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# PEDIATRIC CATARACT

Ophthalomology

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## ABSTRACT

Paediatric cataract is a treatable leading cause of childhood blindness, fighting childhood blindness is a priority of vision 2020: The right to sight, which is a global initiative aimed at reducing the world's burden of avoidable blindness. Early diagnosis and surgical intervention is critical to prevent irreversible amblyopia. Most of the paediatric cataract are idiopathic, autosomal dominant mode of inheritance has also be seen or it can also be associated with certain syndromes like Downs syndrome, Nance-horan syndrome, Lowes syndrome, Noories syndrome. Pathology underlying the development of paediatric cataract is problem in biosynthesis of lens proteins like crystallins, membrane protein, cytoskeletal protein and aggregated crystallins. Certain preoperative evaluation like assessment of visual acuity, head posture, nystagmus, plan for ultrasound bio microscopy, optical coherence tomography, keratometry, and axial length measurement which is required for calculation of IOL power. It is beneficial for bilateral total congenital cataract to undergo surgery at 6 months of age. Surgical technique includes extraction of cataract and managing the posterior capsule which is important to prevent postoperative posterior capsule opacification. Primary or secondary IOL implantation can be done, an alternate technique called as optic capture is developed to prevent posterior capsular opacification. Certain postoperative complications like visual axis opacification, glaucoma, postoperative reaction and retinal detachment are commonly seen.

## KEYWORDS

## INTRODUCTION:

Paediatric cataract is a treatable leading cause of childhood blindness. It is responsible for approximately 10-20% of blindness in children worldwide[1]. Fighting childhood blindness from congenital cataract is a priority of vision 2020:

The right to sight, a global initiative aimed at reducing the worlds burden of avoidable blindness[2]. Critical period of vision development, binocular interaction and stereopsis range from 2 to 6 months of age, management of paediatric cataract and IOL placement is well defined above the age of 2 years[3]. Early diagnosis and surgical intervention is critical to prevent irreversible amblyopia[4]. Poor IOL power predictability, increased inflammation, post-operative complications, technical difficulty and post-operative visual axis opacification are the main concerns of paediatric cataract surgery[5].

## Etiology:

Mostly paediatric cataracts are idiopathic, autosomal dominant mode of inheritance is also seen, can be associated with certain syndromes like nance horan syndrome, Downs syndrome, Lowes syndrome, Noories disease. Metabolic cataract due to accumulation of galactilol leading to oil droplet cataract, TORCH infections, peters anomaly, traumatic cataract and uveitic cataract[6].

## Pathogenesis:

Lens proteins like crystallins, membrane protein, cytoskeletal proteins and aggregated crystallins . Any problem in the biosynthesis of these proteins may cause cataract. Major phospholipid associated with human cell membrane is sphingomyelin, any problem altering lipid composition and distribution may cause cataract[7].

## Cataract Morphology[7] :

### Cataract involving whole lens:-

| | |
|---|---|
| Total cataract | Downs syndrome, metabolic, rubella, sporadic, syndromes |
| Congenital morgagnian cataract | |
| Disc like and membranous cataract | Trauma, rubella, Lowe syndrome, lenticonus |

### Central cataracts :-

| | |
|---|---|
| Lamellar or Zonular cataract | AD, Galactosemia |
| Central pulverulent cataract | AD |
| Nuclear cataracts | AD, rubella, oil droplet cataract, galactosemia |
| Oil droplet cataract | AD |
| Cortical cataract | AD |
| Cerulean, floriform | AD |

### Anterior cataract :-

| | |
|---|---|
| Anteror polar cataract | |
| Dot like | AD, aniridia, RB |
| Anterior pyramidal | |
| Anterior subcapsular | Uveitis, trauma, atopic, skin , alport |
| Anterior lenticonus | Sporaic, AR,AD,Microcornea |

### Posterior cataract :-

| | |
|---|---|
| Mittendrofs dots | |
| Posterior lenticonus | Catarct forms a capsule bows and lens fibres are distorted , Xlinked and AD |
| Posterior cortical cataract | AD |
| Posterior subcapsular | Myotonic dystrophy, Turners syndrome, Fabrys syndrome. |
| Punctate lens opacities | Downs syndrome, Lowes syndrome, Nance horan, Fabrys disease |
| Sutural cataract | |
| Wedge shaped cataract | |
| PHPV | |

## Pre-operative Evaluation :

### History :

Evaluation of child with cataract begins with a detail history including prenatal history, family history, maternal drug use and febrile illness with rash and birth history especially birth weight, since low birth weight maybe associated with idiopathic bilateral congenital cataract. A developmental history should be carefully assessed and if required review should be sought to exclude metabolic or systemic related 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 PFV, lenticonus, lentiglobus[4].

