



NCC Pediatrics Continuity Clinic Curriculum: **Health Maintenance III** *Faculty Guide*



Objective: To recognize the most common procedures used in pediatrics for Sensory Screening (hearing and vision), as part of the [2021 AAP Periodicity Schedule](#).

Pre-Meeting Preparation:

Please read the following enclosures, corresponding to the screening procedures:

1) Sensory Screening:

- a. Vision: “Pediatric Vision Screening” (*PIR, 2013*)
- b. Hearing: “Pediatric Hearing Screening”, OR "Hearing Screening in Primary Care" (*PIR, 2021*)

2) **Bring a hand-held ophthalmoscope or penlight to clinic, if you have one.**

Conference Agenda:

- Review “Health-Maintenance III Quiz”
- **Complete the following group activities for each screening procedure:**
 - 1) Vision Screening: *Encourage residents to do the maneuvers on each other, before completing the picture-examples.*
 - a. Snellen Eye-Chart
 - b. Corneal light reflex
 - c. Cover-Uncover
 - d. **SpotVision** Conversion Charts (*Faculty—review interpretation with residents*)
 - 2) Hearing Screening:
 - a. Conventional audiometer
 - b. Tympanometry

Extra-Credit: ☀️ Encouraged for PGY2 & 3 ☀️

- Vision: [Red Reflex Testing](#); [Vision Screening](#) (*AAP Clinical Practice Guidelines*)
 - **“Pediatric Vision Screening”** (*ppt by Dr. Erika Beard-Irvine*)
 - [“Instrument Based Vision Screening: Update and Review](#) (*Contemp Peds, 2014*)
 - ["The effectiveness of the Spot Vision Screener in detecting amblyopia risk factors"](#) (*J AAPOS, Dec 2015*)
- Hearing: [Hearing Screening Clinical Report](#); [Hearing Screening Policy Statement](#) (*AAP*)
 - **“Pediatric Hearing Screening”** (*ppt by Candice Ortiz Au.D.; play as slide-show*)

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).

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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



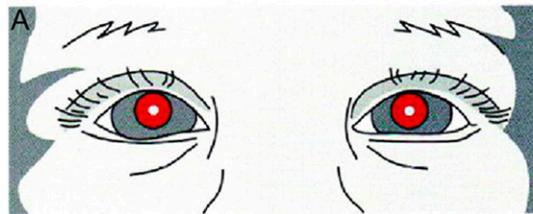
Figure 1. A. A provider performing red reflex testing approximately 2 to 3 ft from the infant. B. The provider's view of bilateral red reflex viewing both eyes simultaneously.

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.

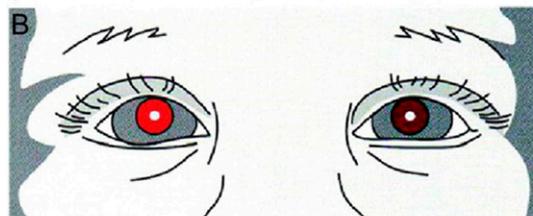
Visual Acuity Testing

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

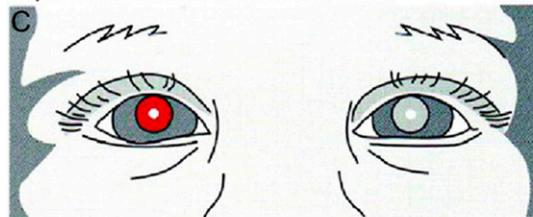
↓ **NORMAL**—Child looks at light. Both red reflections are equal.



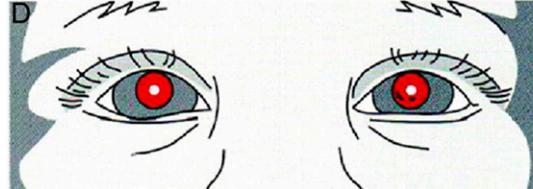
↓ **UNEQUAL REFRACTION**—One red reflection is brighter than the other.



↓ **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.

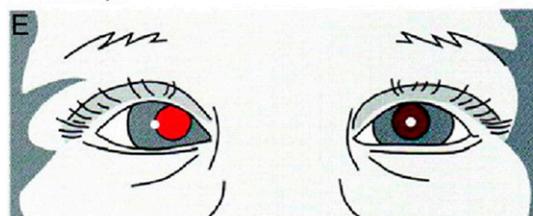


Figure 2. Red reflex examination. A. Normal: Child looks at light. Both red reflections are equal. B. Unequal refraction: One red reflection is brighter than the other. C. No reflex (cataract): The presence of lens or other media opacities blocks the red reflection or diminishes it. D. Foreign body/abrasion (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.

