5 Secondary IOL in Congenital Cataract Surgery

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5.1 Introduction

Primary intraocular lens (IOL) implantation can be done at any age, including early infancy. However, in the first 6 months of age, the preferred approach is to leave the child aphakic and implant an IOL at a later date when the child is older and the remaining refractive growth is easier to predict. There is a higher incidence of reoperations for visual axis opacification when IOLs are implanted in the first 6 months of life compared to leaving the child aphakic [1]. These results have resulted in more children being left aphakic when operated in infancy. Most of these children will eventually undergo secondary IOL implantation. For the initial aphakia after infantile cataract surgery, timely refractive correction is necessary to preserve monocular vision and binocularity and to protect against stimulus deprivational amblyopia. Conservative management may include correction with glasses or contact lenses, although these measures are not possible for all children. Aphakic spectacles are an option for bilateral aphakic children but often undesirable for social and optical reasons.

5.2 General Considerations

Secondary implantation of an IOL is generally recommended when traditional spectacle or contact lens correction of aphakia is unsuccessful or becomes a barrier to the child's development and activities of daily living. If sufficient capsular support for capsular bag or ciliary sulcus fixation is present, we offer secondary IOL implantation soon after we notice contact lens/spectacle wear becomes difficult or fails.

5.3 IOL Fixation Sites

Most commonly, secondary IOL implantation in our patients is performed between the ages of 4 and 6 years, a period when contact lens compliance can be difficult to maintain and yet there is still hope to reverse amblyopia. Due to relatively slower growth of eye after this age, IOL power calculation is also more predictable. We use SilSoft contact lens or aphakic glasses to treat aphakia prior to secondary IOL placement. SilSoft contact lenses are well tolerated in very young children but the material is less well tolerated as children get older. The silicone contact lenses develop more deposits and the lens coat-

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ing breaks down more quickly when worn in older children. Rather than change to a non-silicone contact lens material, some parents will opt to have a secondary IOL implanted at that point.

If capsular support is not available, every effort to correct aphakia using spectacles or contact lens should be exhausted before deciding on secondary IOL implantation. Risk must be minimized but amblyopia must not be left untreated or undertreated. Nowhere is that balance more difficult than when there is inadequate capsular support in the face of contact lens and aphakic spectacle intolerance.

The technical success of secondary implantation depends mainly on how much capsular support was left behind at the time of primary cataract surgery. Ultrasound biomicroscopy (UBM) may help to detect residual capsular support and image the ciliary sulcus when viewing it directly is difficult. Three hundred sixty degree visibility of the fused edge of the posterior and anterior capsulectomy from the previous surgery increases the chances of achieving successful posterior segment implantation of an IOL. In the office, if the posterior capsule is not initially visible at the slit lamp, having the patient look in extremes of gaze while maintaining the slit lamp chin position may allow the capsular remnant to be seen under the iris. Sometimes, examination through a peripheral iridectomy (if present) is useful to look for capsular remnants. There are times, however, when uncertainty about how much capsular support is present remains until surgery when a push–pull instrument or hook can be used to look directly under the iris in all quadrants.

When performing infantile cataract surgery without a primary IOL implantation, an adequate capsular rim should be left for subsequent posterior segment IOL placement. We urge surgeons to plan for the next surgery when performing the initial lensectomy. A 4.5-mm central matching anterior and posterior capsulectomy is ideal. This size is adequate to prevent subsequent closure and reopacification of the visual axis but assures an adequate rim of support when secondary IOL implantation is elected.

5.3.1 In the Bag

If capsular support is available, the IOL should be placed in the posterior segment either in-the-bag or in the ciliary sulcus. The most desirable position for the IOL is within the reopened capsular bag. However, if the capsular leaflets are sealed to one another without re-proliferated cortex (Soemmering ring) to maintain anterior and posterior leaflet separation in the lens equator, ciliary sulcus fixation is an acceptable and safe choice [2].

Despite the above recommendation, poor capsular support is often seen in pediatric aphakic eyes, particularly after removal of congenital cataracts, traumatic cataract, or ectopia lentis. In the absence of sufficient lens capsule support, choosing the best site for the IOL in a child is more difficult and controversial. Sutured or intrascleral fixated posterior chamber IOLs, an open-loop flexible anterior chamber IOL or the Artisan (Ophtec) iris-claw lens are available options. In the absence of capsular support and in the presence of a disrupted iris, sutured transcleral or non-sutured intrascleral fixated posterior chamber IOL are the only viable options.

