Chapter 24
IOL Placement in Developmentally Delayed Patients

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Introduction

Although cataracts in the pediatric population are typically idiopathic, they can also often occur in association with systemic disease. Common systemic diseases and syndromes associated with cataracts include Down syndrome, Lowe syndrome, galactosemia, intrauterine infection including TORCHS infections, Sturge-Weber syndrome, and Nance-Horan syndrome [1, 2]. Children with these disorders commonly have developmental delay, and management of patients with cataracts and developmental delay poses unique challenges at all phases of care. Preoperative assessment is frequently limited, and patients with systemic abnormalities or developmental delay have higher reported rates of complications after cataract surgery [3] and poorer visual outcomes [4].

Preoperative Assessment

The preoperative assessment of patients with developmental delay and cataracts is uniquely challenging. Assessment of visual acuity is often limited by nonverbal status and lack of sustained attention or cooperation. When optotype visual acuity cannot be obtained, preferential-looking tests such as Teller or Cardiff acuity testing should be used to assess visual acuity [5–7]. If cooperation is inadequate to perform a preferential-looking test, then assessment of fixation preference using the induced tropia test can assess for amblyopia [8, 9]. Pattern visual evoked potential can also

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assess for abnormal vision in patients with congenital cataracts [10]. Other clinical findings such as nystagmus or monocular strabismus in the cataractous eye may also provide evidence for the visual significance of a cataract.

In patients where a cataract is suspected, examination of the lens using an upright or handheld slit lamp and a retinoscope should be performed with careful attention to the location and size of the opacity. A central opacity of greater than 3 mm places the patient at higher risk for visual impairment [11, 12]. Cycloplegic refraction should be performed given the high incidence of astigmatism and anisometropia associated with lenticular abnormalities, particularly anterior polar cataracts [13]. In some cases, examination under anesthesia may be required in order to determine if a cataract is visually significant enough to warrant extraction.

Non-surgical Management

The etiology of visual impairment in children with cataracts may have multiple components including direct obstruction of the visual axis, refractive error, and amblyopia, which may be deprivational, anisometropic, refractive, and/or strabismic. In cases of partial cataract where the visual significance is unclear, non-surgical management including spectacle correction, part-time occlusion, and pharmacologic dilation should be attempted [11, 14, 15]. Close follow-up with frequent reassessment of visual function is necessary. For developmentally delayed patients, tolerance of glasses or patching should be assessed preoperatively as this will play a role in intraocular lens (IOL) selection. In cases where there is poor adherence to prescribed treatment, the surgeon may consider adjusting the postoperative refractive target in order to minimize initial refractive error, which may be amblyogenic if uncorrected.

Surgical Planning: Placement of an Intraocular Lens

If the decision is made to perform cataract extraction in a patient with developmental delay, several surgical considerations should be reviewed prior to surgery and discussed with the patient's parents or legal guardian.

One of the most important decisions when planning pediatric cataract extraction is whether to leave the eye aphakic, requiring contact lens correction (see Chap. 11 "Contact Lenses") or aphakic glasses (see Chap. 12 "Aphakic Glasses"), or to implant an IOL. The Infant Aphakia Treatment Study found similar visual acuities, but higher rates of adverse events and additional surgeries in infants aged 0–6 months who received an IOL as compared to those who were left aphakic [16]. Thus the recommendation from the study was that primary IOL implantation in infants <7 months of age should only be performed when contact lens wear might be especially challenging, risking periods of uncorrected aphakia [12]. Patients with developmental delay frequently fall into this category, and thus IOL implantation may be considered in this population at a younger age. Primary IOL implantation in

children has increasingly become more common in children over 1 year old, and for children who were initially left aphakic, secondary IOL implantation can be considered as early as 2–4 years of age when the rate of growth of the eye has slowed or aphakic contact lens or spectacle correction has become more difficult [12].

If an IOL is implanted in a child with developmental delay, postoperative refractive target must be carefully considered. Because growth of the eye during childhood typically results in myopic shift, many established recommendations target hyperopia in order to avoid significant myopia in the long term [17, 18]. However, in patients with poor compliance with spectacle correction, initial high hyperopia creates the risk of uncorrected refractive error and, in cases of unilateral pseudophakia, anisometropia. If there is poor compliance with spectacle correction and amblyopia treatment, then refractory or worsened amblyopia may result. Poor compliance with patching has been reported as the factor most strongly associated with poor visual acuity 7 years following cataract surgery in children [19]. Although published studies vary as to how important postoperative refraction is to visual outcomes [20, 21], if the patient's parents or guardians have significant concerns about compliance with spectacle correction or patching prior to surgery, then a refractive error closer to emmetropia should be considered in order to minimize the risk of amblyopia. If significant myopia or anisometropia develops later, then possible strategies may include contact lens correction, IOL exchange [22], corneal refractive surgery, piggyback intraocular lens, or implantable contact lens [23].

