

PEARLS AND PARADIGMS OF PEDIATRIC CATARACT MANAGEMENT

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Childhood cataract is a priority area for Vision 2020. It accounts for nearly 190,000 of the 1.4 million blind children in the world. It is however important to note that this is a preventable cause of blindness. The prevalence of cataract has been estimated to be 1-15/ 10,000 children. Better understanding and newer techniques in the management of pediatric cataract has resulted in better visual outcome.

OCULAR GROWTH, VISUAL DEVELOPMENT AND TIMING OF CATARACT SURGERY

- At birth the optical apparatus of the eye and optic neural pathways are immature. During first 2 years of life rapid changes occur which lead to development of accurate optics and normal optic neural pathways enhancing perfect vision.¹
- Focused images on immature retina are essential for adequate visual development.^{2,3} The neural pathways depend on transmission of formed images for adequate neuroanatomic development during this critical period.
- **Critical period** :⁴ In general critical period for any function is the time when if deprived of normal stimulus the function's development will be permanently disrupted.
- If there is sensory deprivation in first 2-3 months, nystagmus sets in, this further compromises visual results. This critical period extends from 4wks to 4 months.⁴

Visual function development

The various milestones achieved during visual development are important to prognosticate the visual outcome.

- ◆ Pupillary light reaction 30 weeks
- ◆ Blink (visual stimulus) 2-5 months
- ◆ Central Fixation 2 months
- ◆ Smooth pursuit 6-8 weeks

- ◆ Optokinetic nystagmus
Birth (restricted slow-phase velocity)
Nasal to temporal 2-4 months
- ◆ Saccades 1-3 months
- ◆ Accommodation 4 months
- ◆ Stereopsis 3-7 months
- ◆ Contrast sensitivity 7 months
- ◆ Foveal maturation 4 months
- ◆ Optic nerve myelination 7 months - 2 years

Current literature gives following recommendations for timing of congenital cataract surgery based on studies of visual development:

1. Cataract surgery during the first 6 weeks of life is associated with the best visual outcomes for children with dense unilateral congenital cataracts.^{1,5,6}
2. In children with dense bilateral congenital cataracts, the incidence of poor visual outcomes increases if cataract surgery is delayed beyond 10 weeks of age.^{1,6}
3. The absence of preoperative nystagmus is a better predictor of a good visual outcome than the age at surgery.¹
4. Major form deprivation, even after early surgery, leads to nystagmus. This is mostly manifest latent nystagmus (MLN).⁶
5. The latent period for fixation stability may be as short as 3 weeks.⁶
6. Preoperative congenital nystagmus (CN) can convert to more benign MLN after surgery.⁶

Growth of eye

Axial length :

- 1) During the first 6 months of life, axial length increases by 0.62 mm/month, then 0.19 mm/month from 6 to 18 months and 0.01 mm/month during 18 months to 18 years of age.⁷

- 2) Averaged values from the literature regarding eye size at age 1 year is 20 mm, at 3 years is 22 mm and is presumed 13 year endpoint as 23 mm.⁸

Keratometry:⁹

In premature infants	53.1±1.5D
Neonates	48.4±1.7D
1 month	45.9±2.3
36 months	42.9±1.3D

As seen from above data most of the changes in axial length occurs in first 2 years of life and keratometry becomes stable only by 12-18 months. This makes selecting IOL power more challenging under the age of 2yrs.

PRACTICAL CONSIDERATIONS BEFORE SURGERY

1. Treatment is difficult and tedious. It requires dedicated team effort and most important contributors being parents.
2. Investigations
 - ◆ Unilateral cataracts usually do not require extensive systemic work up, most are not inherited nor associated with syndromes.^{10,11}
 - ◆ Complete ocular examination is sufficient to evaluate for specific causes like PHPV, posterior lenticonus, anterior segment dysgenesis.¹⁰
 - ◆ Bilateral cataracts with no family history or recognizable syndrome should be evaluated as 60% of them can be attributed to specific etiology.^{10,11}
 - ◆ Detailed family history for mode of inheritance.¹⁰
 - ◆ Careful attention to systemic issues like hepatomegaly, failure to thrive, diarrhea.¹⁰

Recommended tests for bilateral cataracts without definite family history:^{12,13}

- ◆ Fasting blood sugar
- ◆ Plasma calcium and phosphorus (Hypothyroidism)
- ◆ Urine reducing substances after milk feeding (Galactosemia)
- ◆ Urine amino acids (Lowe's syndrome)
- ◆ RBC transferase and galactokinase levels

(Galactosemia)

- ◆ TORCH titers

3. Indications for surgery:¹⁰

- ◆ Visually significant cataract more than 3mm in diameter
- ◆ Preventing visualisation of fundus
- ◆ Associated with strabismus or nystagmus.

