## OPTOMETRIC CLINICAL PRACTICE GUIDELINE

Care of the Patient with

# Strabismus: Esotropia and Exotropia



#### OPTOMETRY: THE PRIMARY EYE CARE PROFESSION

Doctors of optometry are independent primary health care providers who examine, diagnose, treat, and manage diseases and disorders of the visual system, the eye, and associated structures as well as diagnose related systemic conditions.

Optometrists provide more than two-thirds of the primary eye care services in the United States. They are more widely distributed geographically than other eye care providers and are readily accessible for the delivery of eye and vision care services. There are approximately 36,000 full-time equivalent doctors of optometry currently in practice in the United States. Optometrists practice in more than 6,500 communities across the United States, serving as the sole primary eye care providers in more than 3,500 communities.

The mission of the profession of optometry is to fulfill the vision and eye care needs of the public through clinical care, research, and education, all of which enhance the quality of life.



# OPTOMETRIC CLINICAL PRACTICE GUIDELINE CARE OF THE PATIENT WITH STRABISMUS: ESOTROPIA AND EXOTROPIA

#### **Reference Guide for Clinicians**

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NOTE: Clinicians should not rely on the Clinical
Guideline alone for patient care and management.
Refer to the listed references and other sources
for a more detailed analysis and discussion of
research and patient care information. The
information in the Guideline is current as of the
date of publication. It will be reviewed periodically
and revised as needed.

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#### **INTRODUCTION**

Optometrists, through their clinical education, training, experience, and broad geographic distribution, have the means to provide primary eye and vision care for a significant portion of the American public and are often the first health care practitioners to diagnose patients with strabismus.

This Optometric Clinical Practice Guideline for the Care of the Patient with Strabismus describes appropriate examination, diagnosis, treatment, and management to reduce the risk of visual disability from esotropia and exotropia through timely care. This Guideline will assist optometrists in achieving the following goals:

- Identify patients at risk of developing strabismus
- Accurately diagnose strabismus
- Improve the quality of care rendered to patients with strabismus
- Minimize the adverse effects of strabismus and enhance the patient's quality of life
- Preserve the gains obtained through treatment
- Inform and educate other health care practitioners, including primary care physicians, as well as teachers, parents, and patients about the visual complications of strabismus and the availability of treatment and management.

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#### I. STATEMENT OF THE PROBLEM

Strabismus, often called "crossed-eyes" or "wall eyes," is a condition in which the eyes are not properly aligned with each other. One eye is either constantly or intermittently turned in (esotropia) or out (exotropia). This ocular misalignment may be accompanied by abnormal motility of one or both eyes, double vision, decreased vision, ocular discomfort, headaches, or abnormal head posture. Although the exact cause cannot always be determined with reasonable certainty, strabismus is usually attributable to refractive, sensory or organic, anatomic or motor, or innervational causes. Any of these factors alone can result in strabismus; however, strabismus may be the result of multiple factors, which, occurring alone, might not cause the disorder. For some individuals, strabismus can result in permanent vision loss. Young children with strabismus often develop amblyopia (lazy eye) and impaired stereopsis (binocular depth perception). Early identification and treatment of strabismic children may prevent amblyopia. The strabismic child with amblyopia has a significantly higher risk of becoming blind by losing vision in the non-amblyopic eye, due to trauma or disease.<sup>2,3</sup>

Normal binocular vision is required for many occupational and avocational tasks, as well as many other activities in daily life. Therefore, prompt diagnosis and treatment of strabismus are critical. Symptoms such as diplopia, headaches, blurred vision, and ocular fatigue may cause individuals with intermittent strabismus who have the potential for normal binocular vision and older persons who are developing strabismus to alter their activities of daily living. Studies comparing binocular with monocular performance on a variety of tasks for a group of normal individuals indicate that strabismus frequently leads to inefficient performance on various educational, occupational, and avocational tasks <sup>4,5</sup>

A student with intermittent strabismus may avoid reading, resulting in poor academic achievement. An employee with intermittent strabismus may suffer fatigue and headaches, resulting in reduced productivity. Strabismus may also be cosmetically displeasing and have a significant psychological impact manifested as low self-esteem, especially in patients whose jobs involve substantial personal eye contact.<sup>6</sup>

Strabismus may also have an adverse effect on family relationships.<sup>7</sup> In addition, delayed development (e.g., reaching milestones such as first walking and using single words) and difficulty with tasks involving visual perception have been found in young children with strabismus.<sup>8,9</sup>

Some types of strabismus, particularly those caused by high refractive errors, are preventable. Esotropias that occur due to an abnormal amount of uncorrected hyperopia are potentially preventable, if the hyperopia is diagnosed and managed prior to the onset of the deviation. More than 30 percent of children with hyperopia that exceeds 4 diopters (D) develop esotropia by 3 years of age. Whereas adult-onset strabismus can result from a neurologic disorder (e.g., brain tumor) or systemic disease (e.g., diabetes mellitus), adequate control of the general health status of a person afflicted with such a condition may lessen the possibility of the individual's developing strabismus.

Preschool vision screenings often include refractive error or visual acuity testing to detect amblyopia, as well as alignment testing to detect strabismus. Some of these screenings require assessment of visual acuity with and without convex lenses, which can identify some children with significant hyperopia. Ideally, these screenings should be conducted early (i.e., before 2–3 years, the peak ages for the onset of accommodative esotropia). As a method of screening for refractive error, photorefraction has gained popularity and may aid in earlier detection of some cases of accommodative esotropia. Nevertheless, because problems encountered in screening young children for refractive errors and strabismus may result in underdetection of vision problems, any child suspected of having strabismus should undergo a professional eye examination.

Remediation of strabismus requires treatment and management by an eye care practitioner, and the results are usually best when instituted early. Preservation of vision and binocular function result from proper diagnosis, treatment and management, and patient compliance. Periodic re-evaluation is important for appropriate patient management.

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#### A. Description and Classification of Strabismus

Proper alignment of the visual axes of the eyes is necessary for normal binocular vision and high-level stereopsis. The tendency for the eyes to deviate from each other can be classified as "latent" when the eyes are kept in alignment by the fusion mechanism and "manifest" when alignment is not maintained by fusion. Latent deviation of the eyes is called heterophoria; manifest deviation is called heterotropia or strabismus.

Strabismus is defined as a manifest deviation of the primary lines of sight of 1 prism diopter (PD) or more. In strabismus, one eye is either constantly or intermittently not directed toward the same point as the other eye when the patient attempts to fixate an object. As a result, an image of the fixated object is not formed on the fovea of the strabismic eye. The convergent (inward) misalignment of one eye is defined as esotropia; a divergent (outward) misalignment, exotropia; an upward misalignment, hypertropia; a downward misalignment, hypotropia. A wheel-like misalignment that is top inward is incyclotorsion, while a wheel-like misalignment that is top outward is excyclotorsion.

Misalignment of the eyes in strabismus can be classified in a number of ways: 13

- Direction (horizontal, vertical, or cyclotorsional)
- Comitant or incomitant (deviation equal in all positions of gaze or varying with the direction of gaze)
- Frequency (constant or intermittent)
- Involvement of accommodative system (accommodative or nonaccommodative)
- State of vergence system, comparing the magnitude of the distance and the near deviation (convergence-insufficiency or divergence-excess exotropia; divergence-insufficiency or convergence-excess esotropia; basic esotropia or basic exotropia)
- Laterality (unilateral or alternating)
- Time of onset (congenital or acquired)
- Size (small, moderate, or large).

The scope of this Guideline includes the diagnosis, treatment, and management of the most common clinical forms of nonparalytic esotropia and exotropia; it does not include vertical, cyclotorsional, or paretic strabismus. See Appendix Figure 3 for the ICD-10-CM classification of esotropia and exotropia. The following sections describe various forms of esotropia and exotropia.

#### 1. Esotropia

A convergent strabismus is termed an esotropia. Most patients with esotropia present before school age, generally between the ages of 2 and 3 years. Esotropia is often constant. In most cases, intermittent esotropia occurs initially in association with accommodative esotropia or decompensated esophoria (a tendency of one eye to deviate inward). The intermittency of accommodative esotropia is attributed to the fluctuating accommodative status of the patient at the onset of the deviation. Without treatment, intermittent esotropia is likely to become constant. The clinical forms of esotropia are discussed below.

#### a. Infantile Esotropia

When esotropia begins in the developmentally and neurologically normal child during the first 6 months of life, it is classified as "infantile." When it occurs after 6 months of age, it is referred to as "early-acquired." True "congenital" esotropia, which is present at birth, is considered extremely rare 14; however, the terms "infantile esotropia," "essential infantile esotropia," and "congenital esotropia" are often used interchangeably. The probable age of onset for infantile esotropia is at 2–4 months of age. 14

#### b. Acquired Esotropia

Acquired forms of esotropia occur at a later age than infantile esotropia. Usually, normal binocular vision has existed prior to the onset of the condition.

• **Accommodative esotropia.** This acquired strabismus is associated with the activation of accommodation. The esotropia is

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attributed partly or totally to either uncorrected hyperopic refractive error and/or a high accommodative convergence/accommodation (AC/A) ratio. <sup>15,16</sup> Accommodative esotropia has a better understood mechanism and a more straightforward treatment or management than any other form of strabismus. It is reduced partly or entirely by correcting the hyperopic refractive error and/or prescribing a near addition.

- Nonaccommodative esotropia. An acquired strabismus that develops after 6 months of age, nonaccommodative esotropia is not associated with accommodative effort. Correcting any coexisting hyperopia and/or prescribing a near addition for children with nonaccommodative esotropia has minimal or no effect on the size of the esotropia.
- **Acute esotropia.** When a convergent strabismus develops suddenly without any apparent etiology in a school-aged or older patient with previously normal binocular vision, it is called acute esotropia. The sudden diplopia that usually occurs in acute esotropia may result from an underlying and potentially lifethreatening disease process, thus it requires immediate evaluation. Its onset can often be traced to a precise hour of a particular day. Causes of acute esotropia are listed in Table 1.<sup>17</sup>
- Mechanical esotropia. A convergent strabismus caused by a mechanical restriction or tightness of an extraocular muscle (e.g., fibrosis of muscle tissue, thyroid myopathy) or a physical obstruction (e.g., blowout fracture) of the extraocular muscles, is classified as a mechanical esotropia. Some patients with Duane syndrome have tightening of the medial and/or lateral recti muscles secondary to the primary underlying neurological miswiring and co-innervation. There is a limitation or absence of abduction, causing an increasing esotropia. The palpebral fissure narrows when the eye rotates inward (adduction). In addition, the patient may exhibit an upshoot or downshoot when the eye adducts.

