Amblyopia: Etiology, Detection, and Treatment

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Amblyopia: Etiology, Detection, and Treatment

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FOCUS QUESTIONS

1. Describe the pathogenesis of childhood amblyopia.
2. How does each of the major predisposing factors induce amblyopia?
3. What are the types of strabismus? What is the most frequently encountered form?
4. How are "cover-uncover" tests performed and interpreted?
5. What are the major causes of cataracts in infants?
6. What are the presenting symptoms and signs of retinoblastoma?

Pediatricians are generally the first physicians to examine babies and children who have ocular diseases. They then refer such patients to pediatric ophthalmologists. This symbiosis between the two professions avails children of the highest degree of medical care. Terminology useful in the evaluation and referral processes is provided in the Table.

There are three major disorders that are crucial for pediatricians to recognize: amblyopia, strabismus, and cataracts.

Amblyopia

Amblyopia, the most common cause of visual loss in American children today, is due to inadequate visual stimulation of the brain during the critical period of visual development. During the first 4 months of life, the eye gradually matures and visual pathways develop. Normal visual development requires the brain to receive equally clear, focused images from both eyes simultaneously to "learn" how to see. The brain's visual pathways remain malleable until approximately 6 years of age. Anything that interferes with the brain's visual learning process will result in the development of amblyopia. Amblyopia is preventable, unique to children, and can be treated effectively only during childhood. However, permanent and irreversible blindness results if amblyopia is not detected and treated during early childhood.

SUPPRESSION

Suppression is the neural mechanism by which a child's brain is able to "shut off" images coming from a misaligned eye or an eye with a blurry image. Uninterrupted suppression results in amblyopia because the brain is not afforded the opportunity to learn to use the suppressed eye. If the suppressed eye is not stimulated by the brain while the visual pathways develop, the eye will have permanent visual impairment.

The duration of visual deprivation and its impact on visual development is related directly to the age of the child when the amblyogenic insult occurs and to the type of insult. Because the visual system is most plastic during the first few months of life, amblyopia will develop most rapidly and profoundly in an infant. For example, in an infant younger than 12 months, constant occlusion of an eye may lead to amblyopia in 1 week. It is reversible if detected and treated immediately. An example of an insult that causes constant occlusion of the visual system is a congenital cataract. In an older child, the visual system is more resistant to the development of amblyopia. The earlier and more prolonged the visual insult, the more profound the amblyopia will be. Thus, the success of amblyopia treatment depends on early diagnosis.

CONSEQUENCES OF AMBLYOPIA

There are several consequences of amblyopia. Not only is vision impaired, but fusion—the central melding of the images perceived by each eye into one three-dimensional picture—is disrupted as well. Fusion is dependent upon receiving clear im-

continued. . .

Table. Definitions of Terminology

TERM	DEFINITION
Abduction	Moving an eye outward.
Accommodation	The process whereby the lens becomes more convex to focus near targets. Accommodation is associated with convergence (ie, a simultaneous inward movement of the eyes).
Adduction	Moving an eye toward the nose.
Amblyopia	Loss of vision due to "disuse."
Anisometropia	Difference in refractive errors of the two eyes.
Corneal light reflex	Straight eyes have symmetrically placed reflections from the cornea over the pupils. Strabismus is characterized by a displacement of the corneal light reflex from the pupil, making the reflexes in the two eyes asymmetric.
Cover test	Diagnostic test for strabismus.
Fusion	The brain's ability to use both eyes together and have depth perception.
Leukokoria	White pupillary reflex.
Orthophoria	Ideal ocular alignment (ie, the eyes are perfectly straight).
Phoria	Latent ocular deviation controlled by fusion.
Red reflex	The reflection of light from the retina that appears bright red in normal eyes.
Strabismus	Misaligned or "crossed" eyes.
Suppression	Child's brain's ability to ignore images from a misaligned eye or an eye with a blurry image.
Tropia	Manifest ocular deviation that cannot be controlled.

ages from each eye simultaneously, and amblyopia disrupts this process so that depth perception is lost.

CAUSES OF AMBLYOPIA

The most common cause of amblyopia is strabismus or "crossed" eyes. Anisometropic amblyopia is caused by a significant focusing problem (ie, refractive error) in one eye that results in a blurred image. The most severe form of amblyopia is deprivation amblyopia, which is caused by any factor that totally occludes vision, especially a cataract.

Strabismus

Strabismus and amblyopia affect 5% of the population.[1] Strabismus implies an improper alignment or crossing of the eyes. Primary strabismus leads to amblyopia through the neural mechanism of suppression, which results because the brain ignores images coming from the crossed eye. Uninterrupted suppression leads to amblyopia, which does not depend on the degree of eye crossing. Another consequence of strabismus is the loss of fusion, resulting in impaired depth perception as well as poor cosmetic appearance.

PHORIA AND TROPIA

Orthophoria is the ideal condition of perfect ocular alignment. Most people have a small heterophoria, a latent tendency for the eyes to drift away from perfect alignment. A phoria is kept latent by fusion; anything that disrupts fusion may elicit a phoria. Although most phorias are benign, a large exophoria (a latent tendency for the eyes to drift outward) may cause eye strain when reading. An esophoria (a latent tendency for the eyes to deviate inward) may be a sign of uncorrected hyperopia or be a precursor of accommodative esotropia.

