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# Infantile Cataracts

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## ■ Introduction

Infantile cataracts are a significant cause of treatable blindness in infants worldwide, accounting for 12% to 15% of visually impaired children in countries as diverse as Finland and India.<sup>1,2</sup> This places an enormous socioeconomic burden, especially on the already strained resources of developing countries.<sup>2,3</sup> Population-based studies have estimated the prevalence between 1 and 13 cases per 10,000 births.<sup>4,5</sup> Most are congenital, but some cataracts may not manifest at birth, but in the first year, making the term infantile cataracts more inclusive. They are evaluated and treated in the same manner. Diagnosing and managing congenital cataracts can be challenging and requires appropriate timing of surgery, amblyopia treatment, refractive correction, and vigilance for the higher incidence of postoperative complications.

## ■ Embryology

Knowledge of the anatomy of the lens is useful in understanding the classification schemes of congenital cataracts. Thickening of the surface ectodermal cells overlying the optic vesicle forms the lens placode or plate at 28 days of gestation. At 29 days gestation, a central depression in the lens plate appears, called the lens pit. Eventually, the central stalk, which connected the lens pit to the surface ectoderm, disappears and the resultant sphere is a single layer of cuboidal cells encased within a basement membrane (lens vesicle). The primary lens fibers elongate

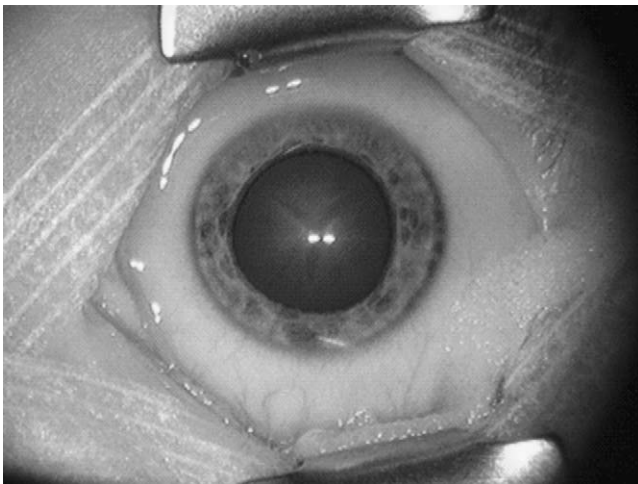
from the posterior surface of the lens vesicle obliterating the lumen. These fibers ultimately form the embryonic nucleus. At 7 weeks gestation, the lens epithelial cells multiply and elongate forming the secondary lens fibers, which comprise the fetal nucleus. The interdigitation of the lens fibers creates the erect Y suture anteriorly and inverted Y suture posteriorly. The lens is nourished by the tunica vasculosa lentis and its remnants can sometimes be seen in healthy adult eyes.<sup>6</sup>

## ■ Morphology

Congenital cataracts can be classified according to morphologic type and often, visual prognosis may vary in accordance with type.<sup>7</sup> Central cataracts include nuclear, lamellar, cortical, sutural, pulverulent, cerulean, and coralliform cataracts. Polar cataracts can either be anterior (anterior polar, anterior pyramidal, and anterior subcapsular) or posterior [posterior subcapsular, posterior lenticonus, posterior fetal vascular (PFV)].

### ***Nuclear Cataracts***

Nuclear cataracts are usually present at birth and those that are visually significant are 3 to 4 mm in size.<sup>8</sup> The area of opacification is located between the anterior and posterior Y sutures in the embryonic and fetal nucleus. The cataracts are bilateral in two-thirds to 80% of cases and many affected eyes are microphthalmic.<sup>9</sup> Bilateral nuclear cataracts represent the most frequent morphology for autosomal dominant inherited cataracts.