## Genetic evaluation:

Genetically based cataracts accounts for 8-29% of congenital cataracts with majority of them being autosomal dominant. Mutations in the genes responsible for the maintenance of lens clarity such as crystalline and connexin gene are most commonly described in the etiology of non-syndromic inherited cataracts. Mutations in alpha-crystallin gene tend to cause nuclear, lamellar, zonular and posterior polar cataract. Anterior polar cataracts are commonly seen with PAX6 mutations with or without aniridia, whereas PITX3 mutations predominantly cause posterior polar cataracts. Genes responsible for major syndromic cataracts include OCRL (Lowe syndrome), GALK117q (Galactosemia), GLA(fabrys disease), and NHS (nance horan syndrome)[8].

## Visual acuity:

First look for fixation. In infants look for visual evoked responses,

catford drum, optokinetic nystagmus and tellers acuity card. Among 1-2 years old children look for worth's ivory ball test, Boerks cany test, 2-3 years old children look for miniature toy test, coin test , LEA symbol test, 3-5 years old test with allens picture card , lippamans HOTV test and letters test can be done. In children aged more than 5 years tumbling landolts broken ring, Snellen's chart and log MAR chart can be used[6].

**Head position and nystagmus:**
Nystagmus develops because child cannot maintain fixation , rarely child might acquire a head position that helps to get maximum vision with minimum amplitude of nystagmus. This sensory type of nystagmus develops by 13 weeks of age when cataract is left untreated[6].

**Anterior segment examination:**
Look for anterior segment abnormalities like corneal opacity, shallow anterior chamber, peripheral anterior synechia in peters anomaly, micro cornea, posterior synechia in uveitis, key hole pupil (iridofundal coloboma) and enlarged ciliary process with vessels on lens in persistent fetal vasculature, look for red reflex with direct ophthalmoscope kept at 30cm and focused on each pupil separately helps in identification of lenticular opacity[6].

**Investigations :**
**Ultrasound bio microscopy:**
To analyse the sulcus, sulcus to sulcus measurement, integrity and the presence of anterior capsule for secondary intraocular lens and lens thickness. It is also important in identifying anterior persistent hyperplastic primary vitreous, posterior capsular defect and posterior polar cataract preoperatively which helps in better management of the case. Ultrasound biomicroscopy also helps us in posttraumatic cases to look forcyclodilalysis, subluxation, and foreign body localization in anterior segment and rupture of posterior capsule. It can also be used for planning the microvitreoretinal entry in anteriorly dislocated lens in case of spherophakia[7].

**Optical coherence tomography:**
It is non-invasive, non-contact technique for imaging the anterior and posterior segment of the eye with a high resolution. The power of lens calculation from extended depth OCT is more accurate. The retinal nerve fibre layer thickness is different in ethnicities, it is also negatively correlated to axial length. The central macular edema can sometimes be missed on clinical examination, which can be captured on OCT. OCT can be used for checking the vaulting of IOL, placement of anterior chamber IOL, angle structures and anomalies[7].

**Axial length:**
Axial length increases rapidly in first 6 months (0.62mm/month) then has a relatively slower growth till 18 months, followed by a slow growth (0.01mm/month). Axial length growth postoperatively is also variable in children, hence the absolute error in children is higher compared to adult population. Axial length measurement has been shown to be better estimated with immersion A-scan than A scan only because of compression of the anterior surface of the cornea. Axial length measurement made with a contact technique are on an average, 0.24-0.32 mm less than measurements made using an immersion technique[7].

