Management of Congenital Cataract

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Abstract

Pediatric cataracts remain a very important and difficult problem to manage, in spite of dramatic advances that have occurred in the field over the past few years. The aim of the present review is to update the reader on advances in the field of pediatric cataract surgery and to present concepts of the three important problems in the management of congenital cataracts: 1) Technical aspects of cataract surgery 2) Changing refraction 3) Unpredictability of functional outcome.

Bilateral Congenital Cataract is the most common cause of treatable blindness in children, world wide. In developing countries, the prevalence of blindness from cataract is higher, about 1 to 4 per 10,000 births.

Etiology

The cause of bilateral congenital cataract in most cases is idiopathic. The most common etiologies include intrauterine infections like Rubella, Toxoplasmosis, Herpes Simplex and Varicella. About one third of cases are hereditary, without a systemic disease. Inherited cataracts differ morphologically within the same pedigree. These are mainly autosomal dominant, but autosomal recessive and X linked traits occur.

Galactosaemia and Hypocalcemia, are metabolic disorders with congenital cataract. Infants with classical galactosaemia develop oil droplet cataracts, and if left untreated, these progress to lamellar and then total cataracts due to accumulation of galactitol in the lens. However if galactose is eliminated from the diet of these children, the cataracts may become transparent again. Hence it is important to test for the presence of reducing substances in the urine after a galactose containing meal [milk] in all infants with cataract. Enzymatic assays and DNA studies can then be used to confirm the diagnosis.

Hypocalcemia leads to seizures, failure to thrive, and irritability in children. Altered permeability of lens capsule results in cataract. These cataracts generally begin as fine white punctate opacities scattered throughout the lens cortex which may then progress to lamellar cataracts. Serum Calcium and Phosphorus levels should be measured in infants with congenital cataracts.

Cataracts are manifested in large number of syndromes with systemic abnormalities like, Trisomy 21, Turners syndrome, Trisomy 13, Lowes syndrome, Alports syndrome, Nance Horan syndrome, Marisolfi syndrome, Marinesco-Sjogren syndrome, etc to name a few. Associated mental retardation is common. Many genes involved in cataractogenesis have been identified.

Investigations

In unilateral cases and in an otherwise healthy infant with one parent involved by the disease, an extensive preoperative investigation may not be necessary to establish the cause for cataract. Antibody titres for rubella, toxoplasmosis, herpes simplex and urine examination for reducing substances should be done in all cases. Further investigations like plasma electrolytes, amino acid studies, enzyme studies and chromosome studies need be carried out only in appropriate cases and with the collaboration of a pediatrician and is often not rewarding.
Morphology

Nuclear cataract is usually present at birth and is non progressive. In cases with dense cataracts present at birth it is usually nuclear. The opacification is located in the embryonic and fetal nuclei between the anterior and posterior Y sutures and is usually very dense in the center. The eyes may be smaller than normal. The cataract is bilateral in 80% of cases, and inheritance can be demonstrated in 30% to 50% of cases.

Posterior cataract in infants and children is commonly associated with PFV (Persistent Foetal Vasculature), and the affected eye is microphthalmic. The retrolental vasculature may be in contact with the lens capsule and may bleed during surgery. Traction retinal detachment and secondary glaucoma are common post operative complications.

Lamellar cataract usually develops after fixation is established, is usually progressive and involves the lamellae surrounding the fetal nucleus peripheral to the Y sutures. Eyes are normal sized with normal corneas, and the cataract is uniform bilaterally and has an autosomal dominant inheritance. Surgery can often be delayed and is undertaken when visual demands are compromised.

Other morphological types like sutural cataract, anterior polar cataract etc have less influence on vision.

Assessment

Visual loss and development of amblyopia depend on the size, location, and density of the opacity. If the opacity is large enough to obscure fundus view through an undilated pupil, amblyopia development can be expected. If the retinal details such as the larger vessels can be distinguished through the central portion of the cataract, conservative treatment can be considered, but occlusion therapy is necessary in unilateral cases and constant follow up to evaluate the monocular and binocular visual behavior should be undertaken.

