Vision Screening Essentials: Screening Today for Eye Disorders in the Pediatric Patient

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Article eye

Vision Screening Essentials:
Screening Today for Eye Disorders in the Pediatric Patient

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Author Disclosure
Dr Tingley did not disclose any financial relationships relevant to this article.

Learning Objectives
After completing this article, readers should be able to:

1. Discuss the role of early intervention for vision-threatening problems.
2. Explain how to screen patients for the most common vision problems.
3. Recognize when to screen for vision problems.
4. Describe who should be screened for vision problems.

Introduction

Screening for eye disorders and vision problems is essential in preparing pediatric patients for the future. For this reason, regular vision evaluations are recommended by the American Academy of Pediatrics (AAP).

Significant problems affecting vision are common in children. Vision problems occur in 5% to 10% of all preschoolers and include refractive error, strabismus, and amblyopia. Strabismus is present in 4% of preschool-age children, and amblyopia affects up to 40% of those having strabismus. Major refractive errors requiring correction occur in 5% to 7% of preschool-age children. Cataracts in children are far less common (0.1% incidence), but vision can be affected seriously without early treatment. A simple screening can check effectively for such problems during a time when treatment is most critical without requiring all patients to have a "gold standard" complete eye evaluation at every age, saving both time and health-care resources.

Screening improves visual acuity. In a randomized, longitudinal study, intensive early screening led to a 60% decreased prevalence of amblyopia and improved visual acuity compared with a one-time screening at 37 months of age. (1) Amblyopia responds to therapy, and results are best when treatment is started early in life. The same study showed a 70% lower prevalence of residual amblyopia after treatment when therapy was initiated before age 3 years. The single most effective screening test for the presence of amblyopia is the determination of visual acuity via noninvasive screening.

The consequences of not finding vision-related problems can include adverse effects on school and social performance as well as adult self-image. Further, uncorrected amblyopia is a risk factor for total blindness if the better eye is injured later.

Common Vision Problems

Refractive error is a term describing focusing problems that usually can be treated with glasses or other optical intervention. Such problems include myopia (nearsightedness), hyperopia (farsightedness), and astigmatism. Most children normally have mild hyperopia and can focus their eyes with minimal accommodative effort. High amounts of hyperopia can make eye alignment difficult when significant focusing effort is required, resulting in accommodative esotropia, a common form of strabismus (Fig. 1).

Strabismus is defined as a misalignment of the eyes, either as a full-time occurrence (tropia) or as a tendency to become misaligned (phoria). Various prefixes are used to describe the relative direction of the misaligned eye's deviation, such as "eso" for an adducting (inward) tendency (Fig. 2) and "exo" for an abducting (outward) tendency. Strabismus often must be differentiated from pseudostrabismus, which is the appearance of a misalignment of the eyes without actual strabismus present (Fig. 3). Pseudostrabismus occurs most commonly when a broad nasal bridge covers the nasal sclera of one or both eyes, resulting in an appearance of esotropia.

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Figure 1. Accommodative esotropia in a child who has a high hyperopia (top). The use of glasses relieves the accommodative effort of focusing, particularly at near objects, resulting in improvement of the induced esotropia (bottom). Photo courtesy of the American Academy of Ophthalmology.

Amblyopia is a loss of visual acuity due to active cortical suppression of the vision of an eye and can occur from a variety of causes. Strabismic amblyopia occurs with misalignment of an eye and suppression of one eye to avoid diplopia. Anisometropic amblyopia occurs when one eye has a different length from the other, resulting in a different focusing ability, that limits the ability of the eye to provide appropriate visual information to the brain. Deprivational amblyopia is a result of not using an eye, as from an extended eyelid closure from a hemangioma or a dense congenital cataract.

Cataract is an opacification of the eye lens. Cataracts affect the clarity and interpretation of the image in the developing eye and can have a significant impact on future visual potential by causing deprivational amblyopia.

Figure 2. Esotropia in a child. Covering the fixing right eye can detect if the left eye is being used equally as well as the right eye. If the left eye holds fixation as well as the right eye, no significant amblyopia is yet present. Additional evaluation as to the cause of the strabismus is warranted. Photo courtesy of the American Academy of Ophthalmology.

