

Pediatric Vision Screening

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Practice Gap

Incorporating vision screening and a basic eye examination in the primary care setting can be challenging. Determining which screening examination to perform and when to refer a patient to a pediatric eye care provider is critical.

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

1. Understand the importance of vision screening and know what conditions can be detected by periodic eye examinations.
2. Describe the components of a vision screening examination at different ages and plan an appropriate evaluation of vision.
3. Recognize the indications for referral to pediatric ophthalmology.

INTRODUCTION

Vision screening is crucial for early detection and prevention of vision loss in young children. Vision screening can be performed by primary care providers, trained laypersons (eg, school-based screenings), and eye care providers. Vision screening techniques are either provider-based (eg, traditional acuity testing, inspection, red reflex testing) or instrument-based. Instrument-based screening can often be performed at an earlier age than provider-based acuity testing and allows earlier screening for risk factors that are likely to lead to amblyopia and poor vision. The American Academy of Pediatrics (AAP) and the American Association for Pediatric Ophthalmology and Strabismus have developed guidelines to help practitioners screen for vision problems at different ages (Table 1).

AUTHOR DISCLOSURE Dr Loh has disclosed no financial relationships relevant to this article. Dr Chiang has disclosed that he has received research grants from the National Institutes of Health and the National Science Foundation, that he is an unpaid member of the Scientific Advisory Board of Clarity Medical Systems, and that he is a consultant/steering committee member of the RAINBOW study for Novartis. This commentary does not contain a discussion of an unapproved/investigative use of a commercial product/device.

THE IMPORTANCE OF VISION SCREENING

Vision screening allows the early detection of preventable vision-threatening or life-threatening conditions. Amblyopia, colloquially called “lazy eye,” is a reduction of best-corrected visual acuity that is not directly caused by any structural abnormality of the eye. It is caused by an abnormal visual experience resulting from strabismus, refractive error, or stimulus deprivation. Amblyopia occurs in 1% to 4% of children (1) and can be caused by visual deprivation (eg, cataract, ptosis, corneal opacity), strabismus (any form of eye misalignment, such as

TABLE 1. Age-Appropriate Methods for Pediatric Vision Screening and Criteria for Referral

METHOD	INDICATIONS FOR REFERRAL	RECOMMENDED AGE				
		NEWBORN-6 MO	6 MO UNTIL COOPERATIVE FOR VISION TESTING	3-4 Y	4-5 Y	EVERY 1-2 Y AFTER AGE 5 Y
Red reflex test	Absent, white, dull, asymmetrical				Evaluate at all ages	
External inspection	Structural anomaly (eg, ptosis, hemangioma)				Evaluate at all ages	
Pupil examination	Unequal size, poor reaction to light, irregular shape				Evaluate at all ages	
Corneal light reflex	Asymmetrical or displaced		*	*	*	*
Instrument-based screening	Failed to meet screening criteria		*	*	*	*
Cover test	Refixation movement			*	*	*
Visual acuity (distance, monocular)		Failure to fixate and follow	Failure to fixate and follow	20/50 or worse in either eye	20/40 or worse in either eye	20/30 or worse or 2-line difference between eyes

Asterisks indicate the method of vision screening that should be performed in that age column.

esotropia or exotropia), high refractive error (eg, hyperopia or myopia), or anisometropia (asymmetry of refractive error between the eyes). Amblyopia is more easily and successfully treated the earlier it is detected, and it becomes impossible to treat after 7 to 9 years of age. Patients and caregivers may be unaware of the consequences of delayed evaluation and treatment. (2) The short window of opportunity to save vision underscores the importance of vision screening to detect amblyopia or its risk factors while treatment is still effective. Vision screening assessments in early childhood reduce the risk of vision loss at age 7 years by more than 50%. (3)