## Table 1 Causes of Acute Esotropia and Exotropia

- Neoplasm
- Head trauma
- Intracranial aneurysm
- Hypertension
- Diabetes mellitus
- Atherosclerosis
- Hydrocephalus
- Multiple sclerosis
- Meningitis/encephalitis
- Myasthenia gravis
- Sinus disease
- Chiari 1 malformation
- Ophthalmoplegic migraine
- Chemotherapy

#### c. Secondary Esotropia

An esotropia that results from a primary sensory deficit or as a result of surgical intervention is classified as a secondary esotropia.

• **Sensory esotropia**. A convergent strabismus resulting from visual deprivation or trauma in one eye that limits sensory fusion is classified as a sensory esotropia. It may result from any number of

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conditions that limit visual acuity in one eye (e.g., uncorrected anisometropia, unilateral cataract, corneal opacity, optic atrophy, macular disease). It occurs most frequently in persons under 5 years of age. <sup>18</sup> Approximately 4 percent of those with esotropia have sensory esotropia. <sup>19</sup>

• Consecutive esotropia. A convergent strabismus that occurs after surgical overcorrection of an exotropia, consecutive esotropia is frequently associated with other oculomotor anomalies (e.g., vertical or cyclotorsional deviations). It may result in amblyopia and loss of normal binocular vision in young children and diplopia in adults.

#### d. Microesotropia

When the angle of esotropia is less than 10 PD, it is classified as microesotropia. This condition often occurs beginning in a child under 3 years of age, and, in some cases, may escape diagnosis by conventional methods. The esotropia is constant and usually unilateral. The terms "microtropia," "microsquint," "minitropia," "monofixation syndrome," and "small-angle deviation" have been used to describe microesotropia.

#### 2. Exotropia

Exotropia, or divergent strabismus, can be subclassified on the basis of its comparative magnitude at distance and near or its frequency.<sup>20</sup>

In basic-type exotropia, the angle of deviation is within 10 PD at distance and near.

In the convergence-insufficiency type, the angle of deviation at near exceeds the angle of deviation at distance by at least 10 PD.

Divergence-excess type exotropia occurs when the angle of deviation at distance exceeds the angle of deviation at near by at least 10 PD.

Although exotropias may be constant or intermittent, most are intermittent. Children with intermittent exotropia often have the

divergence-excess type. Other clinical classifications of exotropia are discussed in the following paragraphs.

#### a. Infantile Exotropia

A divergent strabismus that begins during the first 6 months of life is classified as infantile exotropia. It is less common than infantile esotropia. In infants, some cases of constant exotropia may be associated with neurological syndromes or defects, craniofacial syndromes, and structural abnormalities in the eye.

#### b. Acquired Exotropia

An exotropia occurring after 6 months of age is considered to be acquired rather than infantile.

- **Intermittent exotropia**. In intermittent exotropia, the patient sometimes manifests diplopia, suppression, or anomalous retinal correspondence, and at other times, normal binocular alignment with good stereopsis. The period of strabismus and level of control are variable for each patient.<sup>21</sup> Basic intermittent exotropia accounts for approximately 50 percent of all cases of intermittent exotropia, with convergence insufficiency and divergence excess making up the balance of cases in approximately equal proportions.<sup>20</sup> Intermittent exotropia typically presents between the ages of 1 and 4 years. In the United States, it occurs in approximately 1 percent of children by the age of 7 years.<sup>22</sup> Without treatment over the years, intermittent exotropia may either progress (both in degree and the amount of time it is manifest), stay the same, or, in some cases, improve. 23,24 It rarely deteriorates to constant exotropia and fusion and some fixation at distance is usually maintained.
- **Acute exotropia**. When a divergent strabismus develops suddenly in an older patient who previously had normal binocular vision, it is classified as acute exotropia. This condition can result from an underlying disease process (Table 1) or a decompensating exophoria.

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• Mechanical exotropia. Mechanical exotropia is a divergent strabismus caused by a mechanical restriction or tightness (e.g., fibrosis of muscle tissue, thyroid myopathy) or a physical obstruction of the extraocular muscles (orbital fracture), causing increasing exotropia. Tightness of the lateral rectus muscle may develop secondary to the primary innervational miswiring in a rare type of Duane syndrome. With this type of strabismus, an absence of adduction results in increasing exotropia accompanied by narrowing of the palpebral fissure and retraction of the globe.

#### c. Secondary Exotropia

An exotropia that results from a primary sensory deficit or occurs as a result of some form of treatment for an esotropia is referred to as a secondary exotropia.

- **Sensory exotropia.** A divergent strabismus resulting from a unilateral decrease in vision that disrupts fusion, sensory exotropia may be due to a sensory deficit such as uncorrected anisometropia, unilateral cataract, or other unilateral visual impairment. Sensory exotropia and sensory esotropia occur with equal frequency in children under age 5; however, sensory exotropia predominates in persons older than 5 years. Sensory exotropia occurs in less than 3 percent of all strabismic children.
- Consecutive exotropia. Exotropia that occurs following surgical or optical correction of an esotropia is referred to as consecutive exotropia. This form of exotropia can also occur spontaneously in a formerly esotropic patient. A spontaneous change from esotropia to exotropia over time may be related to amblyopia of the deviating eye, weak binocular function, underaction of the medial rectus, or excessive hyperopic refractive error. When followed long term, the prevalence of consecutive exotropia is reported to be as high as 20 percent for esotropic patients treated with surgery.

#### d. Microexotropia

A constant exotropia of less than 10 PD, microexotropia occurs much less frequently than microesotropia.

#### B. Epidemiology of Strabismus

#### 1. Incidence and Prevalence

The estimated prevalence of strabismus in the general population is from 2 to 5 percent. 26-29 Between 5 and 15 million individuals in the United States may have this condition. Several studies of clinical populations have reported that esotropia appears to occur approximately 3-5 times as often as exotropia in children. 28,30 However, the National Health Survey of individuals 4–74 years of age found a higher prevalence of exotropia (2.1%) than esotropia (1.2%) in the U.S. population. This difference is probably related to the fact that the overall prevalence of strabismus in persons 55–75 years of age (in whom exotropia is more common) is 6.1 percent—substantially greater than for very young children 1–3 years of age (1.9%) or children and adults 4–54 years of age (3.3%).<sup>27</sup> The prevalence of exotropia may be underestimated, because it is most often an intermittent strabismus. For Hispanic/Latino and African American children ages 6–72 months in Los Angeles, California, the prevalence of strabismus was 2.4 percent for the former and 2.5 percent for the latter group. Exotropia was more common than esotropia.<sup>31</sup>

Approximately 50 percent of all childhood esotropias are either fully or partially accommodative. <sup>15,16,32,33</sup> An esotropia is partially accommodative when the accommodative factors of uncorrected hyperopia and/or a high accommodative convergence/accommodation (AC/A) ratio contribute to, but do not account for, the entire strabismus. Nonaccommodative esotropia is the second most common form of childhood esotropia, accounting for approximately 10 percent of all strabismus. <sup>33</sup> Infantile esotropia accounts for approximately 8.1 percent of cases of esotropia, affecting 1 in every 100–500 persons. <sup>19,33</sup> Intermittent exotropia is the most common type of exotropia, affecting nearly 1 percent of the population. <sup>22,34</sup> Exotropia has been reported to be more prevalent among Asian and African American populations than

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among Caucasians.<sup>35</sup> Women comprise 60–70 percent of patients with exotropia.<sup>36,37</sup>

#### 2. Risk Factors

Strabismus is more prevalent in persons with multiple handicaps, occurring in approximately 50 percent of patients with Down syndrome, <sup>38-40</sup> 44 percent of patients with cerebral palsy, <sup>41,42</sup> and 90 percent of patients with craniofacial dysostosis such as Apert-Crouzon syndrome. <sup>43-46</sup> Children born prematurely and of low birthweight have a greater risk of developing strabismus than children born at term. <sup>47</sup>

The prevalence of strabismus is also higher in families in which a parent or sibling has strabismus, ranging from 23 to 70 percent of family members. Whether strabismus itself or the conditions underlying the strabismus are genetic is unclear. The common occurrence of sensorimotor anomalies in the pedigrees of strabismic probands suggests that all siblings of a strabismic child be examined at an early age to rule out the presence of strabismus.

#### C. Clinical Background of Strabismus

Although strabismus can develop at any age, it usually develops during childhood. Most cases begin prior to 6 years of age; the peak age of onset is around 3 years. Strabismus acquired in adolescence or adulthood is frequently either motor or sensory in origin and can be a manifestation of systemic disease (e.g., diabetes mellitus) or neurologic disorder (e.g., brain tumor). Strabismus can also develop in adults following decompensation of a heterophoria. In patients of all ages, trauma to the head or orbit may result in strabismus.

#### 1. Accommodative Esotropia

#### a. Natural History

Accommodative esotropia has an average age of onset of  $2\frac{1}{2}$  years (usual range, 1–7 years)<sup>53</sup>. Cases have been documented beginning as early as 4 months of age and also beginning as late as the teenage years. <sup>16,54</sup> The

esotropia usually occurs when the child first becomes interested in viewing near objects. It is frequently first seen by the parents when the child is tired or not feeling well; in fact, the onset may be precipitated by a febrile illness. As many as half of all children with esotropia either obtain normal ocular alignment or obtain significant but not complete reduction of the esotropia when wearing an optical correction for hyperopia.<sup>33</sup>

#### b. Signs, Symptoms, and Complications

Patients with accommodative esotropia may exhibit any of the following:

- A small to moderate (generally 10–35 PD), often variable, angle of deviation that tends to occur more frequently at near. Depending on the AC/A ratio, the esotropia at near may be larger or similar to the esotropia at distance. Rarely does the magnitude of the esotropia exceed 40 PD. 55,56
- Uncorrected hyperopia (generally 2–6 D) and/or a high AC/A ratio. For patients with normal AC/A ratios (similar distance and near deviations), the average amount of hyperopia is 4.75 D, whereas with high AC/A ratios (near deviation exceeds distance deviation by 10 PD or more), the average amount of hyperopia is only 2.25 D.<sup>57</sup> Anisometropia greater than 1 D increases the risk for the development of accommodative esotropia in hyperopic children, especially in children with 3 D of hyperopia or less.<sup>55</sup> About 5 percent of patients with accommodative esotropia have minimal refractive error and manifest esotropia exclusively for near viewing distances. This type of esotropia, referred to as either nonrefractive accommodative esotropia or convergence-excess esotropia, is attributed to an elevated AC/A ratio.
- Esotropia beginning as intermittent strabismus. When treatment is delayed or not provided, many deviations become constant, and sensory adaptations such as amblyopia, suppression, and anomalous retinal correspondence may develop with loss of binocular vision.

