Cataracts

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In Brief

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Author Disclosure

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Cataracts are a leading cause of preventable blindness, responsible for 5% to 20% of childhood blindness cases worldwide. In the United States, the prevalence of cataracts in children younger than 18 years of age ranges from 1 to 6 per 10,000. A cataract is an opacity of the lens and is considered visually significant when greater than 3 mm and centrally located. The World Health Organization (WHO) defines cataract as a cause of blindness in which sight can be restored through tertiary prevention. Provision of quality surgery within an optimal time frame is one of the WHO's targets for Vision 2020, a global initiative to eliminate preventable causes of blindness.

Cataracts may occur at any age. Approximately 50% of congenital cataracts are idiopathic; the remainder are due to congenital infections, teratogens, genetic or metabolic disease, and ocular disorders. Despite improved prenatal care and immunizations, congenital infections remain a major source of cataracts, particularly in developing nations. Although rubella is the most common infectious cause of congenital cataracts, other congenital infections, such as cytomegalovirus, varicella, herpes simplex, toxoplasmosis, and syphilis, may cause them. Teratogens, such as alcohol and corticosteroids, may also cause congenital cataracts.

Bilateral cataracts are more commonly identified with genetic disease, often related to autosomal dominant traits. Cataracts are associated with numerous genetic syndromes, the most common being trisomies 13, 18, and 21. Although less frequent, cataracts may also be seen in WAGR syndrome (**W**ilms tumor, **a**niridia, **g**enital abnormalities, **r**etardation), neurofibromatosis type 2, CHARGE (**c**oloboma, **h**eart defects, choanal **a**tresia, **r**etardation of growth, **g**enital abnormalities, **e**ar abnormalities) syndrome, Alport syndrome, Lowe syndrome, and Sturge-Weber syndrome. Unilateral cataracts are generally caused by primary ocular disorders in the absence of trauma. These abnormalities include posterior lenticonus, aniridia, microphthalmos, persistent hyperplastic primary vitreous, and anterior segment dysgenesis.

Multiple metabolic diseases are associated with cataracts, the most common being galactosemia. Cataracts in infants born with galactosemia can resolve if the infant is changed to a lactose-free formula soon after birth. Other metabolic conditions that cause cataracts are associated with hypoparathyroidism, hypoglycemia, and copper metabolism disorders. Hence, it is essential to perform an eye examination on children who exhibit failure to thrive and have suspected metabolic disease. After infancy, most cataracts occur from trauma, systemic disease (such as juvenile idiopathic arthritis), or therapeutic intervention (ie, corticosteroid use and radiation).

The major cause of cataract-associated blindness is deprivation amblyopia. There appears to be a latent period during which visual deprivation is reversible before final vision is affected. Therefore, timely identification and treatment of cataracts is essential. Optimally, surgical intervention for congenital cataracts should occur within 6 weeks of birth for unilateral cataracts and 10 weeks for bilateral cataracts. Unfortunately, this time frame often is not met. This short window for intervention makes it crucial for pediatric clinicians to identify and refer any chil-

dren who have abnormal eye findings promptly to a pediatric ophthalmologist. Studies have emphasized the importance of nursery screenings in timely diagnosis and treatment of cataracts. The nursery examination, combined with the American Academy of Pediatrics recommended newborn visit at 3 to 5 days, offers at least two opportunities for diagnosis within the recommended time frame for surgical intervention.

Physical findings that should prompt referral to an ophthalmologist include abnormal red reflex, structural abnormalities, or any irregularity or asymmetry of the pupils. Ideally, the red reflex examination should be performed in a darkened room, with each eye examined individually from a distance of 1 to 2 feet. Immediate referral is warranted for the findings of an absent red reflex, dark spots in the red reflex, or a white reflex. The Bruckner test should follow the red reflex examination. In this evaluation, both eyes are viewed simultaneously using an ophthalmoscope from a distance of 2 to 3 feet, allowing the viewer to note asymmetry of the red reflex that may suggest amblyopia.

After age 3 months, it is important also to test a child's ability to fix and follow objects. The presence of nystagmus or squinting warrants a referral. At 3 years of age, it is reasonable to begin testing visual acuity. In addition, parental observation of any ocular abnormalities and a family history of childhood cataracts are valuable determinants of an infant's risk.

In general, the operative outcome of congenital cataracts is good. Many children can achieve a visual acuity of 20/60. Complications of surgery include, but are not limited to, visual axis opacifications, glaucoma, retinal detachment, and strabismus. Children should have close follow-up evaluation by an ophthalmologist following surgery to ensure optimal visual outcome.

Comment: I still remember reading in college biology class about the experiments in which a cat's eye was sutured shut to identify the critical time for visual development and the resultant harm to visual acuity from light deprivation. Cataracts cause a similar light deprivation. Because the early identification and treatment of cataracts can prevent blindness, pediatricians have an incredibly important role in ensuring that an eye examination is performed in the newborn nursery and in the first postnatal week. At times, seeing the red reflex in a newborn can be a challenge, but strategies such as darkening the room or lifting an infant from a horizontal to vertical position result in the infant opening his or her eyes. Most causes of unilateral cataracts are idiopathic, but when the cataracts are bilateral, it is prudent to search further for a cause, whether genetic, metabolic, or infectious. Of interest, the first genetic mapping of an autosomal disease in humans was accomplished for congenital cataracts. Although congenital cataracts from rubella infection have decreased in frequency in developed countries, they still present a problem in countries where the measles, mumps, rubella vaccine is not universally administered. Researchers are investigating best practices for intraocular lenses and approaches to decrease the postoperative morbidities from cataract surgery, such as glaucoma and the inflammatory response.

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