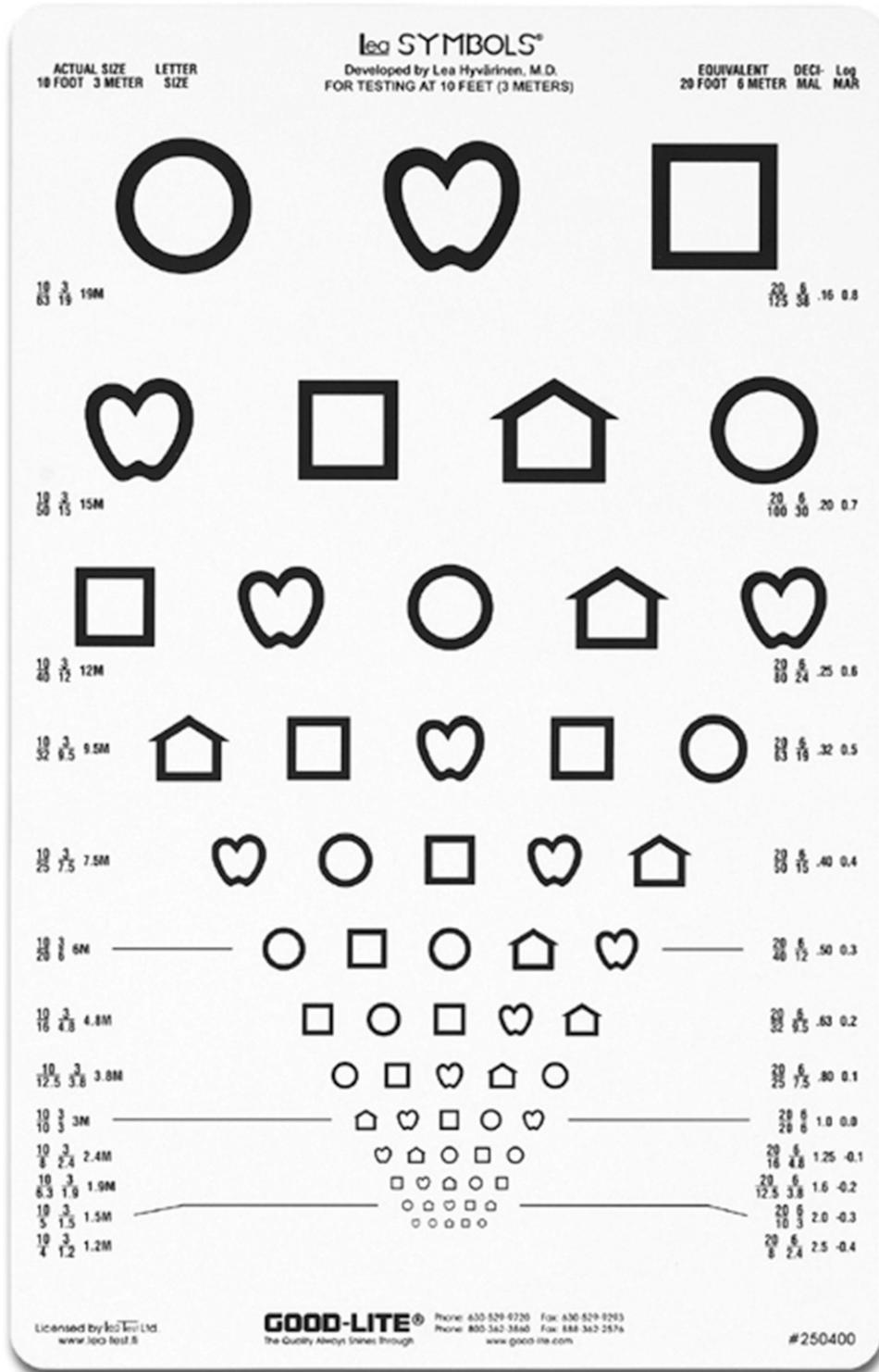


Figure 3. LEA SYMBOLS®. (Reprinted with permission from Good-Lite Co, Elgin, IL)

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	Phot screener	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>.

PIR Quiz

There are two ways to access the journal CME quizzes:

1. Individual CME quizzes are available via a handy blue CME link under the article title in the Table of Contents of any issue.
2. To access all CME articles, click "Journal CME" from Gateway's orange main menu or go directly to: <http://www.aapplications.org/content/journal-cme>.
3. To learn how to claim MOC points, go to: <http://www.aapplications.org/content/moc-credit>.

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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This journal-based CME activity is available through Dec. 31, 2020, however, credit will be recorded in the year in which the learner completes the quiz.



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Pediatric Hearing Screening

(Adapted from <http://www.entcolumbia.org/childscrn.html>)

I. Newborns and Infants

Hearing screening for newborns before they leave the hospital or maternity center is now becoming a common practice. Without such programs, the average age of hearing loss identification is between 12-25 months. When hearing loss is detected late, language development is already delayed.

Screening Techniques for Newborns and Infants

The screening of newborns and infants involves use of non-invasive, objective physiologic measures that include otoacoustic emissions (OAEs) and/or auditory brainstem response (ABR). Both procedures can be done painlessly while the infant is resting quietly.

- *Otoacoustic emissions* (OAEs) are inaudible sounds from the cochlea when audible sound stimulates the cochlea. The outer hair cells of the cochlea vibrate, and the vibration produces an inaudible sound that echoes back into the middle ear. This sound can be measured with a small probe inserted into the ear canal. Persons with normal hearing produce emissions. Those with hearing loss greater than 25-30 dB do not. OAEs can detect blockage in the outer ear canal, middle ear fluid, and damage to the outer hair cells in the cochlea.
- *Auditory brainstem response* (ABRs) is an auditory evoked potential that originates from the auditory nerve. It is often used with babies. Electrodes are placed on the head, and brain wave activity in response to sound is recorded. ABR can detect damage to the cochlea, the auditory nerve and the auditory pathways in the stem of the brain.

What happens if an infant does not pass the screening?

