6
AMBLYOPIA AND STRABISMUS

OBJECTIVES

As a primary care physician, you should be able to recognize the signs and symptoms of amblyopia and strabismus; be able to perform the necessary tests to screen for these conditions; and, if the patient is a child, be cognizant of the need to arrange for prompt ophthalmologic consultation, particularly when intraocular disease is suspected.

To achieve these objectives, you should learn to

- Measure or estimate visual acuity in children
- Detect strabismus by general inspection, the corneal light reflex test, and the cover test
- Perform ophthalmoscopy in a child to rule out any organic causes of impaired vision when amblyopia is suspected
- Explain to parents the need for prompt treatment of amblyopia

RELEVANCE

Amblyopia is a form of treatable visual loss found in approximately 2% of the young adult population of the United States. It can be defined as a loss of visual acuity not correctable by glasses in an otherwise healthy eye. Amblyopia develops in infancy or early childhood and usually can be detected in very young patients, principally by measuring or estimating visual acuity. If detected and treated early, amblyopia can be cured. For best results, treatment should begin before age 5; treatment for amblyopia is rarely successful if initiated past age 10. If amblyopia is not detected and treated early in life, visual impairment from amblyopia persists for the patient's entire lifetime. At least half of all patients with amblyopia also have strabismus, a misalignment of the two eyes.

The pediatrician or family physician will most likely be the first to see a young patient with amblyopia or strabismus and, therefore, will have the principal responsibility for screening. The child's physician must be familiar with the different kinds of amblyopia and strabismus, the close relationship of these two conditions, and how best to detect them.

BASIC INFORMATION

Vision is a developmental sensory function. Vision at birth is relatively poor, but through proper visual stimulation in the early months and years of life, a normal acuity is achieved at about 3 years of age. If this developmental process—the stimulation of the vision-receptive cells in the brain—is prevented because of strabismus, abnormal refractive error, congenital cataract, or some other condition, vision will not develop properly. This is a failure of the developmental process, not primarily an organic abnormality of the eye.

AMBLYOPIA

Amblyopia results from a disruption of the normal development of vision, which distinguishes it from vision loss resulting directly from organic ocular defects, such as cataract, retinoblastoma (a life-threatening tumor of early childhood), and other inflammatory and congenital ocular disorders. It is usually unilateral, but it can (rarely) affect both eyes. Amblyopia does not cause learning disorders.

Amblyopia may develop in young children who receive visual information from one eye that is blurred or conflicts with information from the other eye. To understand how amblyopia may develop in this way, consider that the brain is receiving two stimuli for each visual event: one from a visually aligned (fixating) eye and one from an "abnormal" eye (vision blurred or eye misaligned on another target). The child's brain selects the better image and suppresses the blurred or conflicting image, which results in the faulty development of vision in the amblyopic eye. In other words, the brain continually "favors" the eye with better vision, to the eventual detriment of visual development in the other eye. For this reason, amblyopia is often referred to colloquially as "lazy eye."

A number of predisposing factors can lead to the development of amblyopia. These are summarized below.

Strabismic Amblyopia

A child can develop amblyopia in the context of strabismus (misaligned eyes, Figure 6.1). An adult onset of strabismus generally will cause diplopia (double

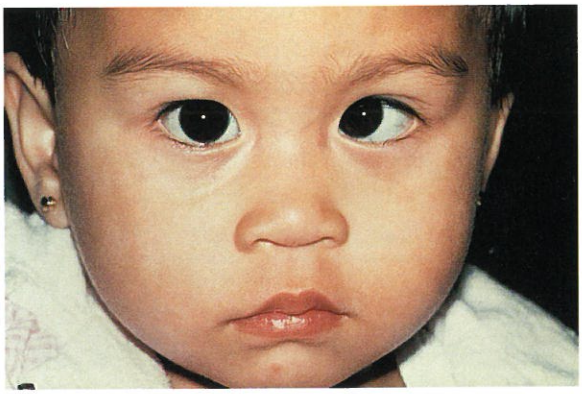

FIGURE 6.1 Strabismus. Strabismus is the most common underlying cause of amblyopia. With constant deviation of one eye, reduced vision occurs. Amblyopia is less likely when the deviation is intermittent or when the two eyes alternate fixation. *(Reprinted from* Understanding and Preventing Amblyopia. A Slide-Script Program. *San Francisco: American Academy of Ophthalmology; 1987:6.)*

vision) because the two eyes are not aligned on the same object. The brain of a child, on the other hand, is more adaptive. In a similar strabismic situation, the child's brain ignores (suppresses) the image from one of the eyes—usually the one that provides the blurrier image. Although such an adaptation overcomes the troublesome symptom of diplopia, this cortical suppression of sensory input from one eye may interrupt the normal development of vision in the higher centers of the brain; this interruption may result in reduced vision, which is amblyopia.

