Pediatric Cataracts

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Introduction

Congenital cataracts are responsible for nearly 10% of all visual loss in children worldwide, and it is estimated that 1 in 250 newborns in the United States has some form of cataract. Cataracts in children can be isolated or can be associated with systemic conditions including chromosomal abnormalities; craniofacial, mandibulofacial and skeletal syndromes; metabolic disorders; congenital infection; dermatologic, central nervous system, musculoskeletal or renal disease; or external factors such as trauma or radiation. In almost all cases of cataract associated with systemic disease, the cataracts will be bilateral (though not all bilateral cataracts are associated with systemic disease). Cataracts can also be associated with other ocular anomalies including persistent fetal vasculature, coloboma, anterior segment developmental anomaly, and aniridia.

Pediatric Cataracts

Onset and Inheritance

Pediatric cataracts can be congenital or acquired. Several inheritance patterns have been identified. When
inherited, familial cataracts are usually autosomal dominant and always bilateral but may be asymmetric. X-linked and autosomal recessive inheritances have been reported but are rare. Not all inherited cataracts are congenital.

**Morphology.** Cataracts can involve the entire lens (total or complete cataract) or can involve only part of the lens structure. The location in the lens and morphology of the cataract provides a great deal of information about its onset, etiology, and prognosis. Any portion of the lens anatomy can become opacified, leading to numerous unique phenotypic presentations, such as oil droplet cataracts seen in galactosemia and Christmas tree–like cataracts in myotonic dystrophy. The clinically most common and important morphologies of partial cataracts follow.

**Anterior polar cataracts (APCs).** These cataracts are common and appear as small white dots in the center of the anterior lens capsule (Figure 1). They are typically 1 mm in diameter but can be smaller or, rarely, larger. They are thought to be a remnant of persistent tunica vasculosa lentis. These opacities are usually not visually significant and are not expected to enlarge or progress, and therefore rarely require surgery. They are congenital, usually sporadic, and can be bilateral or unilateral. Anisometropia is common, so careful refraction and periodic follow-up are indicated.

**Nuclear cataracts.** Nuclear opacities involve the nucleus of the lens (Figure 2). They are typically approximately 3 mm in diameter, but the opacification or irregularity of the lens fibers can extend peripherally. The density of nuclear cataracts is variable and can progress. They can be unilateral or bilateral, and inherited or sporadic. Importantly, eyes harboring nuclear cataracts usually have some degree of microcornea. This is most readily apparent in unilateral cases. These eyes are at increased risk for developing aphakic glaucoma after cataract surgery, and these children need to be monitored carefully throughout life.

**Lamellar cataracts.** These cataracts can be identified by their discrete, round (lenticular) shape affecting one or more of the “rings” in the developing lens cortex. The opacities are larger in diameter than nuclear cataracts, typically 5 mm or more (Figure 3). They are always bilateral, but can be asymmetric in density. Whether symmetrical or asymmetrical, they are potentially amblyogenic. Lamellar opacities are usually acquired and can be...
Like nuclear cataracts, PFV is congenital, and these eyes are nearly always microphthalmic (microcornea) unless glaucoma occurs. Like posterior lenticonus, PFV is almost always unilateral. In severe cases, the lens may
be pushed forward, flattening the anterior chamber and causing secondary glaucoma.

**Posterior subcapsular cataracts (PSCs).** These cataracts are not common in children. When present, they are acquired, bilateral, and tend to be progressive. Secondary causes for the cataracts should be sought such as exogenous or endogenous steroids, uveitis, or retinal degeneration. PSCs can also be seen with delayed onset following radiation of ocular, orbital, or craniofacial tumors. This type of cataract can be seen with neurofibromatosis type II, which may be the first observable manifestation of this systemic disorder.

**Evaluation**

All newborns deserve screening eye examinations, which should include an evaluation of the red reflexes with a direct ophthalmoscope. This test, known as the illumination test, red reflex test, or Brückner test, can be used for routine ocular screening by nurses, pediatricians, and family practitioners. Ability to retinoscope through the child’s undilated pupil can help the ophthalmologist to estimate the visual significance of an axial lens opacity in a preverbal child.