4. Timing of surgery:^{1,5,6,10}

- ◆ Unilateral cataract: before 6weeks
- ◆ Bilateral cataract: before 10weeks

5. Anesthesia related issues:

An anesthetist experienced in pediatric anesthesia especially in cases of very young neonates is needed.

SURGICAL ISSUES

1) Incision:

- a. Clear corneal vs. scleral:¹⁴
 - ◆ Clear corneal not a good choice:
 - ◆ increased risk of endophthalmitis,
 - ◆ more postoperative astigmatism
 - ◆ Scleral incision preferred
 - ◆ but difficult to perform due to low scleral rigidity.
- b. Location:¹⁵
 - ◆ Superior preferred since covered by lid
 - ◆ Temporal is risky in view of possibility of trauma and risk of infection.
- c. All incisions should be preferably sutured^{14,15}
 - ◆ since low scleral rigidity promotes leakage
 - ◆ children are more likely to self traumatize than other populations

2) Management of anterior capsule :

Continuous curvilinear capsulorhexis is the gold standard.¹⁶ It is a critical step to reduce intraoperative complications and long term stable centration of IOLs^{17,18}

Challenges:¹⁶

- ◆ Very thin and elastic capsule need more force necessary to be torn
- ◆ Increased incidence of radial tears and extension to equator.

How to manage:^{15,16,17,18}

- ◆ Use of stain like indocyanin green¹⁶, Trypan blue^{19,20}
- ◆ Use of high viscosity viscoelastics²¹

- ◆ Use of rhexis forceps
- ◆ Regrasping frequently
- ◆ Small rhexis usually adequate and safe
- ◆ Other options:
 - Vitreorhexis: venturi pump, cutting rate 150-300, aspiration 150-250 cc/min¹⁶. Vitreorhexis an alternative approach in children less than 6 years of age.²²
 - Radiofrequency diathermy capsulotomy: High frequency currents of 500 kHz, platinum alloy tip probe heated to approximately 160 degree F producing thermal capsulotomy.¹⁴

Stretching force needed to tear edge is much less, more radial tear during surgical manipulation like removing cataract, inserting IOL.¹⁴ For children aged 6 years and older, manual CCC is the best technique because by that age, capsule is less elastic and capsulotomy is easier to complete.²² Compared to other techniques of anterior capsulotomy, manual CCC produces smoothest, most extensible and mechanically strongest capsulotomy.^{23,24,25}

3) Cortex and nucleus removal :

Multiquadrant cortical-cleaving hydrodissection helps in following ways:²⁶

- ◆ To separate epithelial cells from capsule
- ◆ Reduces surgical time,
- ◆ Reduces amount of irrigating fluid needed
- ◆ Facilitates removal of cortex.

Instruments: Phacohandpiece, vitrectomy tip, automated IA, simcoe's canula, single port aspiration canula with anterior chamber maintainer.

Complete removal of cortex important for¹⁵

- ◆ Reducing postoperative inflammation
- ◆ Reducing incidence of posterior capsular opacification

Vitreous upthrust can be problematic.¹⁵

4) Management of posterior capsule and vitreous :

- ◆ Primary posterior capsulotomy (PPC) with anterior vitrectomy (AV) at the time of IOL implantation for children who are not expected to be candidate for Nd-YAG laser capsulotomy within 18 months of surgery.²⁷
- ◆ Rates of posterior capsular opacification decrease only after anterior vitrectomy not primary

posterior capsulotomy alone in children less than 7years of age.^{16,28,29}

- ◆ Anterior vitreous face (AVF) acts as scaffold for epithelial cells to grow.
- ◆ AVF supports metaplastic pigment epithelial and inflammatory cell growth due to breakdown of blood aqueous barrier
- ◆ Inflammatory response in small children is severe and fibrinous membranes may form on the intact AVF
- ◆ Size of PPC: 1 mm less than size of optic of IOL.³⁰
- ◆ Approach: Limbal approach preferable.³⁰
- ◆ Time: Before IOL implantation preferable.³⁰

Technique of PPC :¹⁶

- ◆ Manual primary continuous curvilinear capsulorhexis: Gold standard
- ◆ Vitreorhexis: When adequate capsulorhexis is difficult due to visibility problems, capsule toughness or elasticity²⁷
- ◆ Can be done after IOL implantation
- ◆ Radiofrequency diathermy: Less mechanical strength and increased vulnerability to surgical manipulation¹⁶
- ◆ Fugo plasma blade

Current recommendations:

- ◆ Primary PPC with AV for children < 6-7 years of age^{28,29}
- ◆ Primary PPC without AV for >7 years of age³¹
- ◆ PC left intact in children above 9-10 years of age³⁰

