Pediatric Cataract Surgery: Consensus And Controversies

Abstract
Pediatric cataract surgery has improved significantly in terms of outcome over the last 2 decades primarily due to improvements in microsurgical instrumentation and techniques and the management of posterior capsule at the time of cataract surgery itself. The essential steps that can ensure a good outcome surgically are adequately sized anterior and posterior continuous curvilinear capsulorhexis, a limited but sufficient anterior vitrectomy and intraocular lens insertion in the capsular bag. Surveillance and care during the immediate post operative period for intraocular inflammation is of paramount importance. Yet, some issues need to be resolved and refined in pediatric cataract surgery such as at what age can intraocular lenses be safely inserted and what is the best power to put in a growing eye. The most important aspect of pediatric cataract surgery remains the aftercare with appropriate long term addressal of residual refractive error, amblyopia and possible complications like secondary glaucoma.

Key words: pediatric cataract, surgery, capsulorhexis, intraocular lens.

Pediatric cataract surgery has come a long way with the outcome improving dramatically over the last 2 decades with advancements in microsurgical techniques and instrumentation. Yet, taken as a whole, management of a child with cataract remains one of the most complex and difficult tasks confronting an ophthalmologist. This article seeks to discuss the status of management as it stands today, both the issues which appear settled and some where the jury is still out.

Timing of surgery:
Regarding the timing of surgery, the general rule is that earlier the visual axis is cleared, the better the visual outcome. However this does not mean that all pediatric cataracts are best removed as soon as they are identified. Weighing the risks with the benefits, the indication for surgery would be cataracts centrally located and greater than 3mm in size that obscures the red reflex on distant direct ophthalmoscopy. In order to assess visual status of these young children with media opacities, it is helpful to quantify visual acuity when possible with Cardiff or Teller acuity cards or Lea symbols. Presence of squint or nystagmus suggests that it is already late for intervention and these children should be taken up for surgery on an urgent basis. Unilateral cataracts need more urgent intervention as compared to bilateral cataracts as they are more amblyogenic. As regards the earliest age to take a child up for cataract surgery, the main consideration to be kept in mind is the anaesthetic risk, which seems to be quite minimal even in the immediate newborns. Current evidence seems to suggest that the window period we have from the amblyopia perspective is 6 weeks from birth for unilateral congenital cataracts and 3-4 months in bilateral cases.

Investigations
Half the bilateral cases are idiopathic in origin and 1/3rd are hereditary without systemic associations, while most cases of unilateral congenital cataract are idiopathic. Hence there is no need to waste precious time and money on unnecessary investigations, especially when they wouldn’t influence management greatly. Unless there are clear evidences to suggest specific diseases or syndromes, it would be judicious to restrict investigations to hemoglobin (anaesthetic considerations) and blood sugar.

Surgery
Pediatric eyes need to be handled a little differently from adult eyes due to the following reasons:
1. Pediatric eyes have reduced scleral rigidity and so tend to collapse more easily during surgery and to leak postoperatively.
2. Anterior capsule is more elastic and therefore needs more force to both fracture and tear. To make matters worse, it has a constant tendency to run out to the periphery.
3. There is more violent inflammatory reaction postoperatively.
which sometimes results in fibrin formation towards the end of surgery itself.

4. Mitotic lens epithelial cell proliferation is much more active in the pediatric eyes and so visual axis obscuration (VAO) by posterior capsule and/or anterior vitreous face opacification is invariable unless managed primarily by posterior capsulorhexis and anterior vitrectomy.

**Technical aspects of surgery:**

**Wound construction:**
A valved incision of the smallest length necessary for instrumentation would help maintain anterior chamber which has a constant tendency to collapse. For the same reason bimanual irrigation aspiration and a 2 port vitrectomy is useful as opposed to irrigation from the same port. It is preferable to suture all wounds in children below 10 years of age.

**Anterior capsule management:**
The anterior capsulotomy size, shape and edge characteristics are crucial for both immediate and long term centration of IOls in the capsular bag5-7. Various options for anterior capsulotomy in pediatric cataract surgery are manual continuous curvilinear capsulorhexis (CCC), vitrectorhexis, radio-frequency diathermy and Fugo plasma blade. Among these manual CCC was found to produce the most extensible opening with best edge characteristics6. Though it is the most difficult to perform and control in pediatric eyes for reasons enumerated earlier, it remains the gold standard for anterior capsulotomy in pediatric eyes (Figure 1). A high molecular weight viscoelastic agent like Healon or Healon GV and capsular staining with Trypan Blue is helpful while performing CCC. While tearing the capsule, the direction of pull on the capsular forceps should be centripetal. Repeated regrasping of the capsule for short tears and aiming for a smaller capsular opening prevent running out of the capsular tear to the periphery (see video - http://www.youtube.com/watch?v=ulU7Q0LSf5Q). The edge of the lamellar cataract often serves as a guide to the ideal size to be aimed for. For the less experienced pediatric surgeon, it is safer to err on the side of a smaller rhexis and then to make it of the appropriate size after IOL insertion as shown in the video (http://www.youtube.com/watch?v=EipthArLSOM). A small rhexis in the postoperative period runs the risk of capsular phimosis and capsular contraction syndrome due to the violent inflammatory response characteristic of children. Manual CCC is especially difficult to perform in very young infants because the capsule is highly elastic. Here vitrectorhexis is a good option as owing to the highly elastic capsule, the edge remains regular and resists radial tearing5. In the older children, the capsulotomy edge of a vitrectorhexis is scalloped and never as regular as a CCC.