**History.** The ophthalmologist should elicit a detailed history of the child’s growth, developmental milestones, feeding and digestive behavior, other developmental anomalies, skin lesions, and family history. A slit-lamp examination of immediate family members can reveal small, previously undiagnosed lens opacities that are visually insignificant but may reveal an inherited cause for the child’s cataracts.

**Visual Function.** The mere presence of a cataract does not imply that surgery to remove it is indicated. That determination requires assessment of the visual significance of the lens opacity.

In infants less than 2 months of age, a normal fixation reflex has yet to develop. Therefore the lack of strong fixation in an infant of this age with a cataract is not necessarily abnormal. In general, anterior capsular opacities are not visually significant unless they occlude the entire pupil, blocking out the red reflex. Central or posterior lens opacities of sufficient density that are greater than 3 mm in diameter are usually visually significant. Opacities that have a significant area of red reflex around them through an undilated pupil or opacities that have clear areas within them will frequently allow for good visual development in infancy and can be observed. Strabismus in unilateral cataract and nystagmus in bilateral cataracts are both late signs that the opacities are visually significant and the optimal time for treatment has passed, though surgery can still result in significant improvement.

In preverbal children greater than 3 months of age, standard clinical assessment of fixation behavior, fixation preference, and objection to occlusion provide additional evidence of visual significance of the cataract(s). Special tests such as preferential looking cards and visual evoked potentials can provide additional quantitative information, but they are generally not necessary to determine the visual significance of a cataract.

In school-age and older children, surgery for bilateral cataract should be suggested when the child’s level of visual function interferes with his or her visual needs. For instance, a child in kindergarten may function well with vision in the 20/70 to 20/100 range, while grade school and high school students have higher visual demands that require intervention at an earlier stage. For unilateral cataracts, vision that cannot be improved past the 20/50 to 20/70 range with optical and amblyopia treatment alone is a reasonable level to suggest cataract surgery.

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**Figure 6** Severe persistent fetal vasculature. Note 360° traction on the ciliary processes by the vascularized retrolental membrane. (Reproduced with permission from Raab EL, Basic and Clinical Science Course, Section 6. San Francisco: American Academy of Ophthalmology; 2010-2011. Courtesy of David A. Plager, MD.)
Ocular Examination. A slit-lamp examination should be performed to classify the morphology of the cataract and to identify any associated abnormalities of the cornea, iris, lens, and anterior chamber. Infants can be held in place at a conventional slit lamp, but a portable handheld slit lamp is a helpful substitute.

If the cataract allows some view of the posterior segment, careful observation of the optic disc, retina, and fovea should be obtained. If no view is present, B-scan ultrasonography can help rule out possible retinal and vitreal pathology.

Work-up. Unilateral cataracts are not usually associated with occult systemic or metabolic disease, and expensive laboratory tests are not warranted.

For bilateral cataracts, if a positive family history of infantile or childhood cataracts can be elicited or examination of the parents’ lenses shows congenital lens opacities, a systemic and laboratory evaluation can be obviated. A basic laboratory evaluation for bilateral cataracts of unknown etiology in apparently healthy children would include:

- Urine for reducing substances and amino acids
- TORCH (toxoplasmosis, rubella, cytomegalic inclusion disease, Herpes simplex) titers and VDRL screening (if not done already in a newborn screen)
- Blood for calcium, phosphorous, glucose, and perhaps red cell galactokinase levels

Any further work-up should be directed by other abnormalities in growth and development and input should be sought from a pediatric geneticist or developmental pediatrician.

Surgical Management

Considerations

Timing. The timing of a surgical intervention in the setting of visually significant pediatric cataracts depends not only on the age of onset of the opacity, but also upon whether or not the condition is unilateral or bilateral. The window of time during which visual deprivation does not induce irreversible harm to the visual system is known as the latent period. A critical period of visual development then follows, rendering a significant visual setback if the visual axis is not cleared.

For dense bilateral cataracts present at birth, a distinct latent period has remained difficult to quantify. Infants with visually significant opacities who undergo lensectomy and anterior vitrectomy at a point after 10 weeks of age have been shown to have an increased likelihood of best-corrected acuity of less than 20/100 when compared to infants whose surgery took place prior to 10 weeks.

