Pediatric Cataract: A short review

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Abstract

With the advent of modern surgical techniques, pediatric cataract has become much more manageable. Intraocular lens (IOL) implantation is the standard care for patients over the age of 2 years. The use of IOL in young infants is still controversial. In addition, there are still unresolved issues, such as the minimum age at which IOL can be safely implanted, IOL power selection and IOL power calculation.

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Introduction
A child becomes bilaterally blind every minute, primarily within developing nations. Of the 1.5 million blind children in the world, 1.3 million live in Asia and Africa, and 75% of all causes are preventable or curable. The prevalence of blindness varies according to the socioeconomic development of the country and the mortality rate of those under 5 years of age. In developing countries the rate of blindness can be as high as 1.5 per 1000 population. Compared to industrialized countries, this figure is 10 times higher. [1-6]

Table 1 summarizes some of the studies regarding prevalence of childhood blindness in various developing countries. As reported by Foster and Gilbert, [2] about 0.5 million children become blind each year (Table 1).

Table 1: Cataract as a cause of childhood blindness

<table>
<thead>
<tr>
<th>Study</th>
<th>Country</th>
<th>Percentage of blindness</th>
</tr>
</thead>
<tbody>
<tr>
<td>Moriarity(1988)[3]</td>
<td>Jamaica</td>
<td>39%</td>
</tr>
<tr>
<td>Rahi et al (1995)[6]</td>
<td>India</td>
<td>12%</td>
</tr>
<tr>
<td>Waddell et al (1998)[7]</td>
<td>Uganda</td>
<td>30.7%</td>
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</tbody>
</table>

Etiology of childhood cataract:

Causes for Bilateral cataract:
1. Idiopathic
2. Familial
4. Craniofacial syndromes
5. Renal
7. Maternal infections (TORCH Complex)
8. Ocular anomalies
   - Aniridia
   - Anterior segment dysgenesis syndrome
9. Toxic (corticosteroids, radiation)

Causes for unilateral cataract:
1. Idiopathic
2. Ocular anomalies
   - Persistent fetal vasculature (PFV)
   - Anterior segment dysgenesis syndrome

- Posterior segment tumors
3. Traumatic

Identification of the etiology is the first essential step in the management of pediatric cataract. Therefore, ophthalmologists need to work with pediatricians to explore the family history and also the possibility of intrauterine infection, metabolic disorders or trauma as a cause of the condition. [7-10]

Morphological classification of pediatric cataract
1. Diffuse/total
2. Polar cataract: anterior polar and posterior polar cataract
3. Nuclear cataract
4. Lenticous: anterior and posterior lenticous
5. Subcapsular cataract: Anterior and Posterior cataract
6. Sutural Cataract
7. Membranous cataract
8. Blue dot cataract
9. PHPV
10. Traumatic

Evaluation of pediatric cataract
1. Family history [autosomal dominant or X-linked]
2. Pediatric physical examination
3. Ocular Examination including:
   - Corneal diameter
   - Iris configuration
   - Anterior chamber depth
   - Lens position
   - Cataract morphology
   - Posterior segment
   - Rule out posterior segment mass
   - Rule out Retinal Detachment
   - Rule out optic nerve stalk to lens
   - Intraocular pressure

Laboratory investigations (esp. for bilateral cataract)
- Urine for reducing substances to rule out Galactosemia.
- Blood levels of sugar, calcium, phosphorous and Red cell galactokinase level.
- Serum ferritin.
- Urine amino acids for Lowe syndrome in case of developmental delay, glaucoma, cataract.

Indications for surgery:
Depends on:
- Effect of cataract on vision “visually significant cataract”, which in turn depend on type, density, location of lenticular opacity and laterality.
- Chances of amblyopia.
  - Unilateral partial cataract:
  - Any central or posterior dense lenticular opacity >3 mm { visually significant}
  - Loss of central fixation
  - Sensory strabismus
- Unilateral complete cataract:
  - Surgery less than 8 weeks
- Bilateral partial cataract:
  - Visual acuity in either eye 20/70
  - Nystagmus
  - Sensory strabismus

Counseling of the Parents regarding poor prognostic factors:
Realistic expectations of visual outcome should be given to the parents’ esp. in following conditions having poor prognosis:
1. Unilateral cataract (Bilateral cataract has good visual outcome)
2. Late presentation
3. Poor initial vision
5. Development of nystagmus in B/L cataract.

