Understanding and Preventing Amblyopia

Speaker Notes

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Understanding and Preventing Amblyopia

Approximately 2% to 4% of the general population has amblyopia, defined as a decrease in best-corrected visual acuity in one or both eyes, with no apparent ocular abnormality on physical examination. Amblyopia is the most common cause of monocular visual impairment in children and young adults. The greatest visual risk lies in the chance of subsequent loss of vision in the better-seeing eye from an unrelated ocular disorder or trauma.

With early detection, amblyopia can be successfully treated and normal visual development can be expected. Although the ophthalmologist is responsible for its treatment, detection of amblyopia is the charge of the primary care physician, who is often the first health professional to encounter a child with a potential eye condition.

The skills and tools for the detection of amblyopia are simple. Understanding and Preventing Amblyopia aims to enable primary caregivers to help reduce the incidence of amblyopia by providing them with an understanding of its predisposing factors, common presenting features, methods of early detection, and guidelines for referral to an ophthalmologist. Treatment rationale and methodology are also discussed; compliance with treatment is crucial to successful outcome, and primary caregivers are in an excellent position to help ensure their young patients (and the affected child’s parents) can maintain the treatment regimens they are given.

Approximate Running Time

45–60 minutes

Suggested Audience

- Pediatricians
- Family physicians
- Internists
- Medical students, interns, residents
- Public health nurses and school health screeners
- State and local meetings of national medical societies such as AAFP, AAP
- Any primary care provider, including nurse practitioners, etc.
Amblyopia is not a new problem. It was described by Hippocrates and has been recognized for over 2,000 years. For normal visual system development, an infant’s visual cortex must receive input from each eye that is of equal clarity and of the same object of regard. If something prevents this from taking place, reduced visual acuity in one or, less commonly, both eyes may result. This resultant poor visual acuity is called amblyopia.

Amblyopia can be defined as a decrease in best-corrected visual acuity in one or both eyes, with no apparent ocular abnormality on physical examination. Traditionally, amblyopia was thought to have no underlying organic cause. Actually, a predisposing factor is always present, but it may not be apparent clinically without the benefit of a complete ophthalmologic examination. Strictly speaking, amblyopia occurs in an eye that is otherwise physically normal. In practice, the term is used to include reduced visual function that results also from structural ocular abnormalities, such as cataracts, corneal opacities, or eyelid ptosis.
Amblyopia is the most common cause of monocular visual impairment in children and young adults. Approximately 2% to 4% of the general population has amblyopia. Commonly, only one eye is affected. The greatest visual risk lies in the chance of subsequent loss of vision in the better-seeing eye from an unrelated ocular disorder or trauma. In such cases, the individual must rely on the reduced visual acuity of the amblyopic eye. An amblyopic eye is a poor source of vision to be held in reserve. Bilateral amblyopia, although rare, may occur, for example, when both eyes have such high refractive errors that focused, clear images cannot be maintained and delivered to the developing visual cortex. Educational and vocational goals may be profoundly limited for individuals with bilateral amblyopia.

Four main factors make screening for amblyopia significant to the busy primary care physician.

1. Amblyopia can be prevented or treated in most circumstances.
2. Early detection is the key to effective treatment.
3. Life-threatening disorders may present as amblyopia.

The responsibility of screening for amblyopia rests with the primary care physician.
Amblyopia can be prevented or treated in most circumstances if detected early enough. Specific predisposing factors for amblyopia can be identified, including ocular media opacities such as cataract; significant refractive errors; and strabismus, or misaligned eyes. Attention to the underlying cause will prevent the development of permanent visual loss.

Early detection is the key to effective treatment. Visual acuity develops rapidly from approximately 20/400 at birth to 20/20 within the first year of life and remains quite plastic within the first decade. Amblyopia results when this maturational process is interrupted. Because the degree of visual system plasticity lessens as the system matures, the earlier amblyopia is detected, the more successful treatment will be. Preferably, detection and treatment should occur by the age of 3 to 5 years. The older the age at detection and the more decreased the visual acuity, the less likely visual function is to improve. Nevertheless, recent evidence indicates that amblyopia may be treatable even into a child’s second decade.