**Figure 1.** *Infant with lamellar cataract.*

### **Lamellar Cataracts**

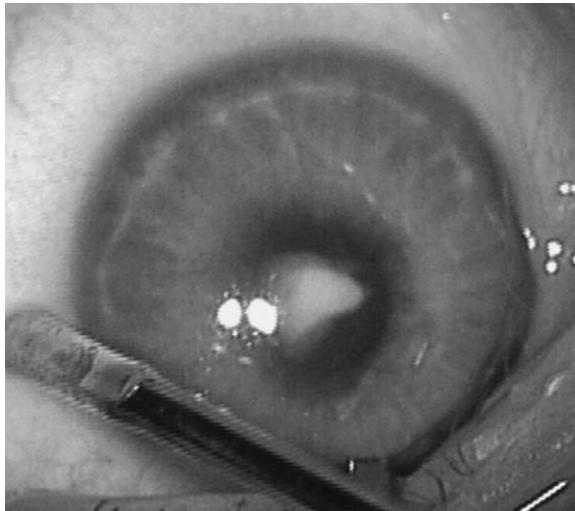
Lamellar cataracts are characterized by opacification of the lamellae surrounding the fetal nucleus and usually carry a better prognosis than other types.<sup>10</sup> They result from an insult to secondary lens fibers during their most metabolically active period.<sup>11</sup> The degree and meridian of opacification can vary greatly. Usually, lamellar cataracts are bilateral, though asymmetry with significant interocular differences can lead to amblyopia. Development is progressive and at a variable rate, and as there is usually an early period of lens clarity, the potential for good vision has already developed. Surgery can be deferred in cases with mild opacities until early childhood.<sup>11</sup> When lamellar cataracts require surgery, they are associated with better long-term visual outcome than other types of cataracts<sup>7</sup> (Fig. 1).

### **Sutural**

Sutural cataracts are opacifications involving the sutures of the lens. They are typically bilateral and stationary and do not impair vision when they are an isolated finding.<sup>8</sup>

### **Pulverulent**

Multiple tiny dots in the embryonic nucleus characterize pulverulent cataracts. They are also autosomal dominant, nonprogressive, and rarely affect vision. When surgery is required, the prognosis of pulverulent cataracts is better than many other types of inherited cataracts.<sup>12</sup>



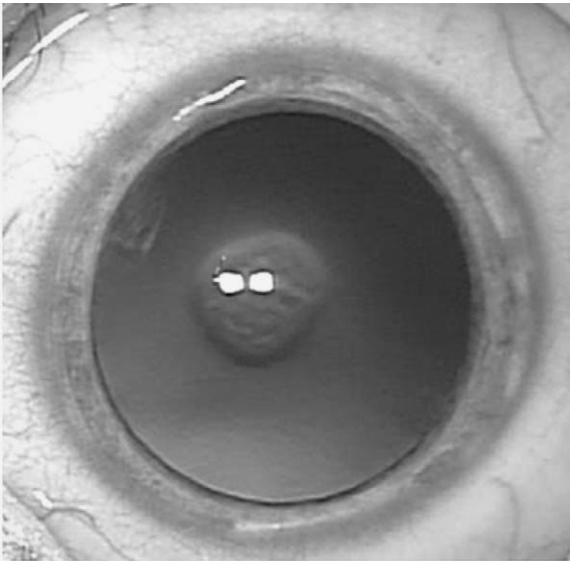
**Figure 2.** *Infant with anterior pyramidal cataract with projection of cataract into anterior chamber.*

### **Cerulean**

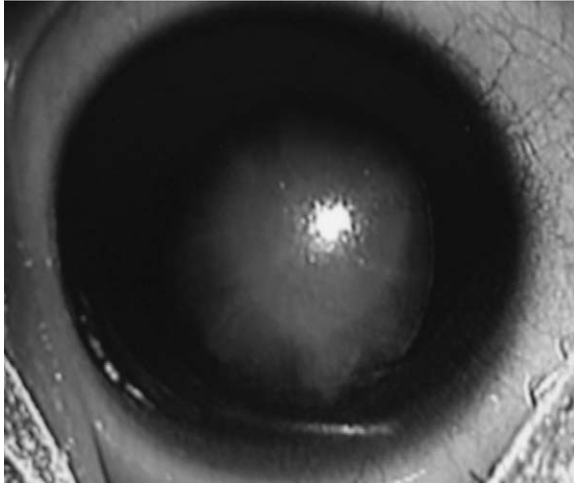
Cerulean cataracts are progressive, bilateral cataracts, which consist of bluish-white opacities scattered through the superficial concentric layers of the fetal and adult nucleus of the lens. Although they are congenital, they do not usually affect vision until adulthood, at which point surgical intervention becomes necessary.<sup>13</sup>