**Lens matter removal**
Being soft, lens material only needs aspiration and none of the nucleus management techniques of adult phacoemulsification surgery. It can be done either with a symcoe cannula or by automated irrigation-aspiration handpiece of the phacoemulsifier. Very rarely only, if the cataract is harder, it becomes necessary to resort to the larger diameter of the phaco probe for irrigation aspiration or actual ultrasound energy.

**Posterior capsule management**
Visual axis obscuration by posterior capsular opacification is the commonest cause of poor outcome following pediatric cataract surgery8-10. Maintaining a clear visual axis continually is of paramount importance in the amblyogenic age group. It is well accepted now that the posterior capsule is best managed by primary posterior capsulectomy and

![Figure 1: Post operative photograph on the 3rd day of a 2 year old boy showing both anterior and posterior capsulorhexis and a PMMA IOL in capsular bag](image1)

![Figure 2: Three month post operative photograph of an 18 month old boy showing opacification of anterior capsular rim, early opacification of posterior capsule and clear visual axis due to a PCCC and anterior vitrectomy](image2)
anterior vitrectomy during the cataract surgery itself (Figure 2, 3). Like anterior capsulectomy, among the various options available, a manual PCCC is the gold standard. The ideal size of the posterior capsulotomy opening is 3.5-4 mm. A smaller opening runs the risk of closing off (Figure 4). Most surgeons perform PCCC before IOL insertion (http://www.youtube.com/watch?v=ulU7Q0LSf5Q and http://www.youtube.com/watch?v=EipthArLSOM) while others prefer to lift the IOL with the non-dominant hand and perform it after the IOL is inserted.

Anterior vitrectomy
In young children, below 6-8 years, a posterior capsulectomy alone would not ensure a clear visual axis. The anterior vitreous face would in such cases invariably be covered by migrating epithelial cells and metaplastic cells in the postoperative period. Therefore a primary limited anterior vitrectomy is recommended in young children. The current recommendation seems to be a PCCC with anterior vitrectomy in children below 3 years, a PCCC alone between 3 and 7 years and leaving the posterior capsule intact in older children and who are likely to cooperate for a YAG Capsulotomy later on11.

Pars plana vitrectomy
Some surgeons prefer to perform anterior vitrectomy through the plane route after IOL insertion. This technique has the advantage of making IOL insertion less risky and also making possible a larger posterior capsulotomy. It is especially useful in resurgeries to remove posterior capsular and vitreous opacifications behind the IOL.

IOL implantation
Primary IOL implantation is the recommended practice in children older than 2 years12-14. In children below 2 years it is controversial. IOLs seem to increase the chances of posterior capsular opacifications (PCO)13-15. Yet with increasing use of Acrysof IOLs, more and more surgeons are implanting lenses in infants as young as 3 months of age. The absolute contraindications to IOLs are microcornea and microphthalmos while relative contraindications include uveitis, glaucoma, aniridia and persistent hyperplastic primary vitreous. The important determinants of outcome related to IOLs are material, position and size of IOLs. PMMA has the longest track record but current opinion seems to favour hydrophobic acrylic16-24. PCOs following Acrysof IOLs set in later and are also predominantly proliferative as opposed to fibrotic in PMMA cases. VAO produced by Acrysof lenses are less severe than those caused by PMMA lenses and so are less amblyogenic25.

IOL sizing was a major issue with PMMA lenses in smaller eyes especially in infants but seems to be a lesser problem with Acrysof. The haptics are extremely flexible and have excellent...
memory and single piece Acrysof lenses adapts well to the smallest capsular bag without becoming decentered25. In the bag placement is the best position to ensure long term stability and centration of IOLs. This is why a well made CCC is crucial to the success of pediatric cataract surgery. Optic capture of IOL in the PCCC, championed by Gimbel26, was introduced to obviate the need for primary anterior vitrectomy. While it produces excellent centration of the IOL, it causes increased inflammatory response27,28 leading to opacification of vitreous face and so is currently not very popular.