Intraoperative Technique

Patients with developmental delay undergoing cataract surgery are at higher risk for postoperative complications [3] and worse visual outcomes [4]. Published rates of surgical complications in children with Down syndrome are higher in non-ophthalmic surgery as well [24, 25]. Furthermore, self-injurious behavior in developmentally delayed patients also poses risks of ophthalmic injuries [26]. Administration of postoperative topical eye drops can be more difficult for caregivers of patients with developmental delay. Certain intraoperative techniques can help to minimize the risk of postoperative complications.

Scleral tunnel incisions should be considered particularly if a larger incision is required, as in the case of intraocular lens implantation (Fig. 24.1). With a scleral tunnel incision, if there is wound leak or dehiscence, there will not be direct communication between the anterior segment and the external environment. Scleral tunnel incisions have been found to have a lower risk of endophthalmitis than corneal wounds following adult cataract surgery [27, 28]. All wounds, including paracenteses, in pediatric cataract surgery should be sutured to minimize risk of wound leak [29]. Scleral wounds, if covered by conjunctiva, may be sutured with non-absorbable 10-0 Nylon suture or absorbable 8-0 or 9-0 Vicryl® (polyglactin, Ethicon, Johnson & Johnson, USA) (Fig. 24.1D). Corneal wounds should be closed with absorbable suture such as 10-0 Vicryl® in order to avoid the need for suture removal under anesthesia postoperatively (Fig. 24.2).

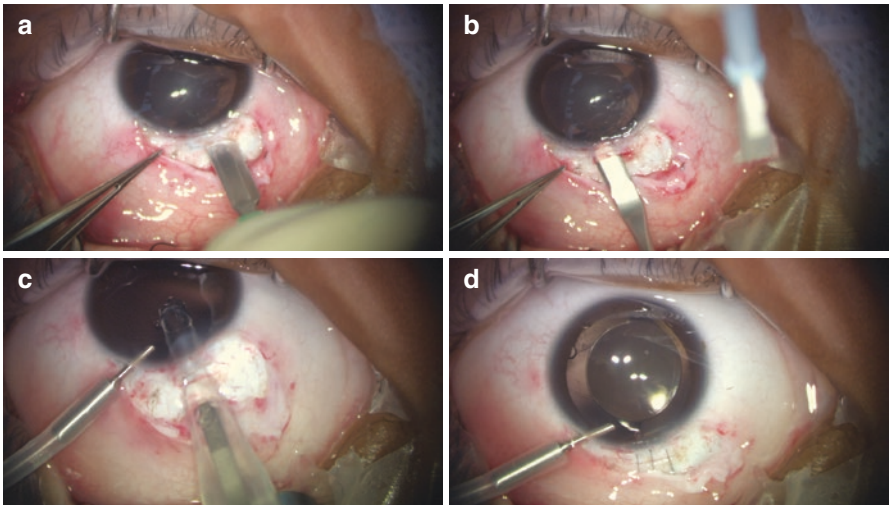

Fig. 24.1 Scleral tunnel incision. In a patient undergoing cataract extraction and IOL implantation, a 6 mm conjunctival peritomy is made (not pictured). A rounded tip blade is then used to make a 4-mm-wide partial-thickness incision into the sclera (**a**), after which a tunneling blade is used to tunnel into the peripheral cornea (**b**). After removal of the cataract, a keratome is used to open the wound into the anterior chamber (not pictured) and the IOL is injected into the anterior chamber (**c**). The wound is sutured with three interrupted 10-0 Nylon sutures (**d**)