### **Polar Cataracts**

Anterior polar cataracts are dotlike opacities on the anterior surface of the lens. These cataracts can be bilateral or unilateral and are stationary.<sup>14</sup> They are frequently diagnosed by the pediatrician or parent as they can be visualized without a slit lamp.<sup>8</sup> It has often been cited that these cataracts are visually insignificant, however, they can cause significant amblyopia and strabismus, necessitating close observation and ultimately surgery.<sup>15</sup> Anterior pyramidal cataracts occur when conical opacities project from the anterior capsule into the anterior chamber and may represent a more severe form of anterior polar cataracts (Fig. 2). They are bilateral, but asymmetric, and can also cause significant amblyopia owing to the size and asymmetry of the opacities.<sup>16</sup> Over one-quarter of patients with anterior lens opacities can have amblyopia, usually secondary to anisometropia.<sup>17</sup> Progress to surgery may be insidious, occurring between 9 and 47 months after initial presentation.<sup>15</sup>



**Figure 3.** *Child with posterior lenticular type cataract.*



**Figure 4.** *Infant with total cataract, completely obstructing the visual axis.*

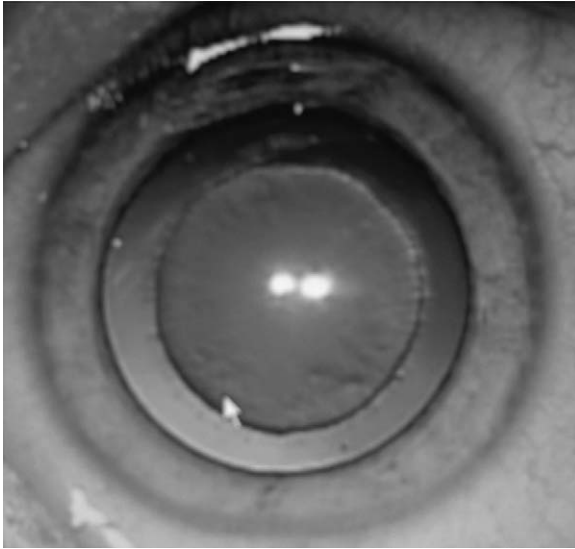
Posterior cataracts also have a wide range of severity and presentations. Some, such as Mittendorf dot (anterior end of hyaloid artery) do not require surgical intervention. Posterior lenticonus is weakness of the central part of the posterior lens capsule, which can be accompanied by cataractous change in the lens cortex<sup>18</sup> (Fig. 3). Persistent fetal vasculature can cause a unilateral cataract owing to abnormal regression of the primary vitreous resulting in a range of ocular abnormalities including microphthalmia, retrolental plaque, persistent hyaloid vessel, and tunica vasculosa remnants.<sup>19</sup> PFV cataracts are associated with worse visual acuity outcomes than other types of unilateral cataracts.<sup>7</sup> Alexandrakis and colleagues<sup>20</sup> found that only 50% of patients achieve vision better than 20/400 after surgery.

### **Total**

These cataracts include complete opacification of the nucleus and cortex. They can be secondary to metabolic disorders, trauma, syndromic, or familial. Surgical intervention is almost always necessary to maximize visual potential (Fig. 4).

### ■ **Etiology**

Congenital cataracts may be caused by intrauterine infections, associated syndromes, or ocular abnormalities or genetic factors.