Visual assessment should be performed using patterns of fixation and supplemented when possible by preferential looking charts, or pattern visual evoked potentials. Measurement of corneal diameter, intraocular pressure, pupillary reflexes, ultrasonography and indirect ophthalmoscopy should be carried out. (Table 1 & Table 2)

Examination Protocol in Paediatric Cataracts

Table 1.

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<tr>
<th>History</th>
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<tr>
<td>1. Duration</td>
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<td>2. F/H of Congenital Cataract</td>
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<td>3. Visual Status: Ambulation in familiar and unfamiliar surroundings</td>
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<td>4. Behavioural Pattern and School Performance</td>
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<tr>
<th>Birth History</th>
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<tr>
<td>1. History and Degree of consanguinity</td>
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<td>2. H/O maternal infection in 1st Trimester</td>
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<td>3. Gestational Age &amp; Birth Weight</td>
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<td>4. Birth trauma</td>
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<td>5. Supplemental O2 therapy in Perinatal period</td>
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Table 2.

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<th>Ocular Examination</th>
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<tr>
<td>1. Visual Acquity and Fixation Pattern</td>
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<td>2. Refraction</td>
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<tr>
<td>3. Cover – Uncover test (Hirschberg’s)</td>
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<tr>
<td>4. Note Nystagmus if any</td>
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<tr>
<td>5. SLIT LAMP EXAMINATION</td>
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<tr>
<td>➢ Associated Congenital Anomalies of iris, lens</td>
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<tr>
<td>➢ Type of Cataract</td>
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<tr>
<td>➢ Iridodonesis / Phacodonesis</td>
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<tr>
<td>6. Tension applanation if possible</td>
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<td>7. Fundus examination if possible</td>
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<td>8. B.Scan USG if there is no fundus view.</td>
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Timing of Surgery

Dense Congenital Cataract: From the available data it would appear that the optimal time to remove a dense congenital cataract in an infant and to initiate treatment is when the child is 4-8 weeks of age. Cataract surgery before 4 weeks of age appears to increase the risk of secondary glaucoma, whereas waiting beyond 8 weeks of age compromises visual outcome. The visual system which is immature at birth has a latent period of approximately 6 weeks before it becomes sensitive to
Fig. 1. Congenital cataract with liquefied cortex which allows the nucleus to move to different positions depending on posture (a) Right way up (b) upside down (c) sideways

Fig. 2. Bilateral symmetrical lamellar cataracts on retroillumination. The acuity is 6/9 in both eyes

Fig. 3. Lamellar cataract with riders, the acuity is 6/24

Fig. 4. Wedge shaped cataracts as part of a lamellar cataract

visual deprivation, and binocular vision first appears at approximately 3 months of age. The pathophysiology of aphakic glaucoma is poorly understood. Its aetiology has been attributed to the damage of the trabecular meshwork by inflammation, the loss of mechanical support of the trabecular meshwork, or a toxic substance gaining access to the trabecular meshwork from vitreous and may present as an early angle closure glaucoma or late open angle glaucoma. It has been reported to occur after both limbal and pars plicata based surgeries.

If the cataract is incomplete at birth, close follow up is advised. Visual acuity should be followed and history about the visual interaction with parents should be noted. Evidence of squint or nystagmus is an indication for immediate intervention. If the child has unilateral partial cataract, occlusion therapy should be considered. 

Counselling of the parents is very important and should be overstressed. It is important to make the parents...
understand that the treatment of the child starts only after surgery. The necessity for regular follow up, need to enforce the constant wearing of glasses, or contact lens despite IOL implantation and the requirement of occlusion therapy after surgery should be emphasized during counselling.