Vision screening and eye examinations within the medical home create frequent and early opportunities to diagnose a myriad of conditions. In the first year of life, causes of deprivation amblyopia are more frequent and result in the most profound vision loss. Conditions affecting infants include corneal opacities, cataracts, ptosis, glaucoma, and retinoblastoma. Deprivation amblyopia in infancy can develop very rapidly. A few weeks of deprivation of visual stimulus in 1 eye from a dense cataract or complete ptosis can result in profound amblyopia that often requires years of numerous hours of patching treatment. Early detection of retinoblastoma can save the child's vision and life. Ocular abnormalities may be the first recognized sign of a systemic disease. For example, blurred vision and bilateral cataracts

in a child can be the first presentation of a neurodegenerative disease, cerebrotendinous xanthomatosis, that if unrecognized causes irreversible cognitive impairment. Early recognition and treatment with an oral medication can prevent lifelong disability. In addition, a crossed eye may be the first sign of vision loss in a baby with optic nerve hypoplasia as part of septo-optic dysplasia. Effective screening by the pediatrician can lead to earlier diagnosis of systemic problems.

The etiologies of amblyopia vary with age. Form deprivation amblyopia is more frequent and profound in infants. A unilateral cataract in a newborn will cause substantial amblyopia if untreated and should be removed within weeks; in contrast, a traumatic cataract in a 6-year-old with previously good vision is much less likely to cause amblyopia, and in the absence of other damage from the trauma, the cataract can be removed nonurgently. In children younger than 3 years, strabismus is the most common cause of amblyopia; in children 3 to 6 years old, strabismus and anisometropia contribute equally. (4) In younger children (eg, up to age 3 years), the examination is more challenging and the disorders can be subtle, but early detection can have a profound effect on the child's vision and future if treatment is initiated quickly. To detect these abnormalities, vision screening should be performed by primary care providers or trained laypersons (eg, school-based screenings) throughout childhood. The combined sensitivity of a series of screening evaluations is higher than a

single evaluation, especially if different methods are used for each screening evaluation. (5) Unfortunately, children present to the ophthalmologist at age 6 to 8 years for their first eye examination with significant amblyopia (frequently, anisometropic amblyopia) whose vision loss would have been preventable if they had been detected earlier and been prescribed glasses at a younger age. The routine assessment of vision in all children cannot be overemphasized; special attention should also be given to children with disabilities. Children with developmental delay can experience a delay in identification of their ocular disease that further impedes successful treatment.

Some children should be referred directly for a comprehensive examination. For example, poor eye contact by a term infant with the caretaker after 8 weeks of age warrants further assessment (8 weeks' adjusted age for premature children). Table 2 lists red flag signs and symptoms for possible eye problems. Special attention should be given to children with a history of a known medical risk factor for vision problems, including prematurity, cerebral palsy, craniofacial abnormalities, Down syndrome, Marfan syndrome, congenital cytomegalovirus, eyelid hemangiomas, Sturge-Weber syndrome, sickle cell disease, and nevus of Ota. Children with medical conditions such as diabetes or juvenile idiopathic arthritis should receive a comprehensive evaluation soon after diagnosis.

PROVIDER-BASED VISION SCREENING

Red Reflex Testing

The red reflex test is the most important screening test for infants and young children. Red reflex testing requires no patient participation and can be performed shortly after birth. A direct ophthalmoscope is used to view both eyes simultaneously from 2 to 3 ft away from the patient (Fig 1).

The red reflex represents reflection of the retina through a clear pupillary axis. Distortion in the red reflex can be caused by an abnormality anywhere in the visual axis (eg, in the retina [retinoblastoma], vitreous [vitreous hemorrhage], lens [cataract], or cornea [scar or infection]). A difference in the red reflex can also be caused by asymmetry in the refractive power of the eye, which can cause amblyopia and be vision threatening. The AAP recommends routine screening for structural abnormalities using red reflex testing. (6) Figure 2 represents examples of normal and abnormal red reflex testing.

Red reflex testing allows for the prompt diagnosis of and referral for leukocoria (white pupil), which occurs when there is an opacity preventing a clear view of the retina. The most concerning cause of leukocoria is retinoblastoma, a life-threatening tumor in children. All patients with abnormal red reflex tests should be referred to pediatric ophthalmology, and concern for leukocoria should be urgently referred.