**Figure 5.** *Infant with autosomal dominant cataract.*

### **Genetics**

Congenital cataract was the first autosomal disease to be genetically mapped in humans.<sup>21</sup> A genetic basis is much more common in bilateral than unilateral cataracts, accounting for over one-quarter of children with isolated bilateral congenital cataracts and only 2% of unilateral cases.<sup>22</sup> Autosomal dominant inheritance is the most common (Fig. 5), though autosomal recessive and X-linked forms have also been isolated.<sup>23–25</sup> A substantial amount of genetic heterogeneity exists in inherited cataracts, both locus and allelic. Loci on chromosomes 1, 2, 3, 6, 9, 10, 13, 12, 14, 16, 17, 19, 20, 21, and 22 have been identified in association with recessive and dominant cataracts.<sup>26</sup> The  $\alpha$ ,  $\beta$ , and  $\gamma$  crystallin genes are the water soluble proteins expressed in lens, which play a critical role in maintaining lens clarity.<sup>27</sup> Fifteen different mutations in crystalline genes have been reported in association with childhood cataract.<sup>25</sup>  $\alpha$  crystallin mutations have been found in a family with congenital lamellar cataracts and zonular nuclear cataracts<sup>28,29</sup> and are also associated with posterior polar cataracts.<sup>30,31</sup> Mutations in  $\beta$  crystallin gene (CRYBB2 on chromosome 22) can produce highly variable phenotypes with the same mutation causing different types of cataracts, indicating the influence of environmental factors on cataract type.<sup>32</sup> Pulverulent, cerulean, nuclear, and sutural cataracts have all been associated with  $\beta$  crystalline gene mutations.<sup>33</sup> The  $\gamma$  crystallin genes (mainly CRYGC and CRYGD) are responsible for many autosomal dominant cataracts—including pulverulent, aceuliform, cerulean, and

lamellar cataracts.<sup>34</sup> Mutations in membrane transport protein (MIP), which control the hydration and transparency of the lens, have also been found in genetic cataracts.<sup>35</sup> Intercellular channel proteins present in gap junction, called connexins, also play a major role in lens clarity. Connexin 46 and 50 genes have been isolated, through linkage analysis in multigenerational families, with autosomal dominant pulverulent cataracts.<sup>36</sup> Developmental regulator genes, PITX3, MAF, and HSF4, which coordinate the development of the lens have also been implicated in cataracts.<sup>37</sup>

### **Infections**

Congenital cataracts have been documented after neonatal infection with varicella, herpes simplex, toxoplasmosis, rubella, and syphilis (TORCHS).<sup>38,39</sup> Before the age of rubella vaccination in the United States, congenital rubella syndrome (CRS) accounted for a significant number of congenital cataracts. In a 30-year prospective study of 34 patients with CRS diagnosed during the rubella epidemic of 1963, 85% suffered from cataracts, and 63% were bilateral.<sup>40</sup> CRS is still a major problem in developing countries and cataracts have been found in as many as 90% of seropositive patients.<sup>41</sup> Fetal infection is most common when the mother is infected during the first trimester and can cause sensorineural deafness, mental retardation, jaundice, microcephaly, glaucoma, and pigmentary retinopathy.

### **Metabolic**

Numerous metabolic deficiencies are associated with congenital cataract formation. One of the most recognized associations is the “oil droplet” cataract with galactosemia. Affected infants possess a central opacity best viewed with retroillumination. Individuals who are deficient in galactose-1-phosphate uridylyltransferase present with jaundice, hepatosplenomegaly, failure to thrive, and cataracts, whereas infants with galactokinase deficiency only present with cataracts.<sup>42</sup> Hypoparathyroidism classically causes multicolor fleck cataracts, mannosidosis has been associated with vacuoles, and hypoglycemia causes lamellar cataracts.<sup>43,44</sup> Disorders in copper metabolism have also been identified in conjunction with congenital cataracts.<sup>45</sup>

### **Ocular Disorders**

Ocular abnormalities account for 2% of bilateral cataracts and 10% of unilateral cataracts.<sup>6</sup> Aniridia, microphthalmos, and anterior segment dysgenesis have all been associated with congenital cataracts.<sup>46,47</sup> Both PFV and posterior lenticonus (discussed above) are developmental abnormalities that typically cause unilateral congenital cataracts.