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- Amblyopia (limited to cases that have become constant and unilateral, cases with clinically significant anisometropia, and cases that are partially accommodative).
- Diplopia or closure of one eye when doing near work.
- Overelevation in adduction, or inferior oblique overaction in one or both eyes, which develops in approximately 35 percent of cases. 58

#### c. Early Detection and Prevention

Children suspected of having accommodative esotropia should be examined immediately. The prognosis for achieving normal binocular vision is excellent, provided management is prompt. Better results usually occur when management is initiated during the intermittent phase. <sup>15,16,59</sup> If unmanaged, the patient may develop a superimposed nonaccommodative esotropia, accompanied by amblyopia, suppression, and anomalous retinal correspondence. <sup>16</sup> Subsequently, lens therapy can be less successful in eliminating the total angle of deviation, and the person can lose binocular vision.

#### 2. Acute Esotropia and Exotropia

#### a. Natural History

Acute esotropia or exotropia generally develops in children older than 6 years, adolescents, and adults. Acute esotropia occurs less frequently than either infantile esotropia or accommodative esotropia. Acute esotropia can be associated with decompensated esophoria, 60 late-onset accommodative esotropia, 61 abducens nerve or lateral rectus palsy, 62 divergence paralysis/divergence insufficiency, 63-65 or acute acquired comitant esotropia. 66-71 Divergence paralysis/insufficiency is an acute esotropia occurring only at distance in adults. More recently, its cause has been attributed to possible anatomical changes in the orbit and/or extraocular muscles associated with aging. 72

Acute exotropia occurs less frequently than acute esotropia. Because isolated medial rectus palsy is rare, acute exotropia is usually associated

with decompensated exophoria, acquired oculomotor nerve palsy, or acquired mechanical exotropia.

#### b. Signs, Symptoms, and Complications

Table 2 presents the signs, symptoms, and complications of the various types of acute esotropia.

#### c. Early Detection and Prevention

The patient with acute esotropia or exotropia should be examined immediately, due to the possibility of an underlying disease process (see Table 1). Consultation with other health care professionals may be needed to determine the underlying cause of abducens nerve palsy, divergence paralysis/insufficiency, or acute acquired comitant esotropia, as well as acute exotropia in those cases in which the clinician is uncertain of the cause.

#### 3. Consecutive Esotropia and Exotropia

#### a. Natural History

Consecutive esotropia is iatrogenic, developing only following surgical treatment of an exotropia. Temporary surgical overcorrection has been recommended to prevent recurring exotropia. Persistent consecutive esotropia, however, can cause amblyopia and loss of stereopsis in young patients, and it can cause bothersome diplopia in adults. Consecutive accommodative esotropia has been reported in hyperopic children following surgical treatment of intermittent exotropia. On the other hand, consecutive exotropia can occur postsurgically or develop spontaneously over time. Predisposing factors for cases that occur spontaneously include hyperopia greater than 4.50 D, amblyopia, and poor fusion. <sup>25</sup>

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#### b. Signs, Symptoms, and Complications

The patient with consecutive esotropia may exhibit these characteristics:

- Constant unilateral esotropia
- Amblyopia and loss of stereopsis, which may occur if the esotropia is allowed to persist in young children
- Diplopia.

The patient with consecutive exotropia may exhibit the following:

- Constant unilateral exotropia at distance and near
- Hyperopia greater than 4.50 D
- Amblyopia and medial rectus restriction, which are common in iatrogenic cases 75,76
- Reduced potential for normal binocular vision and high-level stereopsis
- Diplopia.

#### c. Early Detection and Prevention

In patients who receive surgical treatment for intermittent exotropia and then become esotropic, spontaneous reduction in the consecutive esotropia usually occurs within 2–3 weeks. Persistent small-angle esotropia may develop in 5–10 percent of these patients.<sup>73</sup> Immediate evaluation is essential, because amblyopia may develop in children less than 6 years old; loss of fusion and stereopsis may result at any age.<sup>77-79</sup>

Nevertheless, patients with consecutive exotropia generally do not require immediate treatment unless diplopia develops. Diplopia is more likely to occur when the exotropia develops in adulthood and there is no suppression area on the temporal hemiretina.<sup>80</sup>

#### Statement of the Problem 17

## **Table** Clinical Conditions

| Condition                                    | Deviation<br>in Primary<br>Position           | Motility   | Refraction                                  | Diplopia                      |
|--|---|--|---|-------------------------------|
| Decompensated heterophoria                   | 10-20 PD at<br>distance and<br>near           | Normal   | Similar to non-<br>strabismic<br>population | Yes; at distance<br>and near  |
| Late-onset<br>accommodative<br>esotropia     | 10-35 PD;<br>may be larger<br>at near         | Normal   | Causative (2-6 D) hyperopia                 | Yes; usually<br>more at near  |
| Abducens nerve<br>or lateral rectus<br>palsy | 20-40 PD;<br>larger at<br>distance            | Abnormal;<br>restricted<br>abduction in<br>one or both<br>eyes | Similar to<br>nonstrabismic<br>population   | Yes; usually more at distance |
| Divergence<br>paralysis/<br>insufficiency    | 8-30 PD at<br>distance;<br>4-18 PD at<br>near | Normal   | Similar to<br>nonstrabismic<br>population   | Yes; only at distance         |
| Acute acquired comitant esotropia            | 15-75 PD at distance and near                 | Normal   | Similar to<br>nonstrabismic<br>population   | Yes                           |

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### Presenting as Acute Esotropia

| Fusion                                  | Systemic/<br>Neurologic<br>Disease | Comment   |
|---|------------------------------------|---|
| Yes                                     | No                                 | History of pre-existing esophoria that has become manifest; can be provoked by prolonged occlusion or febrile disease.      |
| Yes                                     | No                                 | Can be prevented by correcting high hyperopic refractive error during childhood.  |
| Yes, may<br>have to<br>use head<br>turn | Frequent                           | Abducens nerve or lateral rectus palsy should always be suspected with acute-onset esotropia.                               |
| Yes                                     | Sometimes                          | Evidence suggests this strabismus may be caused by anatomical changes in the extraocular muscles with aging.                |
| Most of the time                        | Sometimes                          | Patients with related disease may<br>also have poor fusion and other<br>abnormal ophthalmic findings, such<br>as nystagmus. |

#### 4. Infantile Esotropia

#### a. Natural History

Studies indicate that infantile esotropia is not congenital and most likely develops between 2 and 4 months of age, a period during which most infants are becoming orthotropic. <sup>14,81,82</sup> A history of infantile esotropia in parents or siblings of affected patients is common. Usually characterized by a large and constant deviation, infantile esotropia can be associated with cross-fixation (viewing targets in the right field of gaze with the left eye and vice versa), severely interfering with the development of normal binocular vision. It must be distinguished from pseudoesotropia and other early-onset esotropias, including Duane syndrome, congenital abducens nerve or lateral rectus palsy, Moebius' syndrome, and sensory and accommodative esotropias. <sup>83</sup>

#### b. Signs, Symptoms, and Complications

Patients with infantile esotropia may exhibit any of the following:

- A large-angle, usually constant, esotropia (generally 40–60 PD) at distance and near that begins before 6 months of age
- Refractive errors skewed toward low to moderate hyperopia (approximately 50 percent have hyperopia exceeding 2 D)<sup>84</sup>
- Amblyopia (about 40 percent of all cases)<sup>85</sup>
- A high occurrence of various ocular motility disorders, including limited abduction, overelevation in adduction (inferior oblique overaction), and dissociated vertical deviation
- Limited potential for both normal binocular vision and high levels of stereopsis
- Latent or manifest nystagmus
- Monocular nasotemporal-pursuit optokinetic nystagmus (OKN) asymmetry.

#### c. Early Detection and Prevention

Transient strabismic deviations, which are mostly exotropia, occur frequently in neonates, but in most cases have been reported to resolve

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by 3–4 months of age. <sup>81,82</sup> Instability of ocular alignment also occurs in infantile esotropia. More than 25 percent of patients with esotropia at 2–4 months of age experience spontaneous resolution and become orthophoric, usually by the age of 6 months. <sup>86</sup> The resolution of the esotropia is more likely with intermittent, small-angle, variable esotropia and less likely in infants with constant esotropia exceeding 40 PD. <sup>86,87</sup> Any infant with intermittent, small-angle, variable esotropia that persists beyond 6 months of age should be examined immediately. Because strabismus may be secondary to a more serious condition, early examination should be encouraged, especially in the presence of any additional ocular findings (e.g., leukocoria).

#### 5. Infantile Exotropia

#### a. Natural History

Usually characterized by a large, constant deviation, infantile exotropia severely interferes with the development of normal binocular vision. It must be differentiated from the rather common transient exotropias seen during the first few months of life, <sup>14,81,82</sup> as well as from constant exotropia caused by poor vision in one eye. Any exotropia that persists beyond the age of 4 months is abonormal. The development of constant and persistent exotropia in a neurologically normal child 6 months of age or younger is extremely rare, occurring in only 1 of 30,000 patients. <sup>88</sup>

#### b. Signs, Symptoms, and Complications

Patients with infantile exotropia can be characterized by the following:

- A large, constant deviation (generally 30–80 PD) at both distance and near, beginning before 6 months of age<sup>88,89</sup>
- A high occurrence of various ocular motility disorders, including limited adduction, overelevation in adduction (inferior oblique overaction), and dissociated vertical deviation
- OKN and monocular nasotemporal pursuit asymmetry.

