

Management of Congenital Cataract Surgery

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Congenital cataract surgery is a complex issue best left to surgeons who are familiar with its long-term complications and lengthy follow-up. Treatment is often difficult and tedious and requires a dedicated team effort, the most important members being parents. At one point, lensectomy was the standard treatment used in such cases. However, in recent times, small incisions, anterior capsulorhexis, bimanual irrigation/aspiration, and primary posterior capsulectomy and vitrectomy have become preferred treatment options for infantile cataract. Intraocular lens implantation has become the standard of care for the optical rehabilitation of children with cataract from the toddler age group and up.

Anterior Capsule Management:

The anterior capsule in children is very elastic, and therefore it may be difficult to perform a controlled manual continuous curvilinear capsulorhexis (CCC). However, manual continuous curvilinear capsulorhexis remains a gold standard for resistance to tearing and should be accomplished whenever possible. Difficulties of performing manual CCC in infantile eye led researchers and surgeons to search for alternative methods to open the anterior capsule in children. Alternatives to manual CCC currently available include vitrectorhexis, radiofrequency diathermy with a Fugo plasma blade, the two incision push pull technique, and the four incision technique. Wilson analyzed pediatric anterior capsulotomy techniques using porcine model and found that manual capsulorhexis produced the most extensible capsulotomy with most regular and stable edge. In eyes with poor anterior capsule visibility, trypan blue (0.0125%) may be used to stain the anterior capsule. The shape, size and edge integrity of anterior capsulotomy are very important for long-term centration of the IOL.

Management of the Posterior Capsule and Anterior Vitreous Face:

The most frequent and significant problem following pediatric cataract surgery is Visual axis obscuration(VAO).Maintenance of a clear visual axis remains a high priority when planning management of the posterior capsule in the amblyogenic age range. An important question that remains is when should the posterior capsule be left intact? Primary posterior capsulectomy (with or without anterior vitrectomy) are considered “routine surgical steps”, especially in young children. A manual PCCC offers the advantage of a controlled size and strong edges but is more difficult to perform. Many investigators have observed that performing manual PCCC is technically difficult. A potential complication associated with this procedure is the disruption of the Anterior Vitreous Face (AVF). However AVF disruption often goes unnoticed because anterior vitrectomy is a part of the surgical strategy in younger children. The signs of AVF disruption vary from subtle to obvious.These are 1) the presence of vitreous strands in the anterior chamber. 2) The attachment of the vitreous to the capsular flap. 3) Distortion of the capsulorhexis margin. Recently we described a technique to render the vitreous visible to the anterior segment surgeon. On completion of the PCCC, 0.1 ml of a suspension of preservative free triamcinolone acetonide (Aurocort, Aurolab India) was injected for visualizing the anterior vitreous face as well as the presence and extent of vitreous in the anterior chamber. After vitrectomy, if the surgeon hasn't cleared the residual vitreous strands triamcinolone can now be used second time to clearly identify the superficial vitreous gel which may have otherwise gone unnoticed. After IOL implantation and removal of the residual OVD, the surgeon again injects 0.1 ml of the suspension into the anterior chamber. Additional anterior vitrectomy can be performed if vitreous strands were identified in the anterior chamber.

Intraocular Lens (IOL) Implantation:

One of the most important preoperative considerations is whether to implant an IOL or not. Surgeons should be prepared for the common question “*Would you implant an IOL if this were your child?*” The capability of the IOL to offer *constant visual input* is an important advantage for the better visual outcome after pediatric cataract surgery. Use of IOL provides at least a partial optical correction at all times. Because of the advantage it offers, primary IOL implantation has slowly gained acceptance for the management of childhood cataracts. However, as of 2007, use of IOL remains controversial for the management of infantile cataract.The important concerns about primary IOL implantation during infancy are the technical difficulties

of implanting an IOL and selecting an IOL power, and the higher rate of visual axis opacification (VAO). At present, only adult sized IOLs are available, which are often difficult to implant in these small eyes and may cause complications over the long run. Currently available adult sized IOLs are slightly oversized and at times may not fit into small infantile eyes.

The size of the posterior capsulorhexis should be large enough to provide a clear central visual axis, but smaller than the IOL optic, so as to allow stable in-the-bag IOL fixation. Even if the surgeon is not planning to implant an IOL during the primary surgery, it is important to leave behind sufficient anterior and posterior capsular support at the time of cataract surgery to facilitate subsequent IOL implantation. The common practice is to perform posterior capsulectomy and anterior vitrectomy before IOL implantation if the limbal approach has been used whereas, if a pars plana vitrectorhexis is performed, it is done after the IOL is implanted. Further there is no agreement on whether the IOL should be implanted before or after the primary posterior capsulectomy. Some surgeons perform a pars plicata capsulectomy and vitrectomy with the vitrectome after implanting the IOL in the bag. Both PMMA and hydrophobic acrylic foldable IOLs have been widely used in pediatric eyes. However, several studies have now shown that hydrophobic acrylic IOLs are preferable as they offer better uveal biocompatibility and decreased incidence of VAO, with hydrophobic acrylic IOLs causing a delayed onset of PCO. For bilateral cataract during first year, aphakic glasses and/or contact lens use may be a reasonable option; however, for unilateral cataract, we are truly equipoised between whether or not to offer primary IOL implantation at the time of infantile cataract surgery. A large randomized clinical trial - the Infant Aphakia Treatment Study (IATS), in the U.S. and another randomized clinical trial at our centre is currently underway to compare primary IOL implantation to contact lens correction in children undergoing unilateral cataract surgery in the first six months of life.