Screening for the Common Problems

The AAP's vision screening guidelines provide a basis for detecting vision problems at a time when improvement is possible. The methods for screening have been well presented in published sources and are described here briefly. (2)(3) Photoscreening, automated refraction, and electrophysiologic testing may offer opportunities to improve screening methods in the future. (4)(5)

Evaluation of visual function is best initiated at birth with the use of an ophthalmoscope to check if the light reflex in each eye is equal in brightness and color. If a defect in the light reflex is present, the baby might have a cataract, one of the most significant problems with vision that can be detected readily at birth. Variations between the eyes in color or brightness can indicate refractive error in one or both eyes and the need to correct the eyes with glasses early in life (Fig. 4). Retinoblastoma classically causes a white reflex on evaluation (Fig. 5), and this tumor must be considered on any evaluation of the red reflex. However, it is far less common than other causes of a white reflex, including refractive error. Referral to an ophthalmologist for additional evaluation is appropriate if findings are abnormal.

The red reflex test uses the ophthalmoscope with the lens selection on "0" diopters. This device provides illumination directly in line, or coaxial, with the line of sight. Thus, when viewing a normal eye, an orange-to-red light reflection from the fundus is seen filling the

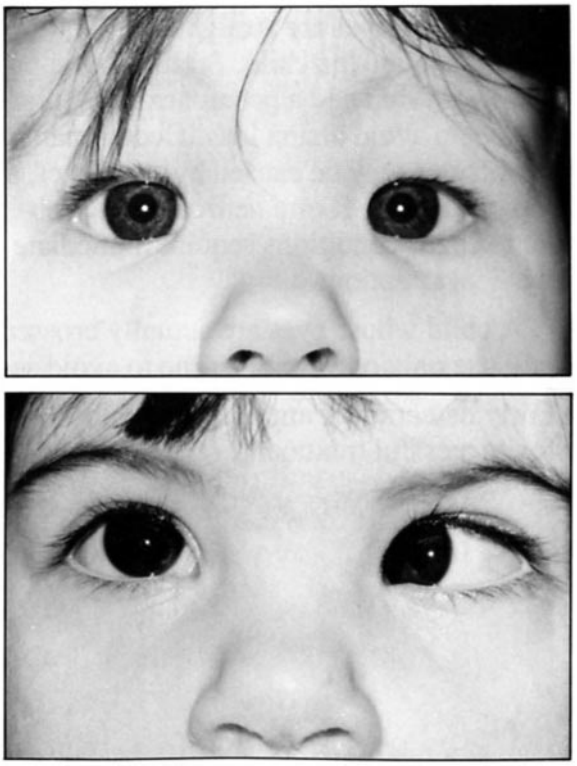

Figure 3. Pseudostrabismus (top) versus true strabismus (bottom). Although the eyes appear misaligned in pseudostrabismus, the light reflection is symmetric in both eyes. In true strabismus, the light reflection is asymmetric. Photos courtesy of the American Academy of Ophthalmology.

Figure 4. Asymmetric red reflexes. This patient shows a variation in color between the eyes, and evaluation revealed a difference between the eyes in focusing, a condition known as anisometropia. In the author's clinical experience, anisometropia is the most common cause of red reflex asymmetry. Anisometropia can cause a significant amblyopia if not detected early by either red reflex testing or visual acuity testing. This child did well with glasses correction. Photo courtesy of the American Academy of Ophthalmology.

pupil. The light should be equal in color and brightness from each eye and fill the pupil completely. Variation between the eyes in any of these aspects is a reason for referral to an ophthalmologist.

Funduscopic evaluation can be difficult to perform without patient cooperation and can be performed more reliably around the age of 3 years per AAP recommendations. Nevertheless, funduscopy can be undertaken in a somnolent infant after the rapid red reflex evaluation is completed.

