoperative success rate in those requiring strabismus surgery. Because amblyopia is uncommon in intermittent exotropia,¹⁶⁸ 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 a 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 also 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.

Convergence Exercises for Convergence Insufficiency Exotropia

Orthoptic therapy may improve fusional control in patients with convergence insufficiency exotropia and with small- to moderate-angle exotropia (i.e., 20 prism diopters or less), with the goal of strengthening fusional convergence amplitudes.^{190,191} Patients 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.¹⁹²⁻¹⁹⁴

Extraocular Muscle Surgery

Surgical correction is considered if the deviation 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 patching. 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 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 bring out the full deviation.

Esotropia and Exotropia PPP:
Section II. Exotropia

In patients with evidence of a high AC/A ratio, the degree of abnormality may be quantified using -2.00 D lenses at distance. A high AC/A ratio is diagnosed when the distance angle exceeds the near angle by at least 10 prism diopters, and there is a significant decrease in the distance angle when -2.00 D lenses are placed over the usual refractive correction. In these patients, a conservative approach may be warranted because of the risk of consecutive esotropia, diplopia, and asthenopia with near fixation.¹⁹⁵



In patients with exotropia and a high accommodative convergence to accommodation ratio (AC/A), treatment with eyeglasses is generally preferred over surgery because of the risk of consecutive esotropia and diplopia after surgery.
(discretionary recommendation, moderate evidence)

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 patients who have undergone early surgery.^{196,197} In one study, alignment before age 7 years, before 5 years of strabismus duration, or while the deviation is intermittent increased the likelihood and quality of stereopsis.¹⁹⁸

Surgery consists of bilateral-lateral rectus-muscle recessions or unilateral-lateral rectus-muscle recession and medial rectus-muscle resection. 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 lateral rectus muscle recession may be done for a small deviation.

Although most 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).^{199,200} One randomized trial (n=36) found that long-term outcome was better after recess-resect than after bilateral recession.¹⁹⁹ 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,^{201,202} but other studies reported a variable and unpredictable outcome following early overcorrection.²⁰³ The duration of follow-up likely influences the report of motor outcomes.^{203,204} When a consecutive esotropia persists for several weeks, placement of temporary press-on prisms that are slowly reduced in power can be helpful. When unsuccessful, additional surgery is often required for the consecutive esotropia. While approximately 80% of patients have good alignment 6 months postoperatively after bilateral-lateral rectus-muscle recession,²⁰⁵ long-term results are less favorable and recurrence is common over time.^{201,206} Outcomes may be improved with a combination of surgical and nonsurgical (orthoptic/occlusion) therapy during management of a child with exotropia.²⁰⁷ Use of an adjustable suture technique (older children and adults) has not been shown to improve outcomes in uncomplicated intermittent exotropia.^{153,208}

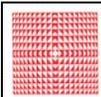
Botulinum Toxin A Injection

Chemodenervation by injection of botulinum toxin type A 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 type A for adults with horizontal, nonaccommodative ocular misalignment, botulinum toxin treatment was less successful (29% vs. 77%) than surgery.²⁰⁹ There is insufficient evidence to make treatment recommendations for botulinum toxin treatment for exotropia.¹³⁵

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 postoperative esotropia are at risk for developing amblyopia and should be followed more frequently. Postoperative esotropia may also precipitate loss of stereoacuity. Prescribing base-out prism in eyeglasses is occasionally useful to alleviate diplopia associated with transient postoperative esotropia. 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. Once visual maturity is reached (i.e., 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), and ocular motility assessment.



Esotropia that persists after exotropia surgery may place the patient at risk for amblyopia, diplopia, and loss of stereoacuity. (*moderate evidence*)

PROVIDER AND SETTING

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

Diagnostic evaluation and treatment by an orthoptist can be a useful adjunct to the ophthalmologic care of patients with exotropia.

