

## Congenital Cataract- Approach and Management Review

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**Abstract:** Childhood cataract remains a challenge to pediatric ophthalmologists despite recent major breakthrough in surgical techniques and instrumentation. Pediatric cataract is one of the major causes of preventable childhood blindness, affecting approximately 200,000 children worldwide, with an estimated prevalence ranging from three to six per 10,000 live births. Congenital cataracts usually are diagnosed at birth. If a cataract goes undetected in an infant, permanent visual loss may ensue. The management of pediatric cataract is a team effort of ophthalmologist, pediatrician, anaesthetist and parents and should be customized depending upon the age of onset, laterality, morphology of the cataract, and other associated ocular and systemic co-morbidities. This review attempts to summarize the available management options to these patients along with some analytical recommendations to optimise the outcome.

**Keywords:** Pediatric cataract, childhood blindness

### I. Introduction

Pediatric cataract is one of the major causes of preventable childhood blindness, affecting approximately 200,000 children worldwide, with an estimated prevalence ranging from three to six per 10,000 live births<sup>1-3</sup>. It may be congenital, if present within the first year of life, developmental if present after infancy, or traumatic. Congenital cataracts occur in about 3 in 10,000 live births<sup>3</sup>. Two-thirds of cases are bilateral and most common etiology is genetic mutation, usually autosomal dominant (AD)<sup>4</sup>; Others etiology are chromosomal abnormalities, metabolic disorders and intrauterine infections<sup>4,5,6</sup>. Etiology of unilateral cases remain unclear but they are usually sporadic.<sup>2,3</sup> Early diagnosis and treatment are of crucial importance to prevent the development of irreversible stimulus-deprivation amblyopia<sup>7,8</sup>. The management of pediatric cataract is a team effort of ophthalmologist, pediatrician, anaesthetist and parents and should be customized depending upon the age of onset, laterality, morphology of the cataract, and other associated ocular and systemic co-morbidities.

#### Work up:



Fig 1. visually significant cataract

#### History-

Evaluation of a child with a cataract begins with a detailed history including family history; an elaborate antenatal history including maternal drug use and febrile illnesses with rash; and birth history, especially birth weight. A developmental history should be carefully assessed, to exclude metabolic or systemic etiologies. A history of onset of lenticular opacities, laterality, and progression is also important. Unilateral cataracts are usually isolated, but they are most commonly found to be associated with persistent fetal vasculature (PFV) or other ocular abnormalities, such as lenticonus/lentiglobus.

#### Examination



**Fig 2** Examination of an infant

Examining a neonate is not only different from adult but also very difficult and demanding job .It can be carried out in the OPD with the child sitting comfortably on its mother lap but many a time examination under anaesthesia is needed. Recent introduction of high quality portable slit lamp has made the examination of a neonate easier (fig 2) .Since a formal estimate of visual acuity cannot be obtained in the preverbals, following features are noted in order to assess the visual significance of the opacity.

- Fixation behaviour, fixation preference and objection to occlusion.
- Density,size,site and morphology of cataract- A red reflex test can be performed in a dark room with a direct and indirect ophthalmoscope to assess the visual significance of the lens opacity. A very dense cataract precluding any view of the fundus, central cataract larger than 3 mm in diameter, unilateral cataract associated with strabismus and bilateral cataract with nystagmus are considered visually significant. Visual acuity in verbal cooperative children should be noted along with glare test in cases of lamellar or posterior subcapsular cataracts. Morphology of the opacity can give important clues to the underlying aetiology and helps in planning their management.
- Associated ocular pathology involving the anterior segment (corneal clouding, microphthalmos, glaucoma, persistent fetal vasculature) or the posterior segment (chorioretinitis, Leber amaurosis, rubella retinopathy, foveal or optic nerve hypoplasia) should be looked for.
- Measurement of intraocular pressures and corneal diameters .

**Investigations:**

**Laboratory workup:**

Extensive laboratory investigations are not usually indicated for unilateral cataracts as most of them are isolated, nonhereditary, and without any systemic associations. In infants with bilateral cataracts,unless there is an established hereditary basis for the cataracts, the investigations should include the following:

<b>1</b>	<b>Serology</b> for intrauterine infections(TORCH)
<b>2</b>	<b>Urine.</b> Urinalysis for reducing substance after drinking milk (galactosaemia) and chromatography for amino acids (Lowe syndrome).
<b>3</b>	<b>Other investigations</b> include fasting blood glucose, serum calcium and phosphorus, red blood cell GPUT and galactokinase levels.Children who have calcium and phosphorus anomalies severe enough to cause cataracts are unwell.
<b>4</b>	<b>Referral to a pediatrician</b> may be warranted for dysmorphic features or suspicion of other systemic diseases. Chromosome analysis may be useful in this context.