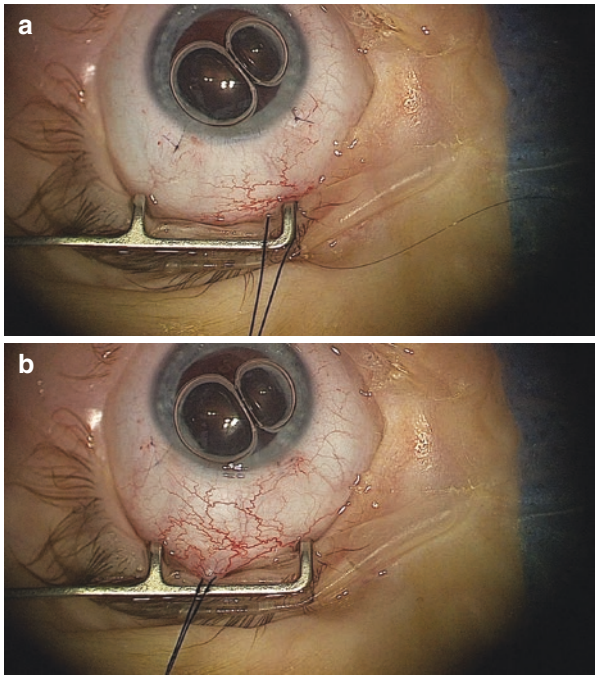

Fig. 24.2 Sutured limbal incisions. In a patient left aphakic, the limbal wounds are closed with 9-0 Vicryl sutures (**a**). Small conjunctival peritomies are closed over the wounds to minimize suture and wound exposure and improve comfort postoperatively (**b**). A filtered air bubble in the anterior chamber is used to improve anterior chamber stability and facilitate postoperative examination (**a**, **b**)

Intracameral or periocular medication delivery is especially valuable in developmentally delayed patients who may have difficulty with topical eye drop administration postoperatively. Intracameral triamcinolone in pediatric cataract surgery has been reported to reduce postoperative inflammation and visual axis obscuration without any increased rate of complication or intraocular pressure increase [30–34]. Intracameral cefuroxime [35, 36] and moxifloxacin [37, 38] have both been reported to be safe and effective in the reduction of post-cataract endophthalmitis in adults and, depending on availability, should be strongly considered in developmentally delayed children undergoing cataract surgery. Though data is limited regarding rates of endophthalmitis in patients with developmental delay, endophthalmitis remains a concern especially in patients who may be prone to eye rubbing or other high-risk behaviors. Postoperative endophthalmitis, while rare following intraocular surgery in children [39], can be a devastating, vision-threatening complication.

A small bubble of filtered air injected with a cannula into the anterior chamber at the end of surgery helps to promote anterior chamber stability (Fig. 24.2) [40]. Additionally, in patients who are uncooperative with examination on the first postoperative day, an air bubble is an easily visible indicator that the anterior chamber is formed and that the surgical wounds are not leaking significantly.

Postoperative Care

In the postoperative period, efforts should be directed toward avoiding trauma to the eye, such as rubbing and self-injurious behavior. In children with self-injurious behavior, a multidisciplinary approach is necessary in order to minimize the risk of postoperative injury or complication. Strategies can include behavioral modification strategies, as well as safety devices such as soft goggles and elbow restraints [26, 41]. Severely delayed or self-injurious children may require inpatient admission for sedation and observation during the critical few days immediately following surgery.

Immediately Sequential Bilateral Cataract Surgery

For some patients with bilateral cataracts and developmental delay, there may be significant concern about the medical risks of two episodes of general anesthesia within a short time frame. If such patients are at higher risk of anesthesia-related complications due to their comorbid systemic conditions, immediate sequential bilateral cataract surgery (ISBCS) can be considered [12, 42, 43]. It should be noted that there is a lack of consensus regarding the risks, benefits, and role for ISBCS. In particular, risks of potentially blinding bilateral complications such as endophthalmitis, toxic anterior segment syndrome, and expulsive hemorrhage must be considered. Thus, an extensive discussion with the anesthesiologist and caregivers is of

critical importance. If ISBCS is performed, extensive precautions in aseptic technique, including separate instrumentation, surgical prep, and surgeon sterility, should be implemented [12, 44].

Case 1

A 15-month-old male presented with intermittent crossing of the right eye for 1 month. His past medical history was significant for autism and developmental delay. He was nonverbal. On exam, he was able to perform Cardiff preferential-looking test, and his right eye has an acuity of 20/160 and his left eye 20/40. He has an intermittent right esotropia of 16 prism diopters with a strong left eye fixation preference. Portable slit lamp examination revealed a posterior capsular speckled opacity with mild obscuration of red reflex in the central 3 mm of the right eye. Left lens was clear. Cycloplegic refraction was +2.00 sphere in the right eye and +2.50 sphere in the left.

Due to the multiple possible mechanisms for amblyopia and esotropia, after discussion with the parents, the decision was made to attempt non-surgical management initially with patching of the left eye 4 hours per day. At 6-week follow-up, Cardiff visual acuity was unchanged. The right esotropia was now constant. The central red reflex in the right eye was more dull than it was previously. The decision was made to proceed with cataract extraction in the right eye. The parents expressed concern about contact lens placement due to the patient's developmental delay and intolerance of patching, and they elected to have an IOL implanted.