COUNSELING AND REFERRAL

Childhood exotropia is a long-term problem that requires commitment from the patient and/or family/caregiver, as appropriate, 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 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.^{166,167}



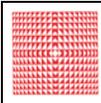
SOCIOECONOMIC CONSIDERATIONS FOR STRABISMUS

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.⁷⁸ Therefore, many authorities recommend early and regular vision screening to detect this and other conditions.

Evidence suggests that many children do not receive the recommended care. In fact, almost 40% of children in the United States have never undergone a vision screening.^{210,211} Children in low-income families, in uninsured families, and in racial and ethnic minority groups may fare worse.²¹⁰⁻²¹³ Studies indicate that in general, African American children and children living below 400% of the federal poverty level receive fewer and less intensive services relative to their white counterparts.^{212,214} There is evidence that these race-ethnicity disparities are reflected in eye care services as well as in other health services.²¹⁴ 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.²¹²

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. It appears from one large study that only about half of children who fail vision screening are seen by eye care providers in follow-up.²¹⁵ Barriers to care may include inadequate information, lack of access to care, and/or financial or insurance coverage difficulties.^{216,217} 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.²¹⁴ 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.²¹⁶

Children with untreated strabismus begin to suffer socially before the age of 6,⁷⁴ have negative perceptions by teachers,⁷¹ and, in general, have reduced psychosocial quality of life.^{218,219} In addition, later employment prospects can be affected by strabismus.²²⁰ In one small (n=140) time trade-off utility study, the majority of adults with strabismus would trade a shortened life expectancy in exchange for no strabismus.²²¹ Treatment studies indicate that appropriate management of strabismus can improve both functional and psychosocial outcomes, even into adulthood.²²²⁻²²⁴



Children with untreated strabismus can have reduced binocular potential and impaired social interactions, and they can be subject to negative perceptions by others, which may affect their psychosocial quality of life. (*good evidence*)

State legislatures have attempted to close the gap in children's eye care by mandating some form of vision screening for children.²²⁵ 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.²²⁵

Optimal eye and vision care for children involves an organized program of vision screening in the primary care and community settings and 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.²²⁶



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 alternate 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.

Esotropia and Exotropia PPP
Appendix 1. Quality of Ophthalmic Care Core Criteria

- ◆ 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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4th Printing: July 2005



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

Esotropia, which includes entities with the following ICD-9 and ICD-10 classifications:

	ICD-9 CM	ICD-10 CM
Nonaccommodative	378.00	H50.00
Accommodative	378.35	H50.43
Alternating	378.05	H50.05
Alternating with A pattern	378.06	H50.06
Alternating with V pattern	378.07	H50.07
Alternating with X or Y pattern (with other noncomitancies)	378.08	H50.08
Monocular	378.01	H50.01–
Monocular with A pattern	378.02	H50.02–
Monocular with V pattern	378.03	H50.03–
Monocular with X or Y pattern	378.04	H50.04–
Intermittent, alternating	378.22	H50.32
Intermittent, monocular	378.21	H50.31–
Unspecified	378.00	H50.00

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 in the ICD-10 CM 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):
 - Right is always 1
 - Left is always 2

**Esotropia and Exotropia PPP:
Appendix 2. ICD Codes**

Exotropia, which includes entities with the following ICD-9 and ICD-10 classifications:

	ICD-9 CM	ICD-10 CM
Alternating	378.15	H50.15
Alternating with A pattern	378.16	H50.16
Alternating with specified noncomitancy not elsewhere classifiable (includes alphabetical patterns)	378.18	H50.18
Alternating with V pattern	378.17	H50.17
Monocular	378.11	H50.11–
Monocular with A pattern	378.13	H50.12
Intermittent unspecified	378.20	H50.30
Alternating, intermittent	378.24	H50.34
Monocular, intermittent	378.23	H50.33–
Unspecified	378.10	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 in the ICD-10 CM 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):
 - Right is always 1
 - Left is always 2