## **Syndromes**

Trisomy 21, has a well-documented association with bilateral cataracts, occurring in 13% to 20% of affected patients. However, the onset of cataracts does not typically occur before the age of 12 years, with only 1.4% of patients being diagnosed in the neonatal period.<sup>48,49</sup> The classic presentation is the cerulean cataract which is not visually significant until the teen years; however, nuclear, cortical, and mixed presentations have also been described. Trisomy 13, Patau syndrome, is characterized by severe cardiac and nervous system defects, which cause death during the neonatal period. Ocular findings include inferonasal iris colobomas with sectoral cataracts in the same quadrant, associated with PFV.<sup>50</sup> Lowe syndrome, or oculocerebrorenal syndrome, is an X-linked disorder of mental retardation, renal tubular acidosis, glaucoma, and bilateral cataracts. Cataracts present early with a mean age of cataract extraction usually around 1 month. Even with early surgical removal, vision is not better than 20/70 and nystagmus often develops.<sup>51</sup>

## **■ Systemic Evaluation**

Unilateral cataracts do not require an extensive systemic work-up, as most are not inherited nor associated with other syndromes. A complete ocular examination is usually sufficient to evaluate for specific causes such as PFV, poster lenticonus, or anterior segment dysgenesis. Bilateral cataracts with no family ocular history or recognizable syndrome should be further evaluated as 60% of them can be attributed to a specific etiology. A detailed family history should be undertaken to elicit autosomal dominant or recessive inheritance, and the parents examined to look for undiagnosed mild cataracts that could be hereditary in nature. Careful attention should be paid to the infant's generalized medical issues. Hepatomegaly, failure to thrive, jaundice, and diarrhea indicate a need to test the urine for reducing substance after milk feeding to evaluate for galactosemia. Metabolic acidosis, congenital glaucoma, and mental retardation would point to Lowe syndrome and the urine should be tested for amino acids. Laboratory screening for TORCHS titers is also often helpful, if this was not already done during the mother's pregnancy. Recognizable physical appearance as with Down syndrome or Patau syndrome provides a quick determination as to etiology of cataracts in some patients.

## **■ Medical Management**

Some congenital cataracts can be managed medically if they do not seem to be visually significant enough to require cataract extraction.

Posterior opacities, such as posterior lenticonus, tend not to require early surgical intervention and can be successfully managed by treating the associated amblyopia. Sixty-seven percent of patients with small posterior lens opacities and lenticonus achieved vision better than 20/40 with only patching and glasses.<sup>52</sup> Alternatives include pharmacologic dilation with cyclopentolate 1% or phenylephrine 2.5% to provide a larger area for viewing around small, dense cataracts. Good vision has been reported when long-term dilation was employed in conjunction with amblyopia therapy.<sup>53</sup>

## ■ Surgical Indications and Timing

Cataract surgery is undertaken when a visually significant cataract exceeds 3 mm in diameter, prevents examination of the fundus, or is associated with strabismus or nystagmus.<sup>54</sup> Surgical timing is crucial in achieving a successful outcome in an infant with cataracts. Deprivational amblyopia is a therapeutic challenge that can prevent good postoperative acuity, especially more so in unilateral cataracts than bilateral cataracts because of ocular rivalry between the cataractous eye and the normal fellow eye. For this reason, it is important to operate on infants before the sensitive period at which point irreversible vision loss can occur from even minor disruptions in visual input.<sup>55</sup> A latent period seems to exist early in the neonatal period in which disturbances to vision do not have a lasting effect.<sup>56,57</sup> For unilateral cataracts, the critical period for surgical intervention is believed to be 6 weeks, with potential for equal outcomes if surgery was performed at any point during the first 6 weeks of life. Infants who undergo surgery in the first 6 weeks of life have been documented to have average visual acuities of 20/40 or better.<sup>58</sup> Cataract surgery after 6 weeks of age is associated with worse contrast sensitivity and vernier outcomes.<sup>58</sup> This latent period is less well characterized for bilateral cataracts. Poorer visual outcomes increase when surgical intervention is delayed past 10 weeks of age and preoperative nystagmus was correlated with worse visual acuity as well.<sup>59</sup>

## ■ Pediatric Cataract Surgery

Cataract surgery in infants requires several modifications from adult surgery. A standard clear corneal incision can be used with minimal risk of postoperative astigmatism.<sup>60</sup> However, some pediatric ophthalmologists prefer a scleral tunnel incision, which reduces corneal scarring, and may be especially useful for intraocular lens (IOL) insertion when a pars plana incision for vitrectomy is planned. All incisions should be sutured given the low scleral rigidity of the pediatric eye, which promotes leakage.