Infantile exotropia is rarely amblyopiogenic because most patients have an alternating fixation pattern; however, patients with infantile exotropia have limited potential for achieving normal binocular vision and highlevel stereopsis.

#### c. Early Detection and Prevention

Although benign transient exotropic deviations frequently occur in neurologically normal children during the first months of life, 81,82 the infant with constant unilateral exotropia requires immediate evaluation to confirm or rule out sensory exotropia. Any infant who has exotropia that persists and who has not previously been evaluated should be examined immediately.

#### 6. Intermittent Exotropia

#### a. Natural History

Most exotropias are intermittent and may be basic, convergence-insufficiency, or divergence-excess type. When intermittent exotropia occurs in younger children, it is usually the divergence-excess type. Often, the parents note that the child is not looking at them properly or tends to close one eye when viewing at distance or in bright sunlight outdoors. Basic and convergence-insufficiency exotropias are more likely in older persons. Intermittent exotropia must be distinguished from constant exotropia (e.g., infantile, consecutive, and sensory exotropia, Duane syndrome type II, and oculomotor nerve palsy).

#### b. Signs, Symptoms, and Complications

Patients with intermittent exotropia may exhibit the following characteristics:

• A significant exotropia at one or more fixation distances. The exotropia may not be apparent until the patient becomes fatigued or inattentive, or after prolonged dissociation. Control of intermittent exotropia can vary throughout the day, sometimes

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even within minutes, changing from exophoria to exotropia and exotropia to exophoria 90

- Minimal or no amblyopia
- Refractive errors similar to those in the nonstrabismic population
- Reduced positive fusional vergence amplitudes and facility at one or more distances
- Levels of stereopsis equal to or greater than 60 seconds of arc when fusing; no stereopsis when exotropic
- Discomfort (e.g., headaches, difficulty reading, and eyestrain) during or following prolonged visual activity
- Closing one eye in bright sunlight
- Diplopia, which usually implies that the intermittent exotropia developed after early childhood
- Suppression and/or anomalous retinal correspondence in the patient who does not report diplopia when the eye is exotropic
- Associated accommodative dysfunction, which is more likely with convergence-insufficiency exotropia. 91

#### c. Early Detection and Prevention

Unlike other strabismic deviations, delay in treatment of intermittent exotropia is not likely to result in permanently worsened visual status for very young children. Nevertheless, professional confirmation is essential at an early age, to differentiate intermittent exotropia from those exotropias that are amblyopiogenic and likely to cause loss of normal binocular vision. If treatment is postponed, the child should be monitored closely. Timely intervention is needed to prevent visual symptoms that may affect school and job performance.

#### 7. Mechanical Esotropia and Exotropia

#### a. Natural History

Mechanical strabismus can be either congenital or acquired. Fibrosis of most or all of the extraocular muscles can be present at birth. In thyroid myopathy, a condition that is usually acquired in adulthood, enlargement of the extraocular muscles, particularly the inferior rectus and medial rectus, causes esotropia and hypotropia.

#### b. Signs, Symptoms, and Complications

Mechanical esotropia or exotropia may be manifested in patients in the following ways:

- Small or moderate size esotropia or exotropia in the primary position 92
- Significantly increased deviation in right or left gaze
- Restricted horizontal versions and ductions
- Frequent compensatory head turns, which usually indicate the absence of amblyopia and the potential for normal binocular vision 93

#### c. Early Detection and Prevention

The patient with mechanical strabismus requires immediate evaluation to distinguish this condition from the more ominous paretic strabismus, which is more likely caused by a disease process.

#### 8. Microtropia

#### a. Natural History

Microtropia frequently results from the treatment of a larger-angle esotropia or exotropia by optical correction, vision therapy, pharmacological agents, and/or extraocular muscle surgery. Microtropia can also occur idiopathically or secondary to anisometropia. 92,94,95,

#### b. Signs, Symptoms, and Complications

The patient with microtropia usually exhibits the following:

- A constant, unilateral esotropia of less than 10 PD at distance and near (constant, unilateral exotropia of less than 10 PD is rare)
- Amblyopia
- Eccentric fixation
- Rudimentary binocular vision
- Anomalous retinal correspondence

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- Deficient stereopsis
- Anisometropia.

#### c. Early Detection and Prevention

Microtropia is usually diagnosed later than the more obvious types of strabismus; however, because of the high incidence of amblyopia, any child suspected of having microtropia should be evaluated immediately.

#### 9. Sensory Esotropia

#### a. Natural History

Most cases of sensory esotropia develop within the first 5 years of life. In this relatively uncommon type of esotropia, congenital or traumatic unilateral cataracts account for nearly 30 percent of all cases. Esotropia develops because an organic visual deficit creates an obstacle to fusion. The degree of monocular visual impairment leading to sensory esotropia ranges from 20/60 to light perception. 18

#### b. Signs, Symptoms, and Complications

Patients with sensory esotropia may exhibit the following:

- Constant unilateral esotropia at distance and near
- High degrees of anisometropia
- Vertical deviations associated with overaction of the inferior and/or superior oblique muscles
- Functional amblyopia superimposed on the organically caused vision loss<sup>96</sup>
- Limited potential for normal binocular vision and high-level stereopsis.

#### c. Early Detection and Prevention

Patients with suspected sensory esotropia should be evaluated immediately to determine the cause for the vision loss and strabismus. For example, there is greater urgency in infants, from whom congenital

cataracts need to be removed within the first weeks of life in the hope of obtaining normal visual acuity and possibly binocular vision. <sup>97,98</sup>

#### 10. Sensory Exotropia

#### a. Natural History

Sensory exotropia occurs in both children and adults, but it is more common in adults. 18,99 Although it occurs more frequently than sensory esotropia, it has the same causative factors. The relative decrease in tonic convergence with age is thought to result in the higher incidence of sensory exotropia.

#### b. Signs, Symptoms, and Complications

Patients with sensory exotropia may exhibit these characteristics:

- Constant unilateral exotropia at distance and near
- High degrees of anisometropia, which can be causative
- Frequently accompanying vertical deviations associated with overactivity of the inferior and/or superior oblique muscles
- Functional amblyopia that may be superimposed on the organically caused vision loss
- Reduced potential for normal binocular vision and high-level stereopsis in patients with early childhood onset.

#### c. Early Detection and Prevention

As with sensory esotropia, the patient with suspected sensory exotropia should be evaluated immediately to determine the cause of the vision loss and strabismus.

#### II. CARE PROCESS

#### A. Diagnosis of Strabismus

The examination of strabismic patients generally includes all areas of the evaluation of a comprehensive adult or pediatric eye and vision examination.\* The evaluation of sensory, motor, refractive, and accommodative functions requires further, in-depth examination. Additional office visits may be required to complete the examination process, especially with younger children.

The evaluation of a patient with strabismus may include, but is not limited to, the following components. Professional judgment and individual patient symptoms and findings may have significant impact on the nature, extent, and course of the services provided. Some components of care may be delegated or referred to other practitioners.

#### 1. Patient History

In addition to gathering information about the patient's general and eye health history, the clinician should also determine:

- Probable time of onset of strabismus
- Nature of the onset (sudden or gradual)
- Frequency of deviation (constant or intermittent)
- Change in size or frequency of the deviation
- Whether the strabismus is unilateral or alternating
- Presence or absence of diplopia and other visual symptoms or signs
- Presence or absence of any compensatory head posture
- History of neurologic, systemic, or developmental disorders
- Family history of strabismus
- Previous treatment, if any, and the type and results of such treatment.

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<sup>\*</sup> Refer to the Optometric Clinical Practice Guidelines on Comprehensive Adult Eye and Vision Examination and Pediatric Eye and Vision Examination.

#### 2. Ocular Examination

#### a. Visual Acuity

Measurement of the visual acuity of each eye with optimum refractive correction helps to establish the presence or absence of amblyopia. In very young children (up to the age of 2 years), who do not respond reliably to subjective visual acuity testing, and in individuals who are nonverbal or otherwise not responsive to a subjective examination, a definite fixation preference with strabismus has been the primary indicator for the diagnosis of amblyopia. Any child with constant unilateral strabismus is vulnerable to developing amblyopia. A definite fixation preference can also occur in the absence of amblyopia, possibly resulting in unnecessary treatment. An alternating fixation pattern, intermittent strabismus, or incomitant strabismus with a compensatory head posture usually precludes the development of amblyopia in the young child. When amblyopia exists, it usually should be addressed before making any effort to establish normal binocular vision.\*

Quantification of visual acuity for children 2 years old or younger can sometimes be accomplished by using preferential looking tests such as the Teller acuity cards. 102 For children ages 3–5 years, visual acuity tests such as the Cardiff cards and the Lea symbol visual acuity test are preferred. The Lea test controls the distance between the symbols or optotypes, making them equal to the width of the symbols. With older individuals, psychometric acuity cards can be used, or standard Snellen visual acuity measurements can be taken. 103 However, the Snellen chart may cause errors and inefficiencies because of the unequal space increments between one level of visual acuity and the next. Presenting isolated Snellen acuity targets may result in underestimation of the degree of amblyopia. 104 The Pediatric Eye Disease Investigator Group has recommended using HOTV letters with surround bars for children 3–7 years old and the Early Treatment Diabetic Retinopathy Study (ETDRS) vision test with surround bars for older children to quantify the visual acuity of strabismic children with amblyopia. 105

#### b. Ocular Motor Deviation

The diagnosis of strabismus, including the direction and frequency of the deviation, may be established by performing a unilateral or coveruncover test at distance and near while the patient fixates a target that controls for accommodation. The alternate cover test with prisms is useful in determining the magnitude of the ocular deviation. After initial measurement of the strabismus in the primary positions of gaze, measurements should be made in all other fields of gaze to determine whether the strabismus is comitant or incomitant. A change of less than 10 PD in strabismus measurements from one visit to the next obtained with the alternate cover test with prisms may be attributable more to test-retest variability rather than real change. When evaluating very young children and nonverbal or otherwise nonresponsive patients who cannot fixate on a target long enough for valid cover testing, the optometrist can estimate the degree of strabismus using the corneal reflex test with prisms (Krimsky test) or without prisms (Hirschberg test). The method of measurement and the presence or absence of refractive correction during the measurement should be documented.

