

ANALYSIS OF VISUAL OUTCOME AFTER SURGICAL MANAGEMENT OF CONGENITAL CATARACT WITH OR WITHOUT IOL IMPLANTATION

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ABSTRACT

BACKGROUND

Early intervention of congenital cataract is posing lots of challenges to the practicing paediatric ophthalmologists, in the form of difficulties in preoperative evaluation and intraoperative techniques. The objective of our study is to analyse the visual outcome in cases of congenital cataract of different morphology and different age group after precise preoperative evaluation and performing cataract extraction with or without IOL implantation with appropriate techniques.

MATERIALS AND METHODS

This is a retrospective analysis of 50 children less than 15 years of age of both sexes undergoing cataract surgery for congenital cataract. Only children with congenital cataract were included in the study. Children less than 15 years of age undergoing cataract surgery due to traumatic cataract and steroid induced cataracts or any other form of cataract were excluded. All types of congenital cataracts with different morphology were included in the study. Some children underwent cataract surgery for unilateral congenital cataract whereas others were operated for bilateral congenital cataract. Eyes with corneal diameter of 10 mm and above were selected for IOL implantation. Others who had microphthalmos, corneal opacity, coloboma iris had lens aspiration alone. All the cases underwent single piece hydrophobic spherical design AcrySof IOL in the bag.

RESULTS

Analysis of visual outcome after surgical management of congenital cataract with or without IOL implantation showed good visual outcome when children underwent early surgical intervention. Among the different types, the commonest encountered was lamellar cataract, which is a bilateral cataract, reported in 26 children. Good visual acuity outcome was noted when children with zonular or lamellar cataract underwent surgery. Children with total cataract, membranous cataract, nuclear cataract had poor visual outcome and suffered from deep amblyopia.

CONCLUSION

Early screening, early diagnosis, early surgical intervention with IOL implantation plays a major role in successful visual outcome in children with congenital cataract.

KEY WORDS

Visual Outcome, Surgical Management, Congenital Cataract, IOL Implantation, Primary Posterior Capsulorhexis.

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BACKGROUND

In Asia, more than one million cases of childhood blindness are due to paediatric cataract¹. Mostly aetiology of cataract in children remains unknown. It can be associated with genetic disorders, intrauterine viral infections especially rubella, systemic syndromes, metabolic disorders, injuries and long-term steroid usage². Congenital cataracts are mostly hereditary in nature, in the form of autosomal dominant, autosomal recessive, X-linked or sporadic.^{3,4} Higher incidence in developing countries is due to consanguineous marriage, large family size and lack of genetic counselling.

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Maternally acquired causes like rubella, toxoplasma, cytomegalovirus, herpes simplex and varicella (TORCH) may be the causes of congenital cataracts.^{5,6} Of these, rubella is the commonest infection affecting the pregnant mother in the first trimester. This has been dramatically reduced due to intense anti-rubella vaccination and health education. The visual outcome of children following the surgical management of congenital cataract is determined by the presence of visual axis opacification postoperatively in the form of membrane, posterior capsular opacification and anterior hyaloid vitreous opacification. Hence, the importance of performing a perfect posterior continuous curvilinear capsulorhexis (PCCC) with or without vitrectomy as a primary procedure is emphasised. Unlike adults, visual axis opacification not only reduces vision but is a major cause for amblyopia. Posterior capsule opacification is almost inevitable^{7,8,9,10} and almost close to 100 percent.¹¹ Age of IOL implantation is always debatable. Less than one year of age and associated co-morbid ocular conditions like microphthalmos, will be a contraindication for IOL

implantation. IOL implantation is always a single-piece acrylic foldable intraocular lens in the bag.