Dense unilateral congenital cataracts have a more well-defined time frame by which the visual axis should be cleared to allow for maximum treatment effectiveness. Birch and Stager have shown a bilinear relationship between the age of the infant at surgery for unilateral cataracts and the ultimate visual outcome. Infants who undergo surgery prior to 6 weeks of age have a better visual potential than those whose surgery and optical rehabilitation are delayed, but very early surgery may increase the risk of aphakic glaucoma.

Wound Construction and Closure. Wound location and architecture, as well as the potential use of an IOL, depend on the age of the patient. Although some pediatric cataract surgeons prefer scleral tunnel incisions, the clear corneal approach has been shown to be equally safe and effective. The advantages of clear corneal incisions include preservation of conjunctival tissue in a population at risk for aphakic glaucoma, as well as increased maneuverability of instrumentation. Most pediatric cataract surgeons do not consider a pars plana/plicata approach to confer any benefit to offset the increased difficulty and potential posterior complications of that approach. Although a temporal approach can be used, the protection that the brow and an intact Bell’s phenomenon confer to children make the superior approach most common.

Regardless of approach, suturing the cataract incision with absorbable suture such as 10-0 Vicryl (polyglactin 910; Ethicon, Somerville, New Jersey) is highly recommended. Frequent eye-rubbing and rambunctious behavior postoperatively in children add to the potential for postoperative wound leak.

Capsular Staining Techniques. When desired, anterior capsular staining can be accomplished with fluorescein sodium, indocyanine green (ICG), and Vision Blue (0.1% trypan blue, DORC International, Zuidland, The Netherlands), but the former 2 options have been largely supplanted by the increasing popularity of the latter. A dye-enhanced anterior capsulorrhexis can prove valuable in numerous settings, such as complete (white) cataracts of infancy, as well as scenarios in which the status of the anterior capsule is poorly defined (traumatic cataract).

Anterior Capsule Management. The pediatric anterior capsule has a highly elastic quality that makes its management very different from capsules in adults. This elasticity is inherent at birth, and tends to decrease with
age. Scleral rigidity is also reduced in the pediatric eye, resulting in a positive vitreous pressure upon entering the eye. The lens is moved anteriorly into an already anatomically shallow anterior chamber, further stretching the anterior capsule.

Although a “can-opener” capsulotomy with a cystotome is a viable option, most experienced pediatric cataract surgeons prefer either continuous-tear capsulorrhexis or vitrectorhexis because they are less prone to radial tear. Vitrectorhexis is most advantageous in children younger than 2 years whose anterior capsules are particularly elastic. A vitrector handpiece is inserted via a 20-gauge incision, while an anterior chamber maintainer is inserted through a second paracentesis. The port is directed downward toward the capsule. Once the capsule is engaged, the handpiece is moved in a circular fashion using a rapid cutting rate to generate a capsular opening slightly smaller that of the intended optic if an IOL is planned (Figure 7).

Manual continuous curvilinear capsulorrhexis remains the gold standard for stability in pediatric cataract surgery; however, the elasticity of the anterior capsule makes the “run-away” capsulorrhexis a risk even in experienced hands. A capsular tear can be generated with a cystotome in a similar manner to adult cataract surgery, but the force vectors applied to the tear are different for children and adults. The more elastic the capsule, the closer toward 90° away from the intended direction of tear should the force be directed. Regrasping the flap frequently after a small progression of the capsulorrhexis is recommended (Figure 8). Modifications of this technique, such as 2-incision push-pull capsulorrhexis, have been developed to increase the reliability of completion of a manual capsulorrhexis.

**ONLINE VIDEO:**
Infant Capsulorrhexis, 1 min 07 sec

**Lens Aspiration.** The nucleus of a pediatric lens is soft but does not require phacoemulsification for removal. The cortex likewise is not rigid and maintains a strong adhesion to the lens capsule that surrounds it.

Depending on the patient’s age and plans for IOL implantation and posterior capsulotomy, a single-port irrigation/aspiration (I/A) handpiece may be used versus a small-incision bimanual technique. Children who will be unable to sit for a YAG capsulotomy within 2 years of IOL implantation, or whose capsule cannot be made clear, will require a posterior capsulotomy and anterior vitrectomy at the time of surgery. These children are perhaps best suited for the bimanual technique, where the anterior capsulotomy, lens aspiration, posterior capsulectomy, and anterior vitrectomy can be performed with the same instrument.