Sometimes the degree of misalignment between the two eyes is very slight, making detection of strabismus and suspicion of strabismic amblyopia difficult. Even with a small angle of strabismus, amblyopia may be quite severe.

Refractive Amblyopia

Amblyopia can result from a difference in refractive error between the two eyes. The eye with the lesser refractive error provides the clearer image and usually is favored over the other eye; suppression occurs and amblyopia develops. Children with asymmetric hyperopia are susceptible, because unequal accommodation is impossible; the child can bring only one eye at a time into focus. Refractive amblyopia may be as severe as that found in strabismic amblyopia. However, the pediatrician or family physician may overlook the possibility of amblyopia because there is no obvious strabismus. Detection of amblyopia must be based on an abnormality found in visual acuity testing.

Form-Deprivation and Occlusion Amblyopia

Form-deprivation amblyopia (amblyopia ex anopsia) can result when opacities of the ocular media—such as cataracts (Figure 6.2), corneal scarring, or even drooping of the upper lid (ptosis)—prevent adequate sensory input and thus disrupt visual development. The amblyopia can persist even when the cause of the media opacity or ptosis is corrected. Rarely, occlusion amblyopia can result from patching of the normal eye.

STRABISMUS

Strabismus is a misalignment of the two eyes, so that both eyes cannot be directed toward the object of regard. Strabismus may cause or be caused by the absence of binocular vision; as with amblyopia, strabismus does not cause learning disabilities.

It is clinically useful to distinguish between concomitant (nonparalytic) and incomitant (paralytic or restrictive) strabismus. Additionally, a number of terms are used to describe and classify strabismus. These distinctions and terms are summarized below.

Concomitant Strabismus

Strabismus is called *concomitant* or *nonparalytic* when the angle (or degree) of misalignment is approximately equal in all directions of gaze (Figure 6.3). The individual extraocular muscles are functioning normally, but the two eyes are simply not directed toward the same target. Most concomitant strabismus has its onset in childhood. In children, it often causes the secondary development of suppression to overcome double vision and thus leads to strabismic

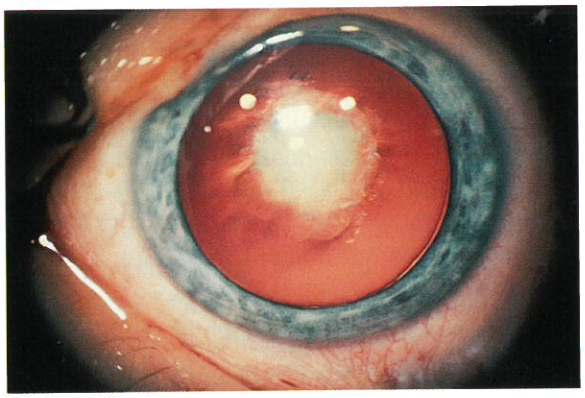

FIGURE 6.2 Opacities of the ocular media. Congenital cataract, if significant, should be removed at the earliest possible age. Good vision can be obtained if the cataract is removed and the refractive error corrected. *(Reprinted from* Understanding and Preventing Amblyopia. A Slide-Script Program. *San Francisco: American Academy of Ophthalmology; 1987:16.)*

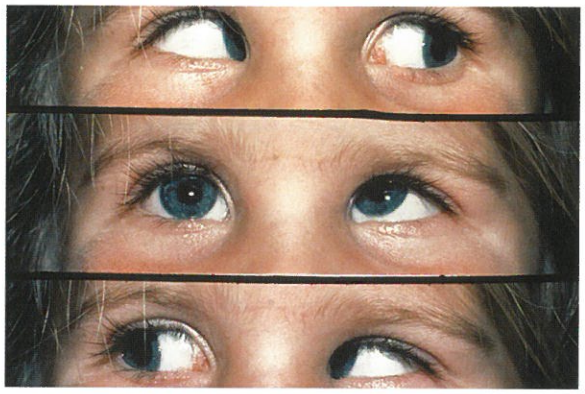

FIGURE 6.3 Concomitant strabismus. In the views presented here, the misaligned eyes exhibit about the same degree of inward deviation (esotropia) in each position of gaze.

amblyopia. Concomitant strabismus in patients under age 6 is rarely caused by serious neurologic disease. Strabismus arising later in life may have a specific and serious neurologic basis. Concomitant strabismus may occur in an adult who loses most or all of the vision in one eye from intraocular or optic nerve disease. A blind eye in an adult will frequently drift outward, while in a child the eye will turn inward.