MATERIALS AND METHODS

This is a retrospective analysis performed in 50 children from birth to 17 years of age involving both sexes in the Department of Paediatric Ophthalmology, Mahathma Eye Hospital, Trichy, India. Only children with congenital cataract were included in the study. Children less than 15 years of age undergoing cataract surgery due to traumatic cataract and steroid induced cataracts or any other form of cataract were excluded. All types of congenital cataracts with different morphology were included in the study. Some children underwent cataract surgery for unilateral congenital cataract whereas others were operated for bilateral congenital cataract. Eyes with corneal diameter of 10 mm and above were selected for IOL implantation. Others who had microphthalmos, corneal opacity, coloboma iris had lens aspiration alone. All the cases underwent single piece hydrophobic spherical design AcrySof IOL in the bag. All the children had either unilateral or bilateral congenital cataract of different morphology for which they underwent cataract surgery with or without IOL implantation. Parents were evaluated for consanguineous marriages, genetic abnormalities, anti-natal history in the form of first trimester fever and rashes (rubella) etc. Preoperatively, vision assessment was performed by fixation pattern and presence of nystagmus and strabismus. Specialised Vision charts like Keeler cards, Cardiff cards, Sheridan – Gardiner charts and Snellen's chart wherever appropriate were used. Examination under anaesthesia (EUA) was performed for children below 8 years of age. During EUA, corneal diameter, corneal clarity, iris and pupillary status, morphology of congenital cataract, IOP measurement, indirect ophthalmoscopic evaluation to rule out retinal and optic nerve pathologies were investigated and documented. In cases of dense total cataract, ultrasound imaging of retina was performed.

Keratometry and immersion biometry was performed to calculate the right IOL power. Even 0.1 mm difference in axial length may result in 0.75 difference in IOL calculation. Since the children's eyes are shorter, axial length error can be magnified to 14 D/mm¹². In all children subjected to congenital cataracts surgery, we used SRK-T formula and under correction of IOLs was performed according to the age¹³. In our study IOL power ranging from +13 D to +30 D was used. These children underwent TORCH TITRE evaluation and evaluation for any metabolic disorders. Postoperative targeted refraction in our series, we always aimed for under correction resulting in mild to moderate hypermetropia in the immediate postoperative period. Wherever significant, these children were given appropriate unifocal lenses for less than 5 years of age and bifocal above 5 years of age. Surgical technique used was anterior limbal approach lensectomy and vitrectomy for aphakic cases. 2.2 mm clear corneal incision with two numbers of 1 mm side port incision was made in children who were subjected to cataract extraction with IOL implantation. Whenever PCCC was performed, it was made 2 mm smaller than anterior rhexis. Hydrophobic spherical design single piece AcrySof IOL was implanted in the bag. All three corneal incisions were sutured using 10 – 0 nylon suture. In cases of 50 children who underwent IOL implantation, 36 children underwent PCCC

with vitrectomy and 14 children did not have posterior capsule procedures. Postoperatively these children were closely monitored for immediate postoperative inflammation and late postoperative visual axis opacification. Children who underwent lens aspiration without IOL implantation, were periodically assessed for glaucoma and retinal complications.

Refraction was performed in all cases in all visits because of the growing nature of paediatric eye. Refraction plays a major role in visual rehabilitation to provide appropriate glasses and for better management of amblyopia management also. All children with aphakia were managed with high plus spectacles. Children with IOL implantation in the school going age group needed either bifocals or progressive lenses. Children with posterior capsule opacification needed either anterior limbal approach or pars plicata membranectomy to avoid amblyopia. Grown up children who were cooperative enough underwent YAG capsulotomy.

Several surgical techniques are available in the form of (a) lensectomy with anterior vitrectomy either through anterior limbal approach or posterior pars plicata / plana approach (b) lens aspiration with IOL implantation with or without PCCC and anterior vitrectomy. The anterior chamber was accessed by clear corneal incision the size of incision was 2.8 mm at 12 o' clock position always. Two side ports of each 1 mm in size were made at 3 and 9 o'clock position. Trypan blue staining of anterior capsule was performed in total cataracts only. Zonular / lamellar cataract and cataracts with good red glow, trypan blue staining was not performed. Anterior continuous curvilinear capsulorhexis (CCC) of 5 mm diameter was performed under viscoelastics. The lens material was aspirated using bimanual irrigation aspiration hand pieces. Primary posterior CCC was performed in required cases at least 1.5 to 2 mm smaller than anterior CCC. Adequate anterior vitrectomy was also performed before implantation of IOL. AcrySof single piece non-aspheric IOL implantation in the bag performed. Corneal incision was sutured using 10-0 nylon and children less than 2 years of age both side ports were also sutured.