Infants who do not pass a screening are often given a second screening to confirm findings and then referred for follow-up audiological and medical evaluations that should occur no later than 3 months of age. These evaluations confirm the presence of hearing loss; determine the type, nature, and (whenever possible) the cause of the hearing loss; and help identify options for treatment. Even if the infant passes screening, certain conditions do not produce immediate hearing loss. Rather, the hearing loss occurs later in the child's development.

II. Older Infants and Toddlers

Infants and toddlers (7 months through 2 years) should be screened for hearing loss as needed, requested, mandated, or when conditions place them at risk for hearing disability. Infants not tested as newborns should be screened before three months of age. Other infants should be screened who received neonatal intensive care or special care, or who display other indicators that place them at risk for hearing loss.

Screening Techniques for Infants, Toddlers and Children

Two screening methods are suggested as the most appropriate tools for children who are functioning at a development age of 7 months to 3 years, visual reinforcement audiometry (VRA) and conditioned play audiometry (CPA). Both of these methods are behavioral techniques that require involvement and cooperation of the child.

- *Visual reinforcement audiometry* (VRA) is the method of choice for children between 6 months and 2 years of age. The child is trained to look toward (localize) a sound source. When the child gives a

correct response, e.g., looking to a source of sound when it is presented, the child is "rewarded" through a visual reinforcement such as a toy that moves or a flashing light.

- *Conditioned play audiometry* (CPA) can be used as the child matures. It is widely used between 2 and 3 years of age. The child is trained to perform an activity each time a sound is heard. The activity may be putting a block in a box, placing pegs in a hole, putting a ring on a cone, etc. The child is taught to wait, listen, and respond.

With both of these methods, sounds of different frequencies are presented at a sound level that children with normal hearing can hear. It is ideal if the child will allow earphones to be placed on his or her head so that independent information can be obtained for each ear. If the child refuses earphone placement or earphone placement is otherwise not possible, sounds are presented through speakers inside a sound booth. Since sound field screening does not give ear specific information, a unilateral hearing loss (hearing loss in only one ear) may be missed.

Alternative procedures, such as otoacoustic emissions (OAEs) or auditory brainstem response (ABR) may be used if the child is unable to be conditioned.

What happens if a toddler does not pass the screening?

A toddler who does not pass the screening should be rescreened or referred for audiologic evaluation. Confirmation of hearing status should be obtained within 1 month, but no later than 3 months, after the initial screening.

III. Hearing Screening in Preschoolers

The goal of screening for hearing loss in preschoolers (ages 3-5 years) is to identify children most likely to have hearing loss that may interfere with communication, development, health, or future school performance. In addition, because hearing loss in this age range is so often associated with middle ear disease, it is also recommended that children in this age group be screened for outer and middle ear disorders (acoustic emittance screening).

Screening Techniques for Preschoolers

- *Conditioned play audiometry* (CPA) is the most commonly employed procedure.
- *Acoustic emittance screening* includes tympanometry, acoustic reflex, & static acoustic impedance:
 - ***Tympanometry*** introduces air pressure into the ear canal making the eardrum move back and forth. A special machine then measures the mobility of the eardrum. Tympanograms, or graphs, are produced which show stiffness, floppiness, or normal eardrum movement. They are classified as type A (normal), type B (flat, clearly abnormal), and type C (indicating a significantly negative pressure in the middle ear, possibly indicative of pathology).
 - *Acoustic reflex testing* measures the response of a tiny ear muscle that contracts when a loud sound occurs. The loudness level at which the acoustic reflex occurs and/or the absence of the acoustic reflex give important diagnostic information.
 - *Static acoustic impedance testing* measures estimate the physical volume of air in the ear canal. This test is useful in identifying a perforated eardrum or whether ear ventilation tubes are still open.

What happens if a preschooler does not pass the screening?

- If the child cannot be conditioned to the play audiometry, the child will be screened using infant-toddler procedures or will be recommended for a more in-depth audiologic assessment.

- If the child did condition and did not pass the screening, then referral for audiological assessment will be made. Hearing status of children referred after screening should be confirmed within 1 month, but no later than 3 months, after the initial screening.

IV. Hearing Screening for School Age Children and Adolescents (5-18 years)

School-age children should be screened for hearing loss as needed, requested, mandated, or when conditions place them at risk for hearing disability. Screening for hearing loss identifies the school-age children most likely to have hearing impairment that may interfere with development, communication, health, and education. School age children with even minimal hearing loss are at risk for academic and communication difficulties.

School age children should be screened at the following times: on first entry into school; every year from kindergarten through 3rd grade; in 7th & 11th grade; upon entrance into special education; upon grade repetition; upon entering a new school system without evidence of having passed a previous screening.

Screening techniques used for school-age students

- **Conventional audiometry**, in which students are instructed to raise their hand (or point to the appropriate ear) when they hear a tone, is the commonly used procedure. Conditioned play audiometry (CPA) is also used.

What happens if a school-age student does not pass the screening?

The student should be reinstructed, earphones repositioned, and rescreened in the same session. If the student does not pass the rescreening, he or she should be referred for audiologic assessment. Hearing status of referred students should be confirmed within one month, and no later than 3 months, after initial screening.