Incomitant Strabismus

Strabismus is called *incomitant, paralytic,* or *restrictive* when the degree of misalignment varies with the direction of gaze (Figure 6.4). One or more of the extraocular muscles or nerves may not be functioning properly, or normal movement may be mechanically restricted. This type of strabismus may well indicate either a serious neurologic disorder, such as third cranial nerve paresis

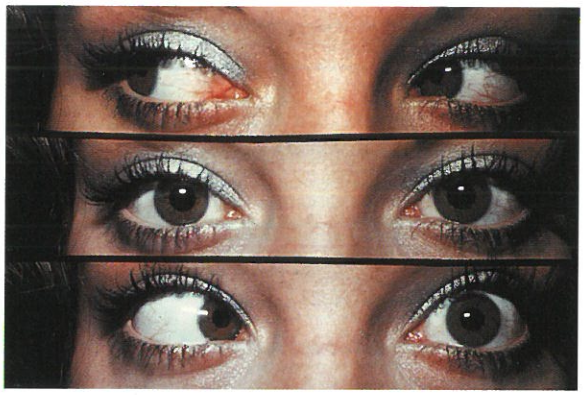

FIGURE 6.4 Incomitant strabismus. The eyes appear straight in right gaze (top) and straight-ahead gaze (middle), but a misalignment is obvious in left gaze (bottom), indicating a paralysis of the left lateral rectus muscle or a restriction of the left medial rectus. These eye positions would be found in a left sixth cranial nerve palsy.

(see Chapter 7), or orbital disease or trauma, such as the restrictive ophthalmopathy of thyroid disease or a blowout fracture.

Heterophoria and Heterotropia

Heterophoria is a latent tendency for misalignment of the two eyes that becomes manifest only if binocular vision is interrupted, such as by covering one eye. During binocular viewing, the two eyes of a patient with heterophoria are aligned perfectly; both eyes are directed at the same object of regard. However, when one eye is covered, that eye will drift to its position of rest. Once the cover is removed, the eye will realign itself with the other eye. A minor degree of heterophoria is normal for most individuals.

Heterotropia is really another term for strabismus. In general, *tropia* refers to a manifest deviation that is present when both eyes are open (no covers). Usually, binocular vision is reduced. Some patients, however, can demonstrate an intermittent heterotropia and thus achieve binocular vision part of the time.

Heterotropia and heterophoria can be subdivided further according to the direction of the deviation involved. In esotropia and esophoria, the deviating eye is directed inward toward the nose. Esotropia is a manifest deviation and is the most common type of deviation in childhood. Exotropia is much more likely to be intermittent than esotropia, with an outward deviation of an eye alternating with alignment of the eyes. Children with this condition suppress double vision when the deviating eye is turned out and achieve some degree of binocular vision when the two eyes are straight. Vertical heterotropias and heterophorias have many different causes, including paralysis or dysfunction of vertically acting extraocular muscles. When vertical deviations are described, the deviating eye (right or left) should be specified. Table 6.1 summarizes the directions of deviation in heterophoria and heterotropia. Figure 6.5 depicts the different kinds of heterotropia.