Statistical Analysis

Data was analysed using SPSS (Version 17.0) software. The frequency (No. of respondents) and percentage (%) of respondents selected for the study were analysed through the software.

RESULTS

The Analysis of Visual outcome after surgical management of congenital cataract with or without IOL implantation was carried out in 50 children of both sexes from birth to 17 years of age involving both sexes in the Department of Paediatric Ophthalmology, Mahathma Eye Hospital, Trichy, India. All the children diagnosed had either unilateral or bilateral congenital cataract of different morphology. Of these different age group tested, the highest number of 13 children with congenital cataract was observed in less than one year of age, followed by the age group 3 to 5, in which 12 children were affected. The children affected with congenital cataract between 8 to 12 years of age and above 12 years were found to be of equal number (7 children). The number of affected children was very less in the age group of 1 to 2 years. The results were tabulated in table -1. The gender analysis showed 32 male children and 18 female children (Figure – 2).

The laterality study revealed 64% of the children were affected by bilateral congenital cataract and 36% of children was observed with unilateral congenital cataract, thus, posing postoperative challenges during rehabilitation. The results were predicted in figure – 2. From this study it was concluded that, the children less than one year of age reporting for congenital cataract treatment was the highest number because of absence of fixation or roving eye movement as noticed by the mother.

The Morphology of congenital cataract in children was recorded as total cataract, Lamellar cataract, zonular cataract, blue dot cataract, membranous cataract, infantile embryonic cataract, posterior polar cataract with or without posterior subcapsular component. Among the different types, the commonest encountered was lamellar cataract which is a bilateral cataract reported in 26 children. The lamellar cataract presented to us with high level of asymmetry. It followed by total and membranous cataracts, in which five children were affected in each. The membranous cataract is usually not identified in routine ophthalmological evaluation unless fully well dilated. The missed membranous cataracts were brought to us with nystagmus after two years of age. The Zonular, blue dot and PCC and PPC cataract were not predominant in our study (Table – 2). The total cataracts were due to intrauterine infections as proven by torch titre especially rubella.

The axial length of eye ball of children affected by congenital cataract was measured and presented in figure – 3. Higher axial length of 20 mm to 22 mm was noticed in 30 children. Twelve children were observed within the range of 22 mm to 24 mm. In four children the axial length in the range of 16 mm to 18 mm was recorded. Three children had above 24 mm. Higher axial length implies pre-existing axial myopia which may be mild or severe and may be associated with fundus pathology. The very low axial length denotes pre-existing hypermetropia. At birth the infant eyes physiologically hypermetropic. According to the axial length and keratometric values IOL power was calculated by immersion biometry and appropriate IOL was used in children with congenital cataract. Among the 50 children, 28 of them underwent cataract surgery with IOL implantation within the power range of 18 to 24D. This embraces normal standard IOL power. Myopic eyes were observed in three children, hence IOL power of lower dioptric values were implanted in those children. These children need to be closely followed up anticipating myopic manifestation, in order to avoid amblyopia. Hypermetropia was noticed in 17 children and these children underwent IOL implantation of higher dioptric values. Two children were diagnosed as posterior polar cataracts with pre-existing large posterior capsular dehiscence, hence they were left aphakic. The results were presented in table – 3.

Visual acuity assessment was started in the one-month post-operative period for all the children. Depending upon the patient’s age group, cooperation status and intensity of amblyopia, visual acuity assessment showed gross variability as shown in the table- 4. Children with severe visual loss of less than 6/60 were due to stimulus deprivation amblyopia in congenital cataracts like embryonic cataract, nuclear cataract and total cataract. Five children manifested nystagmus, as

their embryonic cataract or total cataract was operated after the age of 2 yrs. Children with visual acuity of 6/6, 6/9, 6/12, suffered from congenital zonular cataract or lamellar cataract. These are the cataracts where they are not dense enough to cause dense amblyopia. The visual deprivation induced by them are only partial.

