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Pediatric Vision Screening

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Educational Gap

Although early detection of visual disorders can lead to therapy that will prevent permanent blindness, compliance with screening guidelines of the American Academy of Pediatrics is low.

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

- 1. Be aware of common vision-threatening conditions that can be detected by using basic screening examinations.
- 2. Become familiar with techniques and findings used in vision screening examinations.
- 3. Understand the role of commercial screening tools.

Introduction

Early detection of ocular conditions can allow for assessment and treatment of a visionthreatening or life-threatening condition. Amblyopia, or "lazy eye," can develop if a clear visual image is not projected onto the retina. Amblyopia can be caused by deprivation, strabismus, high refractive error (hyperopia, myopia, or astigmatism), or anisometropia (significant difference in the refractive error between eyes) and can be unilateral or bilateral. The prevalence of amblyopia is estimated to be 1% to 4%. (1)

Many factors may prevent the achievement of universal vision screening, including lack of education of families; language, financial, and state legislative barriers; and a lack of available providers. (2) Primary care physicians are crucial providers for detecting and referring vision-threatening ocular conditions. From the newborn examination, through subsequent health supervision visits, and throughout a child's life, the pediatrician and family physician can perform effective examinations to screen for common and uncommon conditions that may be vision-threatening or even potentially life-threatening. (3) However, vision screening in the primary care office should not take the place of a full eye examination. If a patient cannot be screened effectively after two attempts, a referral should be made to an eye care professional who is comfortable examining children.

Pediatricians and family physicians should have the ability to perform a thorough ocular examination. According to the US Preventive Services Task Force, children under age 5 years should be screened to detect amblyopia, strabismus, and defects in visual acuity. The American Academy of Pediatrics (AAP) guidelines include screening at all health supervision visits, from the newborn period to age 3 years, by using the following components: ocular history, vision assessment, external examination, ocular motility, pupil examination, and red reflex examination. For children ages 3 to 5 years, the AAP recommends age-appropriate visual acuity measurements and direct ophthalmoscopy. (4) Sensitivity of screening examinations increases with age, while specificity remains unchanged. (5)

However, screening is not universal, and compliance with AAP guideline visual acuity screening is low. (6) In addition, there is controversy over who should provide the screening examinations. In 2000, Kentucky passed legislation mandating diagnostic eye examinations by optometrists or ophthalmologists. (7) A survey of primary care physicians found that the percentage of pediatricians who expected to perform screening examinations dropped from 86% to 71% after the mandate, and the percentage of family physicians who expected to screen dropped from 79% to 50%. The concern with this switch to mandated eye examinations is that children may get overlooked by one of the screening programs. In

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addition, we must consider the increased cost of diagnostic eye examinations compared with vision screening programs, given the current health-care crisis.

As discussed, screening from an early age can identify patients who have poor vision. Whether potentially treatable or not, low vision or blindness from amblyopia, nystagmus, or structural abnormalities certainly can affect a patient's reading ability and educational progress. Getting parents and schools involved early and working with low-vision specialists and visual aids can help patients adjust their needs appropriately. Although low-vision services can provide essential educational tools, primary prevention and treatment of potentially vision-threatening conditions is the better goal.

Common Vision Conditions

The age of the patient is an essential consideration in determining a differential diagnosis. From birth to age 1 year, concerning conditions include corneal opacities, cataracts, glaucoma, persistent fetal vasculature, and retinoblastoma. All of these conditions have the potential to cause deprivational amblyopia if not detected and treated at an early age (sometimes within the first weeks after birth). Early detection of retinoblastoma could mean saving a patient's eye or eyes and possibly some vision, along with treating a potentially fatal condition. Other conditions that may be present from birth, such as congenital ptosis or capillary hemangiomas causing mechanical ptosis or unilateral astigmatism, are also risk factors for amblyopia.

From ages 1 to 3 years, more common eye conditions with amblyogenic risk factors include strabismus and refractive errors such as high hyperopia (farsightedness), high myopia (nearsightedness), astigmatism, and anisometropia (significant difference between the refractive errors between the eyes). These disorders can be subtle. Preverbal patients are more difficult to examine, but early detection will have a substantial impact on a patient's future education and life if treatment is initiated promptly.