Cataract extraction with IOL implantation was performed on the right eye. A hydrophobic single-piece acrylic IOL was inserted through a superior scleral tunnel incision into the capsular bag with initial postoperative refractive target of +2.00. Posterior capsulotomy and anterior vitrectomy were performed via a pars plana incision. Prior to closing, cefuroxime and triamcinolone were injected intracamerally. The scleral tunnel was sutured with 10-0 Nylon, and the remaining incisions were closed with 9-0 Vicryl®.

Comment This case illustrates a number of the difficulties in managing cataracts in children with developmental delay. At the patient's initial visit, the patient had a lenticular opacity of borderline size for visual significance, and it was not immediately apparent whether the strabismus was secondary to the cataract or an independent contributory factor to the patient's amblyopia. Therefore, non-surgical management was initially prescribed. On follow-up, progression of the cataract, worsening of the control of the esotropia, and lack of improvement in vision all provided evidence for the visual significance of the cataract, and thus cataract extraction was recommended to the parents.

A thorough discussion with the patient's parents was essential to surgical planning. Because of concerns about aphakic contact lens insertion, the decision was made to implant an IOL. A postoperative refractive target of +2.00 was chosen in

order to minimize anisometropic amblyopia if the patient did not tolerate glasses or patching, with the parents understanding that he may become significantly myopic in the future.

Intraoperatively, the decisions to use a scleral tunnel (as opposed to a clear corneal incision), suture all wounds, and inject intracameral cefuroxime were intended to minimize risk of endophthalmitis, and intracameral triamcinolone was used to help control intraocular inflammation given concerns about compliance with drop administration postoperatively in a developmentally delayed child.

Case 2

A 17-year-old female with CHARGE syndrome presents for her first eye examination in several years. Her past ocular history includes myopia, bilateral iris and inferior retinal colobomas, and chronic total retinal detachment left eye previously deemed inoperable. Her medical history is notable for CHARGE syndrome, cardiac valvular abnormalities, and developmental delay. She is nonverbal.

She was able to do a Teller preferential-looking test and her right eye was 20/100. The left eye had no light perception. On slit lamp and fundus examination, her right eye had an inferior iris coloboma and 3+ posterior subcapsular cataract. There was an inferior retinal coloboma not involving the macula, with no retinal detachment. The left eye had an inferior iris coloboma; fundus exam confirmed total retinal detachment. Cycloplegic refraction in the right eye was −15.00 sphere; the left eye had a poor red reflex.

Cataract extraction with IOL implantation was recommended to the family because of the visually significant cataract in the right eye. At home, the patient does not wear glasses and frequently uses an electronic tablet for communication. Outside the home, she does wear glasses and enjoys looking at distant objects. Therefore, it was decided to target a postoperative refraction of −2.00. Cataract extraction was performed in similar fashion as in Case 1. However, because of her history of retinal detachment in the fellow eye, the posterior capsule was left intact, and no vitrectomy was performed.

Postoperatively, refraction in the right eye was −2.25. Teller visual acuity was 20/40. The parents found the child to have improved visual alertness around the house.

Comment In this case, the visual significance of the cataract in the right eye was clear. Due to the patient's monocular status and developmental delay, an extensive discussion with the patient's parents regarding the risks, benefits, and alternatives of surgery was essential. As in Case 1, a clear understanding of the patient's activities of daily living and tolerance of glasses assisted in the preoperative planning. Because the patient did not wear glasses at home and performed frequent near work with an electronic tablet, the postoperative refractive target was −2.00, with plans to prescribe myopic spectacle correction for distance when the patient is outside the

house. Because of the patient's older age, concern for postoperative visual axis obscuration was lower. Therefore, in light of the patient's inferior retinal coloboma and history of retinal detachment in the fellow eye, the decision was made to not perform posterior capsulotomy or anterior vitrectomy in order to minimize any potential risk of retinal detachment in the patient's only seeing eye.

The management of cataracts and IOL placement in children with developmental delay poses numerous difficulties. Preoperatively, cooperation and examination are frequently limited; postoperatively, complication rates may be higher, and adherence to treatment may be poor. Thorough and careful examination, extensive discussion with the patient's parents or guardians, thoughtful preoperative planning, and the adoption of certain intraoperative techniques can increase the likelihood of positive outcomes in this challenging patient population.

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