Ophthalmoscopic evaluations can detect congenital glaucoma, nystagmus, and other visually significant disorders. Direct ophthalmoscopy uses the light on the dimmest setting that still allows visibility for the examiner and requires the examiner to be close enough to the eye so his or her fingers graze the patient's cheek. The clinician should evaluate the structures that are visible throughout the range of lenses from the high plus lenses (black or green numbers) to evaluate anterior structures of the eye to the progressively more minus lenses (increasing red numbers). This procedure allows evaluation of eye structures from anterior to posterior (Fig. 6). Direct ophthalmoscopy can evaluate the retina well and determine the presence of dilated disc vessels of active retinopathy of prematurity (ROP) (Fig. 7). Referral for additional evaluation is recommended for preterm infants to determine the presence of significant ROP and also is needed for patients who have a family history of amblyopia, strabismus, retinoblastoma, retinal degeneration, or systemic disease affecting the eye.

Vision Testing Methods

Defects in visual function may be found at the first evaluation. Ideally, a vision test should allow an examiner to confirm the resolution achieved at the cortical level at any age when the visual system is in use in regular activities, allowing determination of whether each eye is working well. However, technology in visual science has not yet reached that level of sophistication. Currently, clinicians can judge the response of each eye to the same visual stimulus, evaluating for any asymmetry.

A light stimulus should elicit equal wincing behavior from each eye in an alert infant. Infants occasionally fixate

Figure 5. Asymmetric red reflexes, with a yellow right eye reflex and red left eye reflex. This finding led to the early diagnosis of retinoblastoma growing in the right eye. Photo courtesy of the American Academy of Ophthalmology.

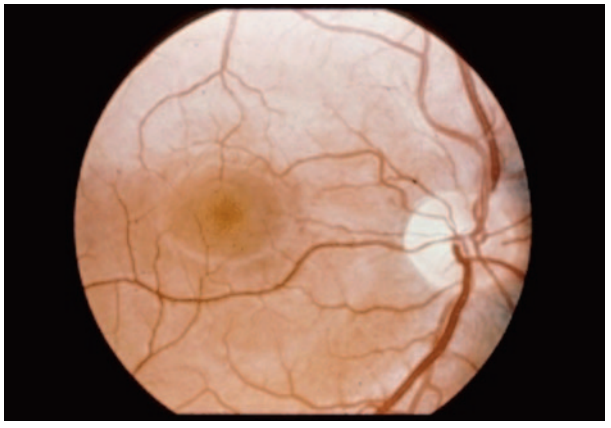

Figure 6. Normal anatomy of the right fundus. Photo courtesy of the American Academy of Ophthalmology.

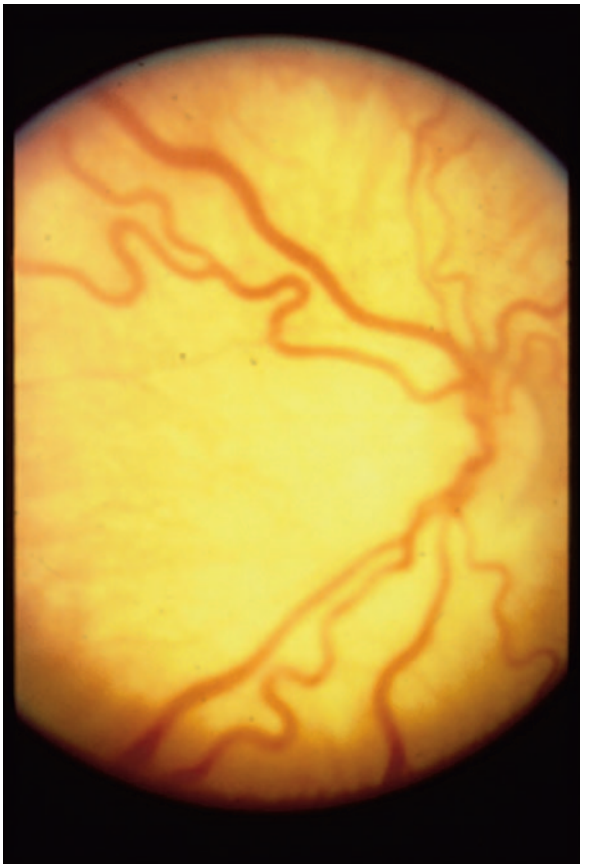

Figure 7. Retinopathy of prematurity (ROP). Note the dilated veins and tortuous arteries in this right eye of an infant who has active ROP. This is known as plus disease and is associated with rapid disease progression. Photo courtesy of the American Academy of Ophthalmology.