HOW TO EXAMINE AND INTERPRET THE FINDINGS

Pediatric vision screening is important for detecting not only amblyopia and strabismus but also congenital cataract, glaucoma, retinoblastoma, and other vision- or life-threatening conditions. Regular screening by the pediatrician or

TABLE 6.1 Summary of Heterophoria and Heterotropia

Prefix	Name of Disorder		Description
	-phoria (latent)	-tropia (manifest)	
eso-	esophoria	esotropia	inward deviation
exo-	exophoria	exotropia	outward deviation
hyper-	hyperphoria	hypertropia	upward deviation
hypo-	hypophoria	hypotropia	downward deviation

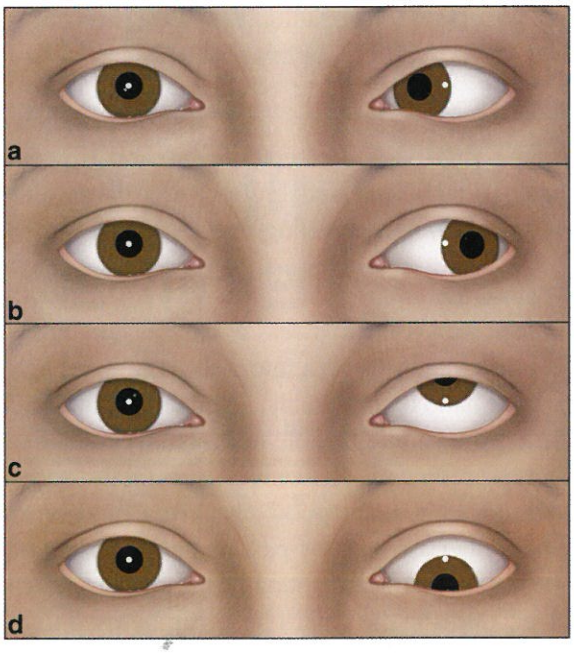

FIGURE 6.5 Types of heterotropia. Note the corneal light reflex. **a.** Esotropia (inward). **b.** Exotropia (outward). **c.** Hypertropia (upward). **d.** Hypotropia (downward). *(Illustration by Christine Gralapp.)*

family physician helps ensure that the child's vision is developing normally or, if it is not, that early treatment is instituted. At a minimum, all children should undergo an evaluation to detect eye and vision abnormalities during the first few months of life and again at about age 3.

Visual acuity testing is important for detecting amblyopia as well as refractive error, which can lead to amblyopia in young children. Strabismus may be detected by general inspection, the corneal light reflex test, or the cover test. Additional tests are important for general eye screening in children of all ages: pupillary reactions are important in assessing normal eye function and health; direct ophthalmoscopy is required to detect media opacities by eliciting a red reflex and to examine the fundus for retinal abnormalities. These techniques are discussed later in the chapter.

AMBLYOPIA TESTING

Amblyopia can be detected by testing the visual acuity in each eye separately. Although there is no specific Mendelian pattern of inheritance, strabismus and amblyopia sometimes cluster in families. Restoration of normal visual acuity can be successful only if treatment is instituted during the first decade of life, when the visual system is still in the formative stage. Techniques for measuring

or estimating visual acuity (or visual function) and detecting amblyopia vary with the child's age, as described below.

Newborns

True visual acuity is difficult to measure in newborns. However, infants' general ocular status should be assessed through corneal light reflex testing, evaluation of the red reflex, pupillary testing, and, if possible, fundus examination.

Infants to 2-Year-Olds

With infants, it is possible only to assess visual function, not visual acuity. To test for amblyopia in infants (from a few months to about age 2), cover each eye in turn with the hand or, preferably, an adhesive patch and note how the child reacts. The infant should be able to maintain central fixation with each eye. If amblyopia is present, the child will likely protest—vocally or by evasive movements—the covering of the "good" eye. Visual function, including ocular motility, may be further assessed by passing an interesting object, such as a ring of keys, before the baby and noting how the infant watches and follows the moving object. Moving the child's head can be used to demonstrate full ocular motility if not otherwise documented by following movements.

Age 2 to 4 or 5

A picture card (Figure 6.6) may be used to test visual acuity in children between 2 and 3 years of age. At age 3 (or before, if the child can follow directions and communicate adequately), visual acuity should be tested with the tumbling E chart (Figure 6.7). In the tumbling E test, the child is asked to point with his or her fingers to indicate the direction of the "arms" of the E. Use of an adhesive patch is the best way to ensure full monocular occlusion and accurate acuity measurement in children at these ages (Figure 6.8).