**USG-B scan-**

It is mandatory in every case of total cataract for posterior segement evaluation.

**Ultrasound biomicroscopy**

It can be informative in children with anterior-segment developmental anomalies and PFV and also in the assessment of posterior capsular support while considering secondary IOL implantation.

**Visual evoked potential**

For preverbal, less cooperative, or developmentally delayed children, Flash VEPs may be useful in complete cataracts to establish the gross integrity of the visual pathways.

**Examination of the family members**

A dilated slit-lamp examination of the parents and any sibling should be performed, which may reveal previously undiagnosed lenticular changes indicative of an inherited cause for the child's cataract . Female carriers of X-linked conditions associated with males presenting with congenital cataracts may show lenticular changes.

## II. Management

### Non surgical management

Visually insignificant lenticular opacity like peripheral lens opacities, punctate opacities with intervening clear zones, and opacities less than 3 mm in diameter can be observed closely and successfully managed by treating the associated amblyopia by patching and glasses. In small central opacities, a larger area of clear visual axis can be achieved by pharmacological dilatation. While conservative treatment can be used, it must be done so with caution, and ancillary tests such as glare testing must be utilized to ensure that the lens opacity is not visually significant.

### Surgical management

Indicated in all visually significant lenticular opacities. A critical period for visual development has been described in the first 6 weeks of life, during which the vision is subcortically mediated and the infant is relatively resistant to amblyopia. Timing of surgical intervention is crucial and the main considerations are as follows<sup>7-10</sup>.

**1. Bilateral dense** cataracts require early surgery when the child is 4–6 weeks of age to prevent the development of stimulus deprivation amblyopia. If the severity is asymmetrical, the eye with the denser cataract should be addressed first.

**2. Bilateral partial** cataracts may not require surgery until later if at all. In cases of doubt it may be prudent to defer surgery, monitor lens opacities and visual function and intervene later if vision deteriorates

**3. Unilateral dense** cataract merits urgent surgery (possibly within days) followed by aggressive anti-amblyopia therapy, despite which the results are often poor.<sup>11</sup> The timing of intervention should be balanced by the suggestion that early intervention (<4 weeks) may result in an increased risk of subsequent secondary glaucoma. If the cataract is detected after 16 weeks of age then the visual prognosis is particularly poor.

### Surgical technique of choice :

Phacoaspiration with primary posterior capsulotomy with or without anterior vitrectomy and in the bag implantation / optic capture of IOL.<sup>12-16</sup>

### Peculiarities of child's eye

Pediatric eyes are different from adult eyes in following aspects making the surgical management technically demanding

- 1 .Small size
- 2 .Thin sclera with decreased rigidity
- 3 .Increased vitreous pressure
- 4 .Elastic capsule with unstable anterior chamber.
- 5 .Changing axial length and corneal curvature
- 6 .Severe inflammatory response after surgery.
- 7 .longer life span after cataract removal, with a potential for irreversible visual loss due to amblyopia.

**Therefore, presurgical evaluation of a growing child's eye is a little complex.**

### Biometry and IOL Power calculation<sup>14-17</sup>

Choosing an appropriate IOL power to prevent the unexpected refractive change in a pediatric case is still a challenge though there is increasing consensus towards IOL implantation even in infants after the age of 6 months. Poor patient cooperation and poor fixation most of the time mandates biometry to be performed under sedation or general anesthesia (Fig 3). Immersion biometry has been shown to be more predictable than the contact method for IOL-power calculation.



**Fig 3** Examination under Anaesthesia

There is a general tendency of partial undercorrection at the time of surgery to balance the postoperative anticipated myopic shift of the growing eyeball. The choice of IOL power should be individualized based on the child's need and refractive status of the other eye in unilateral cases. Different formulas have been used to calculate the desired IOL

power in children, with variable results. Various factors in children younger than 2 years like, smaller axial lengths, shallower anterior chambers, and changes in corneal curvature and corneal thickness may lead to higher prediction errors. The recent report of IATS ( Infant Aphakia Treatment Study ) recommended Holladay 1 and SRK/T formulae for infant eyes. However, at 5 years, refractive errors ranging from +5.00 to -19.00D were detected. Inability to predict axial elongation in infantile eyes was the primary reason for such a wide range of refractive errors.

Refraction-based formulae by Hug, Khan and Algeed for estimation of secondary posterior-chamber IOL power provide comparable results to those obtained by standard biometry-based formulas, and can be useful in difficult situations when standard biometry cannot be performed or when it is not available in the operating room. Many surgeons use an empiric rule whereby children under the age of 3 months are left +8.00 D, 3 months to 1 year +6.00 D, 1-2 years +4.00 D, 2-3 years +3.00 D, 3-5 years +2.00 D, 5-7 years +1.00 D, and 7 years and after +0.5 D until the age of 11 years. These numbers may be modified in unilateral cases, depending upon the refraction of the other eye, so as to cause minimal anisometropia 2 years after surgery.