momentarily on a high-contrast object or a human face and should do so equally well from either eye. Unfortunately, newborn visual ability is difficult to determine with accuracy. Current understanding of visual acuity in healthy newborns, based on a variety of techniques, suggests acuity of 20/400 at 1 month of age. (6) Mean visual development milestones are listed in Table 1. (7) Both eyes begin to work together on the same

Table 1. Normal Vision Development

Function	Age
Visual fixation present	Birth
Fixation well developed	6 to 9 wk
Visual following	3 mo
Accommodation	4 mo
Stereopsis	4 mo

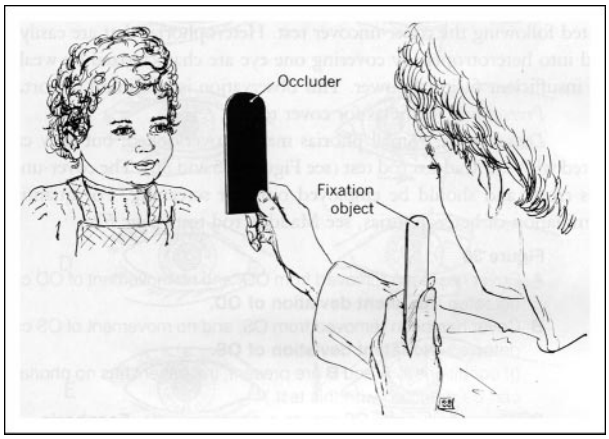

Figure 8. Cover test examination. Reprinted with permission from von Noorden G, ed. *Atlas of Strabismus*. 4th ed. Philadelphia, Pa: Elsevier; 1983.

object at 6 to 9 weeks. Tracking or visual following with both eyes can occur earlier but should be present by 3 months age.

Age-appropriate acuity testing uses instruments that require increasing cognitive ability, ranging from recognizable cartoon pictures (Allen test) or symbols of readily identifiable shapes (LH test) for ages up to 3 years, to choosing which of four letter shapes are presented (HOTV test) or which direction a letter E is pointing (tumbling E test) for ages up to 5 years, to Snellen letters or numbers for ages 5 years and older. The best test for checking acuity is the highest level that the child can complete. Testing is performed at the appropriate distance for the chart used (usually 10 ft), with a line of figures presented at one time. One eye is tested, with the examiner making all efforts to occlude by a patch or wide eye cover the opposite eye at all times, thus avoiding inadvertent "peeks." Differences of two lines of visual acuity between the eyes or vision less than 20/40 acuity in either eye require referral for additional evaluation.

Cover Testing: Identifying Tropias and Phorias

The terms tropia and phoria refer to degree of eye deviations. Tropias define a condition of full-time eye misdirection; phorias define a tendency for the eye to turn when disturbances in binocularity occur, such as when one eye is covered. The cover test is used to distinguish the two conditions. Cover testing one eye at a time can detect a tropia in the other eye because by definition, a tropia is present at all times and does not allow simultaneous alignment of both eyes on the same target. A phoria is detected by observing the covered eye just as it is uncovered; a phoria is present only when binocularity is disturbed.