V. Risk Factors for Hearing Loss in Children

- Parental, caregiver and/or health care provider concerns regarding hearing, speech, language, and/or developmental delay based on observation and/or standardized developmental screening.
- Family history of permanent childhood hearing loss.
- Infections associated with sensorineural hearing loss including bacterial meningitis, mumps.
- In utero infections such as cytomegalovirus, herpes, rubella, syphilis, and toxoplasmosis.
- Neonatal indicators - specifically hyperbilirubinemia at a serum level requiring exchange transfusion, persistent pulmonary hypertension of the newborn associated with mechanical ventilation, and conditions requiring the use of extracorporeal membrane oxygenation (ECMO)
- Syndromes associated with progressive hearing loss such as neurofibromatosis, osteopetrosis, and Usher' s syndrome.
- Neurodegenerative disorders, such as Hunter syndrome, or sensory motor neuropathies, such as Friedreich's ataxia and Charcot-Marie-Tooth syndrome.
- Head trauma
- Recurrent or persistent otitis media with effusion for at least 3 months.
- Anatomic disorders that affect eustachian tube function
- Reported exposure to potentially damaging noise levels or to drugs that cause hearing loss.

Health Maintenance III Quiz

1. At what ages does the AAP recommend **hearing screening**? Do we perform hearing screening in our clinic? If so, how? **Newborn testing (e.g. OAE on the MICC), followed by yearly risk assessment until age 4**, at which time audiometry may be started. (Routine screening then occurs at 4-6 yrs, 8 yrs, and 10 yrs). *We do hearing screening in clinic; refer to audiology (5th floor America Building) if screening failed (may walk-oin for repeat screen x 1 prior to audiology referral.)*

2. At what ages does the AAP recommend **vision screening**? Do we perform vision screening in the clinic? If so, how? **Risk assessment starting as newborn. Screening to be performed starting at age 3 yrs** (3-6 yrs; 8 yrs; 10 yrs; 12 yrs; 15 yrs; 18 yrs). *We use SpotVision Screening at well-baby/well-child visits, starting at 12 months— annually unless some abnormality, then repeated at the next well visit. Snellen Chart used in adolescent clinic and older children in peds clinic on case by case basis.*

3. Please fill in the appropriate sensory screening tests for each of the following age-groups:

Age-Group	Hearing Screen	Vision Screen
Neonate	OAE (otoacoustic emission) ABR (auditory brainstem response)	Red reflex Inspection Fix & follow with each eye Corneal light reflex
Toddler	VRA (visual reinforcement audiometry) CPA (conditioned play audiometry)	Visual acuity (figures) Corneal light reflex Cover-Uncover Red reflex/inspection
Preschool	CPA Tympanometry Static acoustic impedance Acoustic Reflex	Visual acuity (Tumbling E) Corneal light reflex Stereoaucuity (Random Dot E Game) Red reflex/inspection
School-Age	Conventional audiometry	Visual acuity (Snellen Chart) Corneal light reflex Cover-uncover Stereoaucuity Red reflex/inspection

4. Please define the following terms:

a. Myopia = “**Nearsightedness**”. Refractive error where distant objects appear blurred.

b. Astigmatism = A type of refractive error where **vision is blurred** due to either an irregularly shaped cornea or lens which prevents light from focusing properly on the retina. This causes blurred vision at any distance.

c. Strabismus = **Misalignment of the eyes**, including tropias and phorias. Onset is between 18mo and 6yrs. Distinguish from psuedostrabismus, which is the appearance of misalignment, for example due to a broad nasal bridge covering the nasal sclera.

- d. Esotropia = *Full-time occurrence* of eyes to deviate **inward** (adducting)
- e. Exophoria = *Tendency* of eyes to deviate **outward** (abducting)
- f. Amblyopia = **Loss of visual acuity** due to active cortical suppression of the vision of an eye. Can be due to strabismus or deprivation (e.g. congenital cataract).
- g. 20/40 OU = Myopic vision in BOTH eyes. 20/40 means that **when you stand 20 ft away from the chart, you can see what a “normal human” can see when standing 40 ft from the chart.** 20/200 is the cutoff for legal blindness in the US (i.e. you can see what a normal human can see when standing 200 ft from the chart; your vision is worse).

5. Match the following vision problems in children with the appropriate screening method:

Problem	Test(s)
Refractive Error	<u>Visual acuity testing (figures, Snellen)</u>
Strabismus	<u>Corneal light reflex, Cover-Uncover test</u>
Cataract	<u>Red reflex testing (cataract = opacification of the lens)</u>

6. Which of the following patients is at increased risk for hearing loss? **All 4 patients—see “Section V: Risk Factors for Hearing Loss”.** *Encourage residents to share patient scenarios of their own, in which they recommended close audiology follow-up.*

- a. 8mo former 37+4 week infant, readmitted for bilirubin of 28.5
- b. 3yo with Trisomy 21
- c. 18mo female with OME at 12mo, 15mo, and 18mo well-baby visits
- d. 14yo with Neurofibromatosis Type II

7. Please list the visual acuity criteria for optometry referral for the following age groups:

- a. 3 y.o. 20/50 or worse or 2 lines difference between the eyes
- b. 5 y.o. 20/40 or worse or 2 lines difference between the eyes
- c. > 5 y.o. 20/30 or worse or 2 lines difference between the eyes



Ask residents to reflect upon their own practices: Are you making appropriate referrals? How has SpotVision screening changed your referral practices? (Remember, optometry is a self-referral clinic; patients do not need a consult in AHLTA)