Life-threatening disorders may present as amblyopia. For example, the presenting signs and symptoms of retinoblastoma, such as reduced visual acuity or strabismus, may not be clearly distinguishable from amblyopia during a quick screening examination. Any child presenting with such symptoms warrants further examination, because a delay in diagnosis may threaten life or health.
The responsibility of screening for amblyopia rests with the primary care physician, who is often the first health professional to encounter a child with a potential eye condition. Ultimately, the ophthalmologist is responsible for its treatment, but the detection of amblyopia is the charge of the primary care physician. The skills and tools for its detection are simple, and early diagnosis may profoundly influence patients’ lives.

**PREVENTING AMBLYOPIA**

A better understanding on the part of children’s health providers of amblyopia’s origin, detection, and treatment may reduce the incidence of amblyopia. Prevention relies on a familiarity with (1) the factors predisposing to amblyopia; (2) common presenting features; (3) methods of early detection; and (4) treatment rationale.

**Predisposing Factors**

Amblyopia develops if the visual image projected on the central retina is constantly unclear or obstructed during the critical period of early visual development. Comparing the eye to a camera, a poor image on the retina can occur as a result of three major factors:

- Poor clarity of the visual system or blockage of the light pathway (e.g., ocular media opacities, ptosis)
- Poor focus (e.g., refractive error)
- Poor aim (e.g., strabismus)
Media opacities such as cataract or corneal scar block or irregularly diffuse the light entering the eye. This disruption of light rays results in a poorly focused image on the retina, predisposing the patient to amblyopia. The developing visual cortex “prefers” the focused image from the better-seeing eye, suppressing or ignoring the less-distinct image from the eye with the media opacity. The greater the interference, the longer it is present; and the younger the child, the more rapidly and more deep the amblyopia may become. For instance, cataract removal must be performed at a very early age to allow normal visual development within the first few months of life. An infant born with complete cataracts is unlikely ever to achieve visual acuity better than 20/40 unless removal of the cataracts and optical correction occurs within the first 2 months of life.

Refractive error may lead to amblyopia in one or both eyes. When the refractive errors in the two eyes are not equal (termed “anisometropia”), only one eye can receive a focused image, because the visual system focuses both eyes equally and simultaneously. Amblyopia then develops in the eye that is not in proper focus with the fellow eye.
If both eyes have a high amount of farsightedness, nearsightedness, or astigmatism, the individual may be unable to clearly focus images at any distance. Because the image on each retina is blurred at all times, neuronal connectivity in the visual cortex develops without having received a finely focused image from either eye, and bilateral amblyopia develops. Bilateral amblyopia occurs more commonly with hyperopia (farsightedness) than with myopia (nearsightedness).

Strabismus is the most common cause of amblyopia. Deviation of one eye, such as occurs with constant esotropia, results in suppression of the deviating eye. The foveas of the two eyes are pointed toward different objects. The brain “selects” the object viewed by the straight eye and suppresses or ignores the object viewed by the deviating eye. Amblyopia results as a consequence of this conflicting information. Foveal stimulation is important because the fovea is the region of the retina responsible for critical central vision. When visual input from the deviating eye is ignored, amblyopia develops.
Common Presenting Features

If only one eye is amblyopic, often the child and the parents will be unaware that vision is reduced. The child appears to be seeing normally because vision is unimpaired in the non-amblyopic eye. Unilateral amblyopia commonly becomes noticed through one of four ways:
- Failed vision test
- Strabismus
- Consequence of parental concern
- Concern because of a positive family history of amblyopia or strabismus

Vision screening examinations are often initially performed on children between 3 and 5 years of age at the primary care physician’s office, schools, or day care centers. Such tests are very useful, but they have their limitations because the child may peek or the occluder or patch used during testing may not completely cover the eye. Such poor technique may allow amblyopia to remain undetected. An adhesive patch during testing works best.