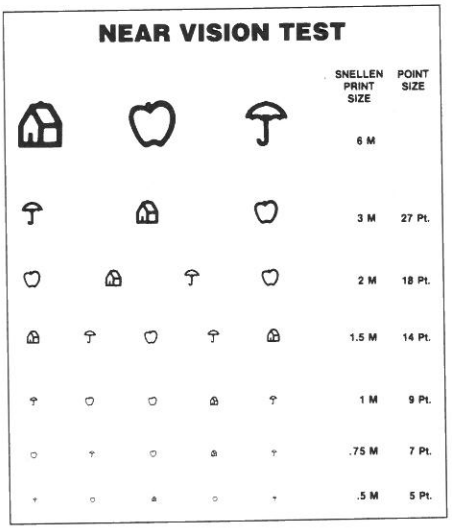

FIGURE 6.6 Picture card. This figure shows one type of picture card used to test visual acuity in young children. To use the picture card, the examiner familiarizes the child with the pictures at close range. Each eye is then tested individually at a testing distance of 6 meters (20 feet) from the child by asking the child to name the various objects.

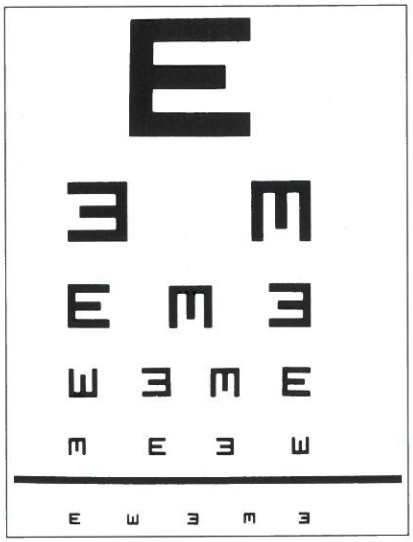

FIGURE 6.7 Tumbling E chart. Visual acuity testing in children should be done at 6 meters (20 feet) with charts such as the tumbling E shown here. The child indicates the direction of the "arms" of the E by pointing with his or her fingers.

Vision should be rechecked annually once visual acuity has been determined to be normal in each eye. Young children may not quite reach 20/20 acuity; this is no cause for concern as long as vision is at least 20/40 and both eyes are equal. A recent advance in early detection of amblyogenic factors is photoscreening. A computerized camera takes photographs of the child's undilated eyes. Refractive errors, strabismus, anisometropia, and media opacities are visible in the photos. This technique permits screening of preverbal children and those unable to cooperate with other types of testing. Photoscreening is not a substitute for accurate visual acuity measurement, but it can provide significant information about factors that may lead to amblyopia.

Age 4 or 5 and Up

The Snellen chart may be used to test visual acuity in children age 4 or 5 and up who know the letters of the alphabet (see Chapter 1).

STRABISMUS TESTING

Strabismus testing for children (and adults) consists of general inspection, the corneal light reflex test, and the cover test, all described below.

Children up to age 3 or 4 months may exhibit temporary uncoordinated eye movements and intermittent strabismus. However, if occasional deviation persists beyond this age, a referral to an ophthalmologist should be made. Constant deviations should be referred at any age.

Epicanthus (Figure 6.9), in which epicanthal skin folds extend toward the upper eyelid and brow and the nose bridge is flat, may give an infant the appearance of esotropia, especially if the head or eyes are turned slightly to the right or left. As the child's head grows and the nose bridge develops, the

FIGURE 6.8 Measuring visual acuity. This becomes possible by age 2–4. An adhesive patch to cover the eye not being tested helps to facilitate the exam as well as prevent "peeking." *(Reprinted from* Understanding and Preventing Amblyopia. A Slide-Script Program. *San Francisco: American Academy of Ophthalmology; 1987:12.)*

epicanthus becomes less obvious. This may be mistakenly interpreted as the child outgrowing presumed strabismus; however, a child does not outgrow a true strabismus. The cover test and evaluation of the corneal light reflex will distinguish between pseudostrabismus (epicanthus) and true strabismus. However, it is important to keep in mind that strabismus can also occur in the presence of epicanthus.

General Inspection

For infants and older children, a general inspection may reveal an identifiable deviation of one eye. Having the patient look in the six cardinal positions of gaze (Figure 6.10) may reveal whether the deviation is approximately the same in all fields—indicating concomitant strabismus—or is significantly different in one field of gaze—indicating a possible incomitant strabismus. Involuntary eye jerks known as *nystagmus* may be detected in primary or other fields of gaze.