Age of the Children (in years)	Frequency of Children with Congenital Cataract	% of Children With Congenital Cataract
0 to 1	13	26
1 to 2	2	4
2 to 3	3	6
3 to 5	12	24
5 to 8	6	12
8 to 12	7	14
> 12	7	14
Total	50	100

Table 1. Age Wise Analysis of Children Undergoing Cataract Surgery for Congenital Cataract

Cataract Type	Frequency of Morphologically Different Congenital Cataract	Percentage of Morphologically Different Congenital Cataract
Total Cataract	5	10
Lamellar Cataract	25	50
Zonular Cataract	4	8
Blue Dot Cataract	2	4
Membranous Cataract	5	10
Infantile Embryonic Cataract	6	12
PCC& PPC	3	6
Total	50	100

Table 2. Morphology of Congenital Cataract

IOL Power D	Frequency of IOL Power	% of Children
13	1	2.08
14	2	4.17
18 / 18.5	2	4.17
19 / 19.5	3	6.25
20 / 20.5	4	8.33
21 / 21.5	3	6.25
22 / 22.5	7	14.58
23 / 23.5	8	16.67
24	1	2.08
25	8	16.67
26	3	6.25
27	3	6.25
30	3	6.25
Total	48	100

Table 3. Analysis of The Dioptric (D) Power of Intraocular Lenses Used

Postop Visual Acuity	Frequency of Children	% of Children
Keelar-Snellen Equivalent of 6/60	2	4
Hm	1	2
Fixing of Light & Following of Toys- Snellen Equivalent Of 6/36	10	20
Cardiff - Snellen Equivalent of 6/60	4	8
Cake - decorations Snellen Equivalent Of 6/60	4	8
Snellen Acuity 6/602/60	1	2
Snellen Acuity 4/60	1	2
Snellen Acuity 6/60	4	8
Snellen Acuity 6/36	3	6
Snellen Acuity 6/24,6/24P	6	12
Snellen Acuity 6/18	5	10
Snellen Acuity 6/12, 6/12P	4	8
Snellen Acuity 6/9	3	6
Snellen Acuity 6/6	2	4
Total	50	100

Table 4. Analysis of Visual Acuity of Children with Congenital Cataract in 3 Months Post-Op

