Chapter 23
Pediatric Cataract Surgery in the Abnormal Anterior Segment

Benjamin Jastrzembski and Asim Ali

An abnormal anterior segment can create many difficulties in pediatric cataract surgery. Issues most commonly encountered include corneal opacities, iris abnormalities, large or small anterior segments, and eyes with prior corneal or glaucoma surgery. This chapter will describe techniques to address the most common scenarios in the above categories.

Corneal Opacities

Corneal opacities seen in association with a number of conditions pose a challenge to the pediatric cataract surgeon. Causes of corneal opacification that are common in the context of pediatric cataract include acquired causes such as traumatic or infectious corneal scars, failed corneal grafts, and congenital causes such as Peters anomaly and endothelial corneal dystrophies. It may be beneficial to perform keratoplasty prior to cataract surgery or perform the procedures at the same time (triple procedure). Such procedures will typically be performed by a corneal surgeon, and the details are out of the scope of this discussion. However there may be situations where it is desirable to avoid keratoplasty due to a high risk of graft rejection/failure or when it is prudent to observe the visual results of cataract surgery first. In such situations, it is helpful to have techniques to improve visualization in the anterior chamber.

B. Jastrzembski
Hospital for Sick Children, University of Toronto, Toronto, ON, Canada

Boston Children's Hospital, Harvard Medical School, Boston, MA, USA
e-mail: Benjamin.Jastrzembski@childrens.harvard.edu

A. Ali (✉)
Hospital for Sick Children, University of Toronto, Toronto, ON, Canada
e-mail: asim.ali@sickkids.ca

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Adjusting the patient positioning and the tilt of the operating microscope before the case may improve the surgical view in many of these cases. Some operating microscopes also allow adjustment of the intensity of oblique field and coaxial illumination in order to improve visualization during different surgical steps. This feature may also be useful when there is an underlying poor red reflex from vitreous abnormalities, such as a tumor and vitreous hemorrhage, or in a vitrectomized eye. With focal opacities or scars, simple rotation of the eye with grasping forceps at the limbus can improve visualization of instruments in the anterior chamber. Parallax induced by such movements also helps in the judgment of the depth of an otherwise obscured feature in the anterior chamber. Trypan blue dye improves visualization of the capsule but may also worsen the view in the context of a poor corneal endothelium as it stains damaged endothelial cells [1]. It should be used judiciously, with the smallest volume needed to produce good contrast. Staining of the endothelium is minimized with the use of an injection of an air bubble or viscoelastic first, but in the latter technique, care needs to be taken to make sure that trypan blue contacts the lens capsule directly or else there will be poor staining. Triamcinolone is used to visualize any vitreous in the anterior chamber, but its use should also be judicious as a large volume of particulate matter can worsen the surgeon's view. During capsulorrhexis, the oblique field illumination may be decreased to improve the red reflex (especially in eyes with prior vitrectomy or compromised zonules).

A very helpful adjunct in pediatric cataract cases with corneal opacities is the use of an endoilluminator, which is a tool commonly used in vitreoretinal surgery (see Case 2). These endoilluminators can come standard in vitrector sets and are especially useful for junior surgeons. They can provide oblique illumination if held near the limbus outside the eye or more effectively can be used inside the eye to tangentially illuminate the field, forming shadows that enhance depth perception [2]. This may require an additional limbal incision. It is helpful to decrease the illumination of the operating microscope when the endoilluminator is used to reduce the light scatter off the opacity back to the surgeon.