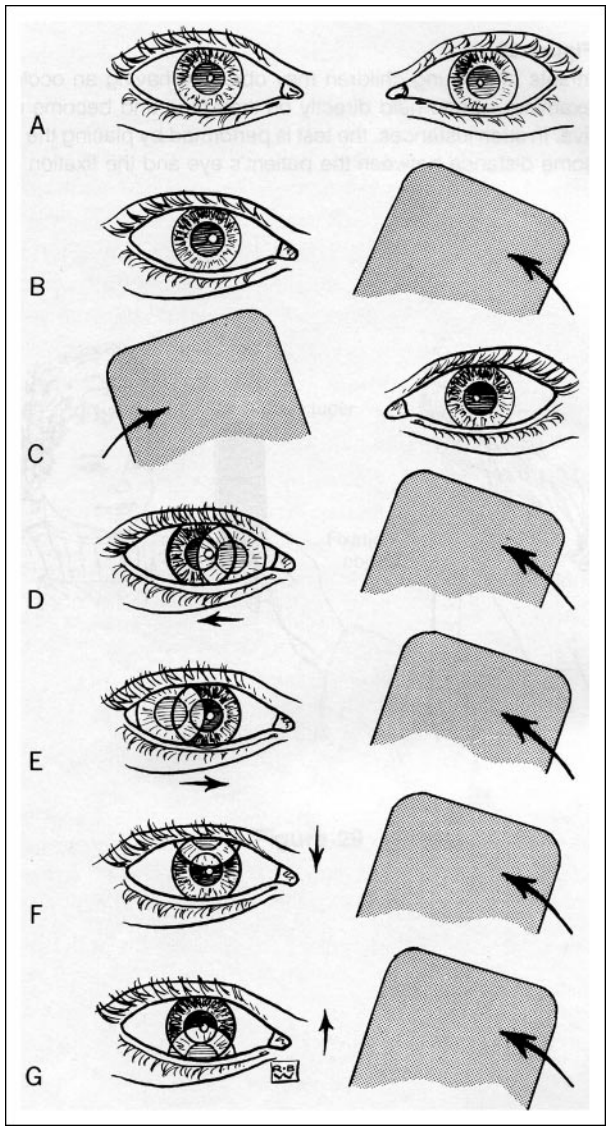

Figure 9. Cover test findings. A. Normal, B. Normal, C. Normal, D. Right esotropia, E. Right exotropia, F. Right hypertropia, G. Right hypotropia. Reprinted with permission from von Noorden G, ed. *Atlas of Strabismus*. 4th ed. Philadelphia, Pa: Elsevier; 1983.

The cover test is performed by covering one eye at a time and observing the eyes for movement as the eye is being covered while the patient observes a fixation target, such as a human face, a small toy, or an attractive picture (Fig. 8). When a child is observing the target and the cover is placed, the uncovered eye should not move; it should not have to move to look directly at the object of

Figure 10. Pseudostrabismus. The light is centered in the pupil of each eye. This is normal eye alignment regardless of the asymmetry in the white sclera visible. Photo courtesy of the American Academy of Ophthalmology.

regard. If the uncovered eye does have to move to look at the target, a tropia is present and is named according to the position of the original deviated position. However, if only the covered eye moves back from a deviated position when uncovered to fixate again on the target, a phoria is present. If the affected eye is deviated toward the nose, an esotropia or esophoria is present. If the eye is deviated away from the nose, it is manifesting an exotropia or exophoria. If the eye is deviated upward, a hypertropia or hyperphoria is present, and if the eye is deviated downward, it is affected by a hypotropia or hypophoria (Fig. 9).

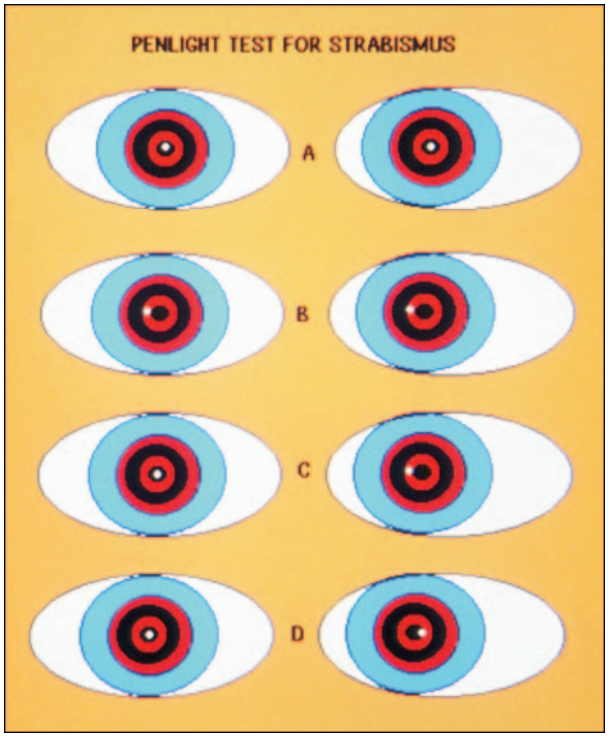

Figure 11. Corneal reflex test. A. Normal, B. Normal, examining light is directed from patient's right side. C. Left exotropia. D. Left esotropia.