#### c. Monocular Fixation

The method of choice for evaluating monocular fixation is visuoscopy, using an ophthalmoscope with a calibrated fixation target. The practitioner should determine whether eccentric fixation is present and, if so, assess its characteristics: location, magnitude, and steadiness. When there is no foveal reflex, entoptic testing, such as Haidinger's brushes or Maxwell's spot, can be useful in the assessment of monocular fixation in older children.

#### d. Extraocular Muscle Function

To determine a reasonable prognosis and management approach, it is important to establish whether the ocular deviation is comitant or incomitant. Direct observation of any abnormal head position can aid the evaluation of comitancy. In addition, version and duction testing can provide objective determination of ocular muscle imbalances in various positions of gaze.

<sup>\*</sup> Refer to the Optometric Clinical Practice Guideline on Care of the Patient with Amblyopia.

The evaluation of both versions and ductions should be performed without spectacle correction. Common extraocular muscle disorders include limited abduction, limited adduction, and overelevation in adduction or overdepression in adduction (overaction of the inferior oblique or superior oblique muscles). The presence of an extraocular muscle palsy or mechanical restriction should be noted and may require additional clinical testing, such as the Maddox rod test, the three-step test in which vertical strabismus is imposed, and the forced duction test for differential diagnosis.

#### e. Sensorimotor Fusion

The ability to determine the presence of fusion potential by sensory testing may be limited by the patient's age and cognitive ability. Tests such as the Worth 4-dot test at distance and near and tests for stereopsis may be used. Among commonly used measures of stereopsis are the Randot and Preschool Randot stereo tests. More detailed sensory testing (e.g., the Bagolini striated lenses, Hering-Bielschowsky afterimage, and synoptophore) can be used to evaluate retinal correspondence in older children and adults.

All sensory testing should be performed while the patient is wearing his/her optimum refractive correction. In addition, performing sensory testing while the patient wears prisms to compensate for any nonaccommodative component of the strabismus can also help determine sensory fusion potential. Once normal sensory fusion has been established, motor fusion can be quantified using a prism bar or rotary prisms for the patient with intermittent strabismus or a stereoscope for the patient with constant strabismus.

#### f. Accommodation

When feasible, an evaluation of accommodative function, including tests of monocular accommodative amplitude (push-up or minus lens method), accommodative facility (plus/minus flipper method), and accommodative response (dynamic retinoscopy), should be performed.

#### g. Refraction

An accurate objective measurement of refractive error is essential, because it is often an important etiologic factor in the development of strabismus. The patient's refractive condition generally should be evaluated under both noncycloplegic and cycloplegic conditions.\* The instillation of 1 drop of 1% cyclopentolate hydrochloride twice at 5-minute intervals followed by retinoscopy 30–40 minutes later is usually adequate. In cycloplegic retinoscopy, it is best to occlude the eye not being refracted and have the patient view the retinoscope to avoid being off axis. For examining young children, hand-held lenses or lens bars may be preferable. Repeated refractions are frequently required during the course of strabismus treatment and management.

#### h. Ocular Health Assessment and Systemic Health Screening

Ocular health should be evaluated to rule out coexisting or causal congenital anomalies or disease associated with strabismus. Pharmacologic dilation of the pupil is generally required for thorough evaluation of the ocular media and the posterior segment.

#### **B.** Management of Strabismus

Management of the strabismic patient is based on the interpretation and analysis of the examination results and overall evaluation (see Appendix Figure 1). The goals of treatment and management may include (1) obtaining normal visual acuity in each eye, (2) obtaining and/or improving fusion, (3) eliminating any associated sensory adaptations, and (4) obtaining a favorable functional appearance of the alignment of the eyes. The significance of normal ocular alignment for the development of a positive self-image and interpersonal eye contact cannot be overemphasized.<sup>67,110</sup>

Refer to the Optometric Clinical Practice Guideline on Pediatric Eye and Vision Examination.

#### 1. Basis for Treatment

The indications for treatment and management and the specific types of treatment and management need to be individualized for each patient. In determining a course of therapy, the optometrist should consider the following:

- Age of the patient at the onset of strabismus
- Current age of the patient
- Overall health status of the patient
- Patient's developmental level and anticipated compliance with therapy
- Concerns of the patient and/or parents
- Symptoms and signs of visual discomfort
- Visual demands of the patient
- Comitancy of the deviation
- Size and frequency of the strabismus
- Presence or absence of fusion
- Presence or absence of amblyopia.

#### 2. Available Treatment Options

The treatment and management of strabismus may include any or all of the following procedures.

#### a. Optical Correction

Regardless of the cause of the strabismus, the goal for strabismic patients, especially very young patients, is to allow binocularity to develop. The best optical correction that allows equally clear retinal images to be formed in each eye is generally the starting point for all treatment and management. However, overcorrection or undercorrection of the refractive error may be prescribed in some instances to affect the angle of strabismus.

Hyperopia may be either partly or totally causative in as many as 50 percent of all cases of esotropia. <sup>15,16,32,33</sup> Generally, when clinically significant amounts of hyperopia are present, the total amount of lens

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power needed to achieve ocular alignment is prescribed. Anisometropia and astigmatism should also be fully corrected. The full prescription of previously uncorrected refractive errors is usually well accepted by younger children; however, if sensory fusion is difficult or if the patient is unable to adapt to a full prescription, undercorrection of refractive errors may be prescribed initially.

Whereas a full correction of refractive error is often prescribed for esotropia and hyperopia, the presence of exotropia and hyperopia may require a more conservative approach. For preteens and teenagers, reduction of the full refractive correction can be attempted if the strabismus is still comfortably controlled. For adults, the refractive correction should be prescribed to the extent tolerated by the patient.

The clinician should continue to re-evaluate the prescribed lenses periodically to assess the effect on the angle of deviation and fusion. The patient should be advised that changes in the lenses may be needed during treatment and management.

#### b. Added Lens Power

Lenses can also be used to take advantage of the AC/A ratio to help obtain or maintain binocular vision. A bifocal lens prescription may be used for the patient with fusion potential or when full plus acceptance at distance cannot be attained initially. Periodic follow-up is required to determine the efficacy of this treatment and management.

Bifocals are often prescribed for the patient with esotropia who has a high AC/A ratio, to eliminate or decrease the angle of strabismus at near to an amount controllable by compensating divergence. Added convex lens power may also be indicated when the esotropic deviation is larger for near than for distance, or when the ocular alignment at distance with the hyperopic correction permits binocular vision but an esotropia remains at near. In that case, the optometrist may prescribe the minimum added convex lens power to allow fusion at near.

For young children, wide-segment bifocals that either bisect the pupils or intersect the lower lid margin are generally prescribed. For older

children and teenagers who still need bifocals, the optometrist can prescribe standard bifocals or progressive-addition lenses. 111 or, as fusional vergence ranges increase. fit the patient with contact lenses 112,113

Added minus lens power (e.g., an undercorrection of hyperopia or an overcorrection of myopia) can be temporarily prescribed in young children for intermittent exotropia that measures the same for distance and near or is larger for distance. 114 With this correction, the patient uses the added accommodative convergence response to stimulate the fusional vergence system. Fusional vergence sometimes increases to the extent that the added minus lenses are no longer needed. Research has shown that approximately 70 percent of patients whose intermittent exotropia was managed with added concave lens power develop improved fusion. 115, 116 Added minus lens power is contraindicated in patients whose exotropia is associated with accommodative insufficiency or who are presbyopic. 91

Management with added minus lens power should be discontinued when the frequency of the exotropia remains unchanged despite the wearing of added minus lens power, or when fusion at near becomes disrupted. A large lag in accommodation, as measured by dynamic retinoscopy, may indicate that the patient is having accommodative difficulties at near with the added minus lens power. In such cases, multifocal lenses may be prescribed. Concern that using added concave lens power might cause large increases in myopia has not been substantiated. 117,118

Patient compliance in wearing prescribed lenses is crucial to the success of any treatment or management plan. The lens prescription must be acceptable to the patient and worn as directed.

#### Prisms

Ophthalmic prisms can aid in the establishment or maintenance of sensory fusion, by moving the image of the target of regard onto or closer to the fovea of each eye. 119 Prisms are generally prescribed for patients with strabismic deviations of less than 20 PD who are capable of fusion. 120 The presence of amblyopia, deep suppression, and/or

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anomalous retinal correspondence generally contraindicates the use of prisms. The maximum prism power that can be incorporated into spectacle lenses is approximately 10–12 PD in each lens (if low-powered lenses). Press-on plastic (Fresnel) prisms to promote binocular vision in early-childhood strabismus, or to alleviate diplopia in late-onset strabismus, are especially helpful in treating larger angles of strabismus. 108 Although as much as 30 PD can be prescribed for each lens using Fresnel prisms, the larger amounts tend to decrease visual acuity, increase chromatic dispersion, and decrease contrast. Disruptive prisms (i.e., overcorrecting or inverse) may be prescribed to eliminate anomalous retinal correspondence. <sup>121</sup> In addition, inverse prisms may be used to improve the cosmetic appearance of the strabismic patient who has a poor prognosis for attaining normal binocularity and is not interested in surgery. 122

The prescription of prisms less than the strabismic angle may allow patients who have some fusional vergence ranges to maintain some active motor fusion. 108 Relative to the magnitude of the deviation, less prism is generally needed for exotropia than for esotropia.

Prisms may also be used to reduce or eliminate mild compensatory head postures in patients with incomitant strabismus. Older patients who have diplopia in association with acquired extraocular muscle palsy, muscle restriction, or phoria decompensation also may benefit from prisms.

Patients managed with prisms need periodic evaluation to determine treatment efficacy. Some patients, especially those with esotropia, manifest increased angles of strabismus while wearing the prisms. 123,124 This change may represent an uncovering of the total deviation, some of which was kept latent by motor fusion or may represent decreased sensory fusion. 108 In the latter case, removal of these prisms usually allows a return to the original angle of strabismus within a few days.