Strabismus is the most common underlying cause of amblyopia. With constant deviation of one eye, reduced vision will occur. Amblyopia is less likely when the deviation is intermittent or when the two eyes alternate fixation.
When adults develop strabismus, double vision (diplopia) is an associated presenting feature. In infants and children, subjective diplopia is rare. This distinction stems from differences in adaptation of the immature visual system of children compared to adults. A child’s brain suppresses the image from the deviating eye; the child effectively sees one image. With constant suppression, amblyopia develops. In adult-onset strabismus, the mature visual system is unable to compensate in this way, leading to perception of a double image with resultant diplopia. Onset of true diplopia in a child may indicate a sudden misalignment of a previously normal visual system and should prompt immediate evaluation and referral. Conditions such as a cranial nerve palsy may be the cause. Other causes of reported diplopia in children include recognition of physiologic diplopia or malingering.

Parental concern may initiate the evaluation for amblyopia. Parents may note that their child’s eyes “look different.” Careful examination may reveal such serious concerns as a white pupillary light reflex (leukocoria, left) or an abnormally large cornea, indicating possible congenital glaucoma (right). There are many causes of leukocoria, including retinoblastoma, retinal detachment, and cataract.
Visual problems, including amblyopia or strabismus, in other family members may increase parents’ concern for their child’s vision. Although strabismus and amblyopia do not demonstrate a specific mode of inheritance, a positive family history is often present in children with amblyopia.

If both eyes are amblyopic, an infant may present with such features of poor vision as a failure to fix and follow an object, wandering eye movements, rhythmic shaking of the eyes (nystagmus), or unusual habits such as eye-poking or hand-waving. In rare cases, poor vision may present as clumsiness or ataxia in the toddler. In comparison to unilateral amblyopia, bilateral involvement is uncommon; however, any concern warrants immediate referral to an ophthalmologist because of the potentially severe visual loss from bilateral amblyopia.

**Early Detection**

The role of the primary care physician is fundamental to the early detection of amblyopia through careful assessment of (1) the red reflex, (2) visual acuity, and (3) ocular alignment.
**Red Reflex Assessment**

**SLIDE 23**
Assessment of the red reflex allows the examiner to evaluate potential causes of amblyopia: media opacities, high refractive errors, and strabismus. The cornea, anterior chamber, lens, and vitreous must all be clear to allow a view of the retina. If the eye is unusually hyperopic or myopic, the red reflex may be very dim unless the ophthalmoscope’s high-power lenses are used.

**SLIDE 24**
An asymmetric red reflex may indicate a significant visual problem, such as strabismus, unequal refractive error, or media obstruction.

**SLIDE 25**
**Step 1.** Use an ophthalmoscope with a bright light source, such as a halogen bulb. **Step 2.** Dial in a +2 to +4 lens, identified by a black numeral, and select the largest round, white light aperture.
Step 3. While looking through the instrument, illuminate both eyes simultaneously and examine the eyes for clarity of the red reflex at a distance of 1 to 2 feet. Compare the patient’s eyes for dissimilarities in the quality of the reflex. Such a difference may indicate unequal refractive errors, a media opacity, or strabismus. It is important to remember that often it is dissimilarity between the two eyes that leads to unilateral amblyopia.

Step 4. Now direct your attention to each eye separately, using your right eye to examine the patient’s right eye and your left eye to examine his left eye. Change the light source to the small, round aperture. As you move closer to the patient, gradually reduce the power of the lens until you are able to see the details of the patient’s fundus. It may be necessary to decrease the illumination if the child seems uncomfortable with the intensity of the light.