FIGURE 6.9 Epicanthus. An extended lid fold and a relatively flat nose bridge may give the false appearance of an esotropia. *(Courtesy Robert T. Lee, MD.)*

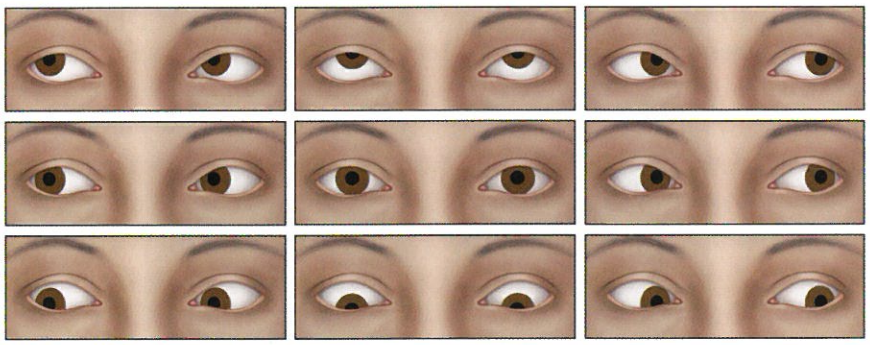

FIGURE 6.10 Positions of gaze. Eye movement can be evaluated by directing the patient to turn the eyes in six cardinal positions of gaze: up/right, right, down/right, down/left, left, and up/left. Also shown are the upward gaze, primary gaze, and downward gaze (middle column). *(Illustration by Christine Gralapp.)*

The patient may assume an abnormal head posture (ie, a tilt or turn to one side) to reduce the nystagmus and improve the visual acuity or to obtain binocular vision in cases of congenital cranial-nerve palsy. All infants or children with nystagmus should be examined and followed up by an ophthalmologist.

Corneal Light Reflex

Observation of the corneal light reflex constitutes an objective assessment of ocular alignment. Certainly in newborns and often in young children, it may be the only feasible method of testing for strabismus.

The patient is directed to look at a penlight held directly in front of the eyes by the examiner at a distance of 2 feet. The examiner aligns his or her eye with the light source and compares the position of the light as reflected by the cornea of each eye (Figure 6.11). Normally, the light is reflected on each cornea symmetrically and in the same position relative to the pupil and visual axis of each eye. In a deviating eye, the light reflection will be eccentrically positioned and in a direction opposite to that of the deviation. The size of the deviation can be estimated by the amount of displacement of the light reflex, but this is a relatively gross estimate.

Cover Test

The cover test (Figure 6.12) is easy to perform, requires no special equipment, and detects almost every case of tropia. It can be used on any patient over the age of 6 or 7 months. To perform the test, have the patient look at a fixation point, such as a detailed or interesting target (eg, a toy) or the Snellen chart. Note which eye seems to be the fixating eye. Cover the fixating eye and observe the other eye. If the uncovered eye moves to pick up the fixation, then it can be reasoned that this eye was not directed toward the

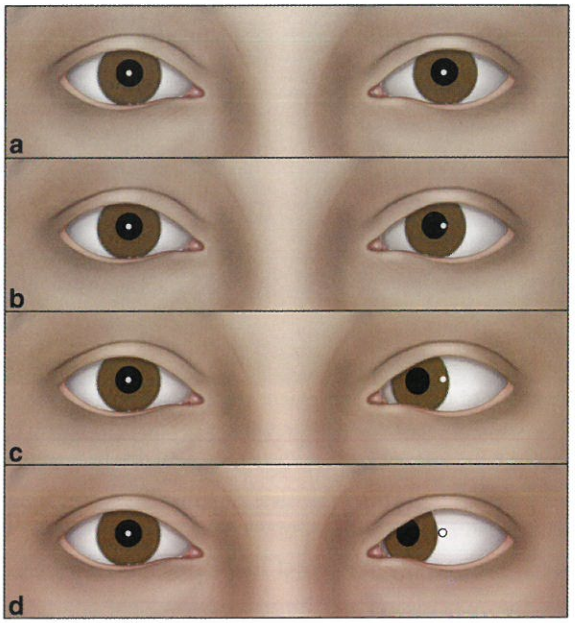

FIGURE 6.11 Corneal light reflex. The position of the light reflection indicates **(a)** a normal alignment, **(b)** a small esotropia, **(c)** a moderate esotropia, and **(d)** a large left esotropia.*(Illustration by Christine Gralapp.)*