#### Vision Therapy

Vision therapy or orthoptics involves active training procedures to improve the patient's fixation ability and oculomotor control, to help eliminate amblyopia, to improve sensory and motor fusion, and to

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increase facility and the range of accommodation and vergence responses. <sup>125,126</sup> Used alone or in conjunction with refractive correction, added lens power, prisms, or surgery, these vision therapy procedures are adapted to the individual patient and modified as the patient achieves binocular vision.

Indications for treating strabismus with vision therapy vary, depending on the type of strabismus and the patient's sensorimotor fusion status. Vision therapy is successful in the treatment of many forms of strabismus. The prognosis is most favorable for patients with intermittent strabismus, especially intermittent exotropia, who have sensorimotor fusion at some point in space and those with recently developed strabismus. Pevertheless, some patients with constant or longstanding strabismus may also be successfully treated with vision therapy, especially when there is fusion potential. 125,126,130,131,133-135

The optometrist may prescribe active vision therapy or refer the patient to an optometrist who has advanced training or clinical experience with strabismus. The time required for therapy depends upon the type of strabismus, the presence or absence of associated visual adaptations and/or visual anomalies, and patient compliance. Office treatment usually requires 24–75 hours of therapy. Patients are usually treated for 30–60 minutes once or twice a week in the office. In addition, home treatment may also be prescribed, often requiring 20-60 minutes per day. During office visits, the optometrist reviews home treatment and prescribes appropriate changes as the patient shows progress with therapy.

#### e. Pharmacological Agents

Pharmacological therapy\* historically provided a potentially useful treatment option for the management of some patients with strabismus. Anticholinesterase miotics such as echothiophate iodide (Phospholine Iodide®) served as temporary alternatives to corrective glasses and

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bifocal lenses for children with accommodative esotropia. <sup>109,136</sup> These drugs act by reducing the patient's accommodative effort and decreasing the associated accommodative-convergence mechanism. <sup>109</sup> The initial dose is usually 0.125% Phospholine Iodide<sup>®</sup> (1 drop q.d.), tapered downward to a level that maintains the desired result. A trial period of up to 8 weeks is needed to determine whether a reduction in esotropia has been obtained. <sup>136</sup>

Treatment with a pharmacological agent has proven less effective and less desirable than using corrective glasses and bifocal lenses, because of the possibility of both local and systemic adverse effects. Such treatment is rarely used in contemporary practice and should be considered for only those patients with accommodative esotropia who cannot wear glasses due to facial deformities, for children who continually remove, lose, or break their spectacles, or for other special cases.

However, pharmacological agents that dilate the pupil are frequently used in amblyopia therapy for strabismic patients.\* Referred to as pharmacologic penalization, atropine 1% is put in the non-amblyopic eye on either a daily or weekend only basis. The child wears his/her spectacles while undergoing this treatment. By dilating the pupil and inhibiting accommodation in the non-amblyopic eye, the atropine drops force the child to use only the amblyopic eye for near viewing. Augmentation by using a plano lens for the non-amblyopic eye may enhance the treatment effect by forcing use of the amblyopic eye for near and also for distance. Studies have concluded that atropine, although somewhat slower, is as effective as patching for young children who have moderate amblyopia. 137

#### f. Extraocular Muscle Surgery

The clinician should consider all aspects of the nonsurgical treatment of strabismus before recommending surgery. Surgical consultation is appropriate for patients whose strabismus is cosmetically objectionable,

<sup>\*</sup> Every effort has been made to ensure the drug dosage recommendations are appropriate at the time of publication of the Guideline. However, as treatment recommendations change due to continuing research and clinical experience, clinicians should verify drug dosage schedules with product information sheets.

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as well as for patients who may not display the intellectual, motivational, or physiological characteristics (including fusion potential) that warrant consideration of other treatment.<sup>126</sup>

In general, surgery for esotropia may be considered when the manifest deviation exceeds 15 PD in the primary position at both distance and near while the patient is wearing the full refractive correction. For patients with exotropia, deviations exceeding 20 PD in the primary position are possible candidates for surgery. Patients with smaller deviations usually should not be considered for surgery, except when adults have acquired symptomatic deviations that do not respond to nonsurgical therapy. Patients with totally accommodative esotropia should not be considered for extraocular muscle surgery, because of the risk of inducing consecutive exotropia. 16,138

Strabismic surgery should be considered for patients whose best-corrected strabismic angle remains too large for binocular comfort and acceptable cosmesis. Strabismus surgery can restore the ocular alignment to normal, or, at least, closer to normal. Expansion of the binocular field of vision occurs for esotropia while reduction of the binocular field occurs for exotropia after surgery. Preoperative and/or postoperative vision therapy should be considered, when appropriate, to enhance functional vision outcomes. <sup>125,126</sup>

The timing and urgency for surgical referral depend upon the type of strabismus, the age of the patient, and the likelihood of improving fusion. Ideally, children with infantile strabismus requiring surgical correction should undergo surgery prior to 2 years of age. Several studies have shown that a better chance of developing rudimentary binocular vision with limited stereopsis when surgery is performed at an early age and when the duration of ocular misalignment has not been extensive. Binocular vision is best achieved when the postsurgical alignment is within 10 PD of orthotropia, whereas a residual deviation of 4 PD or less is usually required to achieve stereopsis. Multiple surgeries are often needed to obtain this result. Possible complications following surgery include diplopia, undercorrection, overcorrection, chronic inflammation of the conjunctiva, excessive scar tissue, lost muscle(s), perforation of the globe, endophthalmitis, anterior segment ischemia, retrobulbar

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hemorrhage, conjunctival pyogenic granulomas, and corneal dellen. <sup>146-148</sup> Surgery is rarely performed at such an early age for other childhood strabismic deviations (e.g., intermittent exotropia). The overall success rate for surgical therapy for horizontal strabismus is approximately 60 percent, when success is defined as an ocular deviation 10 PD or less at 6 weeks postsurgery. <sup>149</sup>

#### g. Chemodenervation

The injection of botulinum toxin type A (Oculinum, Botox®) has been used as either an alternative or an adjunct to conventional incisional surgery in selected strabismic patients. 150-152 The toxin selectively binds to nerve terminals and interferes with the release of acetylcholine. thereby functionally denervating muscles injected with small amounts of the drug. The dose-related but temporary paralysis of an extraocular muscle leads to a change in eye position, followed by some degree of contracture of the opposing muscle. This change has been reported to result in long-lasting and permanent alteration in ocular alignment. Although one injection is often sufficient to produce positive results. one-third to one-half of patients may require additional injections. Transient ptosis and vertical strabismus may develop after chemodenervation. This technique has been most successfully used in patients who have acute abducens nerve palsy and in adults with smallangle deviations. Success with certain types of pediatric strabismus has also been reported. 153

#### 3. Management Strategies for Strabismus

#### a. Accommodative Esotropia

After the diagnosis of accommodative esotropia has been confirmed, correction of the amount of hyperopia needed to obtain ocular alignment should be provided. If present, amblyopia should be treated. The clinician may prescribe active vision therapy procedures for the development and enhancement of normal sensory and motor fusion.

Possible sequelae of treatment for accommodative esotropia are:

- Alignment at both distance and near with corrective lenses.
- Alignment at distance with corrective lenses, but persistence of esotropia at near. The remaining esotropia at near is usually treated or managed with additional near lens power. 15,16,108
- Esotropia persisting both at distance and near while the patient wears corrective lenses. Cycloplegic refraction should be repeated to determine whether additional hyperopia should be corrected.
- Esotropia persisting both at distance and at near and cycloplegic refraction revealing no additional hyperopia, showing the presence of a residual nonaccommodative esotropia. Other treatment options (e.g., prisms, vision therapy, surgery) should be considered. Nonaccommodative esotropia exceeding 15 PD may require extraocular muscle surgery. Surgery is performed with the intent of reducing or eliminating the nonaccommodative component, not the accommodative component, of the esotropia. The patient will continue to need to wear corrective lenses. 16,154

Some patients who at first achieve ocular alignment with corrective lenses may subsequently develop additional esotropia and risk losing binocular vision. This is more likely to occur in the following circumstances:

- Onset of accommodative esotropia during the first year of life
- Delay between the onset of the esotropia and the initiation of treatment
- Large increases in hyperopia
- Incomplete treatment (undercorrection of hyperopia, part-time wearing of corrective lenses, frequently losing glasses)<sup>16</sup>
- Elevated AC/A ratio.

Children with treated accommodative esotropia should be evaluated at intervals, according to the schedule in Appendix Figure 2. New or different findings may alter the frequency of follow-up care needed.

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#### b. Acute Esotropia and Exotropia

Once the cause of the esotropia or exotropia has been determined, prisms may be used to correct small and moderate deviations (except in late-onset accommodative esotropia) to eliminate diplopia and re-establish binocular vision. For larger and transient deviations, the optometrist can prescribe Fresnel prisms.

Vision therapy may be prescribed to expand fusional vergence amplitudes and facility. Surgical consultation may be considered for deviations that have become stable over time, when the angle of deviation exceeds 15–20 PD, and when the strabismus is cosmetically displeasing.

Associated mostly with abducens nerve palsy, and to a lesser extent divergence paralysis/divergence insufficiency and acute acquired comitant esotropia, acute esotropia may change over time. Complete remission is more likely when the cause is associated with vascular or ischemic disease (e.g., diabetes mellitus) and less likely when related to trauma. Patients with acute esotropia or exotropia should be followed as indicated in Appendix Figure 2, and therapy should be altered when necessary.

#### c. Consecutive Esotropia and Exotropia

Persistent consecutive esotropia following surgery for intermittent exotropia should be treated aggressively in young children, using lenses, prisms, and vision therapy to prevent possible amblyopia and loss of binocular vision. Older patients with consecutive esotropia following surgery frequently have diplopia and usually require similar treatment.

Consecutive exotropia that is spontaneous and optically induced can be treated by reducing the power of the hyperopic lenses. This is generally done in younger patients when the exotropia exceeds 20–25 PD. For older patients, reduction in the hyperopic correction may result in accommodative asthenopia, and alternative treatments may be needed.