**IOL power selection**

This is an area where pediatric surgeons have different opinions and protocols. In general, an IOL power which produces emmetropia in the immediate post operative period will eventually end up causing myopia as the eye grows in axial length. Younger the child, more the myopia. To eventually aim for emmetropia, the child will have to be left hypermetropic immediately after surgery, which is amblyogenic and which has to be managed appropriately with reducing power of plus lenses. There are many tables29-31 available for IOL power selection, but it is not easy to predict the rate of growth of an individual eye and its final refractive outcome. It is influenced by several factors like normal growth rate, age at surgery, visual input, presence or absence of IOL, laterality and genetic factors. A recent study32 showed that the residual refractive error after cataract surgery does not influence the myopic shift. Some surgeons advocate aiming for emmetropia, the child will have to be left hypermetropic immediately after surgery, which is amblyogenic and which has to be managed appropriately with reducing power of plus lenses. But this is likely to lead to significant myopia eventually. Predicting rate of growth and the resulting refractive change remains a major challenge of pediatric cataract management.

**Contact lens**

In children who are not implanted with an intraocular lens due to any cause, contact lens is a good option for visual rehabilitation. But compliance and safety are major issues in developing countries with the result that more and more pediatric ophthalmologists are implanting IOLs at a younger age.

**At what age can IOLs be safely implanted?**

As mentioned earlier, the current recommendation is still for IOLs after 2 years of age where the axial length of the child’s eye approaches 90% adult dimensions. There have been a few studies comparing visual rehabilitation with contact lenses and intraocular lenses, with the largest being that of the Infant Aphakia Treatment Study Group (IATS)33. In a randomized, multicenter (12 sites) clinical trial, 114 infants with a unilateral congenital cataract were assigned to undergo cataract surgery between 1 to 6 months of age either with (57 infants) or without (57 infants) primary IOL implantation. Contact lenses were used to correct aphakia in patients who did not receive IOLs. Grating visual acuity tested at 1 year of age by a masked examiner showed no statistically significant change between the 2 groups. Additional surgeries were required more frequently in the IOL group and the authors concluded that caution should be exercised when performing IOL implantation in children aged 6 months or younger given the higher incidence of adverse effects and the absence of improved short term visual outcome compared with contact lens use. Nevertheless several things should be noted in this study as highlighted in the discussion by the authors themselves. One, the contact lens group had a very high compliance rate (average more than 80% of waking hours). This was ensured in the study by providing free contact lenses and patches and frequent monitoring of compliance by telephone calls. This is difficult to duplicate in societies like ours and so what this study estimates is the efficacy (benefit under ideal conditions) rather than effectiveness (benefit under usual conditions). Secondly, assessing the risks and benefits of IOL implantation at 1 year of life may lead to premature conclusions. The real benefit of IOL implantation may occur later, especially if children in the contact lens group become less compliant with contact lenses as they become older. If this is true, it is possible that the children in the IOL group will have an increasing visual advantage with their pseudophakic correction alone as they become older and approach emmetropia. The study plans to retest the visual acuity of these children when they are aged 4 years using the Amblyopia Treatment Study–HOTV acuity test.

The main difference between the 2 groups was in relation to number of additional surgeries and the adverse effects. The main adverse effects noted are lens proliferation in the visual axis and pupillary membranes which were the reason for the additional surgeries. There was no significant difference in the incidence of secondary glaucoma. Cataract surgery in infancy is itself a risk factor for glaucoma34-36, but IOL doesn’t seem to increase the risk. In fact, some studies37 even suggest that IOLs have a protective effect against secondary glaucoma. We can look forward to the IATS study which is supposed to evaluate the children again at 5 years for secondary glaucoma.

So on current evidence, in the debate on IOLs versus contact lenses in the very young, what is in favour of contact lenses is the lesser incidence of post operative VAOs while this advantage might be offset by a lower visual acuity related to poor compliance issues.

**Postoperative treatment and follow up**

A good job on the surgical table is less than half the job done in case of pediatric cataracts. In fact the treatment would
have just started with the surgery. The immediate post operative period is vital because if the inflammation is well controlled then, the anatomical results remain good. It is therefore important to see the child very frequently during the first few post operative days and to be aggressive with topical and, if necessary, systemic steroids.

The next main concern is amblyopia for which appropriate spectacles and patching in unilateral cases is to be started as soon as possible. These children need to be followed up regularly and monitored for vision and change in refraction. Aphakic and pseudophakic glaucoma is a well recognised complication and can occur at any time after cataract surgery. Therefore these children need to be monitored for life.

Conclusion
Surgical management of pediatric cataracts is different from adult cataracts. The reduced scleral rigidity, elastic lens capsules and positive vitreous pressure make surgical manipulations more difficult. The high rates of posterior capsular opacifications make PCCC and anterior vitrectomy mandatory in the younger age group. Ocular growth makes selection of IOL power a difficult choice. However outcomes have improved greatly in the last few decades and with better microsurgical instrumentation and techniques and improved understanding of pediatric eye growth, we can hope for more.

References
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