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Clinical Audit of Visually Significant Congenital and Developmental Cataract

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Abstract

Purpose: To do a clinical audit of the patients of congenital and developmental cataract and to find out morphology of visually significant cataract and the average age of presentation for treatment.

Methods: A retrospective analysis of 57 eyes of 30 pediatric patients ≤18 years with congenital and developmental cataract was done. Results: Out of 30 children, 13(43.33%) of them had congenital cataract and 17(56.67%) had developmental cataract.90% of children had bilateral cataract.

Average age of presentation was 5.6 months in the age group ≤ 12 months, 5.8 years in the age group >12 months. Majority of the patients in our study were from rural area. Most common morphological type of cataract noted was zonular in 70% (21 patients) followed by total cataract. The most common presenting complaint that brought medical attention was leukocoria in 17 children (56.67%), diminution of vision in 12 children (12%) and squint in one child.

Conclusion: Our study showed that there was delayed presentation of visually significant cataract. To ensure optimal visual gain after cataract surgery in children, early diagnosis of the visually significant cataract is the key.

Keywords: Congenital cataract, developmental cataract, early diagnosis, visually significant cataract

Introduction

Childhood cataract is a major cause of blindness and visual impairment worldwide.[1] The prevalence of childhood cataracts which is a treatable cause of blindness is 1–15 cases per 10,000 live birth in developing countries and in India it contributes about 7.4–15.3% of childhood blindness.[2] Childhood cataract is said to be congenital if present within the first year of life, developmental if present after infancy.[3]

Early diagnosis and treatment of pediatric cataract is important to optimize visual outcome and to minimize stimulus deprivation amblyopia. It has been seen that there is delayed presentation of such patients for treatment purpose in developing countries like India. Delayed presentation can be due to various factors like cultural, socio- demographic and health system.

Not all pediatric cataracts require surgery, only those which are visually significant do so. Since a subjective assessment of visual acuity cannot be obtained in very young children, greater reliance must be placed on morphology of cataract, to ascertain whether the cataract is visually significant or not.[4] The morphology of cataract gives clue to the age of onset, visual prognosis, heritability and etiology.[5] In young infants with poor fixation, presence of strabismus or nystagmus[6], dense central opacities larger than 3mm diameter[7] and the presence of a diffuse opacity precluding a view of the fundus under cycloplegia[8] are considered as visually significant cataract.

The purpose of our study was to do a clinical audit of the patients of congenital and developmental cataract presenting at our institute for treatment and find out morphology of visually significant cataract and the average age of presentation for treatment of such patients.

Materials And Methods

A retrospective analysis of 30 pediatric patients ≤18 years with cataract presenting to ophthalmology OPD of tertiary health care institute was carried out. Inclusion criteria was patients who were diagnosed to have congenital and developmental cataract. We excluded cases who presented with traumatic and complicated cataract. Patient's demographic details, history, clinical ocular examination were documented as per predefined proforma. Cataract was defined as congenital if present within one year of life and developmental if present after infancy.

Demographic details included age at presentation, sex, eye involved, laterality, type of presentation, family history were recorded. Examination included detailed anterior segment examination using slit lamp bio-microscope (wherever possible), dilated fundus examination was done with indirect ophthalmoscope and 20D lens and / or an ultrasound B-scan if the media was not clear and for estimation of axial length. The study protocol was approved by the Institutional Ethical Committee and adhered to the tenets of the Declaration of Helsinki.

Results

57 eyes of 30 children with congenital and developmental cataract were included in the study. Amongst which 43.33% had congenital cataract and 56.67% had developmental cataract. On examination 90% had bilateral cataract while the remaining had unilateral cataract.

Table 1 describes the demographic details including age of presentation of children to hospital. The overall average age of presentation was 3.5 years. **Average** age of presentation was 5.6 months in the age group ≤ 12 months, 5.8 years in the age group >12 months. Majority of the patients in our study were from rural area.