Development of strabismus in U/L cataract and nystagmus in B/L cataract are late signs that the opacities are visually significant and optimal time for surgery is past.

TIMING OF SURGERY:
- In cases of Unilateral dense cataract or any central or posterior dense lenticular opacity >3 mm are considered visually significant and need early surgery < 8 weeks of life to prevent visual deprivation amblyopia. [11-13]
- In bilateral cataract, if one has already been operated, the other eye must be operated within 2-3 weeks to prevent amblyopia.

Surgical options
There are a variety of surgical options, depending on the age of the patient and the personal experience of the surgeon.
1. Lens aspiration
2. Lens aspiration with anterior vitrectomy
3. Lensectomy

The standard surgery for infants less than 6 months old is lens aspiration with primary posterior capsulectomy and anterior vitrectomy. Whether IOL should be implanted in infants less than 6 months old is still controversial, although there are increasing numbers of surgeons opting for intraocular lens (IOL) implantation for patients less than 6 months old. The ongoing USA multicentre trial – Infant Aphakia Treatment Trial – may provide an answer to this issue. There is increasing evidence that IOL implantation is safe for patients under 2 years old. It has been suggested that this should be the standard of care for patients over the age of 2 years. [15-18]

MANAGEMENT OF APHAKIA
1. Aphakic glasses: are suitable mode of aphakic correction in the following:
   - In older children.
   - In bilateral aphakia.
   - Conditions where IOL can not be inserted.
   - In children who are intolerant to contact lens.
2. Contact Lens:
   Extended wear contact lenses are used especially in very young children. Contact lenses should be prescribed as early as possible after the surgery. They can be given in bilateral as well as unilateral aphakia. In infants 2.5-3 D overcorrection is done for clear near vision. The choice among contact lenses for use in pediatric aphakia in most cases is silicone material.
3. IOL Implantation:
   At present IOL implantation has become the standard treatment at least for children older than 2yr of age. The IOL implantation in children in less than 2yrs is still debatable, although there is increasing number of surgeons opting for IOL implantation for children less than 6m but the complication rate particularly re-opacification of the visual axis is much higher in infants than the older children.

The other complicating factors peculiar to infants include the following:
- The eyes are smaller & soft making the surgery technically challenging
- Most of the growth in axial length & corneal curvature is yet to occur making the optimal IOL power difficult to determine.
- Performing the anterior CCC is also a surgical challenge as the capsule is much more elastic and notorious & difficult to control. [19]

Significant improvement in surgical tech such as CCC, hydro dissection, in the bag fixation of IOL & the management of posterior capsule in the form of PPC (Primary posterior capsulectomy) with anterior vitrectomy are used today for pediatric cataract.
Contraindications for IOL implantation:
- Microphthalmia
- Aniridia
- Recurrent Uveitis
- Rubella Cataract
- Glaucoma
- Inadequate capsular support

IOL Power calculation
In older cooperative children, standard biometry may be performed. In younger & uncooperative patients, axial length & keratometry should be done before surgery under GA.

Refraction & IOL (Dalen et al 1997)
- Child < 2yrs- Do biometry & undercorrect by 20% or use axial length only
- Child 2-8yrs- Do biometry & undercorrect by 10%

<table>
<thead>
<tr>
<th>AXL mm</th>
<th>Power (D)</th>
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<tbody>
<tr>
<td>17</td>
<td>28</td>
</tr>
<tr>
<td>18</td>
<td>27</td>
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<td>19</td>
<td>26</td>
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<td>22</td>
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In order to reduce rates of IOL exchange and resurgery, they suggested that infant should receive 80% of the IOL power needed for emmetropia while in age group of 2-8yrs; IOL power should correct 90% of aphakia.