A heteroptropia is a manifest ocular deviation that cannot be controlled. The prefixes "eso-," "exo-," "hyper-," and "hypo-" signify the direction of an ocular misalignment, whereas the suffixes "-phoria" and "-tropia" indicate whether the deviation is latent or manifest.

Strabismus consists of several different ocular deviations that have a large variety of causes (Fig 1). Esotropia, or convergent strabismus, is an inward crossing of the eyes. Exotropia, or divergent strabismus, gives a "wall-eyed" appearance. Hypertropia is an upward deviation of the eye, and hypotropia is a downward misalignment of the eye.

PSEUDOSTRABISMUS

Pseudostrabismus may be created by the presence of broad epicanthal folds and a flat nasal bridge, features commonly seen in infants. The mistaken impression that esotropia is present may be given if the medial aspect of the sclera is obscured by the broad epicanthal folds. A pseudostrabismus and a true strabismus are distinguished by observing symmetrically placed corneal light reflexes and a negative cover test in pseudostrabismus.

ESOTROPIA

Esotropias account for more than 75% of all cases of childhood strabismus. Among the types of esotropia in children are infantile esotropia, sixth nerve palsy, Duane syndrome, accommodative esotropia, and sensory deprivation esotropia.

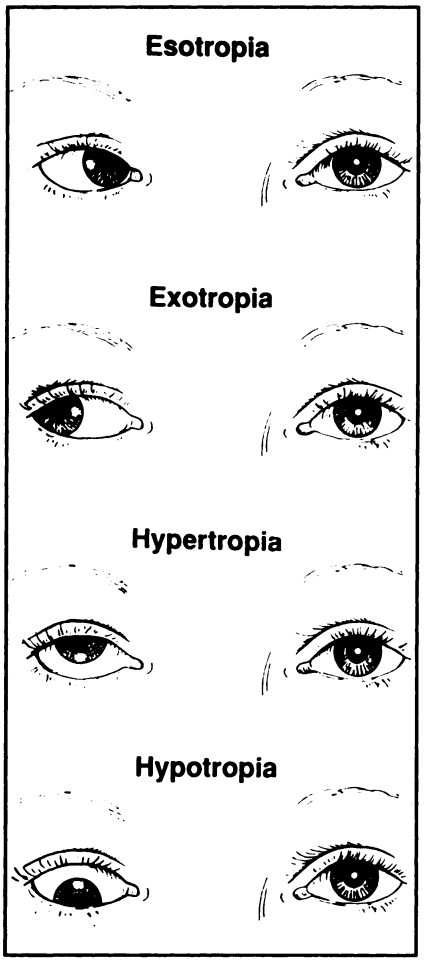

FIGURE 1. Types of strabismus. Note displaced corneal light reflex in misaligned eye.

Infantile esotropia

Infantile esotropia has an early age of onset, less than 12 months of age. It is caused by a poorly understood innervational defect in the ability to use both eyes together and is characterized by an esotropia with poor abduction. Although the angle of deviation usually is quite large, bringing it to attention early, at times the degree of deviation may be more subtle.

Several fixation patterns may be seen in esotropia. There may be alternate fixation, where sometimes the right eye is turned in while the left eye fixates and at other times the left eye turns in while the right eye fixates. If the eyes demonstrate easy, spontaneous alternation of fixation, there is alternate suppression, and amblyopia is less likely to be present because each eye has a chance to develop vision. A less desirable fixation pattern is one of a constant fixation preference whereby only one eye is always turned in. This results in constant suppression and ambly-

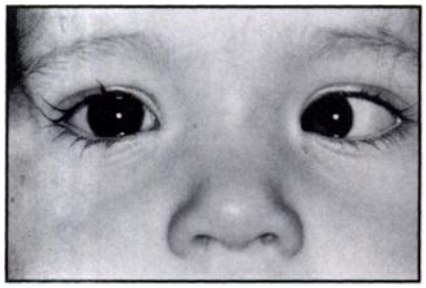

FIGURE 2. Infantile esotropia.

FIGURE 3. Congenital esotropia and cross fixation.

opia of the constantly turned in eye (Fig 2).

Another fixation pattern unique to infantile esotropia is cross fixation (Fig 3) in which the infant fixates the right side of the world with the left eye and the left side of the world with the right eye. Both eyes simultaneously are turned inward and there is alternation of suppression. Sometimes the child may develop a fixation preference for one eye despite cross fixation, resulting in amblyopia in the nonpreferred eye (Fig 2).

Sixth nerve palsy

The apparent lateral rectus weakness in infantile esotropia manifested by poor abduction (ie, weakness of moving either eye outward) may simulate a bilateral sixth nerve palsy. Infantile esotropia can be differentiated from congenital sixth nerve palsy, which is much less common, by demonstrating the presence of lateral rectus function. This is done by spinning the infant, thereby stimulating the labyrinth system, which produces abduction nystagmus in the case of esotropia but not in the case of a true sixth nerve palsy.