Visual Acuity Assessment

In assessing visual acuity, it is always important to consider parental input. Parents may be reluctant to volunteer concern that visual function may be poor, but their intuition must be sought. Other relatives, such as grandparents, may have greater experience with children and may provide valuable information. In short, taking a history is an important step in visual acuity assessment.
Infant visual function is particularly important to assess, since the critical period of visual development occurs within the first year of life. During your evaluation, ask the parent if the baby seems to be seeing; in other words, does the infant fixate on and follow the parents, toys, or other objects of interest? Examine the baby’s visual behavior. By 1 month of age, the baby should regard your face steadily without any noticeable constant strabismus. Intermittent strabismus may occur in normal children up to 4 to 6 months of age. Rhythmic shaking of the eyes (nystagmus) or inability to hold fixation, even briefly, is always a concern.

By the 2-month checkup, each eye should be tested separately for its ability to fix and follow. Ask the parent to hold his or her hand over one of the baby’s eyes and then the other, repeating your assessment of the fixation and following response. An interesting target of high-contrast coloration or movement should be used because a baby’s visual resolution is still low. As an alternative, you may use your thumb as an occluder, taking care to keep it close to the eye or apply an adhesive patch to assess each eye. The fixation should be steady. Any abnormal movement of the eyes should alert the examiner to possible nystagmus or strabismus, requiring further investigation. Similar examinations should be repeated at the 4- and 6-month checkups.
If the baby has extremely poor vision in both eyes, hand-waving or eye-poking may be first noticed at the age of 6 weeks to 2 months. Poor vision can be confirmed by watching the baby’s response to a bright light. Failure to blink the eyes indicates that the baby has profoundly reduced vision. Irregular “roving” eye movements or constant or intermittent nystagmus also indicate reduced visual function.

There is no simple test to quantify an infant’s visual acuity that is available for use in an office setting. The ophthalmologist may use preferential looking tests or more involved testing methods for such quantification, but these are fairly time-consuming and are not intended as screening techniques for amblyopia.

Measuring visual acuity in the office becomes possible with patients between the ages of 18 months and 3 years. At this age, acuity generally can be tested more easily at near using a picture chart; however, many cooperative 3-year-olds can be tested with distant targets. Although near testing does not identify the child with myopia, amblyopia will not develop as long as a clear image is being focused on the retina for near objects. The toddler’s world is, in large part, within a 5-foot radius, making the early recognition of small amounts of myopia less important.
Various types of acuity charts are available for the preliterate child at both distance and near. Many children enjoy picture charts, and recognition of one favorite symbol per line makes testing proceed more quickly. The single E chart, also called the preliterate or illiterate E chart, has enjoyed traditional popularity, but it requires gross motor coordination and involves repetition of a single task.

The standard Snellen chart may be used successfully with many children in the 4-to-5-year range. Most charts are designed for use at 20 feet, or 6 meters. Children cooperate more readily if the examiner points to specific letters, rather than having the child read the entire line.

Testing of binocular depth perception is also used as a screening test for amblyopia in some physician offices. However, this should not substitute for visual acuity testing. The depth perception, or stereopsis, test is often difficult for the child of less than 5 years of age to perform. However, some younger children may respond reliably to stereopsis testing.
Ocular Alignment

The final skill to be mastered in amblyopia screening is testing for alignment of the eyes. The corneal light reflex can be used even in the neonatal period to check the ocular alignment. When a penlight is held 1 to 2 feet from the child’s face, a centered corneal reflex should be simultaneously present in each eye. This test is best done by asking the child to focus on a small target, such as a picture applied to a tongue blade.

In normally aligned eyes, the corneal light reflex will be centered in the pupil. Esotropia, or “crossed eye,” is a nasal deviation of one or both eyes. As a result of this deviation, the corneal light reflex is decentered temporally with respect to the pupil.

In contrast, exotropia is an outward turning of the eye that results in an abnormal corneal reflex that is nasally decentered on the pupil or iris.
A vertical offset of the eyes will also result in asymmetric localization of the corneal light reflex, with the reflex in the deviating eye below the pupil center (hypertropia) or above the pupil center (hypotropia) while the reflex in the fixating eye will be centered in the pupil.

If a deviation is present, note which eye appears to be the deviating eye. Recheck the reflexes periodically during the remainder of the examination to determine whether one eye is constantly deviated or whether the strabismus is alternating.