APPENDIX 3. VISUAL ACUITY TESTING CHARTS

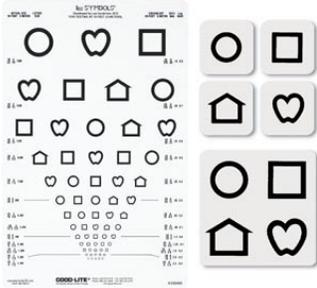
The World Health Organization (WHO) and the National Academy of Sciences Committee on Vision have made similar recommendations about optotype choice and arrangement on visual acuity testing charts.^{85,89} Optotypes should be clear, standardized, of similar characteristics, and should not reflect a cultural bias. Each line should contain five optotypes. Spacing between the optotypes should be proportional: the horizontal spacing between individual optotypes should be equal to the size of the optotype and the vertical spacing between lines should be the height of the optotypes in the lower line. Optotype sizes should generally be presented in 0.1 logMAR decrements. This arrangement leads to an inverted pyramid design for wall charts.

Visual acuity testing charts used with children that meet these recommendations include⁸⁵ LEA Symbols (Good-Lite Co., Elgin, IL), Sloan letters,⁹³ Sloan numerals, Tumbling E, and HOTV. The Snellen chart is less desirable because the individual letters are not of equal legibility and the spacing of the letters does not meet WHO/Committee on Vision standards.^{85,94-96}

Several symbol charts have serious limitations for young children. These include Allen figures,⁹⁷ the Lighthouse chart, and the Kindergarten Eye Chart.⁹⁸ In these charts, the optotypes are not standardized and are presented in a culturally biased fashion.⁹⁹ Although the Tumbling E chart meets WHO/Committee on Vision recommendations, it is less desirable because it requires spatial orientation skills not mastered by all children. Other visual acuity charts are being developed to overcome these limitations, including the Handy Eye Chart and the Compact Reduced logMAR chart.^{227,228}

Table A3-1 lists details of design of visual acuity testing charts that are commonly used.

TABLE A3-1 VISUAL ACUITY TESTING CHARTS

Chart	Meets WHO* ¹ NAS ¹ Recommendations	Attributes/Challenges
<p style="text-align: center;">LEA Symbols⁹¹</p>  <p>Reproduced with permission from Good-Lite Co., Elgin, IL.</p>	<p>Yes</p>	<p>Attributes:</p> <ul style="list-style-type: none"> • Optotypes of similar legibility • Inverted pyramid design with five optotypes per line (at visual acuities better than 20/100), proportional spacing between optotypes, and 0.1 LogMAR decrements in optotype size

**Esotropia and Exotropia PPP:
Appendix 3. Visual Acuity Testing Charts**

TABLE A3-1 VISUAL ACUITY TESTING CHARTS (CONTINUED)

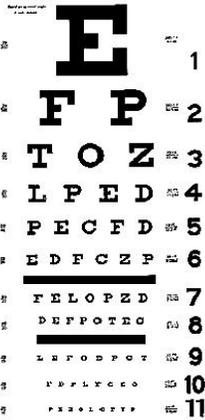
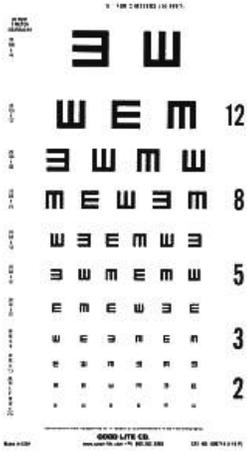
Chart	Meets WHO*/NAS ¹ Recommendations	Attributes/Challenges
<p style="text-align: center;">Sloan Letters⁹³</p>  <p>Reproduced with permission from Good-Lite Co., Elgin, IL.</p>	<p style="text-align: center;">Yes[†]</p>	<p>Attributes:</p> <ul style="list-style-type: none"> • Optotypes of similar legibility • Inverted pyramid design with five optotypes per line, proportional spacing between optotypes, and 0.1 LogMAR decrements in optotype size
<p style="text-align: center;">HOTV</p>  <p>Reproduced with permission from Good-Lite Co., Elgin, IL.</p>	<p style="text-align: center;">Yes[†]</p>	<p>Attributes:</p> <ul style="list-style-type: none"> • Optotypes of similar legibility • Inverted pyramid design with five optotypes per line, proportional spacing between optotypes, and 0.1 LogMAR decrements in optotype size
<p style="text-align: center;">Snellen Letters²²⁹</p>  <p>Image is in the public domain.</p>	<p style="text-align: center;">No</p>	<p>Challenges:</p> <ul style="list-style-type: none"> • Optotypes are not of similar legibility • Variable number of optotypes per line • Nonproportional spacing between optotypes • Nonstandard optotype size decrements