External Examination

The external examination of the eyes, eyelids, and face is an important part of the visual system screening in a primary care office. A simple penlight examination of the eyelids can reveal ptosis, capillary hemangiomas, and port wine stains, which are risk factors for amblyopia and systemic diseases. Careful inspection of the globe size is important in screening for pediatric glaucoma, which can cause either unilateral or bilateral ocular enlargement. Ptosis requires prompt identification because it can cause amblyogenic astigmatism even if the lid itself does not block the visual axis. Substantial ptosis obstructing the visual axis requires urgent referral in any child. For subtler ptosis, referral to an ophthalmologist is appropriate for children too young to assess vision using either optotypes (figures or letters of different sizes used to test vision acuity) or instrument-based screening. In older children who pass a

TABLE 2. Signs and Symptoms of Potential Vision Loss

SIGN/SYMPTOM	POTENTIAL EYE PROBLEM	PEDIATRIC PROVIDER ACTION
No eye contact in an infant >8 wk old corrected age	Decreased vision, delayed vision maturation	Vision screen and refer to ophthalmologist
Head tilt or face turn	Strabismus, nystagmus, high astigmatism	Vision screen and refer to ophthalmologist
Unable to comply with vision screening	Decreased vision	Refer to ophthalmologist
Tearing	Congenital nasolacrimal duct obstruction, glaucoma	Age <1 y without other signs of glaucoma (enlarged corneas, photophobia, blepharospasm), vision screen Age >1 y refer to ophthalmology
Photophobia	Congenital glaucoma, inflammation	Vision screen and consider referral
Squinting	Refractive error, strabismus	Vision screen and consider referral

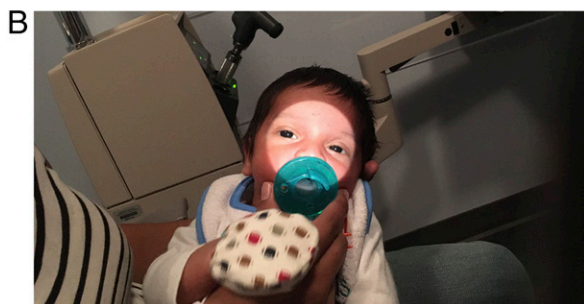


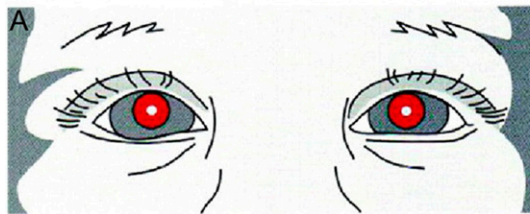
Figure 1. A. A provider performing red reflex testing approximately 2 to 3 ft from the infant. B. The provider's view of bilateral red reflex viewing both eyes simultaneously.

vision screening with subtle ptosis, referral to ophthalmology is not required. Ptosis combined with miosis, which is an asymmetrically constricted pupil, may represent Horner syndrome and would require an ophthalmology evaluation for neuroblastoma in children. In addition, an enlarged globe (buphthalmos) is caused by elevated eye pressure. Any enlarged eye, especially with a history of tearing and photophobia, should be referred immediately for concern of pediatric glaucoma. Careful inspection by an astute primary care provider is crucial to the early identification of vision-threatening and potentially life-threatening problems.

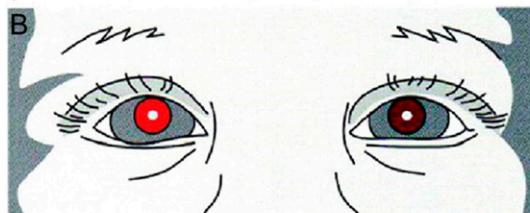
Visual Acuity Testing

What is normal visual acuity? Normal acuity changes with age because visual acuity improves as children grow. Infants,

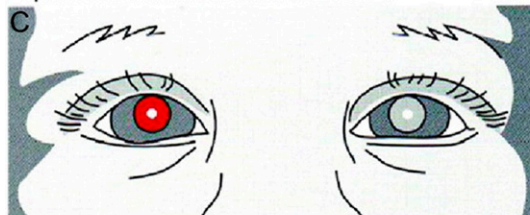
↓ **NORMAL**—Child looks at light. Both red reflections are equal.