In a child with an abnormal head turn or tilt, it is important to place the head in a normal position to check for strabismus, since the child’s preferred head position may hide certain forms of strabismus. Conversely, an ocular misalignment may exist only in one direction of gaze. It is important to assess movement and alignment in horizontal and vertical gazes.

There is no direct correlation between the angle of strabismus and the depth of amblyopia. In some cases of profound amblyopia, the angle of strabismus may be so small that detection is not possible using only the corneal light reflex, emphasizing the importance of accurate visual acuity testing.
Referral Guidelines

Based on the examination techniques outlined, prompt referral is indicated in the following circumstances:

- **When there is any difficulty obtaining a clear red reflex in either eye.** The difficulty may be something as profound as retinoblastoma or as simple as myopia.
- **When the parent or physician is concerned about visual function for any reason; when nystagmus is present; or when unusual habits such as hand-waving or eye-poking are observed.**
- **When visual acuity is poorer in one eye than in the other or, in an older child, when acuity has diminished from a previous measurement.** Establishing a specific cutoff for referral of mild, bilaterally reduced acuity is difficult. Acuity of 20/30 may be normal in a fidgety 4-year-old but is cause for concern in the cooperative 8-year-old. Any patient with a visual acuity in the range of 20/40 to 20/50 or poorer should be referred.
- **When strabismus is noted on examination.** If the deviation is constant or onset is acute, the child should be referred immediately, as certain life-threatening disorders may present as strabismus.

### Referral: Immediate

- Poor red reflex in one or both eyes
- Concern about visual function by parent or doctor
- Asymmetric or diminishing visual acuity
- Constant or acute-onset strabismus
In an infant, referral for strabismus on a less emergent basis should occur whenever an intermittent strabismus is noted or if parents are still concerned about strabismus at age 6 months despite your observations of straight eyes. Special attention should be given to children with syndromes and systemic diseases, since the incidence of strabismus is higher in these children, and congenital eye abnormalities that could result in reduced vision may be present.

In an older child, reduced visual acuity in one or both eyes should prompt referral. In general, an acuity of 20/40 or worse in a child should be of concern. However, any asymmetry of visual acuities or concerns about reduced acuity at any level warrant ophthalmologic evaluation.

**Treatment Rationale**

The responsibility for treating amblyopia rests with the ophthalmologist, but the pediatrician or other primary care physician should understand the rationale of treatment. Treatment involves clearing the ocular media, focusing the image, and, if visual acuity has not developed normally, initiating amblyopia therapy. In most instances correcting the underlying abnormality, such as unequal refractive errors, is not adequate to restore normal visual function. Abnormal processes of visual cortical development must be reversed. In the presence of strabismus, amblyopia therapy can be initiated before ocular alignment is corrected.
Clearing the Media

Mechanical or surgical procedures are often indicated. For example, congenital cataracts, if significant, must be removed at the earliest possible age. Good visual acuity can be obtained if the cataract is removed before the patient reaches 1 to 2 months of age, although the prognosis is poorer in unilateral cases.

Severe congenital ptosis (drooping of the upper lid) that occludes the pupillary axis can prevent normal visual development, making surgical repair or taping of the lids necessary early in life. Less complete ptosis may intermittently occlude the pupil or induce corneal irregularities and, therefore, warrants careful assessment for findings of amblyopia.

Focusing the Image

Infants and children with significantly unequal refractive errors or high bilateral refractive errors require glasses or contact lenses to achieve good vision and avoid amblyopia. The ophthalmologist can objectively quantify an abnormal refractive error. Although parents are often anxious that their infant will not tolerate glasses, any infant who has a visual problem that can be corrected optically does extremely well. Although glasses may be a seemingly mild intervention, knowledge that their infant or toddler needs them may be extremely traumatic to parents.
Contact lenses may be used when there is a large refractive error that would necessitate thick glasses, or when a large difference exists in the refractive error of one eye in comparison to the other. Advances have been made over the past decade in the use of contact lenses in infants and children. However, technical difficulties exist with contact lens management, and infants and toddlers typically do very well with glasses. Any child wearing contact lenses must be carefully supervised because of possible complications related to lens wear, and a back-up pair of eyeglasses is essential.