Sensory Screening Group Activities

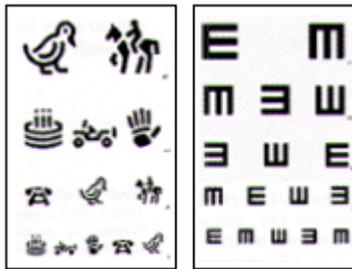
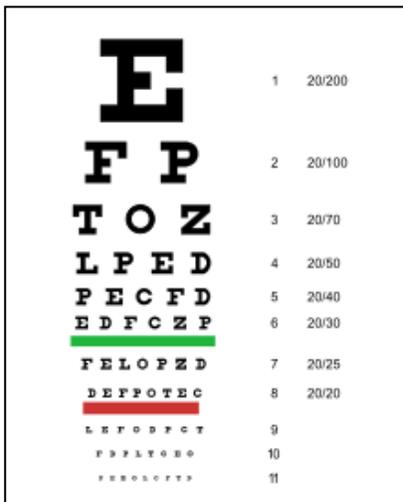
(Adapted from <http://www.aafp.org/afp/980901ap/broderic.html>)

* Remember to add the Snellen chart, audiometer, and tympanometry to your Procedure Log.

Vision Screening

1. Visual acuity (Snellen chart)

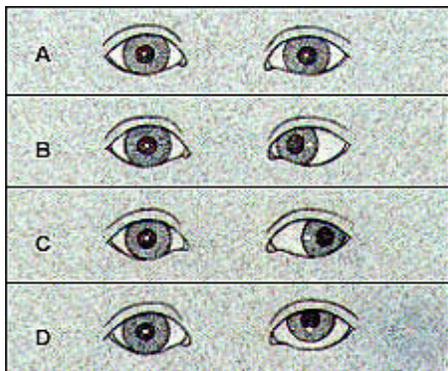
- * Ensure that Snellen chart is **10 or 20 ft away** from where the patient stands.
- * Have the patient cover one eye and read aloud every letter in the chart. If the patient misses only one letter, have the patient continue reading the next line.
- * Record the last line the patient reads accurately, and note what the vision is. (Visual acuity measures are marked on the Snellen Chart)
- * Ask the patient to repeat the process with the other eye, and then with both eyes uncovered.
- * Record the visual acuity for each eye and with both eyes uncovered. Remember— **OD** = oculus dexter (R eye); **OS** = oculus sinister (L eye); **OU** = oculus uterque (both eyes)



Other Pediatric Vision screen charts, used for preliterate young children or older children with MR. (L) Allen object recognition; (R) Tumbling E chart (Not used).

2. Corneal light reflex (Hirschberg Test)

- * Hold a penlight about 3 ft (1m) from both eyes. Note the position of the corneal reflection.
- * The reflection should fall in the same location in the cornea of each eye, even when the eyes move. Displacement of the corneal light reflection in one eye suggests strabismus.



© 1998 Marcia Hartsok

How would you interpret these findings?

- A: **Normal alignment**
- B: **L esotropia (the light reflex is outwardly displaced)**
- C: **L exotropia (the light reflex is inwardly displaced)**
- D: **L hypertropia (the light reflex is downwardly displaced)**

3. Cover-Uncover test

(For demo, see: <http://www.youtube.com/watch?v=TxEQWtlXtrI&feature=related>)

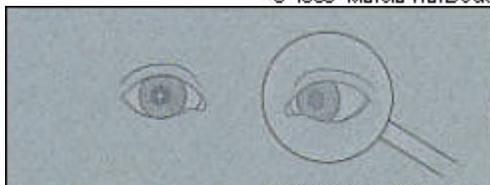
Example 1: Unilateral Cover-Uncover Test:

- * Direct the patient to focus on an interesting object about 10ft (3m) away.
- * For testing of the R eye, cover the L eye and observe the R eye for “fixation” movement.
 - If no movement, the patient does NOT have a R eye tropia.
 - If the R eye moves inward after the left is covered, the patient has a R eye EXOtrofia.
 - If the R eye moves outward after the left is covered, the patient has a R eye ESOtrofia.
- * For testing of the L eye, cover the R eye and observe the L eye for “fixation” movements.
- * Cover each eye for approximately 3-4 sec, and repeat 3x for each eye.

Example 1: Unilateral Cover-Uncover Test for ‘Tropias’



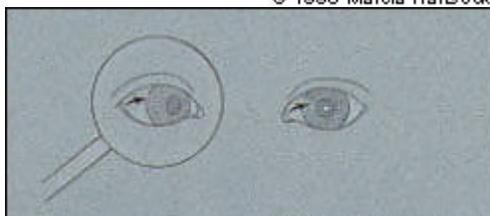
A. Observe the corneal light reflex at rest, the L eye shows **esotropia (corneal light reflex is outwardly displaced).**



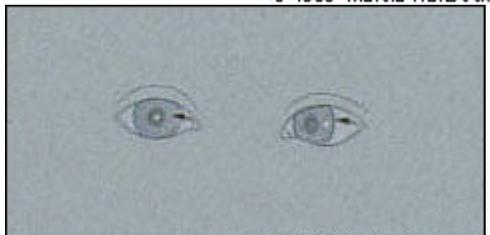
B. Cover the L eye. What happens? **No movement of either eye. The uncovered R eye maintains fixation.**