The anterior lens capsule of the infant is much more elastic and requires greater force to initiate a tear than that of an adult. Techniques for creating a capsulotomy in infants include manual continuous curvilinear capsulorrhexis (CCC), can opener capsulotomy, a “push-pull” capsulotomy (Nischal), mechanized capsulotomy (vitrectorhexis), diathermy capsulotomy, and Fugo plasma blade capsulotomy. The use of capsular dyes such as indocyanine green and trypan blue can aid in the visualization of the anterior lens capsule in cases of dense white congenital cataracts.<sup>61</sup> CCC remains the “gold standard” for capsulotomy and most ophthalmologists will employ this technique in children older than 2 years of age. CCC creates the cleanest edge and is most resistant to tearing when evaluated in porcine eyes, however, the positive vitreous pressure and elasticity of the capsule makes it a difficult technique to master. Using high molecular weight viscoelastics to flatten the lens capsule, planning for an opening smaller than 5 mm, and generating vector forces toward the center of the rhexis, can assist in completing a CCC in young patients.<sup>62</sup> Vitrectorhexis is preferable in children younger than 2 years, in whom a CCC is challenging owing to the highly elastic nature of the lens capsule. With a cut rate of 150, the vitrector instrument is introduced into the eye through the limbal opening, engages the capsule with aspiration, and then cuts, creating several overlapping openings, which coalesce into a round opening. The push and pull technique for anterior capsulorrhexis uses 2 stab incisions 5-mm apart outlining the diameter of the rhexis. The proximal flap is pushed toward the distal incision and the distal flap is pulled toward the proximal incision. Retrospective data over a 5-year period, showed that this technique can produce a reliable CCC in older patients (mean age of 70 mo).<sup>63</sup> The Kloti diathermy uses high-frequency current and creates a thermal capsulotomy. The Fugo blade plasma knife is a portable system, which cuts using a thin filament charged by a plasma cloud. It is able to create a base almost as smooth as CCC when examined by scanning electron microscopy.<sup>64</sup>

The pediatric cataract is much softer than in adults and phacoemulsification power is not necessary for lens removal. Cataract extraction can be accomplished with simple aspiration using either an ocutome or bimanual irrigation-aspiration handpieces. An anterior chamber maintainer can be used instead of the irrigation handpiece for continuous irrigation.

Management of the posterior capsule is extremely important in pediatric cataract surgery. Infants have high rates of posterior capsule opacification and the amblyogenic effect is more profound at younger ages as well.<sup>65</sup> Parks was the first to introduce the concept of a posterior continuous curvilinear capsulorhexis to prevent visual axis obstruction after congenital cataract surgery.<sup>66</sup> The posterior capsule can be removed either via a limbal or pars plana approach, at the time of cataract surgery

with a mechanized vitrector or a capsulorhexis, or postoperatively with the neodymium:YAG laser. The ideal opening in the posterior capsule is approximately 3.5 to 4 mm in size, slightly smaller than the anterior capsulotomy.<sup>67</sup> When a primary posterior capsulotomy and vitrectomy is not performed, especially in children younger than 6, development of a posterior capsule opacity is almost certain with rates of PCO development ranging from 43% to 100%.<sup>63,67,68</sup> Most surgeons advocate combining posterior capsulotomy with anterior vitrectomy because the anterior vitreous can serve as a scaffold for the migration and proliferation of lens epithelial cells, causing opacification even if the posterior capsule itself has been removed. Rates of secondary cataract are much lower when a posterior capsulotomy is combined with anterior vitrectomy, especially in young children.<sup>68</sup> Younger age at time of surgery is also associated with earlier development of PCO and the need for repeated interventions for PCO.<sup>69</sup> As a result, posterior capsulotomy and anterior vitrectomy is recommended in all infants and children under age 5 (to 6) years, irrespective of whether an IOL is being implanted.<sup>70</sup>