IOL power calculation (used by us)
Do biometry & undercorrect according to age:

<table>
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<tr>
<th>Age (yrs)</th>
<th>Undercorrection (D)</th>
</tr>
</thead>
<tbody>
<tr>
<td>2-3</td>
<td>3-4</td>
</tr>
<tr>
<td>4-5</td>
<td>2-3</td>
</tr>
<tr>
<td>6-7</td>
<td>1-2</td>
</tr>
<tr>
<td>8-10</td>
<td>0.5-1</td>
</tr>
<tr>
<td>&gt;10</td>
<td>Same power</td>
</tr>
</tbody>
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IOL lens material –
Both single piece PMMA & foldable acrylic lenses have been widely used in pediatric cataract surgeries over years. IOL with overall diameter 11-12mm with optic size 5.5-6mm are suitable. Foldable IOL is preferred over rigid IOL. Among the foldable IOL’s used for children hydrophobic acrylic IOL’s are suitable. [20]

Postoperative Complications:

Posterior Capsule Opacification (PCO):
PCO is a frequent complication after cataract surgery in children. The reported risk of PCO in children can be as high as 95%, and its occurrence prevents visual rehabilitation [21-23]. The incidence of capsular opacification is related to the age of the patient and degree of postoperative intraocular inflammation [24]. The PCO could either be managed surgically with pars plana anterior vitrectomy and capsulectomy or with laser addition. Sutureless pars plana vitrectomy through self-sealing sclerotomies has been successfully performed in children with PCO [25].

Use of the 25-gauge transconjunctival sutureless vitrectomy system (TSV) would obviate the need for dissecting scleral tunnels, and may also increase the ease of instrument manipulation in children with small eyes and palpebral fissures [26].

Because the pars plana is not well developed in young children, the site of sclerotomies differs according to the age of the patient, as shown in Table 2 which is derived from the literature, the authors’ experience and on an anatomical basis. [27-28]

<table>
<thead>
<tr>
<th>Age of patients</th>
<th>Site of sclerotomies (posterior to limbus)</th>
</tr>
</thead>
<tbody>
<tr>
<td>3 months</td>
<td>1.0 mm</td>
</tr>
<tr>
<td>4-6 months</td>
<td>1.5mm</td>
</tr>
<tr>
<td>7-12 months</td>
<td>2. mm</td>
</tr>
<tr>
<td>&gt;12 months</td>
<td>2.5 mm</td>
</tr>
</tbody>
</table>

Acrylic IOL has been related to a significantly lower PCO rate than older IOL types, especially PMMA lenses. [29]

The use of a 3-mm to 4-mm primary posterior capsule opening and anterior vitrectomy has been advocated to prevent posterior capsule opacification in children 5 years old or younger [26]. Prevention is the first step in management. Besides the prevention of PCO with PCCC and anterior vitrectomy to decrease the migration of LEC across the anterior hyaloids face. [30]

Inflammation:
Since postoperative intraocular inflammation is much more severe in children after cataract surgery and the formation of fibrinous membrane is not unusual, subconjunctival steroids and antibiotics are frequently used at the end of surgery, together with intensive topical
steroid therapy postoperatively. Topical prednisolone acetate 1% every hour to 4 hourly is commonly used occasionally in conjunction with oral steroids. Mydriatic eye drops are important in preventing posterior synechiae and reducing the chance of pupillary block. [31]

Glucoma:
Aphakic glaucoma has emerged as a serious long-term problem after lensectomy in infancy. A more recent study reported a decreased incidence of open-angle glaucoma in pseudophakic eyes compared to aphakic eyes after cataract surgery. [32-34]

Rhegmatogenous Retinal Detachment:
The occurrence of rhegmatogenous retinal detachment after pediatric cataract surgery is one of the late complications. There is scant literature regarding this issue.

The reported average interval between lens operation and retinal detachment is 23 to 34 years. [35]

Conclusion
There are increasing numbers of pediatric cataract surgeons who opt for IOL implantation rather than leaving the infant aphakic. The generally accepted minimum age of IOL implantation is 1 year to 2 years and more surgeons are implanting IOL for infants between 6 months and 1 year old. For those under 6 months old, IOL implantation is still highly controversial and is currently being studied in the Infant Aphakia Treatment Trial. Surgery is only one part of the entire management of the pediatric cataract patient. Participation in the visual rehabilitation of the child involving parents, ophthalmologists, pediatricians and optometrists cannot be overemphasized.

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Competing Interests
None declared.

References

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