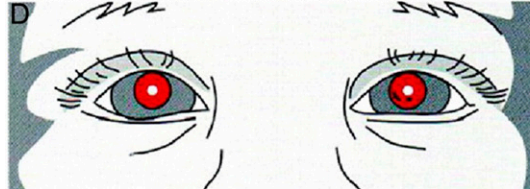
↓ **UNEQUAL REFRACTION**—One red reflection is brighter than the other.



↓ **NO REFLEX (CATARACT)**—The presence of lens or other media opacities blocks the red reflection or diminishes it.



↓ **FOREIGN BODY/ABRASION (LEFT CORNEA)**—The red reflection from the pupil will back-light corneal defects or foreign bodies. Movement of the examiner's head in one direction will appear to move the corneal defects in the opposite direction. (Parallax)



↓ **STRABISMUS**—The red reflection is more intense from the deviated eye.

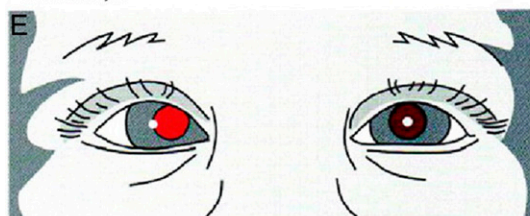


Figure 2. Red reflex examination. A. Normal: Child looks at light. Both red reflections are equal. B. Unequal refraction: One red reflection is brighter than the other. C. No reflex (cataract): The presence of lens or other media opacities blocks the red reflection or diminishes it. D. Foreign body/abrasion (left cornea): The red reflection from the pupil will backlight corneal defects or foreign bodies. Movement of the examiner's head in 1 direction will appear to move the corneal defects in the opposite direction. E. Strabismus: The corneal light reflex is temporally displaced in the misaligned right eye, indicating esotropia. (Reprinted with permission from American Academy of Pediatrics; Section on Ophthalmology; American Association for Pediatric Ophthalmology and Strabismus; American Academy of Ophthalmology; American Association of Certified Orthoptists. Red reflex examination in neonates, infants, and children. *Pediatrics*. 2008;122(6):1402. Image courtesy of Alfred G. Smith, MD © 1991.)

o to 2 months of age, should blink in response to bright light and have equal pupillary responses. Additional signs of normal vision in infants include the “eye-popping reflex.” For the first year of life, children with normal vision manifest an eye-popping reflex where the eyelids retract on turning off the lights. It can be clinically helpful to determine that an infant has at least light perception vision when parents are concerned that their baby cannot see. The eyelid retraction disappears when ambient light is turned on, and the phenomenon is thought to be a form of primitive startle reflex. (7) Fixation and tracking should improve and become reliable around 6 to 8 weeks of life. Premature children may have some delay in their visual development and may not fix and follow until their adjusted age reaches 8 weeks. Newborns may have intermittent strabismus (either eye turning outward or inward), but this should resolve by 2 to 4 months of age. Any constant strabismus is considered abnormal at any age, and intermittent strabismus after 4 months warrants referral. From age 6 months to 2 years, children should be able to fix and follow an object monocularly and have normal alignment. After age 3 to 5 years, subjective vision can usually be measured using eye charts. Video 1 demonstrates a technician checking vision in



Video 1. Video of a technician checking vision monocularly in a 2.5-year-old girl using matching LEA SYMBOLS® optotypes and an eye patch.

a 2.5-year-old girl using matching LEA SYMBOLS® (Good-Lite Co, Elgin, IL) optotypes (standard symbols such as letters or pictures). A practical tip for checking vision in very young children is to begin the “matching game” before covering each eye to confirm understanding and to encourage participation before introducing the eye patch.