C. Uncover the L eye. What happens? **No movement of either eye (the L eye maintains its esotropic position).**



D. Cover the R eye. What happens to the L eye? **Moves outward. The previously esotropic eye takes up fixation.**

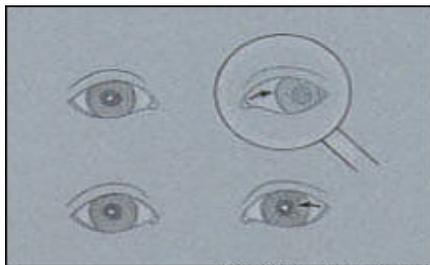


E. Uncover the R eye. What happens to the L eye? **Moves inward, back to its original esotropic position.**

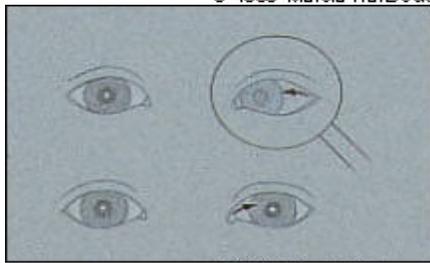
Example 2: Alternating Cover-Uncover Test:

- * Direct the patient to focus on an interesting object about 10ft (3m) away.
- * For testing of the R eye, cover the R eye for 1-2 sec, then move to cover the L eye for 1-2 sec. Observe the R eye *as it is being uncovered* to detect “re-fixation” movements.
 - If no movement, the patient does NOT have a R eye phoria.
 - If the R eye moves inward, *as it is being uncovered*, the patient has a R eye EXOphoria.
 - If the R eye moves outward, *as it is being uncovered*, the patient has a R eye ESOPhoria.
- * For testing of the L eye, cover the L eye, then move to cover the R eye. Observe the L eye *as it is being uncovered* to detect “re-fixation” movements.

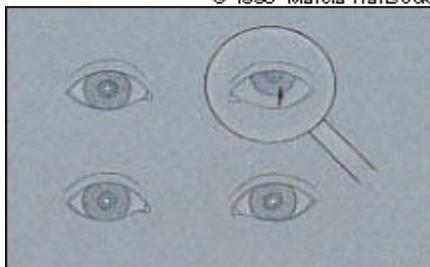
Example 2: Alternating Cover-Uncover Test for ‘Phorias’



A. Cover and uncover the L eye. What happens? The L eye moves inward as it is being uncovered, returning to a central position. This patient has a L exophoria.



B. Cover and uncover the L eye. What happens? The L eye moves outward as it is being uncovered, returning to a central position. This patient has a L esophoria.



C. Cover and uncover the L eye. What happens? The L eye moves downward as it is being uncovered, returning to a central position. This patient has a L hyperphoria.

And finally, what's this?



pseudostrabismus

Conversion Chart: Refractive State to “estimated” Visual Acuity^{[1][2]}

Myopia		Hyperopia			
<i>Nearsighted</i>		<i>Farsighted</i>			
Minus (-) Sphere		Plus (+) Sphere	Plus (+) Sphere	Plus (+) Sphere	
<i>Ages: All</i>	Estimated Visual Acuity	<i>Ages: 5y to 15y</i>	<i>Ages: 25y to 35y</i>	<i>Ages: 45y to 55y</i>	Estimated Visual Acuity
-0.5	20/30-40	+2.00	+1.25	+1.00	20/20
-0.75	20/50	+3.00	+1.75	+1.25	20/25
-1	20/60	+3.25	+2.50	+1.50	20/30
-1.25	20/70	+3.75	+3.00	+1.75	20/40
-1.5	20/100	+4.25	+3.50	+2.00	20/50
-2.5	20/200	+4.75	+4.00	+2.50	20/70

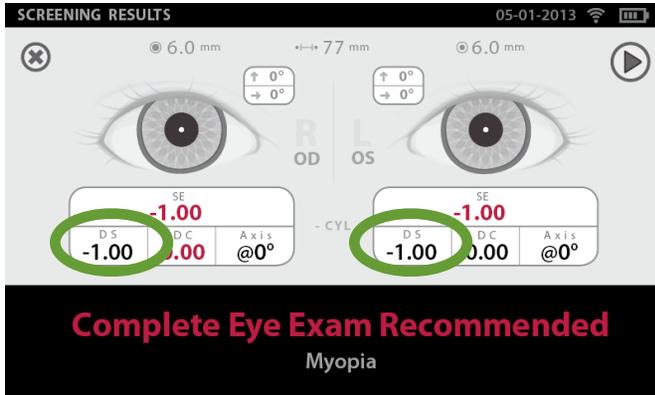
[1] Spherical results are based upon minus (-) cylinder convention.

[2] Source: “Composite Chart of Refractive State to V.A.” Derived from Peter’s multiple tables. *Peters, H.B. (1961): The Relationship between Refractive Error and Visual Acuity at Three Age Levels. A.A.A.O., 38:4.*

Not Recommended for conversion of screening results for children screened for amblyopic risk factors

Instructions for: Conversion Chart Refractive State to “estimated” Visual Acuity

1



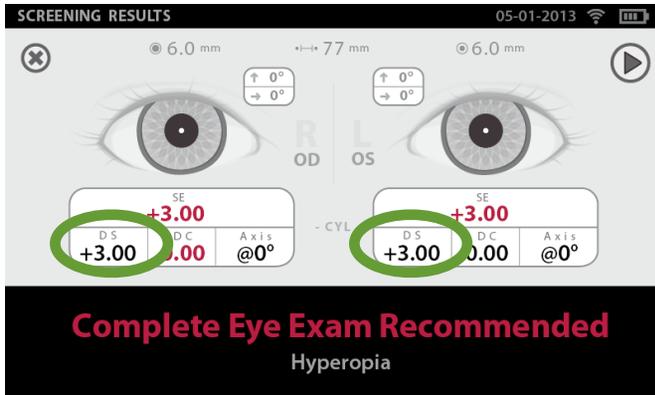
>Example 1

>DS Result in minus (-), use “myopia” columns

>Myopia conversions are for all ages

>Conversion for this example indicates an “estimated” visual acuity of 20/60 (OD) and 20/60 (OS)