#### **Type of IOL<sup>14,15</sup>**

**Hydrophobic acrylic square edge IOLs** are considered best to be used in pediatric cases because of greater biocompatibility, foldable design, and lower rate of PCO formation and are currently IOL of choice for most pediatric cataract surgeons. The indications for multifocal IOLs in children are debatable. Refractive shift during eye growth as well as amblyopia due to loss of contrast sensitivity associated with multifocal IOLs are the main concerns of most pediatric cataract surgeons. Although limited studies in children have shown improved stereopsis and spectacle independence with the use of multifocal IOLs, studies with long-term follow-up are warranted.

#### **Surgical Difficulties with Recent Innovations<sup>19-23</sup>**

Cataract surgery in children is challenging, because of decreased sclera rigidity, increased capsular elasticity, thicker corneas, increased risk of trauma from eye rubbing, less compliance with activity restriction, and most importantly the effect of postoperative astigmatism on amblyopia

#### **Wound<sup>22</sup>**

Conventionally scleral tunnel incision was preferred by most pediatric cataract surgeons because it was thought to induce less postoperative astigmatism but recent trend is towards clear corneal approach. Clinically insignificant difference exists between the two types of incisions, with spontaneous regression of astigmatism over a period of time. Superior incisions are more preferred as compared to the temporal approach, probably in view of less risk of injury and postoperative endophthalmitis. To prevent wound leakage due to the reasons mentioned, suturing of all the wounds, including paracentesis with either a 10-0 or 9-0 Vicryl or nylon suture, is recommended. Absorbable sutures are preferred, in order to avoid a second visit. Research has shown that the use of 10-0 Vicryl caused astigmatism, but that this astigmatism dissipated after 6 weeks.

#### **Capsulotomy<sup>23-27</sup>**

Continuous, smooth, and well-centered anterior capsulotomy is a prerequisite for safe lens implantation. For proper centration of the IOL, the anterior capsulotomy should be smaller than the IOL optic (4-5mm).<sup>24</sup> Elasticity and thickness of the anterior capsule in young children makes manual CCC the most difficult technique, with a steep learning curve. Use of high viscosity ophthalmic visco surgical devices and a soft shell technique After coating the corneal endothelium with dispersive viscoelastic, the cohesive viscoadaptive agent is filled in the anterior chamber and Balanced salt solution is used below the viscoadaptive agent, away from the incision, and creating a surgical operating space with low viscosity. Anterior capsule staining is done using trypan blue by gently "painting" the anterior capsule. It makes capsulorhexis, hydrodissection and later, OVD removal, much easier. Currently used techniques for pediatric anterior capsulotomy includes<sup>25-28</sup>

- **Manual continuous curvilinear capsulorhexis (CCC)**-Gold standard technique and produce the most extensible capsulotomy and the smoothest edge with scanning electron microscopy evaluation in a porcine model. Multiple anterior capsulorhexisotomies are advocated by some recent studies to prevent anterior-capsule phimosis.
- **Vitrectorhexis**- Done with vitrector and it is not robust compared to manual CCC but preferred by some surgeons in very young infants.
- **Plasma blade (Fugo blade)**, diacapsutom, and pulsed-electron avalanche knife, have been suggested to minimize zonular tension and prevent peripheral CCC extension, but have not become popular.
- **Femtosecond laser assisted anterior and posterior capsulotomies**

#### **Techniques for cataract extraction**

Soft consistency of pediatric cataracts makes phacoemulsification unnecessary. Lens removal can be conducted through an anterior approach by manual irrigation and aspiration or through the pars plana using a vitrector. In rare cases with membranous cataracts or retrolenticular membranes in PFV, intraocular scissors are required. An anterior-chamber maintainer used for continuous irrigation prevents the collapse of the anterior chamber during the surgery and aids in pupillary dilatation with the use of 1:1,000 epinephrine added to the balanced salt solution.<sup>28-30</sup> The use of heparin in the balanced salt solution and intracameral recombinant tissue-plasminogen activator has been shown to reduce the fibrinous reaction and pigment deposits on the IOL but their routine use is not yet established. Recently, the use of a transconjunctival 25-gauge vitrectomy system has been gaining popularity for lens removal in pediatric patients.