The technique of in-the-bag secondary IOL implantation in children was reported in 1999 by one of us (MEW) [3]. Our updated experience with this technique was published in 2005 and 2011 [4, 5]. In-the-bag fixation is more consistently achieved in eyes made primarily aphakic during early infancy. This young age at surgery leads to a higher tendency for an exuberant Sommering ring formation. The Sommering ring lens cortex fills the capsular bag equator and prevents the anterior and posterior capsule remnants from sealing to one another and closing the capsular bag remnant. Removing the contents of the Sommering ring and replacing them with an ophthalmic viscosurgical device (OVD), allows in-the-bag IOL placement to occur more predictably. An odds ratio analysis revealed that within our study group, eyes originally operated before 6 months of age were 8.7 times more likely to receive a secondary in-the-bag than those patients who were remained aphakic after the age of 6 months [5]. In a subsequent study, we noticed

that all of the eyes that received in-the-bag IOL insertion had a cataract extraction within the first 4 months of life [4].

During surgery, it is important to assess whether it is possible to reopen the capsular bag leaflets. The key is to locate one area in which the anterior capsule edge is not strongly adherent to the posterior capsule [6]. Using the entry point, an OVD can be very useful in the separation of the capsular layers. A combination of dissection techniques using intraocular scissors, a micro vitreoretinal knife, and, most commonly, the 25-gauge vitrector handpiece are used to create a new anterior capsule edge and reopen the capsular bag for 360°. The Soemmering ring cortical material is then removed by bimanual irrigation/aspiration and a secondary IOL is placed into the capsular bag.

5.3.2 Ciliary Sulcus Fixation

If in-the-bag fixation is not possible, ciliary sulcus fixation is an acceptable alternative if technically possible [2–5, 7–9]. Awad et al. [10] performed UBM on the ciliary sulcus in 10 eyes after secondary IOL implantation. The structure of the sulcus in the implanted eye appeared similar to the sulcus in the contralateral normal eye. However, ciliary sulcus-fixated IOLs are more prone to develop pupillary capture, pigment dispersion, ciliary body erosion, decentration, unstable loop fixation, and lens tilt than IOL with bag fixation [11]. A study of adult eyes reported that even more than 2 years after cataract surgery, anterior chamber flare counts in eyes with sulcus IOL fixation were significantly higher than in eyes with in-the-bag fixation [12]. The three-piece Acrysof (Alcon, Fort Worth, Tx) is appropriate for sulcus fixation as well as in-the-bag positioning. It does not have a propensity to cause pigment dispersion when placed in the ciliary sulcus, although it does decenter at times unless optic capture is done. Non-angulated single-piece acrylic IOLs are not usually recommended for ciliary sulcus fixation. However, we have found that the Rayner C-*flex (570-C)* IOL is an exception. It is well tolerated by pediatric eyes when it placed in the ciliary sulcus. Optic capture through fused anterior and posterior capsule and haptics in the ciliary sulcus is another viable option for keeping the three-piece Acrysof IOL centered and stable. At the time of this writing, the Rayner C-*flex (570-C)* is our preferred IOL for sulcus placement when optic capture is not also being done. The three-piece acrylic lenses are still used by us in the ciliary sulcus but mostly when we are planning either monocapsular or bicapsular optic capture.

In a patient with bilateral aphakia, we recommend implanting the eye with the worst capsular support first. If it is not feasible to safely achieve implantation, bilateral aphakia can still be chosen. This approach can help avoid an IOL in one eye and aphakia in the other. While corneal tunnel incisions are usually utilized for secondary IOL implantation, a scleral tunnel incision should be considered when the capsular support is limited. After the posterior synechiae are severed, a change to a PMMA IOL, if available, may be warranted to provide a stable bridge across a large posterior capsule opening. This change can be accomplished more easily from a scleral incision.

5.3.3 ACIOL

In absence of capsular support, the implantation of an open-loop anterior chamber IOL (AC IOL) is the simplest surgical procedure for surgical aphakic correction. Modern, flexible AC IOLs have a much lower incidence of complications compared to the poorly tolerated rigid closed-loop AC IOLs of the past. The current designs of AC IOLs remain a viable alternative to sutured IOLs when there is an absence of capsular support. However, AC IOLs are used with caution in children. The most common complication seen in children relate to sizing of an angle-supported AC IOL in an eye that is still growing. Lens rotation over time (lens is too short after growth) can cause ovaling of the pupil and iris entrapment. A lens that is too long can cause corneal contact and iris atrophy. Correct sizing for AC angle width is critical to prevent IOL rotation