Duane syndrome

Duane syndrome most commonly appears as a congenital esotropia with marked limitation of abduction of one eye. This condition results from the congenital agenesis of the sixth nerve nucleus and paradoxical innervation of the lateral rectus muscle by fibers from the third nerve. Duane syndrome is characterized by an esotropia when the head is held straight as the child looks forward. On attempted abduction there is widening of the eyelid fissure and the eye rarely moves out beyond midline. On adduction of the involved eye, there is narrowing of the eyelid fissure and retraction of the globe into the orbit. Children who have Duane syndrome frequently keep their heads turned to the side to avoid the side with limited abduction and keep their eyes aligned. Amblyopia of the affected eye may develop if the abnormal head posture is not assumed because the brain will suppress images coming from the misaligned eye.

Accommodative esotropia

Accommodative esotropia usually has an age of onset of approximately 12 months to 7 years. This condition usually is accompanied by hyperopia, where the visual images are in focus behind the retina. In the presence of this condition, the child learns accommodation, the process whereby the ciliary muscle allows the lens in the eye to become more convex and bring the image behind the retina into focus on the retina. Accommodation normally is associated with convergence, or a turning in of the eyes. An innervational defect with accommodative esotropia results in an overconvergence with accommodation.

Accommodative esotropia frequently begins as an intermittent esotropia, most commonly observed for near fixation. Eventually, the esotropia becomes constant for both near and distance fixation. Amblyopia very commonly occurs in this condition.

Sensory deprivation esotropia

Sensory strabismus results from any primary cause of visual loss. If vision is poor in one or both eyes, fusion cannot occur and ocular alignment is lost. The ocular deviation usually seen due to visual loss is esotropia. The rarest, but most dangerous, cause of visual loss and sensory strabismus is retinoblastoma.

Retinoblastoma is the most common primary intraocular malignancy of childhood, occurring in one of every 15 000 births. The most common initial sign is leukokoria, a white pupillary reflex that looks like a "cat's eye reflex" (Fig 4). Strabismus is the second most common sign of retinoblastoma. Less commonly, retinoblastoma is indicated by a red painful eye. Leukokoria is a portentous sign because all of the entities in the differential diagnosis of leukokoria produce significant visual loss. Retinoblastoma is the most ominous diagnosis because it is a malignant tumor whose mortality approaches 100% once the tumor spreads beyond the eye to the central nervous system and bone marrow. The overall survival rate is 92% if the tumor is appropriately treated before metastases.

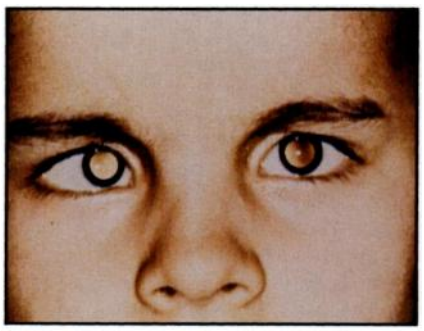

FIGURE 4. Bilateral retinoblastoma with leukokoria and esotropia. Photograph courtesy of Dr David H. Abramson.

EXOTROPIA

The onset of exotropia is often between infancy and 4 years of age (Fig 5). It is much less common than infantile esotropia. It usually begins as an intermittent deviation that is more prominent when the child looks into the distance. Once the deviation becomes constant, binocularity is lost and amblyopia may develop if there is a fixation preference for one eye because images from the constantly deviating eye will be suppressed by the brain.

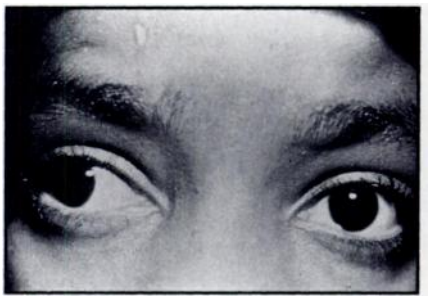

FIGURE 5. Infantile exotropia.

VERTICAL STRABISMUS

The most common vertical ocular misalignment during childhood is congenital superior oblique palsy. The etiology of this condition is unknown. The child exhibits an abnormal head posture that allows him or her to achieve binocularity and fusion by avoiding the hypertropia of the paretic eye that occurs when the head is held straight (Fig 6). An abnormal head posture frequently implies the presence of an ocular misalignment.

Ametropic and Anisometropic Amblyopia

After strabismus, the second category of disorders that lead to the development of amblyopia are focusing problems (refractive errors) of the eye (Fig 7). There are three types of refractive errors: Myopia, hyperopia,

continued. . .

FIGURE 6. Child with head posture characteristic of congenital superior oblique palsy. Photograph courtesy of American Academy of Ophthalmology.

