An Approach to Amblyopia

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Introduction

Hi everyone, my name is Crystal Liu and I am a senior medical student at the Cumming School of Medicine in Calgary, Canada. In conjunction with Dr. Bo Bao and Dr. William Stell, I have created this podcast to provide you with an approach to amblyopia.

After listening to this podcast, the learner should be able to:

1. Define amblyopia
2. Develop a differential diagnosis for a child with reduced visual acuity
3. Recognize the clinical signs and symptoms of amblyopia
4. Discuss management of patients with amblyopia
5. Discuss indications for referral to a pediatric ophthalmologist

Clinical Case

Let's start with a case. Robert is a 4-year-old boy who comes to the pediatric community clinic with his mother. His mother tells you that she is concerned about decreased vision in Robert's left eye, found on a screening exam at day care. However, there have been no noticeable vision problems at home. He is growing normally and reaching his developmental milestones. His past ocular history and medical history are unremarkable. There is no known family history of amblyopia or strabismus. On presentation, his visual acuity is 20/30 in the right eye, and 20/160 in the left. He has no relative afferent pupillary defect. Eye movements are normal. His mom is concerned about his reduced visual acuity and is wondering if you have any suggestions of what to do next. What is on your differential for a child with decreased vision in one eye?

Introduction to Amblyopia

Amblyopia (often called "lazy eye") is a condition in which there is a reduction in visual acuity of one eye or occasionally both eyes (1). Essentially, the eye does not see as well as we are expecting, although the structure and function of the retina and optic nerve are perfectly normal. Amblyopia is quite common, occurring in 1 to 4% of children. (2) The reduction in vision can result from abnormal visual system development early in life, due to strabismus, hyperopic refractive error, or structural obstructions. (1) Risk factors for amblyopia include a history of prematurity, small size for gestational age, first-degree relative with amblyopia, and neurodevelopmental delay. (2) Early detection and treatment of amblyopia can restore vision to its full potential, making regular vision check-ups an important component of well-child check-ups.

Differential Diagnosis

Aside from amblyopia, other causes of reduced visual acuity can include any primary disorders affecting the visual pathway including the retina, optic nerve, and brain lesions. (2) The presence of leukocoria, afferent pupillary defect, or abnormal color vision would suggest an alternative diagnosis. Functional vision loss may also be seen in children with anxiety or emotional disturbances. Often, they are accompanied by other complaints such as headache, photophobia, or diplopia.

Pathophysiology

In children, the visual cortex undergoes continual development until about seven years of age. (2) During this period of neuroplasticity, having clear and focused visual stimuli is incredibly important to stimulate the normal development of the visual cortex; so, disturbances in vision can lead to amblyopia. Fortunately, however, this plasticity of the visual system in kids is also what allows for reversal of amblyopia. We will talk more about treatment later in this podcast. Amblyopia can be classified on the basis of the underlying cause of the visual disturbance. There are three major causes: strabismic, refractive, and deprivational. (2)

First let's talk about strabismic amblyopia. This is the most common type, accounting for approximately 50% of cases. (2) In strabismus, there is an abnormal alignment of the eyes. As a result of the misalignment, at most viewing distances the visual fields of the two eyes are non-congruent, so that corresponding points in the two images are not represented at corresponding points in the visual cortex. To avoid diplopia, the visual input from one eye is suppressed by developmental mechanisms that keep neural connections from that eye from forming in the cortex. Over time, the long-term suppression of input from that eye leads to permanent lack of those connections, and unilateral amblyopia.

The second most common cause is refractive, occurring in approximately 15 to 20 percent of amblyopia cases. (2) When the refractive error in one eye is greatly different from that in the other (high ametropia), unequal image clarity forces the brain to use input from just one eye for seeing. As in the case of strabismus, then, inputs to visual cortex from the eye that sends poorly focused images to the brain – because of hyperopia, severe myopia, or astigmatism – will fail to develop normally, resulting in refractive amblyopia.

The third type, deprivational amblyopia, is the least common, accounting for less than 5 percent of cases, (3) but it is the most severe. This type of amblyopia can be due to a structural abnormality that obstructs the visual axis, such as congenital cataracts, retinoblastoma, ptosis, corneal scarring, or vitreous hemorrhage. (3) The mechanism underlying failure of visual inputs from the affected eye to form in the visual cortex is assumed to be the same as that for the other types of amblyopia, discussed above. Deprivational amblyopia can result in permanent visual impairment if not treated urgently.