2



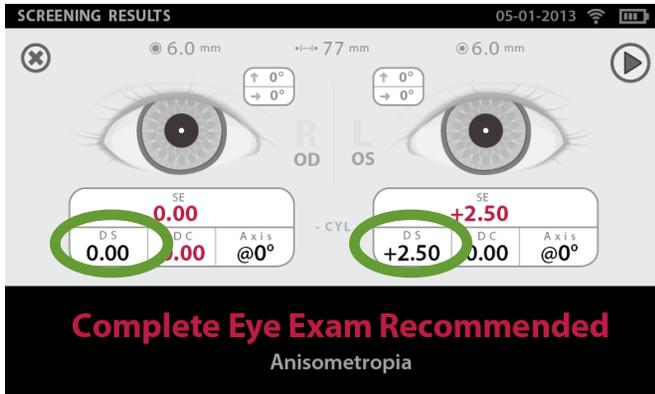
>Example 2

>DS Result in plus (+), use “hyperopia” columns

>Hyperopia conversions require age selection

>Conversion for this example indicates an “estimated” visual acuity of 20/40 (OD) and 20/40 (OS) for a 25 to 35 years of age subject

3



>Example 3

>Conversion for this example indicates an “estimated” visual acuity of 20/20(OD) and 20/20 (OS) for a 5 to 15 years of age subject

>This example indicates a 2.50 diopter variance in refractive power, identified as “anisometropia.”

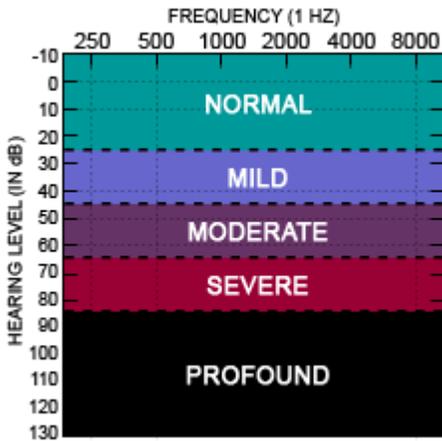
>“Complete Eye Exam Recommended” is properly identified for the subject even when the conversion indicates 20/20

Visual Acuity conversions not intended as an alternative to the recommendation presented on Spot.

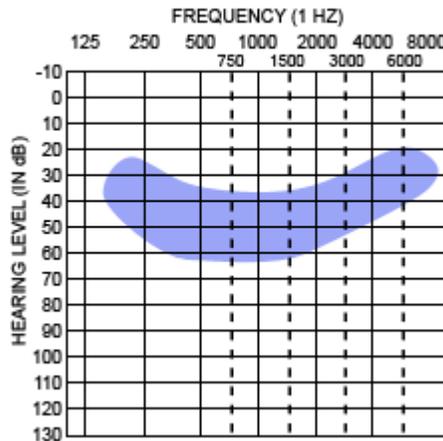
Hearing Screening

1. Conventional Audiometer

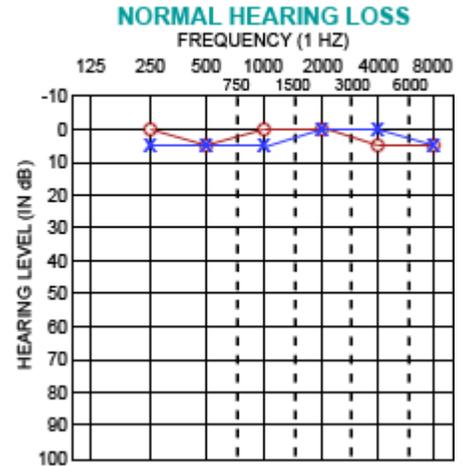
Conventional audiometry uses air conduction testing: different sounds go into the ear canal, through the middle ear, to reach the inner ear. **An audiogram is a graph that shows the softest sounds a person can hear at different pitches or frequencies.** An “O” is used to represent the R ear responses and an “X” for the L ear. The closer the marks are to the TOP of the graph, the softer the sounds that can be heard.



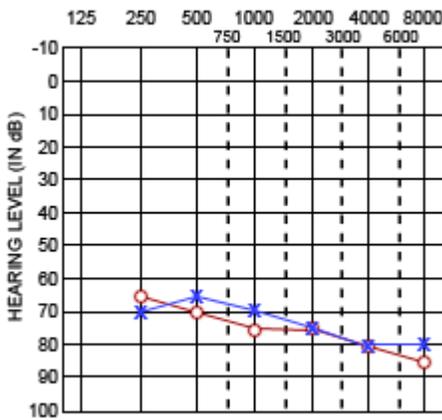
Different degrees of hearing loss



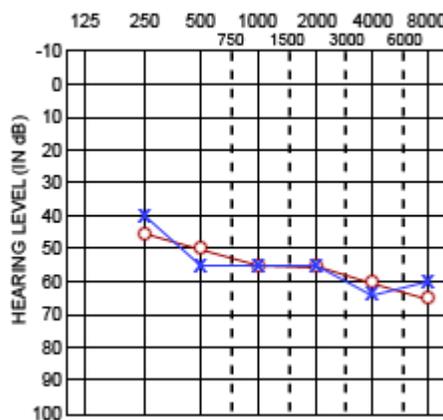
Range of pitch and loudness for most of the “speech sounds”