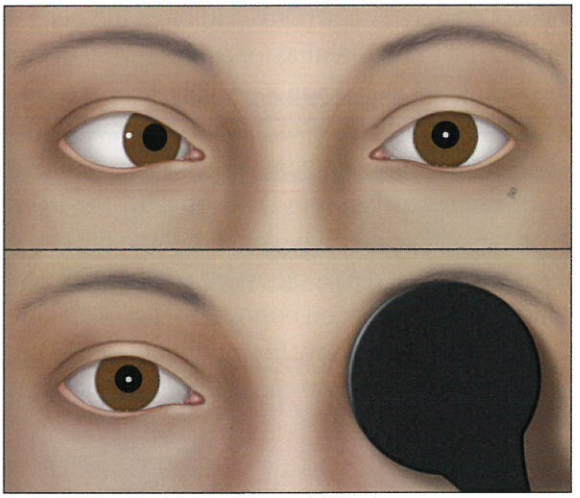

FIGURE 6.12 Cover test. The cover test can be used to screen for strabismus. The results depicted here indicate a right esotropia. When the left eye is covered, the right eye moves outward to pick up fixation. (When the left eye is uncovered, the left eye moves outward to pick up fixation and both eyes assume their original positions.) *(Illustration by Christine Gralapp.)*

object of regard originally (ie, when both eyes were uncovered). If the eye moves inward to fixate, then originally it must have been deviated outward and hence is exotropic. If the eye moves outward to fixate, then it was deviated inward and is esotropic. If the eye moves up or down, then it is hypotropic or hypertropic, respectively; the deviating eye must be specified in a hypertropia or hypotropia. Of course, each eye must be tested separately because there is no way of knowing which eye may be expressing the deviation.

No shift on cover testing means there is no tropia, but a phoria could still be present. A phoria is detected by alternate cover testing. Each eye is alternately occluded and the examiner observes the uncovered eye for a refixation shift. The patient has an esophoria if the uncovered eye moves outward to fixate, and an exophoria if the eye moves inward to fixate.

A very small-angle deviation may be difficult to detect by evaluating the corneal light reflex or performing the cover test. For this reason, visual acuity testing is important in all cases of suspected strabismus for detection of amblyopia.

OTHER TESTS

The following tests are part of general screening for all children.

Pupillary Testing

Abnormal pupillary responses may indicate neurologic disease or other ocular defects. Pupillary testing is discussed in Chapter 1; for further discussion and specific techniques see Chapter 7.

Red Reflex

Light is reflected off the fundus as red when it is examined through the ophthalmoscope from a distance of about 1 foot (Figure 6.13). Media opacities appear in the red reflex as black silhouettes. Leukocoria ("white pupil") is a white reflex that may signify the presence of cataract or retinoblastoma (Figure 6.14).

All infants and children should be evaluated for the red reflex; pupillary dilation may be necessary to achieve a red reflex (phenylephrine 1.0% and cyclopentolate 0.2% in infants, readily available in combination as Cyclomydril). If the examiner cannot elicit a red reflex, the infant or child should be referred to an ophthalmologist urgently.

Ophthalmoscopy

A careful ophthalmoscopic examination of both eyes through dilated pupils is mandatory for any patient with reduced vision or with strabismus. In this

FIGURE 6.13 Evaluation of amblyopia. Assessing the red reflex allows the examiner to evaluate for two potential causes of amblyopia: media opacities and high refractive errors. The cornea, anterior chamber, lens, and vitreous must all be clear to allow a view of the retina. If the eye is unusually hyperopic or myopic, the red reflex may be very dim unless the ophthalmoscope's high-power lenses are used. *(Reprinted from* Understanding and Preventing Amblyopia. A Slide-Script Program. *San Francisco: American Academy of Ophthalmology; 1987:8.)*

way, the examiner can detect potentially serious intraocular lesions, such as cataract, malignancies such as retinoblastoma, or other abnormalities.

MANAGEMENT OR REFERRAL

The early detection of amblyopia and strabismus is an important responsibility for those involved in infant and child health care. Delayed diagnosis may have serious consequences for visual acuity, eye disease, or systemic disease. If an abnormality is suspected, the patient should be referred promptly to an ophthalmologist.