TABLE A3-1 VISUAL ACUITY TESTING CHARTS (CONTINUED)

Chart	Meets WHO*/NAS† Recommendations	Attributes/Challenges
<p style="text-align: center;">Tumbling E Chart</p>  <p>Reproduced with permission from Good-Lite Co., Elgin, IL.</p>	<p style="text-align: center;">Yes†</p>	<p>Attributes:</p> <ul style="list-style-type: none"> • Optotypes of similar legibility • Inverted pyramid design available with five optotypes per line, proportional spacing between optotypes, and 0.1 LogMAR decrements in optotype size <p>Challenges:</p> <ul style="list-style-type: none"> • Requires spatial orientation skills not mastered by all children
<p style="text-align: center;">Allen Figures</p>  <p>Allen HF. A new picture series for preschool vision testing. Am J Ophthalmol 1975;44:40. Copyright 1957. Reprinted with permission from Elsevier. All rights reserved.</p>	<p style="text-align: center;">No</p>	<p>Challenges:</p> <ul style="list-style-type: none"> • Optotypes are not of similar legibility • Variable number of optotypes per line • Nonproportional spacing between optotypes • Nonstandard optotype size decrements • Optotypes not easily recognized by all children (e.g., telephone)

**Esotropia and Exotropia PPP:
Appendix 3. Visual Acuity Charts**

TABLE A3-1 VISUAL ACUITY TESTING CHARTS (CONTINUED)

Chart	Meets WHO*/NAS† Recommendations	Attributes/Challenges
<p style="text-align: center;">Lighthouse Chart</p> <p>Reproduced with permission.</p>	<p>No</p>	<p>Challenges:</p> <ul style="list-style-type: none"> • Optotypes are not of similar legibility • Variable number of optotypes per line • Nonproportional spacing between optotypes • Nonstandard optotype size decrements
<p style="text-align: center;">Kindergarten Eye Chart</p> <p>Reproduced with permission from Wilson Ophthalmic Corp., Mustang, OK.</p>	<p>No</p>	<p>Challenges:</p> <ul style="list-style-type: none"> • Optotypes are not of similar legibility • Variable number of optotypes per line • Nonproportional spacing between optotypes • Nonstandard optotype size decrements

NAS = National Academy of Sciences; WHO = World Health Organization

* World Health Organization. Consultation on development of standards for characterization of vision loss and visual functioning. Geneva, 4-5 September 2003. Available at: http://whqlibdoc.who.int/hq/2003/WHO_PBL_03.91.pdf. Accessed January 24, 2012.

† Committee on Vision. Recommended standard procedures for the clinical measurement and specification of visual acuity. Report of working group 39. Assembly of Behavioral and Social Sciences, National Research Council, National Academy of Sciences, Washington, D.C. Adv Ophthalmol 1980;41:103-48.

‡ Sloan, HOTV, and Tumbling E charts have chart designs that **do not** meet proportional spacing recommendations between individual optotypes and optotype lines.



RELATED ACADEMY MATERIALS

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Strabismus (2012)

Patient Education Downloadable Handout

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To order any of these products, except for the free materials, please contact the Academy's Customer Service at 866.561.8558 (U.S. only) or 415.561.8540 or www.aao.org/store.



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