If possible, central lens material should be removed last, preventing any inadvertent anterior movement of the capsule and subsequent posterior capsular damage. This is especially true for posterior lenticous cataracts, in which a posterior capsular defect may be pre-existing.

**Aphakia Versus Pseudophakia.** The decision to implant an IOL versus leaving the child aphakic depends on many factors, including age of the child, laterality, cataract
type, eye size, family preference, and comorbidities such as uveitis. In general, IOLs have become the standard of care in children over age 1 year in the absence of other contraindications.

The role of IOL use in infants is more controversial. In addition to the technical considerations of operating on the small, soft eyes of infants, IOL implantation at less than 6 months has also been associated with a higher rate of reoperation in the first 6 postoperative months, often secondary to visual axis opacification. This is most often due to cortical proliferation and Elschnig pearl formation because the cataract surgery was done while the lens was still rapidly growing.

A large multicenter randomized clinical trial to evaluate the visual outcome of children with unilateral congenital cataract when treated with primary pseudophakia versus aphakia and contact lens correction completed enrollment in early 2009. The Infant Aphakia Treatment Study (IATS) will also look at postoperative complications as well as parental stress issues for both treatment modalities, with the goal of better defining the role of IOL implantation in infants. The earliest data from this study indicate equivalent grating visual acuity at 1 year of age between children treated with contact lenses and IOLs after cataract surgery performed prior to age 6 months. In addition, an increase in the rate of intraoperative complications and the need for additional intraocular surgeries were found in the IOL group. Continued follow-up on this patient population will further clarify the risks and benefits to IOL implantation in infants.

**Pseudophakic Options.** The gamut of IOL designs that is available for the adult cataract surgeon are also available when lens implantation is desired in children, although all IOL use in children is considered off-label. Probably the most popular lens implanted in children at this time is the hydrophobic acrylic lens. This soft, foldable IOL can be implanted through a clear corneal incision and has proven to be quite biocompatible in the pediatric eye; however, the broad haptics make this lens a poor choice for the ciliary sulcus.

Multifocal lens technology is not widely embraced in children. The potential advantages of the multifocal lens are lost in children due to continued growth of the eye and the resulting refractive changes that can occur well into the late teens. The disadvantage may be the reduction in contrast sensitivity in the diffractive IOLs and its effect on the developing visual system.

**Refractive Selection.** Surgeons have to consider numerous factors in selection of the appropriate IOL power including age of the child, refraction of the fellow eye and, if possible, family genetics. The myopic shift that occurs in pseudophakic children has been shown to follow a logarithmic regression curve similar to that seen in aphakic children, though the rate of myopic shift appears to be lessened when an IOL is implanted. The variability in refractive change decreases with age, allowing a more accurate prediction of postoperative refraction in older children. The refractive outcome of infants younger than 1 year undergoing IOL implantation is quite variable.

The knowledge of this myopic shift has directed most pediatric cataract surgeons toward planned hyperopia in the immediate postoperative period, allowing for a refractive error to approach emmetropia or minimal myopia in adulthood. Tables such as Figure 9, when used for bilateral cataracts or used in combination with the current refraction of the fellow eye in a unilateral cataract, provide a guideline for the amount of hyperopia desired postoperatively. Variability in refractive outcomes after IOL implantation in children has been demonstrated using all of the commonly used IOL calculation formulas, with increasing variability seen in the youngest and smallest eyes. Postoperatively, the residual hyperopia is corrected with spectacles or contact lenses, which can be adjusted as the refractive growth occurs.

**Posterior Capsule Management and Anterior Vitrectomy.** It is generally accepted that all infants and most toddlers should have a posterior capsulectomy performed at the time of cataract extraction (Figure 10). Although some variability exists among study populations, posterior capsules left intact in many pseudophakic children will develop visual axis opacification within 2 years. Therefore, if the child is felt to be of an age at surgery at

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**Figure 9** Table of suggested goal for immediate postoperative refraction according to age. (Reprinted from Plager DA, et al. Refractive change in pediatric pseudophakia: 6-year follow-up, *J Cat Ref Surg*, 2002;28:810-815.)
which he or she could sit for a YAG capsulotomy within 2 years, then leaving the capsule intact at the time of surgery is a reasonable option. Should the capsule opacify prematurely or the child not be able to tolerate a YAG capsulotomy, a capsulectomy and anterior vitrectomy via pars plana approach can be performed. Regardless of patient age, a posterior capsule that incorporates a dense plaque or is of compromised structural integrity, such as posterior lenticonus, needs to be cleared from the visual axis at the time of surgery.