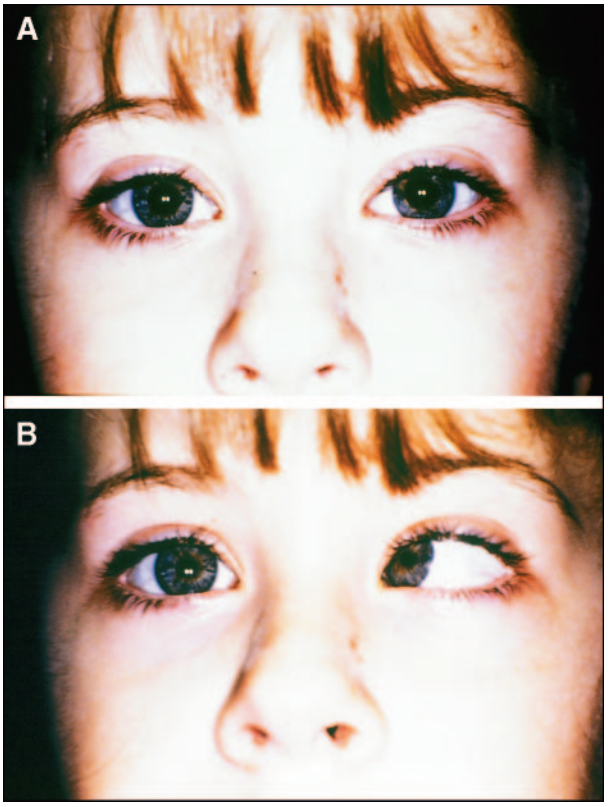

Figure 12. A. Patient evaluated for strabismus by using a penlight at near exhibits a normal corneal reflex test result. B. By using a small target that requires effort to focus, accommodative esotropia becomes apparent. Photo courtesy of the American Academy of Ophthalmology.

The biggest difficulty in preforming a cover test occurs when fixation cannot be sustained reliably for the duration of the test. A visually attractive target is essential to obtaining reliable fixation, and the appropriate object varies by the age of the patient. The face of the examiner can be most attractive to an infant, whereas a small finger puppet or interesting picture might be more attractive to an older child. Results can be interpreted only when fixation is present; a voluntary change in fixation while the eye is covered can be interpreted falsely as either a tropia or a phoria.

Distinguishing Pseudostrabismus

As mentioned, an infant's facial anatomy can be misleading when evaluating the eyes for alignment. The immature nasal bridge often is broad enough to cover the nasal sclera of either eye, and parents may interpret such asymmetry of the amount of white visible about either eye as

Table 2. Pediatric Eye Evaluation Screening Recommendations for Primary Care Providers, Nurses, Physician's Assistants, and Trained Lay Personnel*

Recommended Age for Screening	Screening Method	Criteria for Referral to an Ophthalmologist
Newborn	Red reflex†	Abnormal or asymmetric
To 3 mo	Inspection	Structural abnormality
6 mo to 1 year	Fix and follow with each eye	Failure to fix and follow in cooperative infant
	Alternate occlusion	Failure to object equally to covering each eye
	Corneal light reflex	Asymmetric
	Red reflex	Abnormal or asymmetric
	Inspection	Structural abnormality
3 y (approximately)	Visual acuity‡	20/50 or worse or 2 lines difference between the eyes
	Corneal light reflex cover-uncover	Asymmetric/ocular refixation movements
	Red reflex	Abnormal or asymmetric
	Inspection	Structural abnormality
5 y (approximately)	Visual acuity	20/40 or worse or 2 lines of difference between the eyes
	Corneal light reflex	Abnormal or asymmetric
	Stereoacuity	Failure to appreciate stereopsis
	Red reflex	Abnormal or asymmetric
	Inspection	Structural abnormality
Older than 5 y	Visual acuity	20/30 or worse or 2 lines of difference between the eyes
	Corneal light reflex/ cover-uncover	Asymmetric/ocular refixation movements
	Stereoacuity§	Failure to appreciate stereopsis
	Red reflex	Abnormal or asymmetric
	Inspection	Structural abnormality

