Congenital cataract- a riddle to be solved to prevent childhood blindness

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Abstract

Introduction: Congenital cataract is a significant cause of visual disability in developing countries. It may present at birth or develop within first year of life. Common causes are genetic disorders, intra uterine infections, drug induced and others. Hence management protocol depends from case to case. Timely management of cataract and rehabilitation significantly prevents visual morbidity in children. **Aims and Objectives:** To asses visual outcomes in congenital cataract. **Material and Methods:** A prospective study was done from January 2017 to July 2018, including 40 patients with age ranging from 1 month to 12 years. Detailed history and thorough examination of each patient was done. After fulfilling inclusion criteria we have planned them for small incision cataract surgery (SICS) and phacoemulsification as per the financial status of family. Only cases less than 1 year were left aphakic. IOL power was calculated using SRK-II formula. Post operative visual acuity was noted on 7th day, 15th day and 1 month. Visual rehabilitation in the form of amblyopia therapy, secondary IOL and contact lenses was given. **Result:** It was found that squint (20%) and amblyopia (17.5%) was associated with cataract. All 40 eyes underwent cataract extraction by phacoemulsification (60%) and SICS (40%). Among them 25% cases were left aphakic. Visual improvement was almost similar in both surgeries. Visual rehabilitation in the form of spectacles (45%), contact lenses (5%), occlusion therapy (17.5%) and secondary IOL (22.5%) were given. **Conclusion:** Proper counseling of parents at diagnosis helps in prompt management and improves compliance which significantly reduces visual morbidity in children.

Key words- Congenital cataract, Visual outcome, Visual rehabilitation

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Introduction

Pediatric cataract is one of the major causes of preventable childhood blindness, affecting approximately 200,000 children worldwide, with an estimated prevalence ranging from three to six per 10,000 live births [1-3]. Pediatric cataracts may be congenital if present within the first year of life, developmental if present after infancy, or traumatic.

Common causes are genetic disorders, intra uterine infections, drug induced and others. Early diagnosis and treatment are of crucial importance to prevent the development of irreversible stimulus deprivation amblyopia. The management of pediatric cataract should be customized depending upon the age of onset, laterality, morphology of the cataract, and other associated ocular and systemic co-morbidities.

Manuscript received: 2nd December 2018 Reviewed: 11th December 2018 Author Corrected: 16th December 2018 Accepted for Publication: 19th December 2018 Despite developments in surgical techniques and intraocular lenses, the management of unilateral pediatric cataracts is still clinically challenging. Better visual outcomes are usually obtained with early surgical correction and vigorous amblyopia treatment [4,5,6,7]. Visual system is developed in children as long as sharp, clear and focused images are formed on the retina of both eyes. Otherwise, amblyopia is observed. Therefore, early detection and surgery as well as follow-up visits have significant roles in the restoration of a child's vision in the case of congenital cataract with significant media opacity [8].

Unfortunately, in many cases, even after early surgery, long-term follow-up (at least up to the age of 10) is not properly instituted, and varying degrees of amblyopia have been reported despite wearing appropriate eye glasses, contact lenses, or implantation of intra ocular lenses (IOL) [9].

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The first years of life are crucial for the development of a child's vision and therefore irreversible amblyopia can be induced by blurred and distorted retinal image over that period [10,11].

Material and Methods

Study Design- Prospective interventional study

Sample Size and Duration- A prospective interventional study was done from January 2017 to July 2018, including 40 patients upto 12 years of age.

There was a programme going on by government RBSK (Rashtriya Bal Swasthya Karyakram) to prevent childhood blindness, so we have taken 24 patients out of 40 from this programme.

Methodology- The following clinical variables were analysed: patient's gender, age at diagnosis, presenting symptoms, laterality, morphology, aetiology, presence of other ocular and systemic abnormalities, family history of cataract, follow up time and treatment.