From ages 3 to 8 years, strabismus and refractive errors continue to be significant amblyopic risk factors. As patients age and grow more cooperative, testing for visual acuity becomes more feasible.

Strabismus

Early Detection

Early detection of strabismus or ocular misalignment is essential for the prevention or treatment of amblyopia and allows the possibility of saving binocular vision. If the misalignment is constant, a child's developing brain will ignore the visual input from the misaligned eye to



Figure 1. Accommodative esotropia: A. The patient is esotropic without glasses. B. The eyes are straight with hyperopic correction.

avoid diplopia. If this situation persists, the eye will become "lazy" or amblyopic. If detected and referred early enough, treatment of the amblyopia by using penalization techniques such as patching may improve or resolve the difference in vision between the two eyes. As children get older, treatment is not as effective.

Between the ages of 8 and 10 years, the visual system has developed fully, and therefore decreased vision in one or both eyes cannot be improved, further emphasizing the importance of early intervention. Treatment of the misalignment itself depends on the type of strabismus and can include either using glasses (Fig 1) or performing eye muscle surgery. Surgery may restore binocularity, but it will not treat amblyopia; therefore, amblyopia is treated first with patching or other penalization techniques.



Figure 2. Pseudoesotropia. This patient appears to be turning his right eye in. This is an optical illusion created by wide nasal bridge and epicanthal folds that cover the nasal "white part of the right eye" when the patient turns his head slightly to his right. The symmetric corneal light reflex indicates good alignment.



Figure 3. Asymmetric red reflex. This 7-year-old boy was found to be severely hyperopic in the left eye with no need for glasses in the right eye. This is an excellent example of a patient who might have passed vision screens but should have been referred much earlier for an abnormal red reflex.

Pseudostrabismus Versus True Strabismus

Pseudostrabismus occurs when one eye appears to turn in but is straight on cover/uncover testing. This appearance occurs most commonly in patients with wide nasal bridges and epicanthal folds, giving the child the appearance of esotropia. A true esotropia can be ruled out by using an equal corneal light reflex and normal cover testing (Fig 2).

Examination Techniques

Red Reflex

The red reflex test is the single most important screening tool for infants and young children. Using the direct ophthalmoscope to view both eyes simultaneously is the best way to evaluate the red reflex. The patient's eyes should be viewed through the direct ophthalmoscope from approximately 2 feet away, with a broad beam directed so that both eyes are illuminated at the same time. The patient should be focused on the ophthalmoscope light. Starting with low illumination and increasing the brightness allows the patient to become comfortable with the bright light.

The red reflex will fill the pupil, and the corneal light reflex will also be centered on the pupil (Brückner reflex) if the patient's alignment is correct. The red reflex represents the reflection of the light from the retina. Therefore, abnormalities of the red reflex can be caused by a physical blockage of the normal clarity of the visual axis, such as tear film mucus, corneal opacity, cataract, vitreous hemorrhage or retinal detachment, retinoblastoma, or persistent fetal vasculature. The red reflex also can appear dull in both eyes from a high refractive error (high myopia or hyperopia) or unequal due to anisometropia (high refractive error in only one eye) (Fig 3) or strabismus.

Differences in pigmentation among racial or ethnic groups also may be responsible for variation in the red



Figure 4. Leukocoria.

reflex. Darker pigmented patients will appear to have a darker red reflex. The AAP policy states that all neonates should have a red reflex examination before discharge from the newborn nursery. Urgent referral and direct communication with the accepting ophthalmologist are essential when abnormalities are detected. High-risk patients who have a family history of retinoblastoma, infantile cataracts, glaucoma, or any other ocular disorders that presented early in life should be referred but also should have a red reflex examination before leaving the newborn nursery. Any parental concern raised by suspicion of a white pupil reflex should be referred urgently. (8) If there is ever any concern regarding a child's red reflex



Figure 5. Partial leukocoria. The patient had a congenital cataract viewed through a surgical microscope just before surgery.

status, the most prudent action is to refer the patient for a complete ocular examination.