**Posterior-capsule management**

Maintenance of a clear visual axis is critical for a good postoperative visual outcome after pediatric cataract extraction. If the posterior capsule is left intact, 100% of eyes less than 4 years of age develop significant PCO. The anterior vitreous face acts a scaffold for the proliferation of lens epithelial cells (LECs). Intraoperative measures taken to minimise it are:

**1.Primary posterior capsulotomy (PPC) with anterior vitrectomy**<sup>30,31</sup> in infants and young children or in older children who are poor candidates for possible YAG laser capsulotomy. This step can be facilitated by using trypan blue and triamcinolone acetonide for staining posterior capsule and vitreous respectively. The opening for PPC should be smaller than the anterior capsulotomy. Two-incision push-pull technique has been shown to give consistent results without vitreous loss during the procedure. Primary manual anterior and posterior continuous curvilinear capsulorhexis (ACCC, PCCC) showing a smooth and regular edge<sup>32-34</sup>

**2.Optic capture of the IOL** was believed to obviate the need for anterior vitrectomy and prevent the development of PCO<sup>34</sup>. However, recent studies have proven that anterior vitrectomy is necessary with optic capture even in older children. Moreover, it is a difficult technique to perform with single-piece IOLs without haptic angulation. Posterior vertical capsulectomy with optic entrapment has promising results in children aged 2 months to 8 years, with clear visual axes for 5–12 years postoperatively. Although this procedure is technically challenging, anterior vitrectomy is seldom required.

**3.Subconjunctival or intracameral steroids** are recommended to suppress inflammation in the immediate postoperative period. Intracameral recombinant tissue-plasminogen activator has also been shown to reduce the fibrinous reaction and pigment deposits on the IOL after pediatric cataract surgery, though there is a risk of hyphema.

**Complications:**<sup>35,36</sup>

Cataract surgery in children carries a higher incidence of complications than in adults

**1.Increased inflammatory response** in children can lead to fibrinous reactions, pigment deposits on the IOL, decentration of the IOL, and posterior synechiae. Intensive steroid therapy post operatively is utilised to combat this problem.

**2.Secondary glaucoma** is the most feared complication of pediatric cataract surgery, and is commonly seen in infants.

- Closed-angle glaucoma may occur in the immediate postoperative period in microphthalmic eyes secondary to pupillary block.
- Secondary open-angle glaucoma may develop years after the initial surgery; it is therefore important to monitor the intraocular pressure long-term

Measurement of central corneal thickness in aphakic as well as pseudophakic eyes is essential to diagnose true glaucoma and monitoring for glaucoma is mandatory for any child undergoing cataract surgery early in life.

**3.Visual axis opacification** remains the most common complication after pediatric cataract surgery despite doing PPC, anterior vitrectomy, use of in the bag hydrophobic acrylic IOL. Severe capsular phimosis can also occlude the visual axis. Nd:YAG laser capsulotomy or surgical membranectomy can be performed to clear the visual axis. Sealed-capsule irrigation using either distilled water or 5-fluorouracil has been shown to be successful in reducing the incidence of visual axis opacification in adult eyes by killing the LEC but have potential risk of endothelial loss.

**4.Retinal detachment** is an uncommon and usually late complication

**Vision rehabilitation**

With regard to optical correction for the aphakic child, the two main considerations are age and laterality of aphakia. The modalities for visual rehabilitation after pediatric cataract extraction include the following:

1	<b>Spectacles</b> are useful for older children with bilateral aphakia.
2	<b>Contact lenses</b> provide a superior optical solution for both unilateral and bilateral aphakia. Tolerance is usually reasonable until the age of about 2 years, although after this period problems with compliance may develop as the child becomes more active and independent.
3	<b>IOL implantation</b> is increasingly being performed in younger children and appears to be effective and safe in selected cases. Awareness of the rate of myopic shift which occurs in the developing eye, combined with accurate biometry, allows the calculation of an IOL power targeted at initial hypermetropia (correctable with spectacles) which will ideally decay towards emmetropia later in life. However, final refraction is variable and emmetropia in adulthood cannot be guaranteed.
4	<b>Occlusion</b> to treat or prevent amblyopia is essential. Atropine penalization may also be considered. Compliance with patching has been shown to be associated with good postoperative visual outcomes in unilateral and bilateral cataracts. Educating the parents about the importance of amblyopia therapy postsurgery is crucial.

**Recommendations and Future directions**

Childhood cataract remains a challenge to pediatric ophthalmologists despite recent major breakthrough in surgical techniques and instrumentation. We need to tackle this problem more seriously and sensitively.

1. Immunization programmes for rubella and measles and neonatal screening should be more intensified in developing countries as prevention of disease is always better than its cure.
2. Future research in developing formulas for accurate biometry predicting refractive surprises and techniques for preventing PCO is warranted. Capsule-irrigating devices customized for pediatric eyes, dexamethasone-coated IOLs, are showing promising results but needs to be studied well.

- 3** Evaluation by a geneticist can be helpful in determining the inheritance pattern and to identify associated syndromes. Understanding of molecular genetics have been a major breakthrough achieved in the field of congenital cataracts in these recent years and these informations need to be applied practically by genetic counsellors during marriages.

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