and/or corneal contact or iris entrapment and chronic inflammation. Traditionally, surgeons have used the corneal white-to-white measurement +1 mm as a guide for correct AC IOL sizing. A relatively large incision of at least 6 mm is required for currently available AC IOLs. A surgical peripheral iridectomy should be performed superiorly. A meta-analysis performed as part of the OTAC series revealed that insufficient evidence exists to demonstrate the superiority of a posterior versus an anterior chamber IOL in adults [13]. Anterior chamber intraocular lens implantation is contraindicated in the presence of extensive damage to the iris and anterior chamber angle, preexisting glaucoma, peripheral anterior synechiae (PAS), low endothelial cell count and shallow anterior chamber [14]. With the availability of iris-claw lenses, we no longer implant angle-supported AC IOLs in any children.

5.3.4 Iris-Claw IOL

Iris-claw IOLs, which are lenses placed in the anterior chamber and attached to the iris with small claw-like haptics, have been described to correct aphakia in adults and children, with good visual results (Fig. 5.1). These lenses have been available in most parts of the world for several years and are now being implanted in 20 sites in the USA under a compassionate-use FDA investigational device exemption. The aphakic Artisan IOL (Ophtec, Groningen, The Netherlands) is a peripheral "iris bridge" supported lens. The standard aphakic Artisan IOL is made of Perspex-CQ UV (a type of PMMA) and is 8.5 mm in overall diameter. The optic size is 5.0 mm and the overall body diameter is 5.4 mm. Implantation requires a 5.5 mm incision. Two smaller IOL sizes for microphthalmic eyes are manufactured at 6.5 mm and 7.5 mm in overall diameter with optic sizes of 4.4 mm. These smaller IOLs are not yet available in the USA. The fixation points for all of the Artisan IOLs are in the immobile peripheral iris. The IOL has a central oval shaped optic having an anterior vault (minimizes iris damage) and two fixation arms (haptics) that have a cut in them that provides a claw-grasping mechanism.

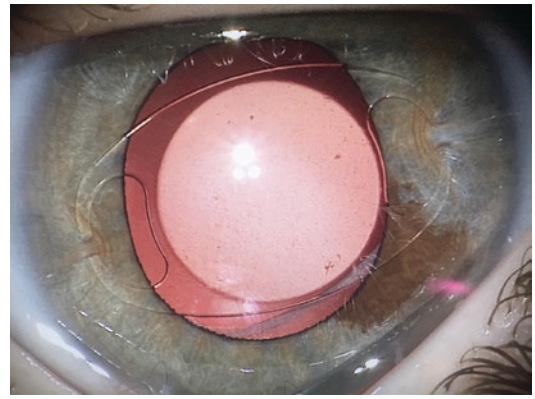

Fig. 5.1 Secondary Artisan IOL in an aphakic child with insufficient capsular support—note there is a small amount of capsule at the bottom of the picture but inadequate for posterior chamber support

Ideal method of iris-claw fixation includes: centrally placed optic, passing of an adequate amount of iris tissue through the claws of the lens, not pulling the iris root, not pulling the sphincter muscle/pupil margin toward the claw, and no injury to corneal endothelium. Advantages of artisan IOL implantation includes: The iris bridge protects the endothelium from touching the PMMA; There are no restrictions to pupil dilation or constriction; Excellent centration stability once fixated; The IOL has maximal visibility, accessibility, and controllability; Virtually cosmetically invisible; Easy to reposition and is reversible and exchangeable; No interference with vascular iris physiology; No sizing is normally needed—one size fits all. Disadvantage of artisan IOL includes: Requires surgical skill to position properly and create the ideal iris bridge but the learning curve is short. Implantation requires an incision of 5.5 mm ideally located at the limbus since the IOL is grasped through the incision when the iris bridge fixations are created. Early astigmatism is high but fades with healing. Wounds need to be sutured.

The Artisan IOL can also be fixated in a retropupillary manner if desired [15]. To accomplish this, the pupil is not initially constricted. The IOL is inserted with the convex side down (upside down) and held behind the pupil with the IOL implantation forceps through the corneal tunnel. As the lens is being inserted behind the pupil, a

miotic should be injected into the anterior chamber to constrict the pupil. The lens is lifted and tilted slightly to show the contour of the claws through the iris stroma. A fins spacula is inserted through a paracentesis to exert gentle pressure on the slotted center of the lens haptic to perform the enclavation. The same maneuver is repeated on the other side. Late dislocation of the lens can occur when one of the enclavation sites becomes dislodged, most often due to trauma. Prevention of this complication involves being meticulous about getting the proper amount of iris enclavated into each claw of the IOL. Late endothelial decompensation is expected to be very rare with the Artisan lens but ongoing long-term monitoring continues.