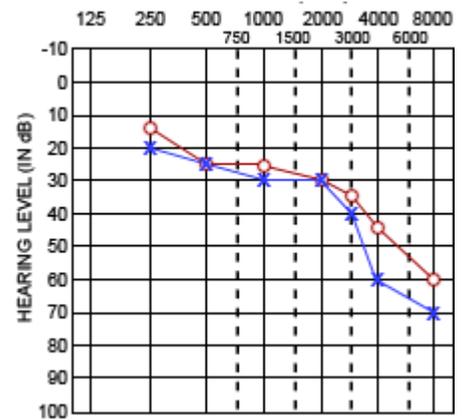
Label the following audiogram examples:



Severe hearing loss



Moderate hearing loss



Mild hearing loss

2. Tympanometry (adapted from: <http://www.aafp.org/afp/2004/1101/p1713.html>)

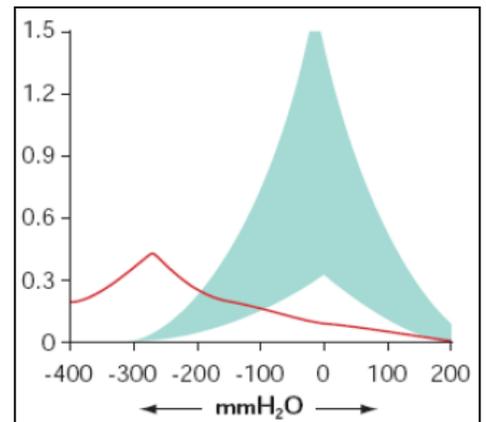
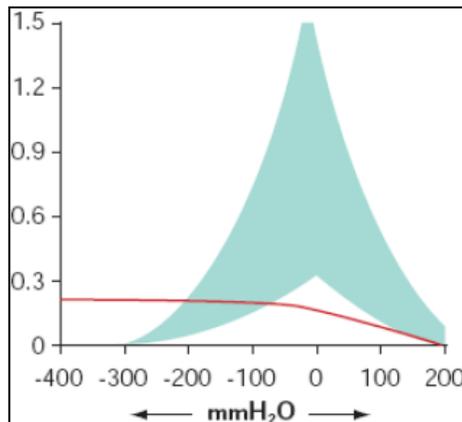
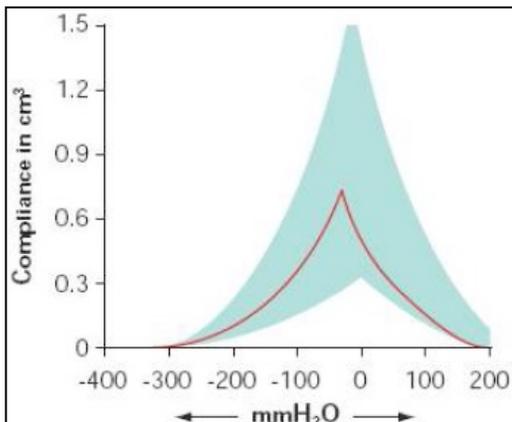


Tympanometry is an examination used to test the condition of the middle ear and mobility of the tympanic membrane and the conduction bones by creating variations of air pressure in the ear canal. A probe is inserted into the canal, permitting a hermetic seal to form. On the probe tip are 3 small holes. Through the 1st hole, we introduce an 85 dB pure-tone sound; through the 2nd hole we measure the sound pressure in the cavity; through the 3rd hole we create and remove pressure in the cavity to get a dynamic measure of the movement of the TM. We can exert positive pressure, pushing the TM away from us, or negative pressure, creating a partial vacuum and pulling the TM toward us. Most testing instruments use +200 mmH₂O to -200mmH₂O.

In basic tympanometry, we insert +200 mmH₂O pressure against the TM, effectively pushing it away from us (into the middle ear space). When we do that, we make it “stiffer”. As we make it stiffer, it reflects more sound back into the cavity, and this allows less energy (“less sound”) through the TM. Then, we begin to remove the pressure in the cavity, a bit at a time. As we do, the TM becomes more compliant, lets more sound through, and the perception is that the sound gets louder. We make a measurement at +200, +100, +50, 0, -50, -100, and -200 mmH₂O. We plot the amount of sound pressure at each of these points to create a tympanogram. The X-axis shows the air pressure, and the Y-axis shows the “static compliance” or mobility of the TM.

There are 5 basic types of tympanograms. **Type A** is normal: there is a normal pressure in the middle ear with normal mobility of the eardrum and the conduction bones. **Type B and C** may reveal fluid in the middle ear, perforation of the TM, scarring of the TM, lack of contact between the conduction bones of the middle ear, or a tumor in the middle ear. **Types AS and AD** represent variations with decreased (s = shallow, stiff) or increased (d = deep, disarticulated) compliance.

Please identify the following tympanometry tracings:



Type A
Due to normal middle ear pressure and static compliance

Type B
Due to no measurable middle ear pressure or static compliance, consistent with middle ear pathology (e.g. MEE, perforation, cholesteatoma)

Type C
Due to significant negative middle ear pressure with normal static compliance, consistent with ET dysfunction/middle ear pathology (e.g. retraction of TM, blockage of ET).