* From American Academy of Ophthalmology Preferred Practice Pattern.
† Physician or nurse responsibility.
‡ Figures, letters, "tumbling E" or optotypes.
§ Optional: Random Dot E Game, Titmus Stereograms (Titmus Optical, Inc, Petersburg, Va.), Randot Sterograms (Stereo Optical Company, Inc, Chicago, Ill.).

evidence of eye misalignment. Pseudostrabismus is the appearance of eye misalignment when, in fact, no strabismus is present (Fig. 10). There is no strabismus if a cover test does not detect any, although sufficient fixation can be problematic when evaluating an infant.

The corneal reflex test is helpful to differentiate strabismus from pseudostrabismus if fixation is poor. The corneal reflex test is performed with an ordinary penlight held more than 18 inches from the child while the child either is observing the light or looking in its general direction. The light reflecting off the cornea, known as the corneal reflex, should be essentially central in the pupil. If the reflex does not appear to be central in the pupil, but is deviated to the periphery of the pupil or further, a strabismus is present until proven otherwise (Fig. 11).

Problems with a penlight test include missing a small or intermittent strabismus and the procedure not being helpful if a child is not alert enough to allow any fixation. Most actual eye deviations in a preverbal child are large, allowing differentiation with this method. Intermittent strabismus can be present, but this phenomenon occurs most commonly in children 2 to 4 years of age, when accommodative effort starts to cause esotropia with near fixation effort, known as accommodative esotropia. Accommodative esotropia can be detected with a corneal reflex test by using a small target for fixation, such as the eyes of a finger puppet or the number on a pencil, while determining the corneal light reflex position (Fig. 12).

When to Screen

Guidelines endorsed by the AAP recommend visual assessment at birth and at all subsequent routine health supervision visits. Anatomy and gross visual assessments should be checked from birth to 3 years of age, about the age when visual acuity can be measured reliably and ophthalmoscopy can be attempted. Vision should be assessed whenever there is a complaint about vision (Table 2).

Who Should Be Screened

All patients should be evaluated at the youngest possible age. Frequent assessments in those younger than age 3 years result in improved visual outcomes. Patients who have a family history of strabismus, amblyopia, congenital glaucoma, or retinal diseases clearly benefit from close observation and referral if screening warrants it. Patients who have developmental delay are at high risk of having a coexistent visual difficulty. Patients who have Down syndrome are at higher risk for vision-related difficulties, including refractive error, strabismus, and cataract. Preterm infants are at risk for ROP, requiring retinal evaluations at 4 to 6 weeks after delivery, as well as at higher risk for strabismus and refractive error later in childhood.

Summary

Vision screening provides children with their best possible vision for a lifetime of use. Screening can be performed in the office and can determine which patients are in need of additional evaluation. Screening is good medicine!

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PIR Quiz

Quiz also available online at www.pedsinreview.org.

Match the clinical situation with the *most* likely diagnosis.

5. The tendency toward inward deviation of the eye when that eye is covered.
6. The refractive state most likely to be found in a 3-year-old child.
7. The most likely result of pronounced ptosis of the right eyelid in a 4-year-old child.
8. Active cortical suppression of the vision of one eye.

A. Amblyopia.
B. Esophoria.
C. Esotropia.
D. Hyperopia.
E. Myopia.

9. The parents of a 1-month-old infant ask you what she can see. You tell them that their daughter's visual acuity is *most* likely:

A. 20/30.
B. 20/80.
C. 20/100.
D. 20/200.
E. 20/400.

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