The posterior capsulotomy can be performed from the limbal or pars plana route. In children left aphakic, the limbal route is the less invasive option, allowing the lens aspiration, posterior capsulotomy, and anterior vitrectomy to all be performed through the same 20-gauge incision. For planned pseudophakia, a pars plana approach after the IOL has been implanted allows for wider access to the posterior capsule and vitreous, making it the preferred approach for many pediatric cataract surgeons.

The location of the pars plana entry wound varies according to patient age. Due to the incomplete development of the pars plana at birth, the sclerotomy must be placed closer to the limbus at younger ages. A distance of less than 2 mm posterior from the limbus in infants less than 1 year of age, 2 to 2.5 mm between 1 and 4 years, and up to 3 mm when older than 4 years have been recommended. An anterior vitrectomy of up to one-third of the vitreous should help decrease secondary obscuration of the visual axis.

Subconjunctival antibiotic and/or steroid injections are optional, as is a hard shield placed over the eye at the end of surgery.

Postoperative Management

After examination on the first postoperative day, a broad spectrum topical antibiotic is prescribed for use 3 to 4 times each day and can be given in the form of a combination steroid-antibiotic compound. Given the robust postoperative inflammation seen in pediatric eyes when an IOL has been implanted, prednisolone 1% should be started at least 4 times per day and titrated to the degree of inflammation that is encountered in the first postoperative weeks. Oral prednisone may sometimes be required, and a dilating agent such as atropine 1% ophthalmic solution can be added to prevent formation of synechiae.

Complications

Surgical Complications. Tears in the posterior capsule can occur at any stage of the procedure, compounded by the fact that inherent defects in the capsule are not infrequent in children, as is seen in posterior lenticonus cataracts. A small rent in the posterior capsule can be rounded by a vitrector, thus eliminating the need for YAG capsulotomy in the future.

The pars plana approach to posterior capsulotomy and anterior vitrectomy is associated with its own set of potential surgical complications, though the rate is very low. A sclerotomy site that is positioned excessively posteriorly can cause posterior segment complications such as retinal detachment or vitreous hemorrhage. Trauma to the capsular bag can occur during creation of the sclerotomy if the microvitreoretinal blade is not inserted in an adequately posterior direction.

Postoperative Complications. The increased tissue reactivity of the pediatric eye predisposes to an increased inflammatory response in the early postoperative period, inducing posterior synechiae, pigmented and fibrous deposits on the IOL, and secondary fibrin membrane formation. Risk factors include microphthalmos and age less than 6 months at the time of surgery when associated with IOL implantation.

Secondary opacification of the visual axis due to proliferation of lens epithelial cells can also occur and is common in infants when an IOL is placed. This opacification is rarer in infants who are left aphakic or in older children even when an IOL is placed (Figure 11).

Aphakic or pseudophakic glaucoma remains a postoperative risk for many children undergoing lensectomy, and the etiology of this condition continues to be poorly understood. The incidence of aphakic glaucoma has been reported to be as high as 41%, with an onset of disease averaging from 3.1 years to 6.8 years after lensectomy.
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Disparity and awkwardness of wearing one aphakic lens, they are a less desirable option for monocular aphakia.

Aphakic contact lenses are a more popular option for the correction of aphakia in infants. Extended-wear silicone lenses such as the Silsoft (Bausch and Lomb, Rochester, New York) contact lens are probably the most common choice of pediatric cataract surgeons, though some prefer rigid gas-permeable lenses. Keratometric values are not required to fit infants with the Silsoft lens, and 1 to 2 diopters of astigmatism can be masked by their wear. These lenses can be worn for 1-week durations and are available in powers ranging from +12 to +32 diopters. Contact lens power can be adjusted easily in the office by retinoscopic over-refraction. Aphakic contact lenses can be a more expensive option than an intraocular lens or glasses for visual rehabilitation in part, because frequent lens loss and power changes, particularly in the first year of wear, are common.