5) Primary IOL implantation :

Age : more than 2 years- well established, efficacious, satisfactory and safe,^{14,32,33}

less than 2 years- Increasing number of surgeons implanting^{33,34}

less than 1 year controversial^{35,36,37,38}

Advantages of IOL:

- ◆ Full time compliance with partial correction in patients that may have difficulty with full compliance¹⁰
- ◆ In correcting unilateral cataracts, better visual outcome than cases treated with contact lenses, especially when contact lens compliance is moderate to poor.³⁸

Problems in infants:^{35,36,37,38,39}

- ◆ Higher rates of postoperative complications
- ◆ Higher rates of reoperations (mainly membranectomies)

Preferred IOL:⁴⁰

- ◆ Hydrophobic acrylic 3 piece or one piece

Advantages:^{40, 41,42,43,44}

- ◆ Ability to inject lens through small wound
- ◆ Haptics unfold slowly, so easy to manipulate in bag even in presence of PCCC
- ◆ Extremely flexible haptics with excellent memory so easy to implant and not prone to deformation
- ◆ One piece construction robust in resisting capsule contracting forces
- ◆ Single piece design adapts to the smallest capsular bag without becoming decentered
- ◆ Lower rates of PCO
- ◆ Lower rates of postoperative inflammation

Size of IOL:⁴⁵

- ◆ Rapid growth phase of lens is complete by 2 years of age, so downsizing of IOL is essential before this age and standard size IOL can be implanted afterwards.
- ◆ For age < 2 years: Overall diameter 10mm
- ◆ For age > 2 years: Standard size 12-12.5mm

Absolute contraindications:^{15,30}

- ◆ Microcornea, microphthalmos, nanophthalmos... eye too small for IOL
- ◆ Uveitis

Relative contraindications:^{15,30}

- ◆ Aniridia
- ◆ Sclerocornea... cornea too flat
- ◆ Congenital Hereditary Endothelial Dystrophy (CHED)
- ◆ Rubella
- ◆ Uncontrolled glaucoma
- ◆ Primary Hyperplastic Primary Vitreous (PHPV)

Contact lenses:

Advantage:⁴⁶

- ◆ Can be changed as refraction of eye changes with growth.

Disadvantage:⁴⁶

- ◆ Contact lenses are impractical for most patients

in the developing world, because the majority live in rural areas where suboptimal living standards and scarcity of clean water makes personal and ocular hygiene difficult.

- ◆ Regular follow up visits to eye care clinics are problematic owing to cost and distance of travel.
- ◆ Contact lenses are expensive and easily lost.
- ◆ Personnel qualified to offer contact lens service are scarce.
- ◆ Need for expensive set up
- ◆ Compliance is difficult
- ◆ Visual outcome better with IOLs in unilateral cataracts^{47,48,49}

Current trend:

- ◆ To consider IOL implantation viable option on selective basis in children younger than 2 years.

Optic capture in PPC :

- ◆ Advantage:^{50,51}
 - ◆ To minimize PCO
 - ◆ Better centration of IOL
 - ◆ Avoiding AV
- ◆ Disadvantage:⁵²
 - ◆ Increased inflammation
 - ◆ Technically difficult

Current status : This technique of optic capture in PPC is less popular.

6) **Secondary IOL implantation :**

Sulcus fixated IOLs:^{53,65}

- As far as possible, prefer in the bag IOL implantation
- Viscodissection helps to dissect synechiae between iris and residual capsule
- Safety of IOL being lifelong in contact with uveal tissue not known
- Foldable IOL are avoided in sulcus, PMMA is preferred.

Scleral fixated IOLs: ⁶⁵⁻⁶⁹

- Has been well tolerated by children but their complications that have been reported are pupillary capture and subluxation

AC IOLs:⁷⁰

- Well tolerated in children if anterior chamber is

developmentally normal.

- Long term safety not known.

Current recommendation :⁷⁰

If there is inadequate capsular support for sulcus fixation, implantation of IOL is not recommended unless every contact lens and spectacle option has been explored fully.

INTRAOCULAR LENS POWER CALCULATION

Difficulties:

- ◆ Changing refraction of eye with growth⁵³
- ◆ Choice of right formula⁵³⁻⁵⁸
- ◆ Lack of fixation under anesthesia may lead to inaccurate keratometry readings⁵⁹
- ◆ Need for hand held automated keratometer
- ◆ Lack of experienced biometrist in OR when A-scan being done⁵³

Principles of choosing IOL power in children:

- ◆ To balance between risk of amblyopia and future large myopic shift⁵³, aim should be not more than 4D myopia when eye fully grows and present hypermetropia not more than 3D.
- ◆ To reduce myopic shift in future, 3 ways to undercorrect as per age:
 - ◆ Dahan et al:⁵⁶
 - 20% undercorrection if the child is less than age 2 years
 - 10% undercorrection for age 2-8 years
 - ◆ Enyedi et al (Target refraction)⁵⁷
 - 1-2 years: +6 to +5
 - 3-4 years: +4 to +3
 - 5-6 years: +2 to +1
 - ◆ Flitcroft et al⁵⁸
 - <1 year : +6
 - 1-4 years: +3
 - 5-8 years: +1
- ◆ Density of amblyopia: Near emmetropic power for deep amblyopia^{59,60}
- ◆ Refractive status of other eye: Anisometropia not more than 3.0D to avoid amblyopia.
- ◆ Higher the power, more undercorrection needed
- ◆ Parental refractive error: Undercorrect more if parents high myopes.
- ◆ If hand held keratometer is not available, based on

axial length IOL power can be chosen as follows:⁶¹

Axial length (mm)	IOL power (D)
17	28
18	27
19	26
20	24
21	22

Choosing right formula :

Problems in using present formulas:⁵³

- ◆ All formulas are derived from considerations for adult eye
- ◆ Axial length in children is short
- ◆ High keratometry values
- ◆ Shallow AC
- ◆ Target refraction significantly different from emmetropes
- ◆ There is no formula derived primarily from characteristics of the child's eye or historical outcome from IOL implantation in children.
- ◆ Considerable variability in refractive outcomes of modern theoretical formulas like SRK II, SRK T, Holladay I and Hoffer Q especially in children under 24 months of age.⁵³⁻⁵⁸
- ◆ Hoffer Q which is considered more accurate for short eyes (axial length < 22 mm) in adults, has been found to overestimate IOL power in children.⁵⁶
- ◆ SRK II formula gives the least amount of variability overall providing fair to good agreement.^{56,57}
- ◆ Also adjustments in A constant and surgeon factor is difficult due to wide range of axial length and K values making refining of IOL power more difficult.⁵³

Current recommendations :⁵³

- ◆ Refractive growth after IOL implantation in infants and children cannot be predicted accurately due to large standard deviation.
- ◆ Any modern formula can be used but more error should be expected.
- ◆ Use of immersion A scan instead of contact for more accuracy
- ◆ Repeated K-readings so that they are reproducible.

VISUAL REHABILITATION AND VISUAL OUTCOME**Factors affecting outcome**

1. Aetiology
 - ◆ congenital rubella syndrome less than optimal outcome- primary prevention needed
 - ◆ Congenital/Developmental cataract
 - Outcome is better for developmental cataract
 - ◆ With/without other ocular comorbidities
 - microcornea, microphthalmos-increased risk of secondary glaucoma
 - uveitis (VKH/ JRA/ non-JRA)- increased risk of secondary glaucoma, amblyopia and complicated postoperative course
 - ◆ PHPV- increased risk of secondary glaucoma, retinal detachment
2. Unilateral versus Bilateral
 - longstanding unilateral cataract develop dense amblyopia
 - ◆ Prognosis is better in
 - lamellar cataract
 - posterior lenticonus
 - PHPV
 - Straight eyes
3. Morphology
 - Outcome poor for total cataract
 - Better for posterior polar, lamellar, posterior lentiglobus
4. Timing of Surgery

If surgery is done in the first 6 weeks life

 - Better contrast sensitivity
 - Better fusion and stereopsis
 - Decreased strabismus
 - Better compliance with patching
5. Intraoperative considerations (PPC/AV/IOL)
 - PPC must be done in children below 6 years.
 - IOL must be given in children > 1 year and in fully developed anterior segment of children < 1 year of age.
 - IOL to be considered in most unilateral cataracts
 - Avoid IOLs in microcornea
 - Wound suturing in children < 7 years of age
6. Associated complications
 - Intraoperative or post-operative complication can

affect outcome adversely

7. Contact lens/Glasses
 - Contact lenses in unilateral cataract
 - Glasses in bilateral cataract
 - IOL best option as better visual acuity, binocular vision, less strabismus
8. Management of amblyopia
 - Key to success
 - Prolonged hours of patching in unilateral cataracts

Poor outcome

- Long time gap between age of onset and age of surgery
- Associated anomalies
- Unilateral cataract
- Post operative complications
- Complications - glaucoma, after cataract
- Poor amblyopia management

Good outcome

- Bilateral cataract
- Older age of onset
- Relatively clear visual axis
- Early surgery
- IOL implantation
- Good amblyopia therapy

FUTURE PROSPECTS**Multifocal IOLs:**^{53,64}

- Multifocal IOLs need highly accurate biometry, astigmatism control and no refractive growth.
- In children, it is difficult to achieve most of these parameters. So caution is advised in considering use of these in children

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