Table 1: Demographic details

Gender	**Number of patients**	**Percentage**
Females	18	60%
Males	12	40%
Total	30	100%
Age of presentation	**Number of patients**	**Percentage**
<12 months	13	43.33%
>13 months	17	56.66%
Total	30	100%
Laterality of cataract		

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Unilateral	3	10%
Bilateral	27	90%
Total	30	100%
Geographical distribution		
Rural	17	56.67%
Urban	13	43.33%
Total	30	100%

Most common morphological type of cataract noted was zonular in 70% (21 patients) followed by total cataract [Table 2]. The most common presenting complaint that brought medical attention was leukocoria in 17 children (56.67%), diminution of vision in 12 children (12%) and squint in one child.

Table 2: Morphology of cataract

Morphology of cataract	Number of patients	Percentage
Total cataract	6	20%
Zonular cataract	21	70%
Zonular cataract with riders	1	3.33%
Posterior subcapsular with blue dot	1	3.33%
Posterior polar	1	3.33%
Total	30	100%

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Table 3 describes the associated ocular abnormalities. The significant variables were microcornea 7 (23.33%), nystagmus 6 (20%), strabismus (16.66%). In 20 cases (66.66%), fundus was not visualized due to dense cataractous changes whereas in 8 cases (26.66%) fundus was seen to be normal on indirect ophthalmoscopy. One case each had rubella retinopathy and retinitis pigmentosa.

Table 3: Associated ocular abnormalities

Associated ocular abnormalities	Number of patients	Percentage
Micro cornea	7	23.33%
Nystagmus	6	20%
Strabismus	5	16.66%
Posterior synechiae	2	6.66%
Frayed pupillary margin	1	3.33%
Rubella retinopathy	1	3.33%
Retinitis pigmentosa	1	3.33%
None	7	23.33%
Total	30	100%

Systemic associated conditions like delayed milestones and congenital heart disease were present in 10% each of children. Malnutrition was present in 2 (6.6%) of them while one (3.33%) child each had lymphangioma and Down's syndrome. In majority of children 20 (66.66%) there was no systemic association found.

Discussion

In this retrospective study, 57 eyes of 30 patients were included. Out of 30 children with cataract in the study 18(60%) were females and 12 (40%) were males. This is in contrast to other studies [1][9][10][11] where male outnumbered female. This reflects health seeking behavior of the population where males are given priority and presented earlier to the hospital as compared to female child in relation to health issues.

Unilateral cataracts were seen in 3 (10%) patients only with bilateral involvement in 27 (90%) cases. This is comparable to other studies [11][12][13][14] where bilateral cataract was more commonly seen than unilateral cataract.

In our study the most common associated ocular abnormality was microcornea (23.33%) followed by nystagmus (20%), strabismus (16.66%), posterior synechiae (6.66%), frayed pupillary margin (3.33%). Nystagmus in 35.8% followed by squint in 14.4%, microphthalmos (4%), microcornea (2%), microphthalmos with coloboma (0.09%), congenital glaucoma (0.04%) [15] were seen by Khanna et al. Microcornea was most common associated ocular abnormality (8%) seen by Nikhil et al [16]

Majority of children (66.66%) had no systemic associated conditions, 10% each had delayed milestones and congenital heart disease, 6.66% were malnourished. One child each presented with lymphangioma and Down's syndrome respectively.

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Down's syndrome[16][17][18], grossly delayed milestones or epilepsy, Marfan's Syndrome[17][18], Galactosemia[16], CNS anomalies like grossly delayed milestones or epilepsy, Lowe syndrome[17], Hallermann- Streiff syndrome and oculocutaneous albinism [18] are the systemic associations recorded by other authors.