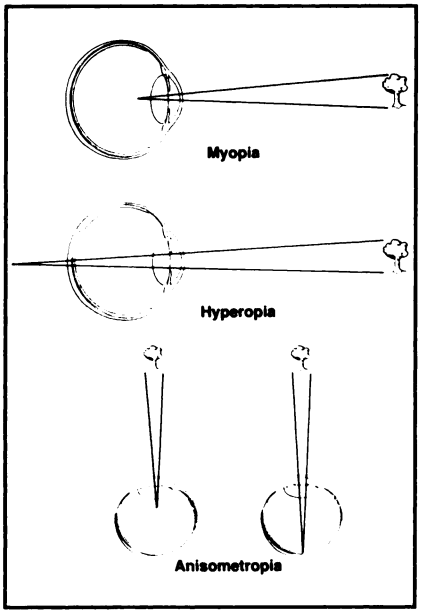

FIGURE 7. Refractive errors and anisometropia.

and astigmatism. In the case of myopia (near-sightedness), the image is focused in front of the retina; in the case of hyperopia (far-sightedness), the image is focused behind the retina; and in the case of astigmatism, light rays in two meridians are out of focus. If images on the retina are significantly out of focus, amblyopia will result.

If the focusing error of the two eyes is significant, amblyopia will develop in both eyes, leading to ametropic amblyopia. If the focusing power of the two eyes is dissimilar, the child will prefer to use the eye with the better focusing power and will ignore images coming from the out-of-focus eye. This condition is known as anisometropic amblyopia. It is more difficult to detect anisometropic amblyopia than ametropic amblyopia because, in the latter case, vision is poor in only one eye and the child will function normally whereas a child who has poor vision in both eyes may demonstrate behavioral clues of poor vision. In the presence of anisometropic amblyopia, the eyes are initially aligned. However, in the case of long-standing amblyopia, a secondary ocular misalignment may occur.

Cataracts and Deprivation Amblyopia

The most severe form of amblyopia is deprivation amblyopia, which results from any factor impeding the transmission of clear images into the eye. Cataracts are one of the most important causes of severe deprivation amblyopia.

A cataract is an opacity of the normally clear crystalline lens, the function of which is to focus light rays onto the retina. Cataracts in children may be congenital or acquired and may be unilateral or bilateral. One of every 250 infants is born with a cataract.[2] However, not every lens opacity or cataract is visually significant. A visually significant cataract diffuses light rays entering the eye, obstructing the transmission of clear images onto the retina and the brain.

ETIOLOGY OF CATARACTS

Four major causes account for most cataracts in children. At least one quarter of the cases are familial, one third are related to a maternal infection during pregnancy, one third are idiopathic sporadic cases, and the remainder are related to particular syndromes and systemic diseases, some of which may be hereditary. The smallest percentage of cataracts is related to trauma. Cataracts also may occur secondary to exposure to medications and ionizing radiation during gestation; these factors also may be associated with other ocular anomalies and diseases.

Familial cataracts most commonly are transmitted as an autosomal dominant trait with almost complete penetrance. Affected patients are usually otherwise healthy. Infants who have sporadic idiopathic cataracts are usually otherwise healthy, and the majority represent the spontaneous mutation of the autosomal dominantly inherited cataract.[3] Thus, more than half of all infants who have cataracts will be otherwise healthy.

The leading embryopathy in the group with congenital infection is rubella. Although the rate of congenital rubella infections has been declining during recent years, rubella still accounts for the majority of cataracts in the maternal infection group. Other infections indicated as causal of congenital cataracts are rubeola, varicella, herpes simplex, toxoplasmosis, cytomegalic inclusion disease, poliomyelitis, influenza, hepatitis, and infectious mononucleosis.

Several syndromes related to cataracts are determined genetically, including chromosomal anomalies such as Down syndrome, Turner syndrome, Patau syndrome, Edward syndrome, and Rubinstein-Taybi syndrome. Lowe oculocerebrorenal syndrome is an x-linked recessive condition characterized by congenital cataracts, glaucoma, mental retardation, aminoaciduria, and renal tubular acidosis. Alport syndrome, a hereditary familial congenital hemorrhagic nephritis, is characterized by progressive cataracts, retinal pigment epitheliopathy, hemorrhagic nephritis, and sensorineural hearing loss.

Other important metabolic disorders associated with cataracts include galactokinase deficiency, galactosemia, hypocalcemia, hypoglycemia, and diabetes mellitus. Galactokinase deficiency is important to recognize because cataracts may be the only clinical manifestation of the disorder, which can be treated with an appropriate diet. Galactosemia results from a deficiency in galactose-1-uridyl transferase and is associated with hepatosplenomegaly, jaundice, lethargy, mental retardation, and failure to thrive. Such cataracts also may be treated with appropriate diet if diagnosed early. Cataracts due to the hypocalcemia of hypoparathyroidism occur in association with long-standing untreated disease. Cataracts in children who have diabetes mellitus develop during late childhood or during adolescence. Children born to mothers who have diabetes or dysthyroid disease also have an in-

creased incidence of congenital cataracts.

Detection of Amblyopia

The diagnostic test for amblyopia is assessment of visual acuity. In infants and preverbal children, this is accomplished by observing the fixation and the following ability of each eye individually. By 1 month of age, the infant is able to fixate steadily on the human face. Blinking or closing the eyes in an infant are indicators of light perception. The presence of pupillary constriction in response to a bright light implies an intact visual pathway to the visual cortex; however, cortical blindness may be present.