Hyperopic spectacle correction is the typical treatment of residual refractive error for both unilateral and bilateral pseudophakia, but in cases of unilateral pseudophakia with large residual refractive errors (greater than +8D), contact lens wear can be considered.

In children younger than 2 years, either aphakic or pseudophakic refractive correction should be purposefully overcorrected by 2 to 3 diopters, allowing the near point to be placed roughly at the child’s fingertips or a slightly greater distance. With toddlers and children more than 2 years of age, refractive correction should be targeted to optimize visual acuity at distance, with a +3.00 bifocal for near tasks.

Amblyopia Therapy

Amblyopia is frequently present in aphakic and pseudophakic children, both as a result of the preoperative deprivation and the postoperative anisometropia in cases of unilateral cataracts. Poor compliance with aphakic spectacles or contact lenses as well as inaccurate refraction in the office further delay or prohibit the patient from obtaining the best visual outcome possible. Part-time patching therapy of the sound eye in unilateral cataract is almost always necessary, but it is frequently employed at some point in the visual rehabilitation of bilaterally affected patients as well. The only role for pharmacologic amblyopia therapy is in the setting of unilateral aphakia when the sound eye is significantly hyperopic, thus inducing a more significant amount of visual distortion than what would occur in a low hyperope or myope.

Visual Rehabilitation

Correction of Refractive Error

The child who is left aphakic may be treated with aphakic glasses or contact lenses. Aphakic spectacles are effective in bilateral aphakia, but due to issues of retinal image disparity and awkwardness of wearing one aphakic lens, they are a less desirable option for monocular aphakia.

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Secondary IOL Implantation

As aphakic children grow older, they sometimes become intolerant of contact lenses or tire of wearing aphakic spectacles. If the eye is adequate size to contain an IOL, secondary IOL implantation may be an alternative. The most important factor in determining the site of implantation within the eye is the degree of capsular support that remains from the primary lensectomy. A preoperative dilated exam may reveal an adequate capsular shelf, on which a secondary IOL can be placed in the ciliary sulcus, but sometimes examination of the sulcus intraoperatively is required before this determination can be made.

If an adequate capsular shelf exists, an IOL can be placed in the sulcus or occasionally in the capsular bag. The broad haptics and lack of angulation of the single-piece foldable hydrophobic acrylic lenses, which work so well for in-the-bag implantation, make this lens unsuitable for sulcus fixation due to the possibility of prolonged iris chafe and chronic uveitis. For some children, especially those operated on in early infancy who have a well-developed Soemmering ring, secondary in-the-bag placement of the IOL is an option.

Eyes that do not have adequate capsular remnants for sulcus fixation need to have a suture-fixated posterior chamber IOL (PCIOL) or placement of an anterior chamber IOL (ACIOL). Neither alternative is ideal, especially for children who have many decades ahead of them in which to develop long-term complications such as suture breakage and dislocation of the PCIOL, or uveitis, glaucoma, and hyphema in association with now-outdated rigid, closed-loop ACIOLs. Newer open-loop ACIOL designs may improve their long-term safety, but no long-term data are yet available to support this. The aphakic iris claw lens seems to have a better long-term record in children, but it is not currently available in the United States.

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**Conclusion**

The diagnosis of pediatric cataracts was once considered a diagnosis strictly associated with poor visual acuity; however, current diagnostic and therapeutic options for the pediatric cataract surgeon have allowed for greatly improved outcomes when employed in a timely fashion. The approach for each individual varies with the age of the patient and morphology of the cataract. Forthcoming studies, such as the IATS, will further ophthalmologists’ ability to best treat these children and minimize vision loss from this condition.

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Clinicians’ Corner

Clinicians’ Corner provides additional viewpoints on the subject covered in this issue of Focal Points. Consultants have been invited by the Editorial Review Board to respond to questions posed by the Academy’s Practicing Ophthalmologists Advisory Committee for Education. While the advisory committee reviews the modules, consultants respond without reading the module or one another’s responses. –Ed.