**Keratometry:**
Obtained using handheld autokeratometer with a manual keratometer in older children under general anaesthesia. Keratometry steeply reduces in the first 6 months i.e. -0.40D/month, -0.14D/month in the second semester, and -0.08D/month in the second year. Corneal curvature reaches adult range at 3 years of age. Girls have steeper cornea than boys. Axial length has a linear relationship with keratometry, as the axial length increases, keratometry decreases[7].

**IOL power calculation:**
There are two methods to calculate the power od IOL , first is ultrasound biometry which is of two types again immersion, applanation , second one being the optical biometry[9].

**Degree of under correction:**
Determining IOL power in congenital cataract surgery is an important issue due to increasing axial length and refractive power of paediatric eyes after cataract surgery and also an additive effect of IOL implantation on eye growth. Paediatric eyes in comparison to adult eye have shorter axial length, steeper cornea with higher keratometry values, small anterior chamber depth, and myopic shift will occur with increasing age. Hence there are recommendations for under correction of IOL power and hyperopic outcome so the patient will be postoperatively hypermetropic which will shift to emmetropia during childhood to reduce the necessity of IOL exchange. As a result there should be an under correction of 20% in infants and 10% in toddlers. However this may make the child prone to amblyopia and make them need corrective glasses or contact lens with a periodical change in power[9].

**IOL power formula:**
IOL power formula are classified into two types regression formulas such as SRK 2 formula and theoretical formulas such as Hoffer and Holladay and SRK/T formula[9].

**Choice of IOL:**
Compared to hydrophilic acrylic lenses, hydrophobic acrylic lenses shows superior reduction rates of posterior capsular opacification and laser capsulotomy. IOL with square edges inhibits lens epithelial cells migration and PCO formation. The perfect IOL would be with a hydrophilic anterior surface and a posterior hydrophobic lens[9].

**Timing Of Surgery:**
It might be safe and more beneficial for bilateral total congenital cataract patients to undergo surgery at 6 months of age. First the average sleeping time with closed eyes of infants aged from 3months to 6months is approximately 15 hours per day, including 5 hours of daytime sleep, which is similar to average sleeping time in new-borns. Second younger infants have a greater risk of experiencing adverse effects during general anaesthesia due to immaturity of cardiovascular, pulmonary, thermoregulatory and gastrointestinal systems, as well as liver and kidneys. Third visual system retains considerable plasticity with significant individual difference even when early blindness extends beyond the critical periods. Performing aggressive interventions to achieve early visual experience at the expense of increased risk associated with aesthesis and postoperative management may be unwarranted especially for those with high plasticity[2].

**Surgery [4]**
**Wound:**
Catarct surgery in children is challenging, because of increased scleral elasticity, thicker cornea, increased risk of trauma from eye rubbing, less with activity restriction, and most importantly the effect of postoperative astigmatism on amblyopia. While scleral tunnel incision was preferred by most paediatric cataract surgeons because it was thought to induce less postoperative astigmatism, recent studies have shown clinically insignificant difference between the two types of incisions with spontaneous regression of astigmatism over a period of time. Superior incision are commonly preferred compared to temporal approach, probably in view of less risk of injury and postoperative endophthalmitis. As the flattening of te cornea occurs along the incisional meridian, there is a tendency toward with the rule astigmatism with the temporal approach in young children. The temporal approach is desirable in deep seated eyes and in children with previous or planned filtration surgery superiorly. To prevent wound leakage due to the reasons mentioned, suturing of all the wounds including paracentesis is recommended.

**Capsulotomy :**
Continues , smooth and well centered anterior capsulotomy is a prerequisite for safe lens implantation currently used techniques for paediatric anterior capsulotomes include vitreorhexis, manual continues curvilinear capsulorrhexis, canopeners, and radio frequency diathermy , other devices such as plasma blade, dicapsulotomy have been suggested to minimize zonular tension and prevent peripheral extension. Elasticity and thickness of anterior capsule in young children makes manual continuous curvilinear capsulorrhexis the most difficult technique. It can be performed using cystitome or forceps. Although manual capsulorrhexis is gold standard, some surgeons prefer to use vitrector for very young children. The two incision push pull technique developed by nischel has been shown to produce a consistent size capsulorrhexis opening with minimal risk of extension or tears. For proper centration of IOL anterior capsulotomy should be smaller than the IOL optic.