Case 1: Pediatric Cataract Surgery Following Penetrating Keratoplasty

A 15-year-old girl with a history of bilateral Peters anomaly with bilateral sequential penetrating keratoplasties at age 2-3 months presented with decreased vision in her right eye. Preoperative examination indicated visual acuity of hand motions in the right eye, horizontal nystagmus, normal intraocular pressure in both eyes, clear corneal grafts, and a white cataract in the right eye. A preoperative B-scan ultrasound demonstrated an attached retina in the right eye. The corneal diameter was 10.5 mm. The family and patient wished to proceed with cataract surgery in the right eye. During the surgery, microscissors were first used to cut iridocorneal adhesions, and then the synechiae between the iris and lens were carefully dissected using micrograspers (Fig. 23.1). In addition to trypan blue for improved visualization, microscissors were needed to complete the capsulorrhexis due to capsular

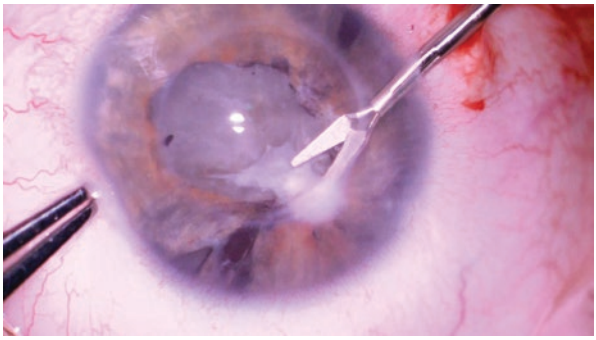

Fig. 23.1 Use of microscissors to cut iridocorneal adhesions during cataract surgery

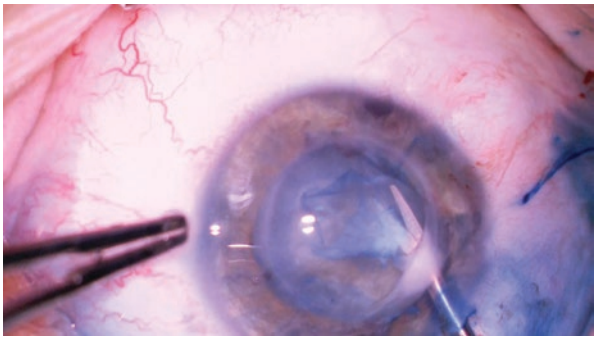

Fig. 23.2 Microscissors for completing capsulorrhexis in case of capsular fibrosis

fibrosis (Fig. 23.2). A one-piece intraocular lens was placed safely in the capsular bag, and the patient had a postoperative visual acuity of 20/250.

Comment The above case illustrates a case of cataract following a penetrating keratoplasty. Careful surgical technique with judicious use of trypan blue allowed for successful creation of a capsulorrhexis. The surgeon was attentive to not using too much trypan, which would compromise the integrity of the corneal graft. Use of microscissors, an instrument not conventionally used for creation of a rhexis, allowed for an opening to be created in the setting of fibrosis of the anterior capsule. Successful in-the-bag implantation of an IOL was possible, and postoperative visual acuity was significantly improved.

Iris Abnormalities

In the case of a cataract following a corneal laceration, there is often an organized membrane from the iris to the old corneal wound, and this scenario may require use of an OVD or microscissors to sever the adhesions and revise the wound prior to addressing the cataract [3, 4]. Similarly, OVD and microscissors are used to break lens-iris adhesions in Peters anomaly. Peripheral anterior synechiae in uveitic or glaucoma patients may also require synechiolysis with OVD during cataract

surgery. Only judicious pushes of OVD are required to achieve maximum force in breaking these adhesions. The use of additional OVD will not increase efficiency and may inadvertently induce reverse pupillary block, increase IOP, or cause iris prolapse. If OVD is unsuccessful, then mechanical means including a cyclodialysis spatula or microscissors are indicated.

The pupil may be persistently miotic in certain scenarios, such as trauma, Marfan syndrome, uveitis or in association with congenital cataract. In these cases, mydriasis may be achieved by using the same array of tools as in adult cataract surgery including iris hooks, ring devices, OVDs, and preservative-free epinephrine in irrigation fluid [4–6]. Care should be taken with the use of ring devices in an eye that is microphthalmic due to size or where a posterior capsulotomy and anterior vitrectomy are to be performed due to risk of posterior dislocation.