By 3 to 4 months of age, the infant should be able to fixate and follow a penlight or a small toy. The absence of steady fixation and following are indications of decreased vision in the eye. Objection to occlusion of one eye is a sign of poor vision in the nonoccluded eye. Lack of response to a bright light, nystagmus, and eye pressing are signs of severe visual impairment in both eyes. Eye pressing or digitalization is the process whereby a blind child presses the eye very hard as a form of self-stimulation. Nystagmus and eye pressing develop in infants with profound bilateral visual loss.

EYE CHARTS

Eye charts utilizing pictures, the E game, or letters are useful in older children (Fig 8). Picture recognition using the Allen cards at 16 in or the projected Allen card pictures at a distance are useful for children aged 2½ and 3 years.

The "illiterate" E game consists of a chart of capital E's presented at a distance, usually 20 ft. The child indicates the direction of the arms of the E pointed to on the chart with his or her fingers. The E game is most useful for children of ages 4 and 5 years. One problem is that children frequently confuse right and left, but this should be ignored if the child can accurately and easily distinguish the horizontal E's from the vertical ones. Up and down confusion is much less common.

An alternative to the E game is the "HOTV" chart, which is a variation of the Sheridan-Gardiner Test. In this test, a chart consisting of a combination of different sized letters limited to H,O,T, and V is presented 10 ft from the child while the child holds a card with the letters H,O,T, and V. The child is asked to match the letter pointed at by the examiner on the chart with the corresponding letter on the held card. This is extremely useful in children aged 3 to 5 years. The standard Snellen letter chart at 20 ft is the best test for school-aged children who are well versed in letter recognition. Today, with early schooling and Sesame Street on television, some children as young as 4 years of age can respond to the Snellen letter chart.

Many children, especially younger ones, have difficulty concentrating on a chart 20 ft away. Therefore, in some cases it may be wise to bring the chart closer. The only disadvantage of doing this is that near vision is always better than distance vision, even with visual impairment.

One crucial aspect of measuring visual acuity is to be certain that the untested eye is well covered. Children frequently will peek from behind an occluder to see better and please the examiner. This is a common pitfall in the diagnosis of amblyopia.

TESTS FOR FUSION

Assessment of fusion and binocular function using the Worth 4 Dot Test may be performed in children as young as 2½ to 3 years, simply by having the child touch the dots on the Worth 4 Dot flashlight while wearing the red-green glasses (Fig 9). The red-green glasses are worn with the red lens over the right eye and the green lens over the left eye. A flashlight with 2 green, 1 red, and 1 white light is viewed at distance and near. If fusion is present, the

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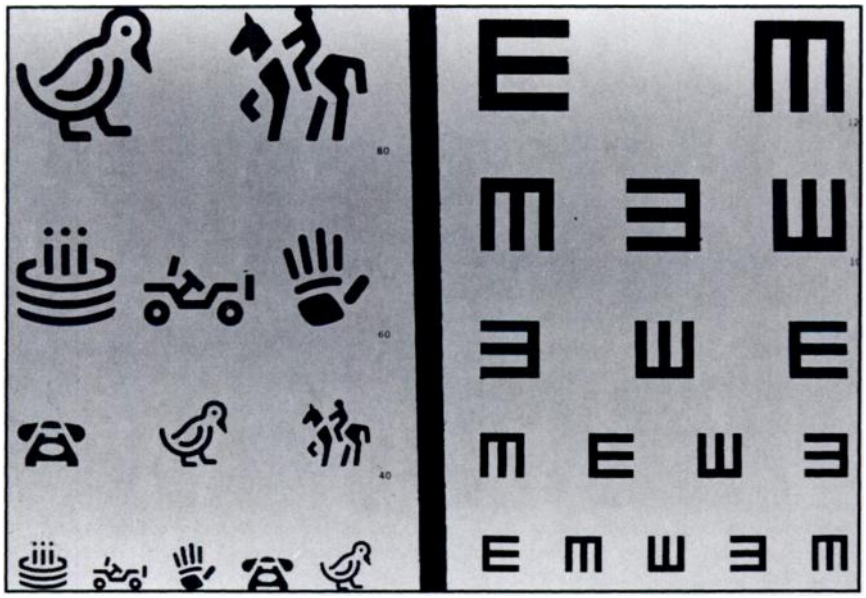

FIGURE 8. Eye charts for young children: Allen cards (left) and "E" game (right).

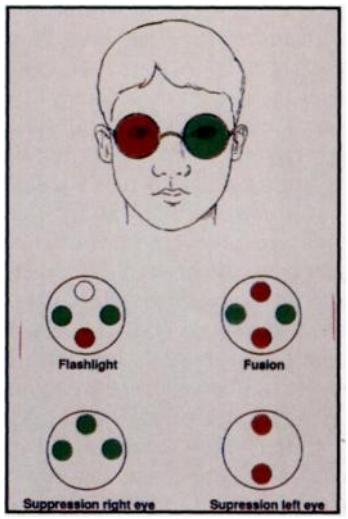

FIGURE 9. Worth 4 Dot test.

child will see 4 lights: 1 red, 2 green, and 1 alternating red or green light. If the right eye is suppressed, only three green lights will be seen because the brain will suppress seeing the red light from the right eye. If the left eye is suppressed, only the two red lights will be seen.