**Amblyopia Therapy**

Patching, or occlusion, therapy is the mainstay of amblyopia treatment. Patching the unaffected or better-seeing eye provides monocular stimulation to the amblyopic eye, promoting more normal visual development. Occlusion therapy is prescribed to improve visual acuity and, as a rule, does not eliminate strabismus.

During patching, visual acuity must be checked carefully at close intervals, as determined by the patient’s age and severity of amblyopia. Follow-up examinations ensure that vision is not being reduced in the patched eye (“occlusion amblyopia”). In general, the patient is followed at intervals calculated at 1 week per year of age, or every 3 to 6 weeks in the older child.
Recent studies have shown that full-time occlusion may not be necessary to achieve good results in many cases of amblyopia. Part-time occlusion may be prescribed, depending upon the patient’s visual acuity, availability for follow-up visits, and receptiveness. Although practitioners’ preferences may vary, several hours per day of occlusion may suffice to treat moderate amblyopia.

To ensure successful patching, the parents should be given a full explanation of its purpose and the importance of compliance. Patching cannot be delayed until a more convenient time—effective treatment of amblyopia is sooner, not later. However, evidence indicates that success in amblyopia treatment may extend into the teen years. Once amblyopia has been reversed, the child must be followed periodically until at least the teen years to ensure that vision remains stable. Occasionally, patching must be reinstituted to maintain optimal visual acuity.
Patching itself requires a major effort from the child, parent, and physician. If the decrease in visual acuity is great, resistance from the child may be significant. Often, a child who must wear a patch feels embarrassed or punished. Positive parental attitude is essential, and reinforcement from the primary care physician is very helpful. Reward systems can be invented, such as the use of calendars with paste-on stars for each day the patch is successfully worn. Some children enjoy drawing pictures on the patch prior to wearing it; others will relate to the use of patches on dolls or stuffed animals.

In addition to the psychological concerns, skin irritation may develop as a consequence of the adhesive. Holes may be punched in the adhesive tape or the position of the patch may be altered to reduce the area of skin contact.

Recent studies have shown that the use of atropine eye drops in the non-amblyopic eye is successful in treating moderate amblyopia. Atropine in the better-seeing eye impairs the child’s ability to use that eye at near by blocking accommodation. As a result, the child must switch fixation to the amblyopic eye for near work. The ophthalmologist should determine if this is an option. If the visual acuity at near in the amblyopic eye is too reduced, the child will not switch fixation from the preferred eye and the therapy will fail.
The ophthalmologist treating a child undergoing atropine therapy will carefully monitor the progress of visual acuity, ensuring that near acuity in the amblyopic eye can support near tasks. Allergic reaction to atropine is rare (less than 1%); uncommon systemic side effects may include dry mouth, flushing, fever, tachycardia, and irritability. The child may complain of light sensitivity. Parents should be alerted that the eye receiving the drops will have a fixed and dilated pupil, as shown in the left pupil in this photograph.

Many other techniques for amblyopia treatment have been tried and have failed. Partial occlusion masks, vitamin therapy, eye exercises, and even a steady diet of white wine and veal have all enjoyed transient popularity. Modern day “cures” should be viewed with skepticism.

In summary, amblyopia is created when an abnormality in the developing visual system is present that results in (1) poor clarity of the ocular media or light blockage, (2) poor focus of the image, or (3) poor aim in one eye.
Amblyopia is detected by assessment of (1) red reflex, (2) visual acuity, and (3) ocular alignment.

Amblyopia is treated by (1) correcting the underlying media opacity, (2) focusing the image, and (3) amblyopia therapy with occlusion or atropine optical penalization.

Elimination of this preventable cause of visual loss depends on effective communication among parents, primary care physicians, and ophthalmologists.
APPENDIX 1

Resources