In case of surgery, the following parameters were assessed: age at surgery, time from diagnosis to surgery, intra and postoperative complications and pre and postoperative visual acuity.

Detailed history and thorough examination of each patient was done.

Ocular Examination-

- 1. Visual acuity and best corrected visual acuity (by preferential looking behaviour in younger (upto 3 year) and Snellen's chart in older children (above 3 year).
- 2. Slit lamp bio microscopy by 90D or 78D lens
- 3. Fundus Examination- by direct ophthalmoscopy and indirect ophthalmoscopy using 20D lens.
- 4. B- Scan ultrasonography to evaluate posterior segment pathology

Results

Table-1: Demographic profile.

Biometry-	IOL	power	was	calculated	using	SRK-II
formula.						

Keratometry was done using bousch and lomb automated keratometer under general anaesthesia. Axial length of eye using A-scan

- 6. Examination under anaesthesia (if required)
- Intra ocular pressure by Sciotz tonometry
- Syringing for patency of lacrimal apparatus.

All routine pre operative blood investigations were sent along with TORCH (Toxoplasma, rubella, cytomegalo virus, herpes virus) profile of mothers and children. Post operative ocular examination and visual acuity was noted on 7th day, 15th day and 1 month.

Visual rehabilitation in the form of amblyopia therapy, secondary IOL and contact lenses was given accordingly.

Inclusion Criteria

5.

- Patients aged less than 12 years.
- Parents who are willing to follow up

Exclusion Criteria

Children with associated ocular condition like

- Traumatic cataract
- Retinopathy of prematurity
- Microphthalmos
- Persistent fetal vasculature
- Children with other systemic diseases like Marfan syndrome, Lowe's syndrome, Galactosemia, Hypothyroidism and those with learning disability were also excluded.

After fulfilling inclusion criteria we have planned them for small incision cataract surgery (SICS) and phacoemulsification as per the financial status of family.

S. No.	Sex	Number of patients	%
1.	Male	20	(50%)
2.	Female	20	(50%)

In our study equal sex distribution was observed.

Table-2 Demographic profile.

S. No	Age group	Number of patients	%
1.	<1 Year	10	25%
2.	1 -5 Years	10	25%
3.	>5 Years	20	50%

In our study 50% patients were age group more than 5 year.

Table-3 Type of surgery

Serial No.	Procedure	Number of patients	%
1.	SICS with PCIOL	12	30%
2.	SICS + Anterior Vitrectomy	4	10%
3.	Phacoemulsification with PCIOL	18	45%
4.	Phacoemulsification + Anterior Vitrectomy	6	15%

In our study 60% cases were of phacoemulsification (PCIOL/Anterior vitrectomy)

Table-4: Pseudophakic/Aphakic.

Procedure	Number of patients	%
PCIOL Implanted	30	75%
Aphakia	10	25%

In our study PCIOL is implanted in most of the cases.

Table-5: Laterality.

Sex	Children with Bilateral presentation	Children with unilateral presentation
Male	14(35%)	6(15%)
Female	12(30%)	8(20%)
Total	26	14

In our study bilateral presentation were more common.

Visual Acuity	Age					
	<1 Year	%	1-5 Years	%	>5 Years	%
Uncooperative	6	15%	2	5%	0	0
FL TO CF	4	10%	4	10%	8	20%
1/60-6/60	0	0	3	7.5%	6	15%
>6/60	0	0	1	2.5%	6	15%
Total	10		10		20	

Pre operative visual acuity were Following light to counting fingers in most of the cases.