Posterior chamber intraocular lens (PCIOL) offer several advantages and many authors recommend them even in eyes lacking capsular or zonular support [16]. Scleral fixation of PCIOL is a method to overcome the lack of capsular support but is technically more difficult and time-consuming compared to ACIOL or Artisan IOL implantation. Due to its anatomic location in the eye, PCIOL is often reserved for patients with low endothelial cell counts, peripheral anterior synechiae (PAS) and cystoid macular edema (CME) [17]. Transscleral sutured IOL fixation can be considered in cases with inadequate capsular support or in cases with compromised anterior chamber structure. Shuaib and colleagues compared the results of transscleral IOL fixation to retropupillary iris-claw lens implantation in cases of pediatric aphakia without capsular support. This study concluded that both techniques yielded comparable visual outcome. Retropupillary iris-claw lens fixation is a shorter procedure and technically easier than sutureless transscleral fixation, but the risk of disenclavation should be considered especially in younger age groups. Scleral fixation is the only option in cases of severe iris damage [15]. Scleral suture fixation can be performed by either Ab externo (passing needle from outside to inside) or ab interno approach (inside to out). The ab externo approach is preferred by many. A disturbing late complication of transscleral suture fixation of IOLs is the spontaneous breakage of the polypropylene suture leading to IOL displacement, especially in young patients [18, 19]. Buckley [20] reviewed the literature on transscleral sutured IOLs in children. He reported the outcome of 33 eyes with an average follow-up of 61 months (range 9–200 months). Twenty-one of the eyes had greater than 3 years of follow-up and 14 eyes (42%) had more than 5 years of follow-up. Four patients had spontaneous suture breakage at 38, 66, 96, and 107 months after implantation. An additional 13 cases of 10.0-prolene suture breakage in children were uncovered by a survey of pediatric ophthalmologists. In addition to slow biodegradation of the suture, there are other factors that may contribute to the higher probability of suture breakage in children. These include globe enlargement with age, continuous eye rubbing, and higher probability of eye trauma due to more active lifestyle. To reduce the chance of suture breakage, it is important to consider the use of multiple sutures on each haptic, thicker sutures [21] (such as the 9–0 polypropylene mentioned above), or a different suture material such as GORE-TEX (W. L. Gore & Associates, Newark DE).

In adult eyes, iris-sutured IOLs have been used successfully to correct aphakia. The application of these techniques in the pediatric age group has been limited. The iris-sutured IOL fixation is preferred by some surgeons over the transcleral IOL fixation techniques because it is technically less demanding [22], and requires a shorter operative time. It also believed that the risk for cystoid macular edema, IOL tilt, and late suture breakage is lower in iris-fixated IOLs [23, 24]. Another potential advantage of iris suturing is a lower risk of suture erosion, and diminished risk of suture wick endophthalmitis [25–29]. Condon and associates suggested that the lower risk of late suture breakage may be the result of the fact that the elasticity of the peripheral iris provides a more forgiving suspension system than the sclera, which, in a transsclerally sutured IOL, is fixated to a rigid IOL spanning the posterior segment [30]. Iris fixation of a PCIOL is an alternative for secondary IOL placement in pediatric aphakic patients without capsular support in the short-term, but should be approached with

caution. Overall, in posterior chamber IOL suture techniques, the complication rate seems to be decreasing over the years, probably due to the increasing experience of surgeons in these difficult techniques. In addition, transscleral suturing techniques are now being replaced by intrascleral sutureless fixation techniques. With these methods, the IOL haptics are externalized and then tucked back into either an intrascleral tunnel beneath a glued scleral flap or tucked back after a cautery-induced mushroom cap has been fashioned at the tip of each haptic.

While secondary IOL power can be estimated using the aphakic refraction [31, 32], the estimate is less accurate than biometry [33]. If biometry cannot be obtained in the clinic setting due to poor cooperation and if biometry is not available in operating room, the aphakic refraction can be used to estimate IOL power. A change of the IOL fixation site may require adjusting the *IOL power*. If a decision is made to place the IOL in the ciliary sulcus, rather than the capsular bag, a decrease in lens power is often necessary. This is due to the fact that as the optic is shifted more anterior (moved closer to the cornea), its "effective power" increases. The amount of this change is dependent on the "base power" of the IOL. The greater the power of the IOL, the greater the difference in its effective power when it is placed in the ciliary sulcus instead of the capsular bag. If an IOL power calculated for the capsular bag needs to be changed to the ciliary sulcus, substract 1.5 D for ≥28.5 D, 1 D for 28.0 to 17.5, 0.5 D for 17.0 to 9.5 D. No substraction is required if power at capsular bag is <9 D (http://www.doctor-hill.com/iol-main/bag-sulcus.htm).