#### d. Infantile Esotropia and Exotropia

Once the diagnosis of infantile esotropia has been confirmed, the clinician should make an effort to determine whether a superimposed accommodative component exists, by evaluating the effect of correcting the hyperopic refractive error on the angle of deviation. In most cases, a large esotropia persists, despite corrective lenses, and repeated cycloplegic refractions show little change in the amount of hyperopia.

Amblyopia, if present, should be treated.\* When amblyopia is suspected in preverbal patients who show strong fixation preference, occlusion therapy may be used until an alternating fixation pattern is established. The acquisition of alternating fixation implies resolution of amblyopia and should also prevent amblyopia regression. Two hours of daily occlusion of the preferred eye can be prescribed initially.\* Children with infantile esotropia who undergo occlusion therapy should be monitored every 4–6 weeks. Maintenance amblyopia therapy may be required for children whose amblyopia has resolved.

When the esotropia is large and nonaccommodative, surgical ocular alignment should be considered. Most ophthalmic surgeons prefer to intervene before 24 months of age, some as early as 6 months, in the hope of establishing binocular vision. However, the binocular vision achieved in these cases is usually not bifoveal, but peripheral, implying a fusion response, (i.e., detectable by the Worth 4-dot test at near but not at distance). Limited stereopsis, when possible, is best accomplished when the postsurgical strabismic deviation is within 4 PD of orthotropia. <sup>144</sup> Approximately 40 percent of treated cases of infantile esotropia achieve some stereopsis. Multiple surgical procedures are frequently needed in such cases. <sup>140,145,162</sup>

The most accepted approach to timing the surgery for infantile esotropia is to perform it after the deviation can be accurately determined, when it is stable on follow-up visits, and after adequate attention has been directed to correcting any accommodative component and treating amblyopia. The development of binocular vision for infantile esotropia

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appears to be related to the age at surgery, the duration of ocular misalignment, and the size and stability of postsurgical alignment. 141-143

There are several possible sequelae to surgical alignment for infantile esotropia: 162

- Recurring strabismus
- Accommodative esotropia
- Recurring amblyopia
- Increasing dissociated vertical deviation and inferior oblique overaction

Due to the instability of visual acuity, ocular alignment, refractive error, and extraocular muscle function, young children who have been treated for infantile esotropia should be evaluated according to the schedule in Appendix Figure 2. New or different findings may alter the frequency of follow-up care.

The treatment of infantile exotropia is similar to those for infantile esotropia. Treatment often includes surgery because of the large angle of constant exotropia. Prisms are usually not helpful in cases with poor fusion potential. Surgery should be considered after (1) the refractive error and fundus have been assessed, (2) amblyopia has been treated, (3) the angle of exotropia is stable on subsequent examinations, and (4) other treatment options are not appropriate or have failed. The likelihood of success in establishing any fusion is less with infantile exotropia than with infantile esotropia. <sup>163</sup>

#### e. Intermittent Exotropia

Individual cases of intermittent exotropia are treated in different ways and often by a combination of treatments. Therapy for intermittent exotropia should include correction of significant refractive error. Usually, the full amount of myopia, anisometropia, and astigmatism should be fully corrected. Hyperopia may be undercorrected for younger patients.

<sup>\*</sup> Refer to Optometric Clinical Practice Guideline on Care of the Patient with Amblyopia.

Added minus lens power may be used temporarily to help facilitate fusion in children with divergence excess or basic intermittent exotropia. The minimum minus lens power (generally 1–3 D) that will allow the patient to maintain alignment easily, as determined using the unilateral cover test, may be prescribed. An overall success rate of 70 percent has been estimated for cases of intermittent exotropia with this treatment modality, which can be used in conjunction with active vision therapy. <sup>115,116,130,131,165</sup>

Compensatory base-in prisms can be used to facilitate fusion. The degree of fusion varies, but generally the optometrist should prescribe enough prism to balance the patient's deviation with his or her fusional vergence amplitudes, so that the patient can maintain alignment without excessive effort. The estimated overall success rate for prism therapy with intermittent exotropia is 28 percent. Prism therapy is often used in conjunction with active vision therapy.

Numerous vision therapy procedures, including but not limited to expanding fusional vergence amplitudes and vergence facility, diplopia awareness, biofeedback, and increasing accommodation are prescribed for small and moderate size deviations. Although success has been reported with home-oriented therapy, more intensive office-based treatment may be required. The estimated overall success rate of orthoptics and vision therapy with intermittent exotropia is 59 percent. 165

Surgical intervention should be considered when, after a reasonable time, other treatment modalities have not been successful and the deviation persists or increases. Surgery is rarely performed in a child under 4 years of age (except in cases of a very large and frequent deviation) because of the increased risk for persistent consecutive esotropia with amblyopia and loss of stereopsis. <sup>78,79</sup> Intermittent exotropia has a high recidivism rate following surgery despite the excellent visual acuity and binocular vision. <sup>166</sup> Approximately 1 in 5 patients will require a second surgery. <sup>167</sup> The overall success rate of surgical therapy with intermittent exotropia has been estimated as 46 percent. <sup>165</sup> It is possibly higher for the subgroup undergoing surgery between the ages of 4 and 6 years.

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The course of intermittent exotropia, when untreated or when treatment recommendations have not been followed, is unclear. Lack of compliance would likely result in the continuation of signs and symptoms. However, retrospective studies that have followed patients over the years indicate that intermittent exotropia does not always get worse. <sup>23,24,168</sup> Some patients' vision improves. Getting worse implies that the frequency and magnitude of exotropia increase. Getting better is defined as the patient's becoming completely exophoric, with reestablishment of continuous fusion. Prospective studies on the natural course of intermittent exotropia and clinical trials to improve the evidence base for its management are needed to better understand this common type of strabismus.

For patients who have been treated (especially those treated surgically), the possibility of recurring intermittent exotropia exists. Patients with a history of intermittent exotropia should receive follow-up evaluations according to the schedule suggested in Appendix Figure 2.

#### f. Mechanical Esotropia and Exotropia

The patient with mechanical esotropia or exotropia may need no specific therapy if there is either minimal or no strabismus in the primary position of gaze and the patient does not experience diplopia. For example, treatment for Duane syndrome, which may have secondary tightness of the medial and/or lateral rectus muscle, is generally restricted to cases in which there is an objectionable compensatory head turn, a large angle strabismus in the primary position, extreme elevation or depression of the eye, or extreme retraction of the globe in adduction. Because head-turning is prevalent, amblyopia is uncommon and high-level stereopsis usually exists. Prisms may be prescribed for slight head turns. For large head turns, surgery may be used, but it does not improve the deficient abduction or adduction.

#### g. Microtropia

Microtropia is a fully adapted strabismus that rarely gives rise to symptoms unless other conditions become superimposed. Its treatment consists mostly of correcting significant refractive errors and any

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coexisting amblyopia. The use of vision therapy and prisms to establish bifoveal fusion and high-level stereopsis has been successful in selected cases of microtropia. <sup>169</sup>

#### h. Sensory Esotropia and Exotropia

Infants diagnosed at birth with sensory esotropia or exotropia due to unilateral congenital cataracts should be surgically treated within the first 2 months of life. Treatment and management may include cataract surgery, optical correction with contact lens, or intraocular lens implants, and occlusion therapy for amblyopia. <sup>97,98</sup> Neutralizing prisms may be prescribed when fusion exists, and, depending upon the size of the deviation, subsequent strabismus surgery may be performed. The attainment of normal binocular vision is generally not a realistic goal.

In older children and adults with acquired sensory exotropia due to dense traumatic unilateral cataracts, fusion may be lost if the cataract remains in situ for more than 2 years, despite subsequent cataract extraction, prisms, vision therapy, and strabismus surgery. Therefore, treatment should not be delayed.

#### 4. Patient Education

The prognosis, advantages, and disadvantages of the various modes of treatment or management should be discussed with the patient and/or the patient's parents, and a plan based on this dialogue should be developed. Patients who suddenly develop strabismus of undetermined etiology should be informed that such an event may be related to a systemic or neurologic disease that would necessitate referral for consultation with, or treatment or management by, another health care provider.

It is important for parents of strabismic children to learn about the condition and the child's risks of developing amblyopia and impaired binocular depth perception. Treatment and management plans formulated in consultation with the patient and parents should be responsive to their preferences. The optometrist should elicit the child's and/or the parents' expectations for outcomes, advise the persons involved, relate the findings, prepare treatment and management plans,

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discuss options, and recommend strategies for successful treatment and management. Parents and children must understand that timely examination and management are critical to reducing the risk for loss of vision and fusion and the development of other symptoms associated with strabismus.

#### 5. Prognosis and Follow-up

The purpose of the follow-up evaluation is to assess the patient's response to therapy and to adjust treatment as needed (see Appendix Figure 2). The schedule of follow-up visits depends upon the patient's condition and associated circumstances. Follow-up evaluation includes monitoring of several aspects of the patient's condition:

- Patient history
- Visual acuity
- Characteristics of strabismus at distance and near
- Fusion status
- Extraocular muscle function
- Refractive error
- Tolerance, efficacy, and side effects of therapy.

#### Conclusion 47

#### CONCLUSION

The optometrist should emphasize the examination, diagnosis, timely and appropriate treatment and management, and careful follow-up of patients with strabismus. Proper care can result in reduction of personal suffering for those involved, as well as a substantial cost savings for persons with strabismus and their families.