History and Physical Exam

The use of vision screening exams, during well-child check-ups in a primary care setting, is important for detection of amblyopia. Since timely intervention of amblyopia is important for reducing its prevalence and severity, the goal is to promptly identify children who can benefit

from intervention. (1) Guidelines of the *American Academy of Pediatrics* recommend vision screening for children during examinations of newborns, infants 6 months, 1 to 2 years of age, 3 to 4 years of age, 5 years of age, and older – and repeat screening every 1 to 2 years after 5 years of age. (3) For a detailed review of ocular history and physical exams, please refer to the podcast titled "Pediatric Eye Exam in Primary Care". Vision screening begins with a thorough history to identify risk factors, including premature birth, low birthweight, family history of strabismus, amblyopia, retinoblastoma, or genetic eye diseases. (1) You might also ask the parent such things as: Does the child recognize faces and objects? Do the parents notice eye deviation, abnormal head posturing, or squinting? During the newborn eye exam, look for red reflex, pupil response, eye deviation and congenital cataracts. For infants from 6 months of age to 3-5 years of age , who are unable to read an eye chart, fixation-reflex testing and occlusion testing can be performed. The goal is to examine for large differences in visual acuity between the eyes, not necessarily for absolute visual acuity. Testing of the fixation reflex can be done by moving a visual target side to side within a child's field of view. Another method involves alternately occluding one eye and then the other, and monitoring the child's response. A child with moderate to severe visual impairment will likely become more irritable when the 'good' eye is occluded, thus indicating an eye preference. As well, ocular alignment tests can be done to screen for strabismus. In a cooperative, verbal child 3 years of age and older, visual acuity testing with eye charts should be attempted. A testing distance of 10 feet is recommended. (4) A greater than two-line difference between the eyes indicates abnormality.

Treatment

Children with amblyopia should be managed by an eye specialist. It is important to remember that the 'weaker' eye (and its retina and retinal output) are just as good as those of the 'good' eye. It is the neural connections in the visual cortex that are abnormal. Management of amblyopia involves penalizing the 'good' eye, allowing input from the 'weaker' eye to work and form connections. The management is a three step approach: 1. Eliminate any visual obstructions; 2. Correct refractive errors; and 3. Encourage use of the amblyopic eye. Treatment for amblyopia should begin as early as possible, as it is most responsive to therapy (i.e., the cortical neuroplasticity needed for recovery is still most operational) before the age of seven. (1) Nonetheless, treatment should be attempted in all children with previously untreated amblyopia, regardless of age. In children with refractive amblyopia, the refractive error must be corrected with glasses or contact lenses. The refractive correction alone can sometimes be enough to treat amblyopia, if vision improves on three to four-month follow-up. If amblyopia persists, we start to encourage use of the amblyopic eye by patching the unaffected eye. The duration of patching typically varies from two hours to all waking hours of the day, depending on the severity of the amblyopia and the age of the child. Adherence to patching is a major management problem, as children often resist wearing the patch. Another method for encouraging use of amblyopic eye is to administer cycloplegic agents to blur the vision in the good eye. A cycloplegic agent, as the name implies ("cyclo" = "ciliary body"; "plegic" = "paralytic"), causes temporary paralysis of the ciliary body – leading to loss of the ability to accommodate. The inability of the 'good' eye to focus provides the amblyopic eye with an advantage and encourages its use, thus promoting the formation of new input connections from that eye in the visual cortex. Atropine is used for children older than two years of age, while cyclopentolate is used for children younger than two years of age. Pharmacologic therapy is second-line, being used mainly after failure of patching.

Indications for referral

Children suspected of having amblyopia should be referred to an ophthalmologist or optometrist for a more complete evaluation. Referral should be considered for:

- Visual acuity worse than 20/40 in a child 3 to 5 years of age or worse than 20/30 in a child older than 5 years of age
- Greater than 2 lines difference in visual acuity between the eyes, indicating ametropia
- Abnormal alignment of the eyes, indicating strabismus
- Abnormal red reflex, indicating obstruction
- Unilateral ptosis or lesions obscuring the visual axis

Conclusion

Now let's go back to the case with Robert, the 4-year-old boy with a history of unilateral vision deficit. You proceed to explain amblyopia to Robert and his family, then refer Robert to an optometrist for correction of refractive error. You discuss with his family the potential for an ophthalmology referral if symptoms worsen. In the meantime, you decide to have the patient follow up with you in 2 months, to see whether his signs and symptoms have improved.

Take-home points

Let's review a few of the key take-home points:

1. Amblyopia is defined as a functional reduction in visual acuity, without abnormality of the retina and/or optic nerve, and can be classified into three types according to its causes: strabismic, refractive, or deprivational.
2. Vision screening for children under the age of 5 is recommended. Early detection and treatment of amblyopia improve the prognosis.
3. Amblyopia should be suspected, given unequal vision via the two eyes. In preverbal children, abnormal vision may be indicated by an abnormal fixation reflex or occlusion test. In older children, formal visual acuity testing with Snellen or tumbling E chart can be done.
4. Management of amblyopia involves penalizing input to visual cortex from the 'good' eye, while encouraging the formation of inputs from the 'weaker' eye, with either patching or cycloplegic drops.

Thank you for listening to our podcast on amblyopia. Shout out to Dr. William Stell for his support and expertise on this subject. Stay tuned for more podcasts to come!

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