Leukocoria

Leukocoria, or a white pupil, occurs when the red reflex appears white rather than the typical red (Fig 4). The most concerning diagnosis on the differential is retinoblastoma. Toxocariasis, Coats disease, persistent fetal vasculature, or a chronic retinal detachment will also appear white and therefore create a white reflex. Cataracts also can cause leukocoria or just an asymmetric red reflex (Fig 5). Because retinoblastoma is potentially fatal, all cases of leukocoria require an urgent referral to determine the cause of the condition.

External Examination (Inspection)

The external eye examination can be performed with a penlight to look for any external structural abnormality of the eyelids and adnexa. Paying attention to the eyelids and the vertical and horizontal fissures can reveal ptosis, capillary hemangiomas, and eyelid colobomas, which are important findings in detecting possible risk factors for obstructive amblyopia and systemic diseases. The sclera also can be evaluated easily by using a penlight. Corneal clarity can be evaluated simply by determining if there is a clear view of the iris and pupil. If there is not a good view of the iris or pupil, there may be a corneal opacity or haze. In newborns and infants up to age 2 months, asymmetry of the eyes or face should be noted. From 3 months on, any face turn or head tilt should be noted, especially if mentioned by the parents.

Visual Acuity

Visual acuity improves with age as does the ability to recognize letters or shapes. At age 0 to 2 months, patients should manifest a blink response to bright light, equal pupillary response, sporadic fixation, and following that becomes more consistent with age. The neonate can have intermittent strabismus with either an eso- or exodeviation of the eyes (eyes turned in or out), which should resolve by 2 to 4 months, after which the deviation is considered pathologic. If the infant has a constant strabismus, he or she should be referred for evaluation.

At age 2 to 6 months, infants should be able to fix and follow an object, such as a light or mother's face, and the eye alignment should be straight. From age 6 months to 2 years, children should have central fixation, reach for toys, and demonstrate good alignment.

From age 3 to 5 years, subjective vision can be obtained. Children should test to 20/40, or better, on ageappropriate charts with one eye occluded (Allen or LEA pictures, tumbling Es, or HOTV letters). There should be no more than two lines of difference between the eyes. Patients become more cooperative after age 5 years. These children should test to at least 20/30 vision with a regular Snellen chart, with no more than two lines of difference between the eyes.

Cover Test and Hirschberg Test

The cover test (Fig 6) and Hirschberg test are used to examine ocular misalignment. The cover test reveals a manifest deviation (tropia). If a patient is fixating on a target with the right eye and the left eye appears to be turned in or turned out when the right eye is covered, there will be a shifting movement of the left eye in the opposite direction from its deviated position as the left eye picks up fixation. This finding is diagnostic of a manifest deviation because the misalignment is constant. When the cover is removed from the right eye, the right eye will either continue to be deviated or it will refixate if there is a right eye preference, indicating a possible amblyopia.



Figure 6. Alternating cover test.

A latent deviation can be induced by using alternate cover testing (Fig 7). This test is performed by moving the occluder directly from one eye to the other without allowing binocular viewing. This maneuver can bring out a phoria (latent deviation) or intermittent tropia (manifest but controlled at times). A manifest deviation is more worrisome due to the higher risk of amblyopia developing. The general pediatrician who is unsure about the proper performance of these tests should ask a local ophthalmologist to demonstrate the appropriate technique.

When there is a constant deviation, the Hirschberg test can be used to estimate the amount of deviation. Using a penlight directed on both eyes, the light reflex is examined to determine if there is an asymmetry. The light reflex should be approximately in the center of the pupil in both eyes when the child is fixating on the light, or in the same spot in both eyes if the patient is fixing on a different target. If the light reflex is displaced nasally, this finding indicates an exotropia (the eye is turned out) (Fig 8). When the light reflex is displaced temporally, this finding indicates an esotropia (the eye is turned in) (Fig 9). This test can be helpful in determining a true deviation versus pseudostrabismus (discussed earlier).