Hypoplastic iris tissue that is floppy and easy to tear can be encountered in congenital cataract cases especially in anterior dysgenesis syndromes like Axenfeld-Rieger and congenital rubella [7, 8]. Care must be taken in these cases to disturb the iris tissue as little as possible to avoid pigment loss and intraoperative iris prolapse. The most common cause of iris prolapse in these patients is overfilling the anterior chamber with OVD when not taking into consideration the smaller volume of the pediatric anterior chamber.

Case 2: Traumatic Cataract Following Open-Globe Injury

A 9-year-old boy presented 6 months after injury to the right eye with a pipe and primary corneal laceration repair done overseas. Visual acuity was count fingers at 2 feet. There was a corneal scar with neovascularization and a loose exposed suture, iris-cornea touch, retained cilia in the anterior chamber, inferotemporal iridodialysis, and a mixed anterior capsular and nuclear sclerotic cataract. In the operating room, the loose suture and intraocular cilia were first removed with forceps (Fig. 23.3). Iris tissue adherent to the corneal scar was dissected using OVD and microscissors. An endoilluminator and movement of globe to induce parallax were used to improve the view of the anterior chamber and facilitate cataract removal

Fig. 23.3 Removal of intraocular cilia during surgery for traumatic cataract

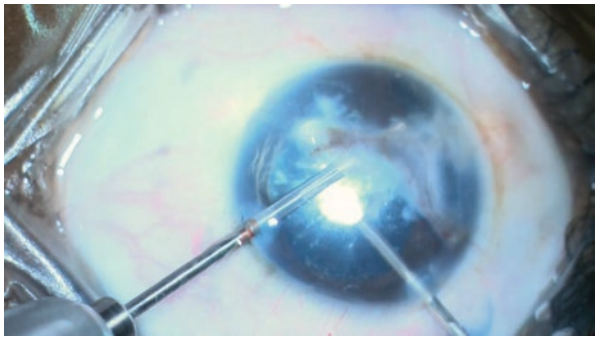

Fig. 23.4 Endoilluminator for improved view during lensectomy for traumatic cataract with corneal scar

(Fig. 23.4). An intraocular lens was placed in the sulcus with posterior optic capture with a power based on intraoperative A-scan of the operated eye and the keratometry values from the fellow eye (the corneal scar precluded keratometry in the operated eye). Postoperatively, the manifest refraction in the operated eye was $-1.00 + 1.50 \times 050$ with a visual acuity of 20/32–2 despite the corneal scar at 1-year follow-up.

Comment Globe trauma and the resultant cataracts can have very poor visual prognosis. Corneal scars and zonular weakness are just two of many reasons the surgical removal of lens opacities can be complex and challenging. That said, when they occur in an older child outside of the amblyogenic age range, the surgeon has the potential to greatly improve vision. Appropriate preoperative planning and strategic intraoperative maneuvers are important. This case introduces the reader to the use of an endoilluminator for improved visualization when corneal scar prevents clear visualization of all lens quadrants.

Abnormally Large or Small Anterior Segment

An abnormal anterior segment may involve an anterior chamber deeper or shallower than the cataract surgeon is accustomed to. In small[1] eyes, attention should be given to identifying the limbus carefully as limbal incisions tend to be positioned too

[1] The classification of small eyes is confusing [9]. Simple microphthalmia is defined as having a short axial length less than 2 standard deviations below the normal for age, typically 17.8 mm in infants and 20.5 mm in adults with no other malformations. Complex microphthalmia indicates the presence of microphthalmia with additional malformations such as iris coloboma, chorioretinal coloboma, persistent fetal vasculature, or retinal dysplasia. Relative anterior microphthalmos is a normal length eye with an abnormally small anterior segment [9, 10]. Microcornea is defined as a corneal diameter of less than 9 mm in an infant and 10 mm in an adult. Microcornea may be a feature of microphthalmic eyes, nanophthalmic eyes, and sometimes even long, myopic eyes [11]. Nanophthalmia is a short eye with a small anterior segment and thick sclera and choroid; there is no agreed-on axial length cutoff for nanophthalmic eyes [10].