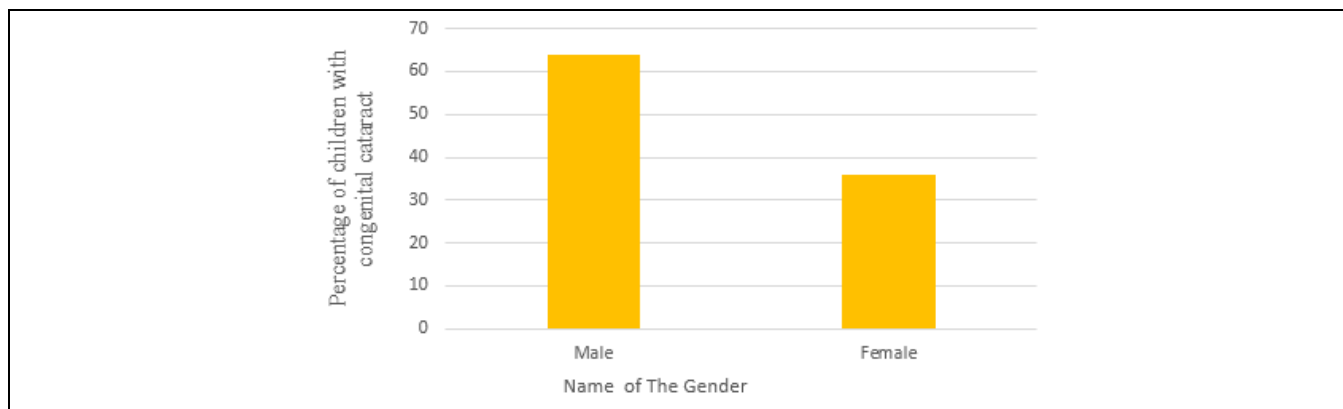


Figure 1. Sex Wise Analysis of Children with Congenital Cataract

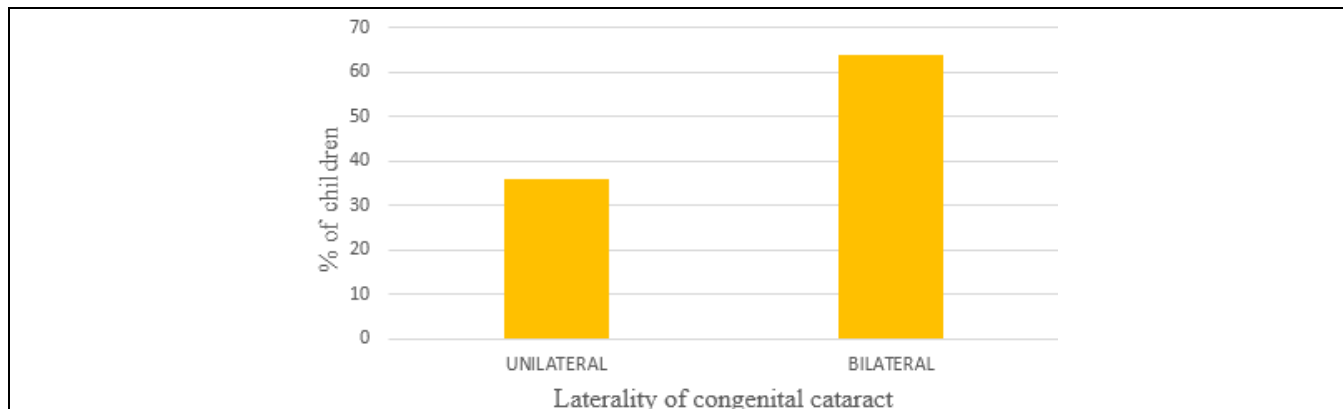


Figure 2. Laterality of Manifestation of Congenital Cataract

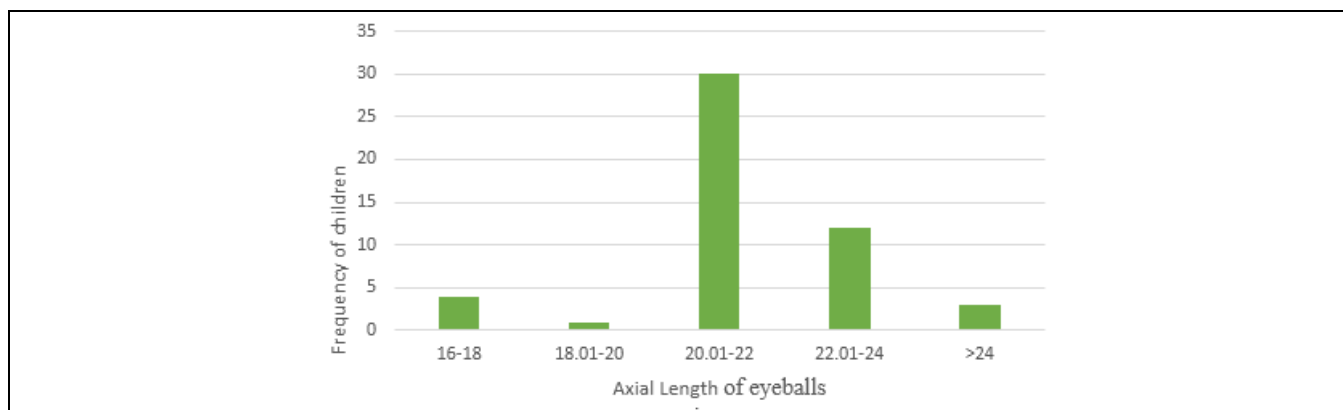


Figure 3. Axial Length of Eye Ball of Children Affected by Congenital Cataract

DISCUSSION

In most of the occasions, children are brought to paediatric ophthalmologists only when the paediatricians or parents notice white reflex in the pupillary area or after manifestation of nystagmus, strabismus and sometimes for not recognising faces or poor fixation of eyes. These are the situations wherein, stimulus deprivation amblyopia sets in very strongly. Hence, even best surgical management and rigorous amblyopia therapy does not improve the vision of the child satisfactorily. The child remains partly sighted rest of the life.