Ocular Motility and Nystagmus

The patient's ocular motility should be evaluated as soon as the child is old enough to fixate and follow an interesting target. Eye movements become smoother as infants get older. Parents may report "funny eye movements," which could indicate a more complex strabismus such as congenital fourth nerve palsy, Brown syndrome, or Duane syndrome.



Figure 7. Alternating exotropia induced by cover testing.



Figure 8. Exotropia. Corneal light reflex in the left eye is nasal compared with the light reflex on the right eye, which is central.

Observing for nystagmus is also important. There are many types and causes of nystagmus, but all children who manifest symptoms should be referred for evaluation of low vision as a cause. If an obstruction to clear visual development, such as a congenital cataract, has not been corrected at an early age, the presence of nystagmus could be a sign of poor vision. Other types of abnormal eye movements, such as opsoclonus, warrant an urgent referral. Opsoclonus can be a presenting sign of neuroblastoma. This condition appears as rapid, involuntary eye movements in all vectors of gaze.

Pupil Examination

Patients should have equal and reactive pupils from birth. It is more difficult to elicit this response in newborns but having the room dark and using a bright penlight often are helpful in distinguishing a pupillary response. The older a patient gets, the more important it is to have him or her focus at a distant target, dim the lights, and check the pupils while standing to the side so the patient does not focus on the examiner and induce accommodation. Any evidence of congenital anisocoria, or pupils of different sizes, also should be referred to evaluate for possible Horner



Figure 9. Esotropia. Corneal light reflex is not as apparent in this photograph, but the room light reflected off the cornea is temporal in the right eye compared with the central position in the left eye.

Age	Evaluation	Indications for Referral
Newborn (0–1 mo)	Examine outer structures of the eye and red reflex before the neonate leaves the newborn nursery	Abnormal red reflex requires urgent consultation History of retinoblastoma in parent or sibling
1 mo-3 y	History Vision assessment; fix and follow External examination Ocular motility Pupil examination Red reflex evaluation	Poor tracking by 3 months Abnormal red reflex Chronic tearing or discharge
3–5 у	History Vision assessment: LEA and Allen figures, HOTV letters, tumbling Es, Snellen chart External examination Ocular motility Pupil examination Red reflex evaluation(photoscreening) Ophthalmoscopy	Strabismus Chronic tearing or discharge Fail vision screen (cannot read 20/40 with one or both eyes or two-line difference between eyes) or photoscreening Uncooperative after two attempts
≥5 y	History Visual acuity External examination Ocular motility Pupil examination Red reflex evaluation(photoscreening) Ophthalmoscopy	Strabismus Cannot read at least 20/30 with one or both eyes or two-line difference between eyes Fail photoscreening Not reading at grade level
At-risk children of any age	History Visual acuity External examination Ocular motility Pupil examination Red reflex Ophthalmoscopy	Retinopathy of prematurity Family history of retinoblastoma, congenital glaucoma or congenital cataracts Systemic diseases with associated retinal dystrophies/degenerations Nystagmus Neurodevelopmental delays

Table. Vision Screening Recommendations

Adapted from American Association for Pediatric Ophthalmology and Strabismus. Vision Screening Recommendations. Accessed online at http://www.aapos.org/terms/conditions/131.

syndrome. The most concerning condition associated with Horner syndrome in children is neuroblastoma.

In addition to the screening examination, taking a complete medical and family history is vital. At birth, it is important to ask about a family history of any congenital eye conditions or blindness from birth. This inquiry is imperative in evaluating for possible heritable eye diseases such as retinoblastoma, congenital cataracts, congenital glaucoma, and aniridia.

Commercial Screening Tools

There has been growing interest in using commercial screening tools in schools and primary care offices. Standard techniques of visual acuity tests measure visual function directly. Patient cooperation, understanding, age, and attention, as well as the skill and patience of the examiner, play a role in the success of testing visual acuity. There are a variety of photoscreeners and autorefractors that objectively detect amblyopia or amblyogenic risk factors and require little patient cooperation.