Children 3 years or older typically can participate in provider-based subjective visual acuity testing. Children who cannot participate in subjective visual acuity testing are considered untestable, and untestable children have been shown to have vision problems more often than testable children. (8) Repeated examination in 6 months is recommended, and inability to assess vision in a 3- to 5-year-old merits referral to an eye care provider. (5) Recommendations for referral based on visual acuity are presented in Table 1.

Recognition visual acuity testing is the gold standard in vision screening and the preferred method for assessing vision to detect amblyopia, especially in older children. Vision is routinely tested at 2 standard distances (10–20 ft for distance vision and 14–16 in for near vision). Vision should be measured monocularly, which involves sufficiently occluding 1 eye with an adhesive patch or occlusive tape. Vision should be checked while the child is wearing any necessary corrective lenses. Young children improve performance if allowed to match optotypes presented on the chart to a handheld card (eg, Video 1). The choice and presentation of optotypes on an eye chart affect the visual acuity obtained. The current recommendations by the American Academy of Ophthalmology are for LEA SYMBOLS® (Fig 3) or HOTV letters to be used as the preferred optotypes for preliterate children. The goal is for the optotypes to be standardized, clear, and without cultural bias. Allen figures, tumbling E charts, and Lighthouse symbols are not standardized and are no longer recommended as preferred optotypes.

Cover and Hirschberg Testing for Strabismus Evaluation

A common concern among parents is for strabismus (misalignment of the eyes). An esodeviation refers to a convergent misalignment of the visual axis. Esophoria is a latent esodeviation that under normal binocular conditions the eyes remain properly aligned. Esotropia is an esodeviation that is not controlled by fusional mechanisms, so the deviation is constantly manifest. Exodeviation is a divergent strabismus that can be latent (exophoria) or manifest (exotropia). Most people have some latent strabismus (esophoria and exophoria) that can be revealed by extensive cover testing and is not amblyogenic, but manifest strabismus is a frequent cause of amblyopia.