Stereoacuity testing using the Titmus Stereo Test may be performed in children aged 4 years and older, although full maturity of stereopsis does not occur until about age 6 years.

Detection of Strabismus

Strabismus may be detected by observing the position of the corneal light reflexes and performing cover tests to identify tropias and phorias.

CORNEAL LIGHT REFLEX TEST

In the corneal light reflex test, the child fixates on a penlight and light is reflected off the cornea. When the eyes are aligned, the corneal light reflexes will be placed symmetrically in the pupils. If a tropia is present, the light reflex will be displaced in the deviating eye. For example, in the presence of an esotropia, the corneal light reflex will be deviated temporally away from the pupil in the deviating eye (Fig 1). In the presence of an exotropia, the corneal light reflex will be displaced nasally in the deviating eye.

COVER TEST

The diagnostic test for strabismus is the cover test. In the cover-uncover test, one eye is covered with a thumb or an occluder while the child fixates upon a particular target, such as a letter on the eye chart or a toy. If the uncovered eye is deviating, it must make a refixation movement to maintain fixation on the object of regard. The cover then is removed and the opposite eye is covered. A refixation movement of the uncovered eye as the opposite eye is covered is diagnostic of a tropia (Fig 10). A refixation movement by both eyes with the cover test implies alternate fixation. If only one eye always deviates, a fixation preference is present. In the presence of an esotropia, the deviating eye will make an outward movement to take up fixation as the fixating eye is covered. In the presence of an exotropia, the deviating eye will make an inward movement to take up fixation as the fixating eye is covered.

A phoria is kept latent by fusion and may be elicited by disrupting binocularity and not allowing the child to view the object of regard with both eyes simultaneously. Binocularity is disrupted by the cover test. If a phoria is present, the eye will deviate under the cover as the cover is brought over one eye (Fig 11). As the cover is removed and binocularity is reestablished, there is a quick refixation movement of the eye that has just been uncovered.

Detection of Focusing Problems

Focusing problems are detected by observing the "red reflex." A dim reflex that is made brighter by dialing in minus or plus lenses in the ophthalmoscope is a sign of a focusing problem. Asymmetry in brightness of the red reflexes may be a sign of anisometropia.

Detection of Cataracts

The morphologies of different cataracts vary (Figs 12-14) and, unless the anterior portion of the lens is involved in the cataract, the cataract

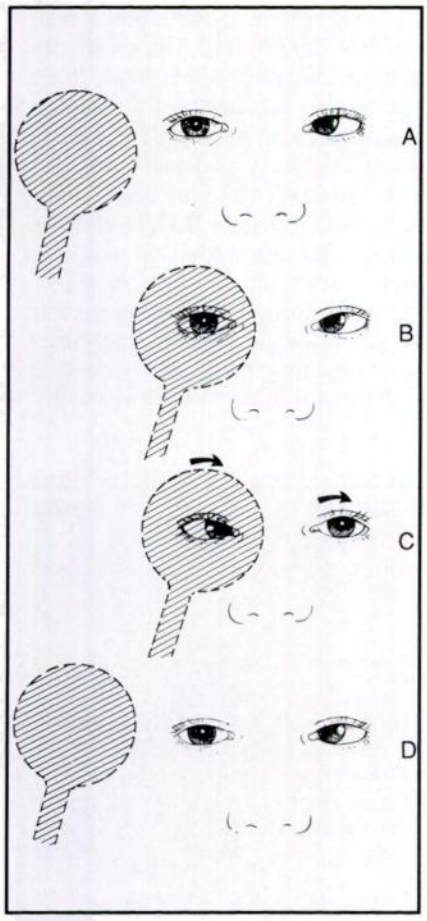

FIGURE 10. Cover test demonstrating a tropia. A) Right eye is fixating and a left esotropia is present. B) Cover is brought in front of right eye. C) To maintain fixation on object of regard, left eye makes a refixation movement outward while right eye turns in under cover. D) If right eye is the preferred eye for fixation, it will straighten out and left eye will turn in again as cover is removed.