The risk factors that should be identified by screening instrumentation include significant anisometropia (>1.50 diopter difference in prescription between the two eyes), manifest strabismus, hyperopia greater than 3.50 diopters, myopia greater than 3.00 diopters, any visually significant media opacity (>1 mm and in the visual axis), astigmatism >1.50 diopters in the regular meridians or >1.00 in oblique axis, and ptosis. (9)

Not all patients who have these risk factors will develop amblyopia. For instance, many infants have a high degree of clinically significant astigmatism that is either eliminated or greatly reduced by age 4 years. (10) Therefore, systems with higher sensitivity but lower specificity will over-refer due to false-positive results. (9)

Many studies have compared the different screening tools. The two most common methods are photoscreening, which involves taking pictures of the red reflex of both eyes simultaneously, and autorefractors, which can estimate the child's refractive error. Photoscreening was shown to be more time-efficient and had a higher positive predictive value than traditional screening techniques in 3- to 4-year-olds. (11) A study of vision in preschool-aged children found autorefractors detected 15% more amblyogenic risk factors than photoscreeners. In addition, depending on the criteria used in the various screening techniques, referral rates can be different. There have been no studies or consensus as to which method should be standard of care. Ease of referral and compliance of parents to keep the referral appointment are obstacles regardless of which method is used. (12)

There is no mechanical substitute at present for an adequate physical examination conducted by an educated primary care physician. Screening programs should be designed to easily identify individuals who have amblyopia or those at risk for developing amblyopia, with the additional concern of keeping the screening inexpensive.

Recommendations

The table highlights the recommendations made by the American Association for Pediatric Ophthalmology and Strabismus for which examinations should be conducted at different ages and also lists guidelines for referral to an ophthalmologist.

Summary

- Pediatricians and family physicians are essential in assessing the health of the eye and in vision screening.
- Newborns should be evaluated before leaving the nursery and referred urgently for abnormal findings on external examination or abnormalities of the red reflex.
- Vision assessment should begin at age 3 years by physicians, nurses, or technicians trained in vision evaluation. Children should be referred to licensed eye care professionals for abnormal findings or poor cooperation after two attempts.
- Photoscreeners can be a valuable addition to routine vision screening, especially in preverbal children.

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- 1. The parents of a 4-month-old girl are concerned that she has strabismus. Using a penlight, you find that the light reflex is centered in the right pupil and falls temporally in the left pupil. Your diagnosis is:
 - A. Left esotropia.
 - B. Left exotropia.
 - C. Pseudostrabismus.
 - D. Right esotropia.
 - E. Right exotropia.
- 2. The mother of a 6-month-old boy brings in several recent photographs that show the infant's right pupil to be whitish. Among the following, the most important question to ask is:
 - A. Did anyone in the family have eye surgery or eye disease as a child?
 - B. Does your infant sometimes look cross-eyed?
 - C. Has a grandparent had a cataract removed?
 - D. Have the infant's eyes looked bloodshot?
 - E. Was the child hit in the eye with a sharp object?
- 3. A 12-month-old boy has developed rapid, horizontal and vertical conjugate eye movements. Among the following, which test is most likely to yield the diagnosis?
 - A. Blood glucose level.
 - B. Sedimentation rate.
 - C. Serum electrolytes.
 - D. Stool culture.
 - E. Urinary catecholamines.
- 4. A 2¹/₂-year-old boy is taken into foster care. He is noted to have strabismus. In your letter supporting intervention, you note that treatment should begin as early as possible but must be undertaken before the visual system has developed fully, ideally before age:
 - A. 3 years.
 - B. 5 years.
 - C. 7 years.
 - D. 8 years.
 - E. 11 years.
- 5. As part of her school entry evaluation, a 4-year-old girl is having her vision tested in your office. If her vision is normal, you expect her to test at least to:
 - A. 20/100.
 - B. 20/60.
 - C. 20/40.
 - D. 20/20.
 - E. 20/15.

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