In our study, the mean age of presentation was 3.5 years. The delayed presentation in our study maybe as majority of our patients were from rural areas. Also Sethu et al [11], Sen et al [12], Shah et al [19] noted major presenting population from rural areas in their studies respectively. They are more likely to have lack of knowledge and awareness regarding the problem – the visual prognosis as well as the facilities available. In other studies causes of delayed presentation included inadequate knowledge of infant's vision[9]; false assumption that child is seeing well and hence non appreciation of need to take the child to hospital (38.54% cases) [10]; low education of parents (16.4% cases).[14]

Age at intervention and hence age at presentation has a significant effect on visual outcome. A majority of children presented late to the hospital according to different studies done in India which were highlighted by Khanna et al [15], Anushree et al [16], Sen et al. [12] The mean age of presentation was 4.4 years, 7.6 ± 4.2 years, 7.78 ± 4.34 years in their studies respectively. The vision in late or untreated congenital cataracts is very poor because of profound deprivation amblyopia, unless there has been some period of visual experience in early life.[5] Early treatment of unilateral and bilateral paediatric cataract is important to optimize vision in life and to minimize amblyopia.[11] Surgery of unilateral congenital cataracts by 4–6 weeks and bilateral cataracts within 6–8 weeks of life can be preventive in development of strabismus, nystagmus and amblyopia.[6] Visual impairment at an early age can hamper personality development, hinder education, and deprive the individual of career opportunities, thus increasing the socioeconomic burden on the family and the community.[21]

The morphology of cataract gives a clue to the age of onset, visual prognosis, may suggest heritability and etiology. Some morphological types have a better visual prognosis than others, with lamellar cataracts and posterior lenticonus doing well and dense central cataracts have relatively poor visual prognosis. [22] The morphology of a cataract is mainly determined by the timing and nature of the insult that caused the abnormality and the anatomy of the lens.[23][24] The most common type of cataract in our series was zonular cataract (70%) followed by total cataract (20%). According to Foster et al prevalence ranged from: lamellar cataract (40%) ;10% each of nuclear and posterior lenticonus; 9% of posterior polar cataract; 3% of total cataract, 2 % each of cortical cataract and anterior polar cataract.[25] Wilson et al. reported nuclear cataract (54%), cortical cataract (25%), total cataract (4%).[26]

In our study maximum patients had partial cataract of zonular variety which was visually significant in whom surgical intervention was needed. The mere presence of cataract does not indicate surgical removal. The morphology of the cataract – type and size can be indicative of its visual significance. As young children especially infants cannot be assessed for visual acuity, it is important to assess morphology of the cataract to decide about the necessity of surgical intervention.

Our study highlighted that delayed presentation to hospital for management remains a significant problem. Visually significant cataract with late presentation added to poor visual prognosis.

The Government of India in June 2017 has introduced Rashtriya Bal Swasthya Karyakram (RBSK) guidelines for timely eye screening of newborns in order to prevent avoidable cause of blindness. According to the guidelines, examination of eyes should be carried out immediately after the birth till discharge if admitted.[27] Strict application of the guidelines will help in early diagnosis and treatment of congenital cataract and hence prevent avoidable blindness and visual impairment.

Brückner's test / red reflex test is an inexpensive tool for rapid screening of the newborn for common sight-threatening entities such as congenital cataract, retinoblastoma, asymmetrical refractive error, squint, corneal opacities and other simulating causes for leukocoria [28] This test should be incorporated as a must know in the curriculum of all primary health care providers as well as pediatricians. This would lead to an inexpensive and simple way to detect a vision threatening at the appropriate time.

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In the digital age which we live in now, smartphones can also be used effectively to screen the red reflex. There are smartphone apps like MDEyeCare and CRADLE for early detection of leukocoria which are red reflex based so that a non – ophthalmologist could also make use of it for early detection of leukocoria. Apart from it's use in detection of congenital cataract, it can also be used in diagnosing life threatening condition like retinoblastoma. Amongst the two, MDEyeCare app is preferred as it detects leukocoria in early stages of disease.[29].

Conclusion

Our study showed that there was delayed presentation of visually significant cataract. To ensure optimal visual gain after cataract surgery in children, early diagnosis of the visually significant cataract is the key. Therefore, it is recommended that programs with red reflex test screening should be used for early detection of pediatric cataract for screening newborns. The Rashtriya Bal Swasthya Karyakram (RBSK) of the government of India is one such initiative. Early diagnosis, prompt management and parental counselling is needed for successful visual rehabilitation.

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