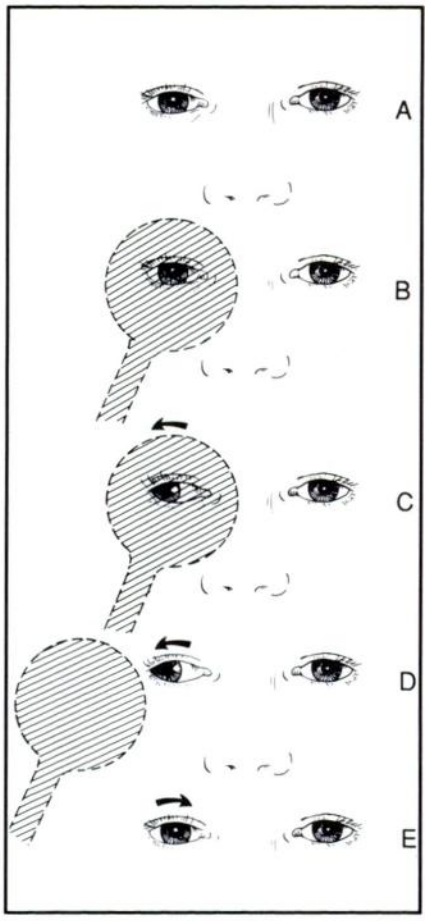

FIGURE 11. Cover test demonstrating a phoria. A) Eyes are straight. B) Cover is brought in front of right eye and disrupts fusion. C) Right eye drifts outward under cover (exophoria). D) Cover is removed and binocularity is reestablished. E) Exophoric eye quickly makes a refixation movement inward to maintain fusion and ocular alignment. Eyes again are straight.

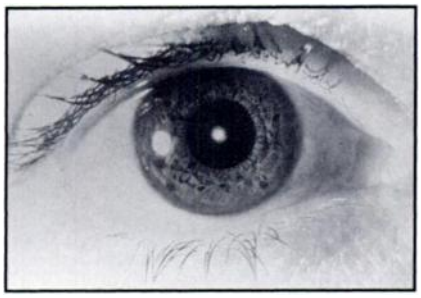

FIGURE 12. Anterior polar cataract. This cataract involves the anterior portion of the lens and may be seen on penlight examination. Although surgery usually is not indicated, such cataracts require careful observation for amblyopia, which may be treated with correction of underlying refractive errors, dilating drops to widen the pupil, and patching of the better eye to encourage visual learning in the eye with the cataract.

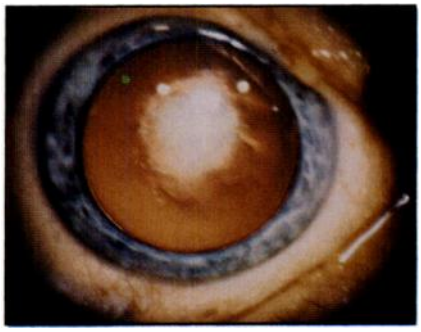

FIGURE 13. Central lens opacity measuring >3 mm. This cataract requires surgery and may be seen only by examining the red reflex with the ophthalmoscope. The bright red reflex is seen surrounding the cataract. Photograph courtesy of American Academy of Ophthalmology.

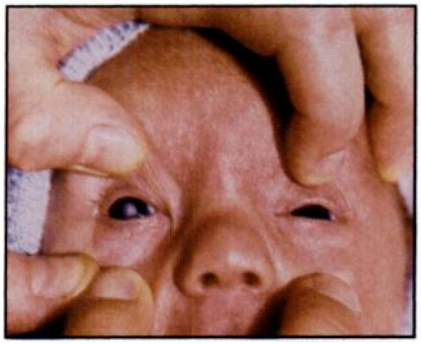

FIGURE 14. Leukokoria. This complete white cataract, which requires surgery, may be visible on penlight examination as well as red reflex examination.

will not be detected by a routine penlight examination. The presence of strabismus and nystagmus implies that severe visual deprivation has occurred in both eyes.

Diagnosis of the cataract depends on the observation of a disturbance of the red reflex of the eye. The red reflex is the reflection of light from the retinas, seen as red pupils when a bright light is directed into the pupil. Evaluation of the red reflex is useful not only in detecting a cataract, but in diagnosing other causes of leukokoria, such as retinoblastoma.

Examination of the red reflex is accomplished by using the direct ophthalmoscope with a bright halogen bulb, selecting the largest, white round aperture and dialing in a +2 or +4 lens. One looks through the ophthalmoscope at the eye from 1 or 2 ft away and directs the light into the pupil as if to look at the fundus. The normal red reflex is bright and clear.

Treatment

AMBLYOPIA

Amblyopia is treated by eliminating the amblyogenic factor, by providing the child with glasses to correct any refractive error, and by patching the preferred eye with an adhesive patch, thereby forcing the amblyopic eye to work (Fig 15).

STRABISMUS

The treatment of strabismus is aimed at eliminating amblyopia and realigning the eyes to restore binocularity. Realignment usually requires surgery, except for special types of strabismus such as accommodative esotropia. Accommodative esotropia is treated with glasses for the hyperopia. Some children may require bifocal glasses to control a deviation on near fixation that exceeds the distance deviation (Fig 16).

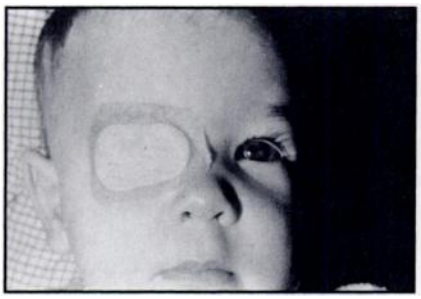

FIGURE 15. Child undergoing amblyopia therapy with adhesive eye patch over preferred eye to stimulate visual development in amblyopic eye. Photograph courtesy of American Academy of Ophthalmology.

FOCUSING PROBLEMS

Focusing problems are treated with glasses. If amblyopia of one eye is present, patching therapy is necessary as well.