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Figure 1
Optometric Management of the Patient With Strabismus:
A Brief Flowchart

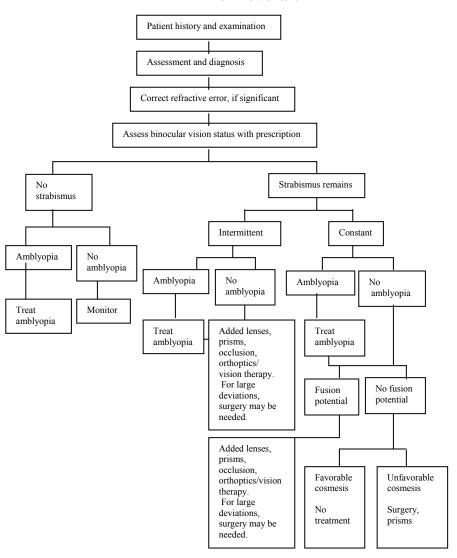


Figure Frequency and Composition of Evaluation and

| Type of Patient                                     | Number<br>of Evaluation<br>Visits | Treatment<br>Options   | Frequency<br>of Follow-up Visits,<br>by Age*                                       |
|---|-----------------------------------|--|--|
| Accommodative esotropia                             | 1–3                               | Optical correction<br>Added lenses<br>Vision therapy                         | < 6 yr: every 4–6 mo<br>6–10 yr: every 6–12 mo<br>≥11 yr: every 12 mo              |
| Acute esotropia and exotropia                       | 1–3                               | Prisms Vision therapy Surgery when applicable                                | Every 3–12 mo  |
| Consecutive esotropia and exotropia                 | 1–3                               | Optical correction<br>Prisms<br>Vision therapy<br>Surgery<br>when applicable | Variable, depending on etiology  |
| Infantile or early-acquired esotropia and exotropia | 1–3                               | Optical correction<br>Prisms<br>Vision therapy<br>Surgery<br>when applicable | <2 yr: every 3 mo 2–5 yr: every 4–6 mo 6–10 yr: every 12 mo ≥11 yr: every 12–24 mo |
| Intermittent exotropia                              | 1–3                               | Optical correction<br>Added lenses<br>Prisms<br>Vision therapy<br>Surgery    | <5 yr: every 4−6 mo 5−10 yr: every 6−12 mo ≥11 yr: every 12−24 mo                  |
| Mechanical esotropia and exotropia                  | 1-3                               | Prisms<br>Surgery  | Variable, depending on etiology  |
| Microtropia   | 1–3                               | Optical correction<br>Prisms and<br>vision therapy<br>when applicable        | Every 3–12 mo  |
| Sensory esotropia and exotropia                     | 1-3                               | Optical correction<br>Prisms<br>Vision therapy<br>Surgery when<br>applicable | Every 3–12 mo  |

<sup>\*</sup>Vision therapy would require additional visits.

#### 2 Management Visits for Esotropia and Exotropia

#### **Management Plan**

Provide refractive correction; treat any amblyopia; use added plus at near if needed to facilitate fusion; prescribe vision therapy to develop/enhance normal sensory and motor fusion when applicable.

Use prisms to eliminate diplopia and re-establish binocular vision; prescribe vision therapy when applicable; in stable deviations exceeding 20 PD, consult with strabismus surgeon regarding extraocular muscle surgery.

Provide refractive correction; prescribe prism and/or vision therapy to prevent amblyopia, eliminate diplopia, and establish normal sensory fusion, if applicable.

Provide refractive correction; treat any amblyopia; consult with strabismus surgeon regarding extraocular muscle surgery.

Provide refractive correction; use added minus lens power or base-in prism if needed to facilitate fusion; prescribe vision therapy; if deviation persists or increases, consult with strabismus surgeon regarding extraocular muscle surgery.

No therapy if strabismus is not present in the primary position of gaze and no diplopia. Consider prisms and/or surgery to treat head turn.

Provide refractive correction; treat any amblyopia; prescribe vision therapy and/or prism if applicable.

Consult with ophthalmologist regarding treatment of any underlying ophthalmic disease; provide refractive correction; treat any amblyopia; prescribe vision therapy and/or prism, if applicable; if deviation persists or increases, consult with strabismus surgeon regarding extraocular muscle surgery.

| Appendix ( | 59 | 70 | Strabismus: | Esotro | pia and | Exotropia |
|------------|----|----|-------------|--------|---------|-----------|
|            |    |    |             |        |         |           |

| Figure 3  |  |  |  |  |
|---|--|--|--|--|
| ICD-10-CM Classification of Esotropia and Exotropia |  |  |  |  |

| ICD-10-CM Classification of Esotropia and Exot  | ropia                | <b>Monocular exotropia with other noncomitancies</b> Monocular exotropia with X or Y pattern |        |  |
|---|----------------------|--|--------|--|
| Strabismus and other disorders of binocular eye movements<br>Excludes: nystagmus and other irregular eye movements (379.50-37 | <b>378</b><br>79.59) | Alternating exotropia  | 378.15 |  |
| Esotropia   | 378.0                | Alternating exotropia with A pattern   | 378.16 |  |
| Convergent concomitant strabismus<br>Excludes: intermittent esotropia (378.20-378.22)   |                      | Alternating exotropia with V pattern   | 378.17 |  |
| Exotropia, unspecified  | 378.00               | Alternating exotropia with other noncomitancies Alternating exotropia with X or Y pattern    | 378.18 |  |
| Monocular esotropia   | 378.01               | Intermittent heterotropia  | 378.2  |  |
| Monocular esotropia with A pattern  | 378.02               | Excludes: vertical heterotropia (intermittent) (378.31)                                      |        |  |
| Monocular esotropia with V pattern  | 378.03               | Intermittent heterotropia, unspecified Intermittent:   | 378.20 |  |
| <b>Monocular esotropia with other noncomitancies</b> Monocular esotropia with X or Y pattern                                  | 378.04               | esotropia NOS<br>exotropia NOS   |        |  |
| Alternating esotropia   | 378.05               | Intermittent esotropia, monocular  | 378.21 |  |
| Alternating esotropia with A pattern  | 378.06               | Intermittent esotropia, alternating  | 378.22 |  |
| Alternating esotropia with V pattern  | 378.07               | Intermittent exotropia, monocular  | 378.23 |  |
| Alternating esotropia with other noncomitancies Alternating esotropia with X or Y pattern                                     | 378.08               | Intermittent exotropia, alternating  | 378.24 |  |
|   | /                    | Other and unspecified heterotropia   | 378.3  |  |
| Exotropia Divergent concomitant strabismus  Excludes: intermittent exotropia (378.20, 378.23-378.24)                          | 378.1                | Heterotropia, unspecified  | 378.30 |  |
| Exotropia, unspecified  | 378.10               | Monofixation syndrome<br>Microtropia   | 378.34 |  |
| Monocular exotropia   | 378.11               | Accommodative component in esotropia   | 378.35 |  |
| Monocular exotropia with A pattern  | 378.12               | Mechanical strabismus  | 378.6  |  |
| Monocular exotropia with V pattern  | 378.13               | Mechanical strabismus, unspecified   | 378.60 |  |

#### Appendix 71

#### **Abbreviations of Commonly Used Terms**

AC/A Accommodative convergence/accommodation

D Diopter

OKN Optokinetic nystagmus

PD Prism diopter

q.d. Daily

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#### Glossary

**Accommodative convergence/accommodation (AC/A) ratio** The amount of convergence of the eyes due to a unit of accommodative response, which is the response AC/A ratio. Clinically, the stimulus AC/A ratio is used when the accommodative response is not objectively measured.

**Alternating strabismus** The ability of a strabismic individual to hold fixation with either eye while both eyes are open.

**Amblyopia** Reduction in best-corrected visual acuity in the absence of any obvious structural anomalies or ocular disease. It is usually unilateral (as in unilateral strabismus or with significant anisometropia) or occasionally bilateral (as with high refractive error of both eyes).

**Anisometropia** A condition of unequal refractive state for the two eyes, one eye requiring a different lens correction from the other.

**Anomalous retinal correspondence** Correspondence of subjective directionalization of the fovea of the fixating eye with an extrafoveal area of the strabismic eye, occurring in strabismic individuals whose onset is in early childhood. The condition is usually clinically abbreviated as ARC (for anomalous *retinal* correspondence) since testing involves retinal locations; another abbreviation, however is AC with the rationale being that directional correspondence takes place in the cortex rather than in the eyes.

**Comitant** The normal condition in which the magnitude of deviation of the visual axes (heterophoria or strabismus) remains essentially the same in all positions of gaze and, in cases of strabismus, with either eye fixating. *Comitant* is used clinically, but *concomitant* is the etiologically correct term.

**Diplopia** A condition in which a single object is perceived as two rather than one.

**Ductions** Ability of the eyes to show a full range of motion under monocular (one-eye) viewing conditions.

**Eccentric fixation** Attempted fixation of a target under monocular viewing conditions in which the image of the target falls on a retinal area other than the center of the fovea.

**Fusion** The process by which stimuli, seen separately by the two eyes, are combined, synthesized, or integrated into a single perception.

**Heterophoria (also phoria)** A latent deviation of the visual axes from the orthophoric position that requires fusional vergence for clear bifixation.

**Incomitant** A binocular anomaly in which the magnitude of deviation of the visual axes is not the same in different positions of gaze or the deviation differs with either eye monocularly fixating. Generally, the magnitude of change must exceed 5 prism diopters for clinical significance. The etymologically correct term is *nonconcomitancy*.

**Inferior oblique overaction** Overelevation of the eye upon adduction, more noticeable on versions. The right eye will abnormally elevate on left gaze, creating a right hypertropia; the left eye will do so on right gaze, creating a left hypertropia.

**Motor fusion** The ability to align the eyes in such a manner that sensory fusion can be maintained.

**Nystagmus** Rhythmic oscillations or tremors of the eyes which are independent of the normal eye movements.

**Primary position** The position of the eye, or eyes, when looking straight ahead, with the head erect and still and facing in the same direction. This is usually designated as *primary position* of gaze.

**Prism diopter (PD)** The customary unit of measurement of the magnitude of deviation of the visual axes in strabismus or heterophoria. One prism diopter is the angle subtended by a deviation of 1 centimeter at a distance of 1 meter.

**Pseudoesotropia** The false appearance of having esotropia when actually no convergent misalignment of the visual axes exists. Common with infants and

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very young children who have flat and broad nasal bridges with prominent epicanthal folds.

**Ptosis** Drooping of the upper eyelid below its normal position.

**Sensory fusion** The ability of the brain to bring together two sensations with the end result of a single percept.

**Stereopsis** Binocular visual perception of three-dimensional space, based on retinal disparity. Clinically referred to as depth perception.

**Suppression** Under binocular viewing conditions, the inability to perceive all or part of objects in the field of vision of one eye, attributed to cortical inhibition.

**Unilateral strabismus** A condition in which only the nonstrabismic eye can maintain fixation while both eyes are open. In young children, a constant unilateral strabismus may cause amblyopia.

**Versions** More than one conjugate movement in which the two eyes move in the same direction.

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