Pre operative examination under short anaesthesia with fully dilated pupils is mandatory before surgery. Examination under the operating microscope or hand held slit lamp biomicroscope is performed to assess the type and degree of cataract. The examinations performed under anesthesia include 1) Tonometry to rule out any associated glaucoma, 2) Measurement of corneal diameter 3) Posterior Segment evaluation with an indirect ophthalmoscope whenever fundus view is possible 4) Performing a B.Scan Ultrasonography in situations where there is no fundus view 5) Keratometry with a hand held keratometer and 6) A.Scan biometry for IOL power calculation.

Surgical Technique in Children

In infants with bilateral cataracts it is advantageous to perform surgery in both eyes at the same time, to prevent an amblyopia in the second eye. If both eyes are operated at the same time, sterility must be maintained during the whole procedure, changing all instruments for the second surgery.

The lens can be approached through the limbus or pars plicata. Although temporal clear corneal incisions are favoured in adults, it may not be a good choice in pediatric cataracts. Most pediatric patients have with the rule astigmatism and temporal incisions may induce further worsening of with the rule astigmatism. Hence a superior limbal or scleral tunnel incision is preferred. Using the limbal approach, a high viscosity ophthalmic viscoelastic material should be used to overcome the vitreous pressure and prevent the shallowing of the ant. chamber. If the pupil is small, flexible iris retractors can be used to enlarge the pupil. Anterior Capsule staining with Trypan blue makes the anterior capsulorhexis easier. If an IOL is implanted the anterior capsulorhexis should be round, smaller than the optic and placed in the center. The capsule is thick and elastic in children, which makes it more difficult to perform a manual continuous capsulorhexis. The capsulorhexis opening tends to be larger than intended. The anterior capsulorhexis can be created preferably with a needle and forceps or it can also be created using a diathermy. Mechanised capsulotomy by a vitrector is easier to perform and is the third option for anterior capsule management. The vitrector should be placed with its cutting port posteriorly in contact with the intact anterior capsule. The cutter should be turned on and suction increased. Cutting rates of 150-300 cuts per minute and aspiration of 150-250 cc/min should be used for vitrectorhexis.

After rhexis most surgeons perform a hydrodissection to separate the lens capsule from the cortical material and to shear the epithelial cells away from the capsule. Hydrodissection has a shearing effect on lens epithelial cells and retards PCO. Multi quadrant hydrodissection helps in wash out of equatorial lens material. For removal of the cortical material, a phacoemulsification hand piece, a vitrectomy tip, or an automated irrigation aspiration device can be used. It is usually possible to remove the nucleus and cortex with irrigation and aspiration and heparin can be used in irrigating solution to minimize the inflammation after surgery. Phaco probe and ultrasound energy is sometimes needed in dense cataracts. The aqualase liquefaction technique using a warm waterstream would probably be helpful in removing these dense cataracts. It is important to remove all the lens epithelial cells to prevent later posterior capsule opacification.

Since the intact posterior capsule opacifies rapidly in children and maintenance of a clear visual axis is necessary to prevent amblyopia, a posterior capsulorhexis is preferred by most surgeons. The posterior capsule is thinner and inelastic than the anterior capsule and a posterior capsulorhexis smaller than the anterior capsulorhexis is performed. Sometimes rhexis is impossible and a vertical posterior capsulotomy with a needle may suffice. If fibrotic parts are found in the posterior capsule, scissors can be used. If persistant hyaloid artery is found adherent to posterior lens capsule, it should be cut with scissors, and cautery is seldom indicated. The IOL should be placed in the bag rather than the ciliary sulcus because of the complications like pupillary capture and IOL decentration after sulcus fixation.
It is debatable whether an anterior vitrectomy should be performed at the primary surgery. Inflammatory reaction in anterior vitreous is severe in children and can result in fibrous membrane formation. This acts as a scaffold for lens epithelial cell (LEC) migration and proliferation. Anterior vitrectomy is necessary in children < 2 years of age along with a posterior capsulorhexis as they are subject to severe posterior capsular opacification and intense uveal inflammation. It may not be necessary in children > 2 years or when you are implanting an IOL which has good biocompatibility with the anterior vitreous face. It can be performed through the pars plana or through limbal incision up to a depth of 2 mm. This technique appears to be a good way of preventing the formation of after cataract.