Advances in IOL technology have made primary implantation a viable option in young children. Primary IOL implantation presents the advantage of full time compliance with partial correction in patients that may have difficulty with full compliance. In correcting unilateral congenital cataracts, IOL implantation may offer better visual outcome than infants treated with contact lenses, especially when contact lens compliance is moderate to poor.<sup>71</sup> However, IOL implantation in infants is controversial and is associated with higher rate of postoperative complications and reoperations (mainly membranectomies).<sup>69</sup> Currently, the preferred choice for IOL material is a hydrophobic acrylic lenses, such as the 3 piece or 1 piece AcrySof IOL (Alcon, Inc Ft Worth, TX). The ability to inject the lens through a smaller wound, lower rates of posterior capsule opacity and associated inflammation make it an attractive choice in the pediatric patient population.<sup>72,73</sup>

Predicting the refractive change in children over time from infancy to adulthood presents another challenge in IOL implantation. A child's eye undergoes a significant amount of growth including elongation of the axial length and flattening of the corneal curvature, especially in the first 6 months of life. This growth creates a substantial refractive change postoperatively.<sup>74</sup> Therefore, most surgeons aim to under correct infants, by choosing an IOL power that will leave the child hyperopic, with the goal of future emmetropia in adulthood. For example, in the Infant Aphakia Treatment Study, infants who are randomized to primary IOL implantation have a targeted postoperative refraction of +8 D for those operated on at <49 days and +6 D for those operated on at 49 to 210 days. The pediatric IOL calculator may be of some use in determining the average rate of refractive change based on age at surgery, axial length, and keratometry measures.<sup>75</sup> Some have suggested

that implantation of an IOL may itself increase postoperative myopia,<sup>76</sup> though this has been negated by more recent studies.<sup>77</sup> Proper selection of an IOL requires use of an accurate IOL calculation formula. IOL formulas are not specifically designed for the smaller anterior chamber depth, increased corneal curvature, and shorter axial length of the pediatric patient and their reliability has been questioned. A comparison of the SRK II, SRK T, Holladay I, and Hoffer Q showed that the Hoffer Q tends to overestimate IOL power especially in children younger than 24 months. However, there was considerable variability in the refractive outcomes using all of the formulas, with the SRK II formula giving the least amount of variability overall.<sup>78</sup>

### ■ Aphakic Correction

Aphakic correction includes contact lenses, aphakic spectacles, and secondary IOL implantation. Aphakic glasses are not optimal for the correction of monocular aphakes because of the induced aniseikonia; however, they are appropriate for bilateral aphakia. A bifocal segment can be added to the glasses when the child begins to require better vision at near distances, at 2 to 3 years of age. Aphakic spectacles have the disadvantage of poorer optics and cosmesis. Contact lenses can either be silicone lenses, which have high oxygen permeability and can be worn for an extended period of time or rigid gas permeable lenses, which are useful for very steep corneas. Contact lenses are usually well tolerated and the power can be changed frequently to match the continuously changing refractive error of the infant eye. When compliance with contact lenses is good, contact lenses achieve the same postoperative visual acuity in monocular cataract patients as IOLs, even in infants operated upon before 6 months of age.<sup>71,79</sup> If a patient is noncompliant or intolerant of contact lenses and spectacles, a secondary IOL can be implanted. Either a PMMA IOL or a 3 piece foldable IOL such as an AcrySof MA60BM (Alcon Inc, Fort Worth, TX) is placed within the reopened capsular bag or ciliary sulcus. Foldable IOLs have higher rates of decentration when placed in the sulcus as compared with PMMA lenses, though they have a higher success of achieving “in the bag” fixation than PMMA.<sup>80</sup>