CATARACTS

The impact on vision and the treatment of the cataract is directly related to its morphology and density. Small nonprogressive opacities, such as an anterior polar cataract (Fig 12),

continued. . .

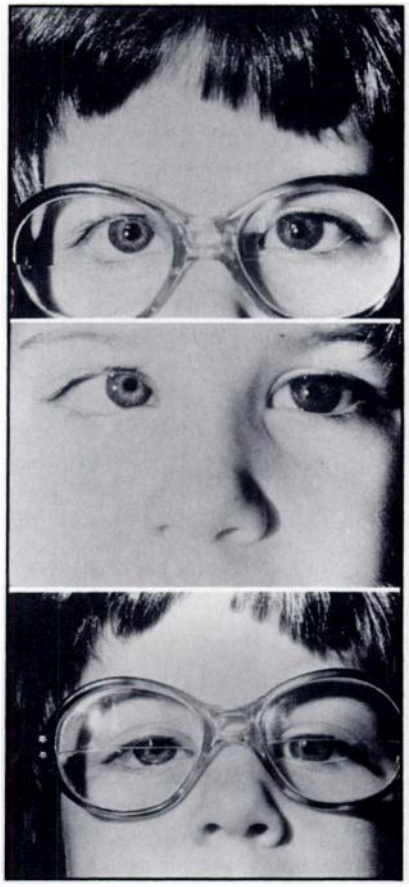

FIGURE 16. Accommodative esotropia treated with bifocal glasses. The child has an esotropia without glasses. The eyes are straight at a distance with hyperopic glasses correction. However, the esotropia on near fixation is corrected with a bifocal segment to the glasses. Photograph courtesy of American Academy of Ophthalmology.

may require observation, correction of refractive errors, dilation of the pupil of the eye with the cataract, and patching of the better-seeing eye with an adhesive patch. Central opacities >3 mm in size require surgical intervention. Examples of cataracts requiring surgical intervention are shown in Figures 13 and 14.

The most important factors for a successful visual outcome of congenital cataracts are: 1) early surgical intervention, within the first 8 weeks of life; 2) immediate accurate postoperative optical rehabilitation; and 3) compliance with aggressive patching therapy for unilateral cataracts.

Surgical treatment involves removing the entire lens. Postoperative aphakia (absence of the crystalline lens) requires treatment by replacing the crystalline lens by another lens that will focus light rays onto the retina to stimulate normal visual development.

Optical rehabilitation may be accomplished with glasses or contact lenses for bilateral aphakia and a contact lens for unilateral aphakia. Optical rehabilitation of unilateral aphakia is limited to the contact lens because of the optical properties of aphakic glasses and contact lenses.

Patching therapy for amblyopia is the other crucial aspect of postoperative management of cataracts. Vision is frequently asymmetric in infants who have bilateral cataracts, which requires patching treatment of the preferred eye. Infants who have unilateral cataracts require intensive prolonged patching of the normal eye after surgery until the age of visual maturity to encourage the brain to use the aphakic eye. This is because the phakic eye always will be preferred over the operated eye.

Timing of Screening

All infants should undergo an assessment of the red reflex at birth and at periodic intervals after birth. An evaluation of visual acuity, ocular alignment, and ocular disease should be performed between 4 and 6 months of age, which is the time during which development of visual acuity and ocular coordination occur. Reassessment of acuity and alignment should be performed routinely at least every 12 months thereafter. Earlier reevaluation should take place if symptoms or signs of decreased vision or strabismus occur and if parental concern is expressed. The entire screening process takes 10 to 15 minutes.

Indications for Referral

Indications for immediate referral to a pediatric ophthalmologist include any disturbance in the red reflex of the eye, asymmetric or diminished visual acuity, constant or acute onset strabismus, and the presence of risk factors (eg, family history of retinoblastoma, congenital cataracts, metabolic disease, and genetic disease) as well as parental concern. Referral also is recommended for children who experience intermittent strabismus as well as associated syndromes or systemic diseases.

Summary

Amblyopia is a preventable cause of visual loss in children that may be permanent unless it is detected and treated early. It may be caused by strabismus, refractive errors, or cataracts. Primary strabismus may lead to loss of vision from amblyopia and the loss of binocularity. Secondary strabismus may be a sign of primary visual loss in one or both eyes. The most serious disorder that may present as secondary strabismus is retinoblastoma. It is imperative to detect retinoblastoma early because of its morbidity and mortality.

Amblyopia is detected by assessing the visual acuity of each eye. Strabismus is detected by using the corneal light reflex test and the cover test. Focusing problems are detected by assessing the visual acuity and the red reflex. Cataracts and retinoblastoma may be detected by examining the red reflex of the eye.

Treatment of amblyopia consists of correcting the amblyogenic factor with appropriate glasses and surgery. The preferred eye is patched with an adhesive patch to stimulate visual development in the amblyopic eye.

The pediatrician plays a crucial role in the early detection of amblyopia, strabismus, and cataracts. The key to successful visual outcome is early recognition by the pediatrician, referral to the pediatric ophthalmologist, and prompt treatment.

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