Simultaneous eye muscle and IOL implantation surgery in patients with strabismus and aphakia has been reported [34]. Combining IOL implantation and strabismus repair may reduce the number of surgical and anesthetic procedures, speed rehabilitation, and offer financial benefit to the patient and third-party payors. However, concern may arise regarding a potential increased risk of postoperative infection, anterior segment ischemia, or excessive discomfort for the patient. Determining the ocular alignment in eyes with poor vision may be difficult. We typically offer secondary IOL implantation surgery first (if indicated), wait for vision to recover, start amblyopia treatment, evaluate for strabismus, and offer strabismus surgery as a separate procedure.

5.4 Complications

Some postoperative complications after secondary IOL implantation are worth mentioning here. We reported that eyes with preexisting glaucoma were more likely to develop an IOP spike during the early postoperative period [35]. We recommend additional topical antiglaucoma medication or oral acetazolamide for children undergoing secondary IOL implantation with the preoperative diagnosis of aphakic glaucoma. Decentration of the IOL was the most common complication reported in our series. *C*linically significant decentration was noted in 4 (5.2%), while dislocation of the IOL was reported in 2 (2.6%) eyes; and pupillary capture requiring repositioning of the IOL in 1 (1.3%) eye. Clinically significant decentration requiring surgical intervention was noted only in eyes with sulcus-fixated foldable IOLs (28.6%; 4/14). None of the 29 eyes with sulcus-fixated polymethylmethacrylate (PMMA) IOL implantation developed decentration. Perhaps the rigidity of PMMA IOLs helped to avoid decentration. Foldable IOLs in the sulcus have been noted to be at risk for decentration and dislocation [36]. Decentration/dislocation was responsible for 21% of explanted three-piece hydrophobic acrylic IOLs in a 2001 survey [37]. All the decentrations were in an inferior direction and occurred more often in the eyes of male patients [5]. Perhaps this is due to the higher incidence of trauma among males. Another possible reason is that male eyes have been noted to have a longer axial length than female eyes. Eyes with an axial length of >23 mm were 4 times more likely to develop decentration if implanted with a sulcus-fixated foldable IOL when compared with eyes measuring <23 mm. We hypothesized that longer eyes may also have a "wider" anterior segment. A wider sulcus-to-sulcus distance may promote IOL decentration [5]. Another recent study has reported decentration in 6% of eyes after sec-

ondary placement of foldable Acrysof IOLs in the ciliary sulcus [38]. Jacobi and associates noted decentration of a scleral fixation of a secondary foldable monofocal or multifocal IOL implant in 19.2% of eyes of children and young adults [39]. We have now begun to use the Rayner C-*flex (570C)* IOL in the ciliary sulcus more often than the Acrysof MA-60 IOL, especially in eyes longer than 23 mm in globe axial length and when optic capture is not easily achieved.

A higher incidence of *cystoid macular edema* with secondary IOL implantation in adults in whom vitreous loss occurred at the time of initial cataract surgery has been reported. Other studies, however, reported excellent results when a careful and controlled vitrectomy was performed with secondary IOL implantation. Although we did not perform angiography, we did not observe clinically significant cystoid macular edema after secondary implantation in our series of pediatric eyes [5].

5.5 Conclusion

In summary, in the presence of adequate capsular support, secondary IOL implantation can be safely achieved in pediatric eyes. In-the-bag fixation is preferred over ciliary sulcus fixation but either is acceptable. When leaving an infant aphakic in the first months of life, the surgeon should plan for the secondary IOL placement by carefully leaving 360° of capsular support and matching anterior and posterior capsulotomy edges at 4 to 4.5 mm centrally. With this plan, an easy and safe secondary IOL surgery can be offered when these children reach preschool or grade-school age. In the absence of available capsular support, scleral-supported IOLs, iris-claw lenses, or ACIOLs can be used, depending on the surgeon's preference, the ocular environment, and IOL availability. Each of these options require careful follow-up over an extended period of time. As opposed to when adequate capsular support is present, these alternative implantation strategies should be performed only after the surgeon and parents are convinced that visual rehabilitation of the patient cannot be achieved with a contact lens or a safer surgical procedure. Because of the possibility of late complications such as dislocation of IOL in these children without capsular support, they should be observed regularly.

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