Another technique involves performing an optic capture, where, the IOL is pressed through the posterior capsulorhexis while the haptics remain in the bag. However this technique does not appear to fully prevent the formation of after cataract and it is reported that anterior vitreous face becomes semiopaque and opacification of anterior and posterior IOL surfaces can occur. Optic capture might be a good technique in some cases since it provides a good centration of the IOL which is necessary after trauma and in incomplete capsulorhexis. However optic capture is difficult or impossible with single piece IOL that does not have angulated haptics. Table 3 gives a synopsis of various types of optic capture that can be performed.

The viscoelastic should be completely removed, and no vitreous should be in the anterior chamber. The sclera is soft and elastic in children and it is hard to achieve a self-sealing incision in most cases. So the incision should be closed by sutures. Endophthalmitis is the most serious complication and prophylactic antibiotics are indicated in all cases.

### Correction of Aphakia

#### IOL Implantation-

Today most children are implanted with an IOL during surgery and the criteria of IOL implantation depend on the child's age and whether the cataract is unilateral or bilateral. It is perfectly safe and acceptable to perform primary implantation in a child older than one year. In children younger than 1 year IOL implantation is controversial. A foldable acrylic hydrophobic IOL is the most biocompatible IOL as of today. A single piece IOL is for in the bag insertion and a 3 piece for sulcus fixation. The use of multifocal IOL in young children have been studied by Jacobi et al and the results are encouraging.

#### Contact Lens-

If no IOL is implanted, contact lenses are given as early as possible to prevent stimulus deprivation amblyopia. Frequent retinoscopy should be performed to decide the power of CL and an overcorrection of +2 to +3D is mandatory. Silicon lenses or soft hydrogels are well tolerated.

#### Spectacles-

In some children with bilateral aphakia, spectacles are better tolerated than contact lenses. In addition, a secondary strabismus may be manipulated by prismatic effect of spectacles. Bifocal glasses should be prescribed when the child is about to start school.

### Factors to Consider When Deciding Between Aphakic Glasses and Contact Lenses

#### Unilateral or Bilateral Aphakia

Aphakic glasses are not suitable for monocular aphakia because of relative magnification differences.

#### Institutional and Parent Compliance Factors

When contact lens care cannot be provided by the institution or when there is poor parental compliance aphakic glasses can be prescribed.

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Table 3. Optic Capture Technique (Courtesy: Trivedi R.H. & Wilson)
Cost factor
Silicone lenses are expensive. The need to change contact lenses frequently due to change in refraction as well as frequent lens looseness adds on to the expenditure in contact lens wearers.

Occlusion Therapy- In unilateral cases occlusion therapy is started as soon as the media are clear and refraction is corrected. In bilateral cases occlusion is sometimes useful if one eye is more amblyopic than the other. These children should be followed up into adulthood.

Postoperative Complications
Secondary opacification of the visual axis (VAO) is common and is treated by an ND Yag laser or surgical intervention. Secondary glaucoma is better prevented than treated. Some cases can be controlled by local medications but surgical intervention is often required. Amblyopia and strabismus, endophthalmitis, retinal detachment, cystoid macular oedema etc are to detected and treated when needed.

Conclusion
Dense congenital cataract requires prompt surgery and the optimum time is at 4 to 6 weeks of age. To remove cataract before 4 weeks appears to increase the risk of secondary glaucoma, while waiting beyond 8 weeks compromises visual outcome. Nystagmus and strabismus are indications for immediate intervention. The treatment regimen consists of surgery within 2 months combined with immediate optical correction. IOL implantation is safe in children older than 1 year of life. Anterior capsulorhexis is mandatory. In all children younger than 2 years the posterior capsule needs to be opened during surgery or soon thereafter by ND Yag.

Anterior dry vitrectomy is recommended often, though not always. Incomplete cataracts are followed up and the timing of surgery depends on the visual status.

References


14. Wilson Jr, Trivedi Rupal, Pandey Suresh-Paediatric Cataract Surgery