### ■ Amblyopia Management

Dense congenital cataracts result in stimulus form deprivational amblyopia, which can be extremely difficult to treat. Postoperative visual outcome depends upon the timely management of amblyopia, especially in monocular cataracts. Vision in unilateral pseudophakes/aphakes is further complicated by ocular rivalry as the fellow eye has a clear retinal image without the need for refractive correction. Usually, patching

treatment of the fellow eye is necessary after cataract extraction in addition to optical correction. Patching compliance is often difficult to achieve in children 18 to 30 months of age and those parents who are able to maintain some form of patching treatment during this age, even for an hour a day, have better long-term results. Compliance with occlusion is very strongly associated with postoperative visual outcome in bilateral and unilateral cataracts, though even more so in unilateral disease.<sup>81</sup> Analysis of monocular cataract patients has found that mean visual acuities of patients who were compliant with patching treatment (>75% patching hours prescribed) were 20/50 as compared with 20/1000 of noncompliant patients.<sup>82</sup>

## ■ Postoperative Complications

Visual axis opacifications or secondary membranes can occur in pediatric patients even if a posterior capsulotomy and anterior vitrectomy are performed at the time of cataract surgery. Reproliferation of cortical lens material occurs universally, usually in the form of Soemmering ring, but occasionally Elschnig pearls or lens material may occlude the visual axis. With a mean follow-up of 76 months, Hosal and Biglan<sup>83</sup> report that 22.5% of eyes that underwent cataract surgery with posterior capsulectomy and anterior vitrectomy suffered from secondary membrane formation. Secondary membrane formation was more common in eyes with an IOL and in younger patients, with surgery before the age of 1 year increasing the risk of membrane formation by 4.74.<sup>65,83</sup> For infants, the rate of patients requiring surgery for secondary visual axis opacification was 70% for pseudophakes and was 0 in aphakes.<sup>79</sup> Restoration of the visual axis can be accomplished by membranectomy/pars plana vitrectomy.

Infants have a more robust inflammatory response than adults resulting in IOL deposits and greater posterior synechiae. In a prospective, nonrandomized, nonmasked trial, Rumelt and colleagues<sup>84</sup> proposed that the addition of low molecular weight heparin to the irrigation solution at the time of surgery decreases inflammation and fibrin in the anterior chamber, though they implanted PMMA IOLs instead of acrylic ones. In a double-masked, randomized trial, intracameral tissue plasminogen activator was found to prevent fibrin formation and pigmented IOL deposits after congenital cataract surgery, though several cases were complicated by hyphema.<sup>85</sup> Further randomized studies are necessary to determine if enoxaparin or tissue plasminogen activator would be valuable in congenital cataract surgery.

Aphakic glaucoma occurs more commonly in patients who have undergone cataract surgery during the first year of life, presenting in 5% to 41% of aphakic eyes. Diagnostic clues include an increase in axial length, corneal diameter, or myopic refractive shift. Initially, studies

seemed to support the idea that IOL implantation was somehow protective of the development of glaucoma. Asrani and Wilensky et al<sup>86</sup> found that pseudophakes had lower rates of glaucoma (0.3%) compared with their aphakic counterparts (11.3%). These results are now thought to be secondary to selection bias as eyes that were “unhealthier” (ie, microcornea, PFV, or other ocular abnormalities) were left aphakic. A retrospective review of 267 pseudophakic eyes and 47 aphakic eyes performed by Trivedi and coworkers<sup>87</sup> found no significant intergroup difference in the incidence of glaucoma. They did find that all cases of postoperative glaucoma occurred in patients who were operated upon before the age of 4.5 months.<sup>87</sup>

Retinal detachment is an uncommon complication of pediatric cataract surgery, which can occur many years after surgery. A retrospective review of 1017 aphakic eyes evaluated rates of retinal detachment when a limbal approach for cataract extraction was used. They found that 3.2% of patients developed a retinal detachment at a mean interval 6.8 years after surgery.<sup>88</sup> Aphakic patients are not at an increased rate of retinal detachment after postcapsulotomy/PAV or secondary membrane surgery.

## ■ Conclusions

Congenital cataracts represent a diverse spectrum of morphologies, etiologies, and clinical presentations. Determination of the visual significance of congenital cataracts depends upon measurements of the size of opacity, cycloplegic refraction, and assessment of the red reflex. Infantile cataract surgery requires several modifications from adult surgery. Surgical technique, choice of aphakic correction, and postoperative complications highly depend upon age of the patient and must be considered in the pediatric patient.

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