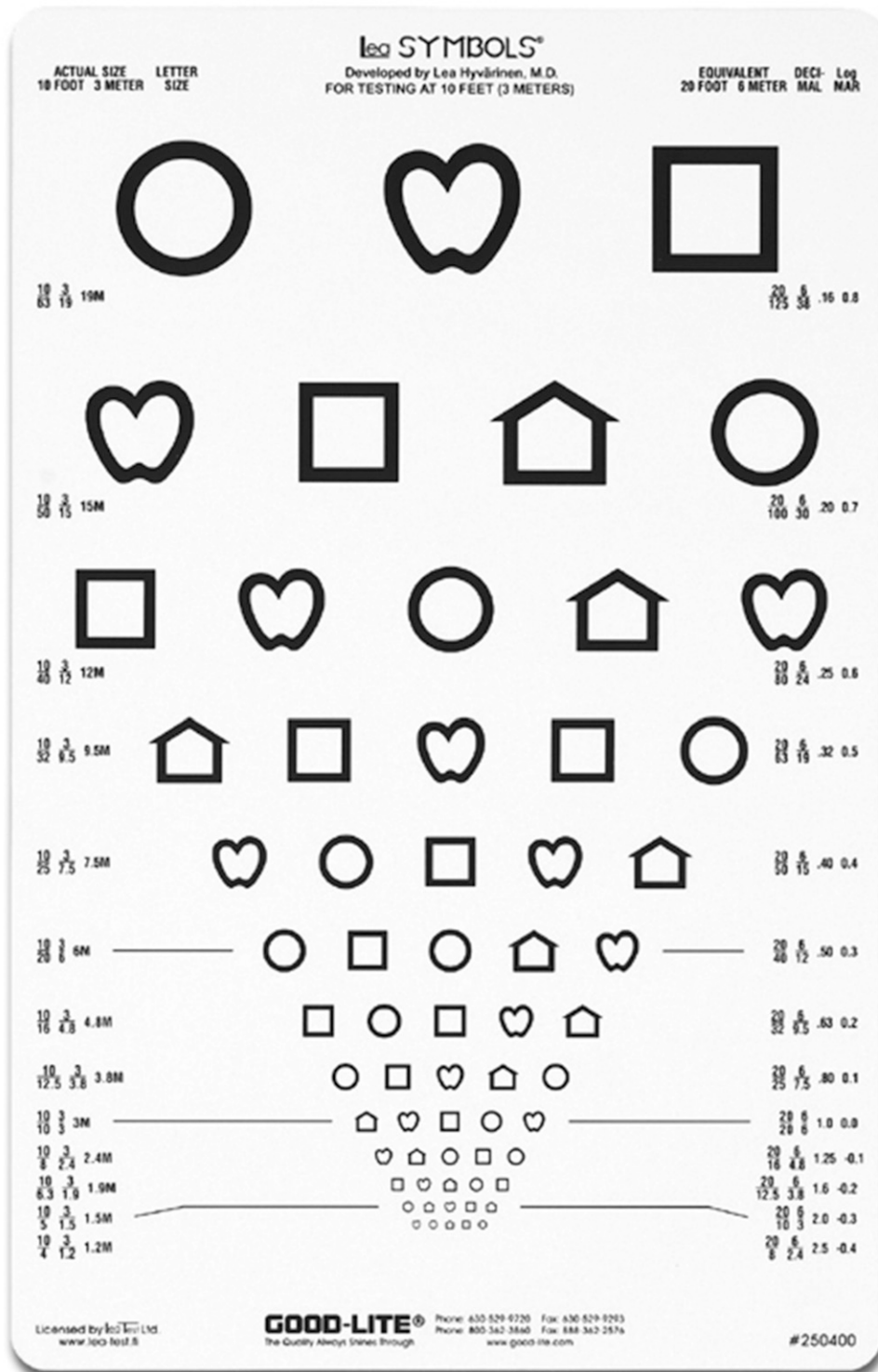


Figure 3. LEA SYMBOLS®. (Reprinted with permission from Good-Lite Co, Elgin, IL)

Examination techniques such as cover testing and Hirschberg testing are crucial for evaluating concern for strabismus. For example, parents can report esotropia but may in fact be appreciating pseudoesotropia. Children with large epicanthal

folds or a flat nasal bridge often have pseudoesotropia (appearance of crossed eyes when the eyes are in fact straight), which may be diagnosed by symmetry of the pupillary light reflex when shining a penlight toward the patient or by cover



Figure 4. Pseudoesotropia with wide epicanthal folds. Despite the left eye appearing crossed because there is less white sclera visible nasally, the well-centered light reflexes confirm pseudoesotropia.

testing (Fig 4). If the primary care provider is confident in the diagnosis of pseudoesotropia, these patients do not require referral to ophthalmology.

The cover-uncover test and the Hirschberg test are used to determine whether there is a manifest misalignment of the eyes. The Hirschberg test is a simple test where a penlight is directed at the patient while he or she is looking straight ahead. The light reflex should be reflected in the center of each pupil if the patient's eyes are straight. If there is a manifest exotropia, the light reflex will be nasal to the pupil. If there is a manifest esotropia, the light reflex will be temporal to the pupil (Fig 5). The Hirschberg test, along with the cover test, can be very helpful in distinguishing a benign pseudoesotropia from a true manifest strabismus requiring ophthalmology referral.

The cover test requires the child to fixate either at near (eg, a sticker on an examiner's nose or a toy held up close) or distance (eg, a television or parent at the end of the room). If the patient is watching your nose and there is a manifest exodeviation of the right eye (ie, the right eye is turning out), the left eye is straight and fixating. The cover test involves covering the fixating eye and watching the deviated eye shift toward central fixation. If the misaligned eye is drifted out,



Figure 5. Esotropia of the left eye.

the eye will shift inward when the fixating eye is covered. If the deviated eye is turned in, the eye will shift outward when the fixating eye is covered. Video 2 demonstrates a child with esotropia whose inward-turned eye refixates outward when his opposite eye is covered.