NEWER APPROACHES:

Manual PCCC via Pars plana approach: Vasavada and coauthors recently introduced a technique of performing manual PCCC via pars plicata. After implantation of the IOL in the

capsular bag all the incisions are sutured with 10-0 nylon and residual OVD is left in the anterior chamber. The pars plicata entry is made 1 - 1.5 mm behind the limbus and an initial puncture is made in the center of the posterior capsule and later a coaxial capsulorhexis forceps is introduced and a flap is generated. The edge of the flap is grasped and then re-grasped every 2 clock hours fashioning the PCCC in a clockwise manner.

Bag-in-the-lens implantation: Tassignon and colleagues reported the outcome of a surgical procedure they called 'bag-in-the-lens' in pediatric cataractous eyes. In this technique, the anterior and posterior capsules are placed in the groove of a specially designed IOL after a capsulorhexis of the same size is created in both capsules. The principle behind this IOL design is to ensure a clear visual axis by mechanically tucking the two capsules into the IOL, thereby preventing any migration of proliferating lens epithelial cells.

Heparin in irrigating solution: Heparin has been used in intraocular irrigating solutions to reduce inflammatory reactions after pediatric cataract surgery. It has been documented that infusion of enoxaparin, a low molecular weight heparin, during pediatric cataract surgery may minimize the postoperative inflammatory response.

Posterior capsulorhexis combined with optic capture of IOL: This technique for pediatric cataracts was first described by Dr. Howard Gimbel. This was in the early 1990s using a single piece PMMA IOL, which were the preferred IOLs in those times. He suggested that performing an anterior and posterior capsulorhexis, and then capturing the IOL optic through the PCCC will cause fusion of the anterior and posterior capsule leaflets, thereby sequestering the proliferating lens epithelial cells. This way, VAO can be reduced and yet anterior vitrectomy can be avoided. Subsequently, this technique had fallen out of vogue. However, now with the introduction of 3 piece foldable hydrophobic acrylic IOLs and better surgical techniques as well as OVDs, optic capture of IOLs is being revived both in children and in adults. In a randomized clinical trial we followed up children below the age of 4 years with in the bag IOL and anterior vitrectomy versus those with optic capture without anterior vitrectomy for 12 month postoperatively. We found that there was no significant difference in the incidence of either VAO, or complications such as glaucoma or inflammation. Thus, this appears to be a promising modality which can prevent or reduce VAO, and yet avoid anterior vitrectomy even in young eyes.

Recently R. Menapace introduced Posterior optic buttonholing (POBH) a safe and effective technique which not only excludes retro-optical opacification, but also withholds capsular fibrosis by obviating direct contact between the anterior capsular leaf and the optic surface.

Summary:

While dramatic advances have occurred in this field over the past 10 years, some technical aspects of surgery, changing refraction and functional outcome continue to pose significant problems. Primary management of the posterior capsule is mandatory depending on the age of the child at surgery. With refinements in surgical techniques, improvisation of IOLs and better understanding of growth of the pediatric eye, in the coming years IOL implantation is likely to become an established mode of treatment of children even in the youngest age group.

Safety of Aphakia vs Pseudophakia following Bilateral Cataract Surgery in Children under 2 years of age : Randomized, Clinical Trial

There are several options for visual rehabilitation following cataract surgery. These include spectacles, contact lenses, or primary intraocular lens (IOL) implantation. However, while choosing the method of visual rehabilitation, it is best to select the option that leads to maximum visual benefit and yet ensures safety of the eye. Although still considered “off label” by the United States FDA, primary IOL implantation is now an accepted practice during pediatric cataract surgery, particularly in children above the age of 2 years. However, in infants and children younger than 2 years, there is still no consensus on whether primary IOL implantation should be performed during cataract surgery. In the past, in a number of studies, it was observed that a high level of postoperative complications occurred when an IOL was implanted.

However, there is paucity of literature on the advisability of implanting an IOL in children below the age of 2 years presenting with bilateral cataract without any associated ocular comorbidities. Therefore in bilateral congenital cataractous eyes operated on in infancy, surgeons choose to leave the eyes aphakic and recommend correction with spectacles or contact lenses instead of performing primary IOL implantation. This prompted us to conduct the present study in a single-centre setting with the goal of generating evidence as to which option is the better choice: implanting an IOL? Or leaving eyes aphakic after bilateral cataract surgery?

Methods:

This is a prospective, randomized, clinical trial of 60 children with bilateral congenital cataract carried out between January 2005 and January 2007. Children with congenital anomalies such as both anterior and posterior persistent fetal vasculature or tractional retinal detachments, aniridia, traumatic cataract, chorioretinal coloboma, microphthalmos (in the present study this was defined as a corneal diameter less than or equal to 9 mm), Down's syndrome, and subluxated cataract were excluded from the study. All the enrolled patients were randomly assigned to one of the following two groups. Group 1 (n=30 patients) comprised 60 eyes without IOL implantation while Group 2 (n=30 patients) comprised 60 eyes with IOL implantation.

We believe that this randomized, controlled clinical trial, will clarify whether IOL implantation, contact lenses, or spectacle treatment is associated with a lower complication rate and better visual outcome postoperatively following the surgical extraction of bilateral congenital cataract during the first 2 years of life.

We will be discussing the results of the 5 years follow-up of this trial.