Since visual development is an ongoing process, any deprivation results in amblyopia, strabismus and nystagmus.¹⁴ When stimulus deprivation amblyopia has set in and the child is more than 8 yrs. of age - when functional growth of the eyes is completed, then restoration of the visual acuity to normalcy is close to impossible.¹⁵ Dense cataracts result in sensory nystagmus. Early intervention minimises stimulus deprivation amblyopia.¹⁶

In our study, children receiving IOL implantation were always under-corrected. So that, in the immediate postoperative period they have significant hypermetropia. The aim of having significant hypermetropia is to combat the 'myopic shift'. At the time of visual adulthood, these children will either achieve emmetropia or mild to moderate myopia. Research has confirmed that lens epithelial cells proliferation over the anterior hyaloid face of vitreous¹⁷ can also cause visual axis opacification (VAO). In spite of Posterior continuous curvilinear capsulorhexis and anterior vitrectomy five children in our series had VAO due to inadequate posterior capsular opening and anterior vitrectomy¹⁸. Gimbel¹⁹ says vitrectomy in children's eyes has a negative impact on ocular development.

In our study five children required Nd: YAG Laser capsulotomy. Both aphakic children and five children who underwent IOL implantation had VAO. The four children below 8 years of age and one child above 8 years of age developed PCO. Vasavada et al.²⁰ studied the effect of acrylic IOL (AcrySof) in children between 2 and 6 years of age. They inferred that, posterior capsule opacification developed in 83 % who did not undergo posterior capsulotomy, PCO in 37 % who had posterior capsulotomy without vitrectomy, 6 % with capsulotomy and vitrectomy.

Vision assessment in children poses a great challenge to any paediatric ophthalmologist. In children belonging to preverbal age vision is assessed by fixation pattern, preferential looking technique, vanishing optotype principle. In verbal age group use Sheridan-Gardiner charts and children above 5 years use Snellen's chart. The most vital preoperative assessment from surgical point of view is to examine the child under general anaesthesia to assess the ocular morphological abnormalities like microphthalmia, corneal pathology, iris and pupillary abnormalities, glaucoma, retinal and optic nerve pathologies. These children undergo corneal diameter evaluation, intraocular pressure measurement, pupillary dilatation assessment, morphology of cataract, B-scan in case of total cataract, corneal curvature keratometry measurement, axial length measurement using immersion biometry and indirect ophthalmoscopic fundus evaluation wherever possible.

IOL Calculation Formula

Most formulas in ophthalmic usage are from data received from analysing adult eyes. The Hofer Q and Holladay 2 formula are more accurate in shorter eyes.^{21,22} There are several studies on this title. Trivedi et al.²³ studied the accuracy of Holladay 2 formula and compared the predicted error to Hoffer Q, Holladay 1 and SRK T formula. Holladay 2 gave the least predicted error followed by Hoffer Q especially in eyes shorter than 22 mm.

Postoperative targeted refraction depends on various factors which still remain unresolved. Immediate correction of postoperative refractive error is very important in preventing amblyopia. Myopic shift later in life may necessitate IOL exchange. Myopic shift results from visual deprivation, axial length elongation, change in the IOL position with the growth of eye ball. Gimbel et al. recommended immediate emmetropia after surgery for better amblyopia management, but the risk of large myopic shift always remains.

CONCLUSION

Congenital cataracts should be screened early to avoid stimulus deprivation amblyopia. Surgical management is the only option. IOL implantation with proper posterior capsule and anterior vitreous management adds to the success of the long-term visual outcome. Subsequent periodic refraction, amblyopia and visual axis management adds to the successful visual rehabilitation of these children.

Technical challenges in the form of appropriate evaluating instruments, surgical techniques and technology, ideal IOL design, material, size plays a major research-oriented approach in managing congenital cataracts.

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