**Technique of cataract extraction:**

Lens removal can be conducted through an anterior approach by manual irrigation and aspiration or through the pars plana using a vitrctor. In case of membrane cataract or retro lenticular membranes in PFV intraocular scissors are required. An anterior chamber maintainer used for continuous irrigation prevents the collapse of anterior chamber during withdrawal of instruments during the surgery.

**Posterior capsule management:**

Maintenance of clear visual axis is critical for a good postoperative visual outcome after paediatric cataract extraction. If posterior capsule is left intact, 100% of eyes less than 4 years of age develop significant posterior capsule opacification. The anterior vitreous face acts as a scaffold for proliferation of lens epithelial cells, therefore it is essential to combine primary posterior capsulotomy with anterior vitrectomy. Posterior polar capsulotomy can be performed either through a limbal or pars plana approach. The opening for posterior capsulotomy should be smaller than the anterior capsulotomy. Staining of posterior capsule with tryphan blue 0.1% may facilitate the procedure of posterior capsulotomy. ppc using the two incision push pull technique has been shown to give considerable results without vitreous loss during the procedure. Optic capture of IOL was considered another surgical technique that would obviate the need for anterior vitrectomy and prevent posterior capsular opacification. Postoperatively subconjunctival or intracameral steroids are recommended to suppress inflammation in the immediate post-operative period.

**Primary V/s Secondary Iol Implantation [10]:**

**Primary IOL implantation:**

Options for optical correction after paediatric cataract surgery are primary IOL implantation, aphakic glasses and contact lenses. One of the most important preoperative consideration is whether to implant an IOL. The capability of IOL to offer constant visual input is an important advantage for better visual outcomes after paediatric cataract surgery. Use of an IOL provides at least a partial optical correction at all times. The important concerns about primary IOL implantation during infancy are the technical difficulties of implanting an IOL and selecting an IOL power along with higher rate of visual axis opacification. A randomised clinical trial evaluating primary aphakia versus primary IOL implantation in children under 2 years of age with bilateral cataracts. At 5 years follow up both groups achieved the same visual acuity. IOL group achieved final visual acuity much faster than the aphakic group. Presence of IOL ensured a much faster visual recovery and better visual rehabilitation. Rate of complications like glaucoma were comparable between two groups. Based on results it is believed that primary IOL implantation in children's less than 2 years of age with bilateral cataracts is safe and effective option. Hydrophobic acrylic IOL are preferable as they offer better biocompatibility and decrees incidence of visual axis opacification. With hydrophobic acrylic IOL causing a delayed onset of posterior capsular opacification. In the bag fixation is most preferred site of IOL implantation although IOL may also be implanted in ciliary sulcus with caution in case of inadequate posterior capsular support.

**Secondary IOL implantation:**

Eyes that are left aphakic are likely to require a secondary IOL implantation. It is important to leave behind sufficient anterior and posterior capsular support at the time of cataract surgery to facilitate in the bag or sulcus IOL implantation once the child and the eye grows.

**Optic capture:**

An alternative technique to stabilise the IOL was introduced with capturing an IOL optic through anterior capsulorrhexis. Haptics were placed in the ciliary sulcus and IOL optic was then placed through the anterior capsulorrhexis to capture the IOL for stable optic fixation. This concept has been used to develop the technique of posterior capsulorrhexis with optic capture through posterior capsulorrhexis to prevent posterior capsular opacification. In optic capture there is fusion of anterior and posterior capsule leads to sequestration of lens epithelial cells with in the capsular fornices and prevents their migration to the intact anterior vitreous face[11].

**Post-operative complications:**

Glaucoma following cataract surgery is observed in 10-25% cases. Visual axis opacification is the most common complication after paediatric cataract surgery with or without IOL surgery and may cause visual deprivation amblyopia. In a thick visual axis opacification, posterior capsulotomy combined with anterior vitrectomy is required. Postoperative reaction is seen more commonly in pre-existing complicated and traumatic cataract. This can result in posterior synechiae formation, leading to seclusion pupillae, iris bombe, and subsequent secondary angle closure glaucoma and visual axis opacification. Retinal detachment is rare but more common in eyes with persistent fetal vasculature[6].

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