1. What laboratory work-up do you recommend when you see bilateral cataracts?

Dr. Lambert: If there is a family history of hereditary cataracts, I generally do not perform a laboratory work-up. In all other children, I perform laboratory testing tailored to their particular clinical findings. The laboratory test that has had the highest yield for me is urine amino acid testing for male infants with bilateral cataracts. The urine amino acids have been elevated in 3 babies I have screened. None of the infants were known to have developmental problems at the time of cataract surgery. On further testing, all of them were found to have Lowe syndrome (oculocerebrorenal syndrome). Early diagnosis is important because of the associated developmental problems and proximal renal tubular dysfunction that needs to be managed by a nephrologist.

I also routinely screen children with cataracts for serum calcium and phosphorus. I screen some infants for galactosemia with a comprehensive galactosemia panel that tests for galactose-1-P-uridytransferase (GALT) enzyme activity, Gal-1 P concentrations, and common GALT mutations. I also screen children with lamellar cataracts for galactokinase enzyme activity.

I do not routinely screen all children for rubella antibody titers because of the high rate of rubella vaccination in the United States, but I do screen infants with mothers who have immigrated to the United States from countries with high rates of congenital rubella syndrome.

Dr. VanderVeen: For bilateral infantile cataracts with no family history of pediatric cataract, most importantly, I consult with the child’s pediatrician to be sure that a complete physical examination has been performed and that there are no signs of other anomalies or systemic disease. If there are any dysmorphic features or additional findings, a genetics consultation is obtained. The
Clinicians’ Corner

child must be cleared for general anesthesia by the pediatrician, and I usually suggest a preoperative chest X-ray, which would detect an unrecognized cardiomyopathy. Laboratory testing, if not already done, may include ToRCHS (toxoplasmosis, rubella, cytomegalic inclusion disease, Herpes simplex, and syphilis) titers and calcium, phosphorus, and glucose levels in the blood. Testing for galactokinase deficiency may be considered if the cataract appears typical for this disorder or if there are other concerns about diet or growth. However, laboratory tests are almost universally unrevealing in our population, and I do not order them routinely.

2. What types of pediatric cataracts are amenable to medical management?

Dr. Lambert: Unilateral anterior polar cataracts only rarely need surgical treatment. However, some of these children develop anisometropic amblyopia, and for this reason they should be followed on a regular basis during early childhood. On the other hand, bilateral anterior polar cataracts are usually larger and may be associated with miosis. I treat some of these children with chronic pharmacological mydriasis. Unilateral posterior polar cataracts can sometimes be managed with part-time patching of the fellow eye until it becomes clear how much they are degrading vision. Likewise, some children with lamellar cataracts can be followed until they are older if the vision is carefully monitored.

Dr. VanderVeen: Small or partial cataracts may be managed medically. A general rule is that if the view to the fovea through the undilated pupil is clear using direct ophthalmoscopy, or if retinoscopy is possible, then the cataract is not (yet) visually significant. Anterior opacities, partial or scattered lens opacities with clear intervening lens, or central opacities <3mm (as long as the pupil is >3mm) can often be watched. Larger or posterior opacities are more amblyogenic. Cataracts are often associated with refractive error or strabismus, so these additional amblyogenic factors must be addressed in addition to deciding about surgical intervention.

3. How often is intraocular bleeding a problem in children with cataracts related to persistent fetal vasculature (PFV)? If you see a stalk from the posterior lens surface to the optic disc on ultrasonography, do you recommend that the surgery be performed in conjunction with a retinal specialist?

Dr. Lambert: On rare occasion, I have had a hyaloid vessel bleed during cataract surgery, resulting in a vitreous hemorrhage. For this reason, if there is a patent hyaloid vessel, I cauterize it with intraocular diathermy prior to extirpating it from the retrolenticular plaque. If the PFV is associated with nonattachment of the retina or the retina cannot be visualized, I refer these patients to a vitreoretinal surgeon for their management. I don’t base this decision on whether a stalk can be seen by ultrasonography, but on the severity of the PFV and the likelihood of retinal involvement.