Ocular Motility and Nystagmus

In addition to alignment, ocular motility is an important part of the eye examination. Parents who observe “funny eye movements” may be the first observers of a complex strabismus syndrome, such as congenital fourth nerve palsy, Brown syndrome, or Duane syndrome. Congenital fourth nerve palsy is characterized by a vertical misalignment of the eyes caused by weakness of the ipsilateral superior oblique muscle. Children typically have a head tilt toward the side of the palsy. Brown syndrome is also characterized by vertical misalignment of the eyes caused by a deficit of the superior oblique muscle where the affected eye cannot elevate in adduction. Duane syndrome is characterized by anomalous innervation of the lateral rectus by the sixth cranial nerve causing limitation in horizontal eye movements as well as retraction of the globe on attempted adduction. If a patient has an esotropia, evaluating motility is important for determining the urgency of the referral. The most common forms of esotropia, including congenital esotropia and accommodative esotropia, typically have normal ocular motility. Limitation of abduction (eye movement away from the nose) can



Video 2. Video of a provider evaluating for strabismus. This is a 7-year-old boy with esotropia whose inward-turned eye refixates outward when his opposite eye is covered.



Video 3. Video of a technician using a vision screener with a 6-year-old boy.

be a sign of a sixth nerve palsy in contrast to congenital or accommodative esotropia with full ductions and no abduction limitation. In addition, the presence of nystagmus or unusual eye movements in an infant or young child can indicate decreased vision or neurologic dysfunction and warrants further evaluation by either an ophthalmologist or a neurologist.

Pupil Examination

The pupils should be equal in size and reactive to light from birth. A dim room and using a bright light elicits the best pupil response, especially in newborns. The pupils normally will decrease in diameter with accommodation and if the child is looking at the examiner at near; the pupil response may be less marked if the pupils are small already during

TABLE 3. Instrument-Based Vision Screening Devices (18)

	DEVICE				
	ISCREEN (14)	PLUSOPTIX S09, S12C (19)	SPOT SCREENER (12)	RIGHTON RETINOMAX (4)	SURESIGHT (20)
Type	Photoscreener	Hybrid	Hybrid	Autorefractor	Autorefractor
Monocular/binocular	Binocular	Binocular	Binocular	Monocular	Monocular
Image interpretation	Vendor	Automated	Automated	Automated	Automated
Conditions screened	Refractive error, strabismus, anisometropia, anisocoria, cataracts	Refractive error, strabismus, anisometropia, anisocoria	Refractive error, strabismus, anisometropia, anisocoria	Refractive error	Refractive error
Cost per machine, \$	4,200	7,595	7,490	12,495	3,999
Cost per test	~\$10	None	None	None	
Time per test, seconds	7	1	1	1	1
Time for results	<1 h	Instant	Instant	Instant	Instant

Data from Colburn J. Comparison of instrument-based vision screening devices. American Academy of Ophthalmology website. <https://www.aao.org/pediatric-center-detail/vision-screening-performance-data-resource-2>. Published 2014. Accessed August 1, 2017

accommodation. Any evidence of anisocoria or pupils of different shapes should be referred to an ophthalmologist.

INSTRUMENT-BASED VISION SCREENING

Provider-based visual acuity assessment depends on child participation and screener experience. With practice, instrument-based vision screening can be fast and require less participation from the child. (Video 3) Instrument-based screening can be very helpful in screening children before reliable subjective visual acuity can be obtained. (9) Recent guidelines released by the AAP in January 2016 recommend instrument-based screening starting at 1 year old and continuing until the child can reliably read the eye chart. (10)