Visual Acuity	Age					
	<1 Year	%	1-5 Year	%	>5 Year	%
Uncooperative	2	5%	0	0	0	0
FL TO CF	8	20%	1	2.5%	2	5%
1/60-6/60	0	0	5	12.5%	8	20%
>6/60	0	0	4	10%	10	25%
Total	10		10		20	

Table-7: Post operative best corrected visual acuity at 1 month

Post operative visual acuity was improved to more than 6/60

Table-8: Fundus evaluation in operated cases

Fundus Appearance	Number of patients
Salt & pepper retinopathy	6
Myopic fundus	5
Normal	29

In our study, normal fundus were found in most of the cases

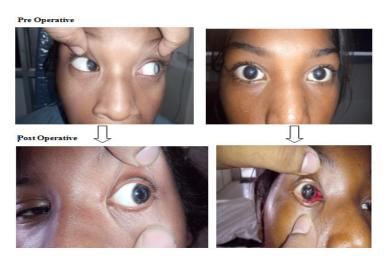
Table-9: Type of cataract

Type of Cataract	Number of patients
Lamellar cataract	5
Zonular cataract	7
Membranous cataract	28

We have found that 5 children came positive for TORCH infections.

Among them one was positive for Rubella (Titre >1.1) and other 4 were positive for both Rubella as well as cytomegalovirus (Titre >1.1).

Visual Rehabilitation- Visual rehabilitation was given in the form of spectacles, contact lenses, secondary IOL, occlusion therapy. Spectacles were the most commonly prescribed mode of visual rehabilitation followed by contact lenses



Discussion

In the present study, the clinical and surgical data of 40 patients with congenital cataract were taken. The youngest child in this study was 1 month old and eldest was 12 years old.

Most of the cases in our study were operated at more than 5 year of age while in the studies of the UK [12,13] China [14] Ethiopia [15] and Kuwait [16] the children were operated at the mean age of 12 months, 8.7 weeks, 5.6 months, 7.21 years, and 12 months, respectively.

Our mean age of surgery was closer to the results from Ethiopia than other countries. Based on the above reports, it should be noted that the diagnosis and surgery of congenital cataract were unfortunately more delayed in some developing countries compared to developed countries. The reason could be due to late diagnosis and therefore late surgery, less severity or peripheral lens opacity with acceptable visual acuity, unavailable subspecialty of pediatric ophthalmologist in the rural regions, poor economic status, or a combination of some the above mentioned etiologies.

The postoperative visual acuity results of 25% of patients having BCVA \geq 6/60 compare favourably with reports from Central India [17] Tanzania [18] and Nepal [19]. There were 26 patients (65%) with bilateral and 14(35%) with unilateral congenital cataract. Lee and Msamati et al. also reported more bilateral cases in their studies [20,21].

Compared with previous studies from developed countries [22,23,24] delay of surgery in this study was still very common because most of our patients came from rural areas where there was a poor primary eye care system. The preoperative visual acuity ranged from perception of light to 6/36. After surgery, 32.5% (13 eyes) had 1/60 to 6/60 vision, 35% (14 eyes) had >6/60 vision. The rest 27.5% (11 eyes) had perception of light to finger counting vision.

Conclusion

The generally accepted minimum age of IOL implantation in pediatric cataracts is 1-2 years. For <1 year old IOL implantation is still controversial. The basic concept is to leave them more hyperopic the younger they are to compensate for the myopic shift. Refined surgical techniques with posterior CCC, anterior vitrectomy, in-the-bag IOL implantation with/without IOL capture helps to reduce incidence of PCO.

Early diagnosis and prompt surgical intervention are extremely important in the management of paediatric cataract, as also adequate visual rehabilitation in the form of spectacles with both distance and near correction. The parents need to be counselled about the importance of postoperative care, follow-up, refraction and compliance of spectacle wear [25]. In our study surgery was performed by single surgeon (unit head) with preoperative and post operative examination. Data collection and manuscript work was done by resident under consultant supervision and guidance.

What this study adds to existing knowledge- Our study emphasize the importance of programmes held by government to prevent childhood blindness so that we could get more number of children in this short period of time. Surgery is only one aspect of the entire management of pediatric cataract patient. Participation in the visual rehabilitation of the child involving parents, ophthalmologists, paediatricians and optometrists need not be over emphasized.

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