anteriorly, increasing visible scarring and making access to the lens more challenging. Sometimes transillumination with an endoilluminator or the use of ultrasound biomicroscopy (UBM) may be helpful to mark the limbus when it is indeterminate. In eyes with a small anterior chamber, the corneal thickness is often greater, and limbal incision with a typical entry angle will form an incision that is too long [10]. A scleral tunnel may be used as an alternative to a limbal incision if an IOL is to be placed. When considering an IOL in a small eye, the surgeon should consider whether the eye is large enough to safely fit the IOL, whether an IOL with sufficient power is available, and the reduced accuracy of IOL calculations for these cases [10]. Leaving the child aphakic and using an aphakic contact lens or spectacles is the preferred approach if there is any question as to whether the small eye can fit an IOL. Corneal diameters may be measured using the traditional white-to-white measurement with calipers, optical biometry, or UBM [12, 13]. Postoperative care is frequently complicated by glaucoma in these cases.

In microspherophakia, weak zonules induce an increased curvature of the lens and a challenge for the cataract surgeon. The increased lens curvature causes a shallow anterior chamber with associated risk of pupillary block as well as high lenticular refractive power [14–16]. In microspherophakia, additional viscous OVD will be necessary to deepen the shallow anterior chamber. A capsular or iris hook at the capsulorrhexis margin may assist in performing the rhexis as the capsule has little zonular support [17]. Capsular tension rings or segment may be used to stabilize the lens and capsule, although the lens diameter is often too small to fit these devices and the IOL [18]. In some cases, it may be best to remove both the lens and the capsule completely if the zonules are so loose as to make the capsule useless as a method of supporting an IOL. IOL calculations in microspherophakia may be inaccurate because of postoperative changes in effective lens position, and the small size of the capsular bag may make it difficult to place an IOL within the bag [14].

In a large eye with a deep anterior chamber, the IOL may become easily decentered due to the large capsular bag and ciliary ring. An anterior capture of a three-piece IOL with the optic located inside of the capsular bag and the haptics located in the sulcus can provide the best centration of the optic [19]. The haptics in the sulcus stabilize the optic rather than relying on zonules that may be stretched and weak to support an in-the-bag IOL. Axial myopia also increases the uncertainty of IOL power calculation and increases risk of a hyperopic surprise, but formulas such as the Barrett Universal II and Haigis have shown excellent performance in eyes with axial length greater than 26 mm [20].

Eyes with Prior Glaucoma/Corneal Surgery

In cataract cases in which the eye already has a previously placed glaucoma drainage device, a few techniques may be used to avoid complications. The tube and patch graft area should be avoided when making the cataract surgery incisions. At the end of the case, the tube should be primed with BSS to ensure no OVD remains inside the tube. A low-flow technique should be used with the vitrector to avoid

massive subconjunctival chemosis from the bleb which can obscure the view. Some surgeons advocate tying off the tube, but in our experience, with low-flow techniques, this is not needed.

In the context of a graft or cornea with limited endothelial cells, reducing the intraocular pressure and total amount of fluid irrigation through the eye is also prudent to minimize further endothelial damage. A dispersive OVD should be used to protect the endothelium during cataract removal, and a cohesive OVD should be used during IOL implantation for its ease of removal. Minimizing wound leaks around incisions decreases the total volume of fluid used.

In summary, an eye with an abnormal anterior segment certainly makes pediatric cataract surgery more complex, but with appropriate planning and creativity, these challenges can be met. This chapter has presented what in our experience are the most common issues we have faced in these eyes and the techniques we have found most useful in addressing them.

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