There are 2 types of instrument-based vision screening: photoscreeners and autorefractors. Neither type measures visual acuity itself but rather measures risk factors for vision loss, including myopia, hyperopia, astigmatism, and strabismus. Children with retinal disease or structural causes for amblyopia may have false-negative screening tests. Amblyopia risk factors were identified in 5% of preschool children participating in 16 photoscreening programs of more than 400,000 children. (11) Approximately 4% of children younger than 6 years have myopia, 5% to 10% have astigmatism, and up to 20% have hyperopia. (12)(13) (14) Photoscreeners are binocular devices that estimate refractive error, media clarity, ocular alignment, and eyelid position. Photoscreeners have been shown to have high sensitivity and specificity when used in community and office settings. (15)(16)(17) Autorefractors monocularly estimate refractive error and are useful for screening for high refractive error and anisometropia. Once a child is old enough to reliably read an eye chart, direct visual acuity should supplement vision screening. Instrument-based vision screening would not detect structural abnormalities causing decreased vision (eg, retinal dystrophies or optic nerve hypoplasia) even if vision was poor because visual acuity is not directly measured. Table 3 compares common

commercially available instrument-based vision screening devices.

VISION SCREENING CODING

Last, *Current Procedural Terminology* codes 99173 and 99174 are specific for provider-based visual acuity screening and instrument-based photoscreening, respectively. The AAP recommends that vision screening not be bundled into the global code of well-child care. Adequate reimbursement for photoscreening must be encouraged to promote widespread adoption of vision screening. Unfortunately, some insurance plans may not cover vision services. The National Eye Institute has information regarding vision services available to uninsured and underinsured children (<https://nei.nih.gov/health/financialaid>).

Summary

- On the basis of expert consensus as well as prospective cohort research, routine vision screening decreases the incidence of vision loss in early childhood. (3)
- Based on consensus, the most important aspects of a provider-based vision screening are red reflex testing, external examination of lids and adnexa, ocular motility, and visual acuity testing.
- Based on some research evidence as well as consensus, instrument-based vision screening can be used to reliably evaluate vision in children. (10)
- Referral to an eye care provider is indicated if a patient does not pass a component of the vision screen or when further diagnostic and management recommendations are required.

References for this article are at <http://pedsinreview.aappublications.org/content/39/5/225>.

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1. A 3-year-old boy is brought to the clinic for a routine health supervision visit. He was the product of a full-term pregnancy and spontaneous vaginal delivery. He has been followed in the same clinic for his health supervision visits and he has had normal growth and development. Which of the following is the recommended method to be used to best assess visual acuity in this child?
 - A. Allen figures.
 - B. Ishihara plates.
 - C. LEA SYMBOLS®.
 - D. Lighthouse symbols.
 - E. Tumbling E charts.
2. You are evaluating a 2-year-old boy with Down syndrome. His mother is concerned that his eyes are turning in. In view of the fact that patients with Down syndrome have epicanthal folds and a wide nasal bridge, you want to differentiate true strabismus from benign pseudoesotropia. Which of the following is the most appropriate next assessment to perform in this patient to determine whether this is a true finding?
 - A. Check for a symmetrical red reflex.
 - B. Check for buphthalmos.
 - C. Confirm normal visual acuity.
 - D. Demonstrate an "eye-popping reflex."
 - E. Perform the Hirschberg test.
3. A 6-year-old girl is found to have left exodeviation on physical examination using the Hirschberg test and the cover-uncover test. Which of the following responses to the cover-uncover test with near focus confirms the strabismus in this patient?
 - A. Cover the left eye and the right eye turns laterally.
 - B. Cover the left eye and the right eye turns medially.
 - C. Cover the right eye and the left eye does not change position.
 - D. Cover the right eye and the left eye turns laterally.
 - E. Cover the right eye and the left eye turns medially.
4. Your practice is considering the implementation of instrument-based vision screening in young children to comply with the 2016 American Academy of Pediatrics guidelines. Which of the following is the primary goal for this screening tool?
 - A. Identify retinal disorders.
 - B. Identify risk factors for vision loss.
 - C. Identify optic nerve hypoplasia.
 - D. Measure intraocular pressure.
 - E. Measure visual acuity.
5. The parents of a 9-month-old child are concerned about their child's vision because there is a strong family history of glaucoma. The child is brought to the office for vision screening and evaluation. The presence of which of the following findings would be most concerning for the diagnosis of glaucoma?
 - A. Anisocoria.
 - B. Photophobia.
 - C. Ptosis.
 - D. Squinting.
 - E. Strabismus.

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