AMBLYOPIA

In children younger than 5, strabismic amblyopia can usually be treated effectively by the ophthalmologist through occlusion of the unaffected eye (Figure 6.15). The child wears an adhesive patch over the good eye, forcing the brain to utilize the previously suppressed eye. In general, the success of occlusion treatment for amblyopia patients between the ages of 5 and 9 will depend on the age of the patient, the degree of the amblyopia, and the persistence of patient compliance with treatment. Treatment is better tolerated by younger children but can be successful in children as old as 10. A treatment program started early in life often must be continued throughout the patient's first decade. Amblyopia treatment by patch occlusion of the unaffected eye must be monitored carefully, especially during the younger years, to avoid causing amblyopia through sensory deprivation of the occluded eye.

FIGURE 6.14 Leukocoria. A cataract is not the only cause of a white reflex. In this child, a retinoblastoma fills the vitreous cavity. Any change from the normal red reflex warrants careful ophthalmic examination.

FIGURE 6.15 Reversing amblyopia. Patching (occlusion) of the eye with better vision may be prescribed to help reverse amblyopia. Compliance and follow up are important. *(Courtesy R. Michael Siatkowski, MD.)*

Treatment of refractive amblyopia consists first of wearing glasses, followed by patching of the better eye if the visual acuity difference persists after 4 to 8 weeks of wear. Equal vision in both eyes is readily achievable with parental cooperation in almost all cases. In general, the earlier the individual with amblyopia is diagnosed and treated, the better the chance of achieving equal vision. An alternative to occlusion therapy with an adhesive patch is the use of dilating drops (atropine 1%) daily to the better-seeing eye. This blurs the vision in the better-seeing eye and forces the child to use the amblyopic eye.

STRABISMUS

The most effective way to support fusion (binocular vision) is to treat the amblyopia and equalize the vision. Glasses can treat some or all of the esotropia in a farsighted, or hyperopic, individual (Figure 6.16) and may decrease the frequency of deviation in a myopic individual with exotropia. However, surgical correction of the misalignment may still be necessary, particularly in those children who develop esotropia before the age of 6 months (congenital esotropia). Even when binocular vision may not be achievable, the impact of a disfiguring strabismus on a patient's self-image is a valid indication for sur-

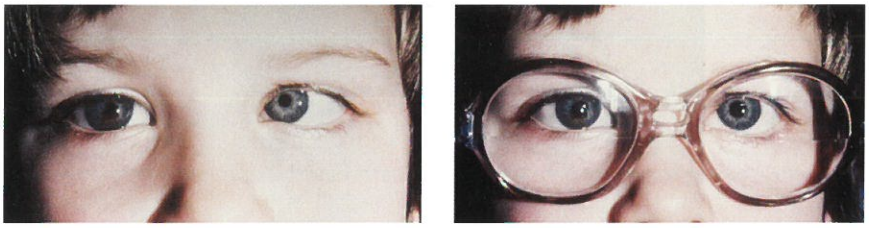

FIGURE 6.16 Esotropia. An inward turning of the eye, esotropia is the most common type of strabismus. About one half of all cases of strabismus are due to a form of esotropia caused by excessive focusing, or accommodation. Accommodative esotropia frequently begins as an intermittent crossing and gradually becomes constant. The age of onset is usually about 2 years, but may be as late as 7. This child is farsighted and without glasses accommodates to see, resulting in crossed eyes. Glasses relieve this accommodative demand enabling the eyes to straighten.

gery. It must be stressed that surgery is not an alternative to glasses and patching when amblyopia is present. "Vision training" has no proven value for the treatment of amblyopia or strabismus.

POINTS TO REMEMBER

- Amblyopia must be detected early and referred to an ophthalmologist to be treated successfully.
- The importance of visual acuity testing in detecting amblyopia cannot be overemphasized. Amblyopia may be present in eyes without strabismus, so the vision in each eye may not be normal even if the eyes appear normally aligned.
- Several serious organic conditions cause strabismus as one manifestation of the disease; therefore, all patients with strabismus should be referred to an ophthalmologist at the time of diagnosis for further testing.
- Children may have cataracts, glaucoma, and retinal diseases, so children with unusually large eye(s), decreased or no red reflex, or poor vision should be referred to an ophthalmologist.
- Vision training has no proven value in the treatment of amblyopia or strabismus.

SAMPLE PROBLEMS

1. A 3-year-old girl is brought to you by her mother, who tells you that she suspects her daughter's right eye is not straight. What steps would you take to determine if a significant problem is present?

Answer: Visual acuity testing should be attempted using the tumbling E chart or a picture card, with each eye alternately covered by an adhesive patch. A difference in visual acuity between the eyes or decreased vision