Dr. VanderVeen: If the stalk is attenuated, there is usually very little bleeding, particularly when the vessel is cut transversely. Persistent pupillary vessels likewise bleed transiently and minimally. The hyaloid vessel or vessels within a PFV plaque can be cauterized prior to cutting in order to avoid significant intraocular bleeding. If the ultrasound or visualization shows a thick hyaloid stalk, or if there is any evidence of optic nerve or peripapillary retinal distortion, then I would refer the patient to a retinal specialist.

4. What is your success rate in terms of visual outcome for children with monocular cataracts who are operated upon within the first month of life versus after 1 month?

Dr. Lambert: I no longer operate on children with monocular cataracts during the first month of life, because in my experience this seems to increase their risk of developing glaucoma. The literature suggests that the visual results are just as good if the surgery is performed when children are 4 to 6 weeks of age versus <4 weeks of age. If the child is compliant with his or her optical correction (contact lenses or glasses if pseudophakic) and patching therapy, my experience is that he or she will generally have an excellent visual result.

Dr. VanderVeen: While there is a suggestion that visual outcomes may be slightly improved by surgery before 4 weeks, there may also be a slightly higher rate of complications. Therefore, when an infant presents...
with congenital cataract(s), I generally plan for surgery when the child is about 1 month old, with the youngest patients being 3+ weeks of age at the time of surgery. I have 10 to 15 eyes that were operated on at less than 4 weeks, and all others (>150) were 4 weeks or older, so it is really difficult to really compare the outcomes. Overall, we have found that microcornea and PFV are risk factors for glaucoma or other secondary complications, rather than younger age at surgery.

5. What are your guidelines for implantation of an IOL at different ages?

Dr. Lambert: I almost always recommend IOL implantation in children 7 months of age or older. For infants 6 months of age or younger with bilateral cataracts, I prefer to leave them aphakic and optically correct them with either contact lenses or spectacles. When they are older I implant secondary IOLs if the parents are having problems with the contact lenses or they are bothered by the cosmesis of aphakic spectacles. For infants 6 months of age or younger with a unilateral cataract, I discuss the pros and cons of IOL implantation with the parents. I explain to them that while the visual outcome associated with IOL and contact lens correction are comparable, IOL implantation is associated with an increased incidence of intraoperative complications and additional intracorneal surgeries to clear visual axis opacities. I then let the parents choose the treatment.

Dr. VanderVeen: I do not generally offer IOL implantation for infants less than 6 months of age, but I do explain the options of an aphakic contact lens versus IOL for those 6 to 12 months of age. Because the parents will have to deal with the contact lens or spectacle wear, and often patching after surgery, I find that active participation in decision-making is better for most families. The parents tend to be happy with whichever choice they make as long as the advantages and disadvantages of each option are explained prior to surgical intervention. Infants more than 1 year of age generally receive primary IOL implantation.

6. Do you have any experience with anterior chamber IOLs (ACIOLs)? Do you advise their use in children?

Dr. Lambert: I do not implant IOLs in the anterior chamber of children. I have examined a few children who had IOLs implanted in their anterior segments by other surgeons and these children seem to be doing okay, but I’m concerned about the long-term effects of ACIOLs on the corneal endothelium and angle of a child who may live for another 70 or 80 years.

Dr. VanderVeen: I have not implanted an ACIOl in a child and have preferred sutured posterior chamber lenses when there is insufficient capsular support for in-the-bag or sulcus implantation. Newer iris-supported lenses may be a reasonable choice, though few have been implanted in pediatric eyes and long-term effects are not yet studied.

7. What is your preferred method for secondary IOL implantation in an eye without adequate capsular support? Do you perform pars plana vitrectomy in these cases?

Dr. Lambert: I have sutured IOLs in the sulcus in the past, but I have not been happy with the results. One of the Prolene sutures suspending the IOL broke in one patient and the IOLs became tilted in other patients. We have an excellent contact lens service at Emory, and in most cases we have been able to successfully fit aphakic children with contact lenses even when they were deemed “contact lens intolerant” by others.

Dr. VanderVeen: Fortunately most children with acquired aphakia have reasonable capsular support so that in-the-bag or sulcus implantation can be accomplished. If there is inadequate support, I would opt for a scleral-fixated, sutured IOL, with previously described standard techniques. I usually create scleral flaps, use 9-0 Prolene suture, and choose an IOL with an eyelet for the suture. Vitrectomy should be performed, as needed, and I use an anterior approach.

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