Original Article

Congenital Cataract: Morphology and Management

Sana Nadeem, Muhammad Ayub, Humaira Fawad

Pak J Ophthalmol 2013, Vol. 29 No. 3

See end of article for authors affiliations

Correspondence to:
Sana Nadeem
Senior Registrar
Ophthalmology Department
Fauji Foundation Hospital,
Rawalpindi

Purpose: To evaluate the morphology of congenital cataracts presenting to us and their subsequent surgical management and visual rehabilitation.

Material and Methods: A total of 46 eyes of 28 patients in the age range from 3 months to 25 years with unilateral or bilateral congenital cataract (diagnosed at any age), with no other associated ocular pathology of the anterior or posterior segment, no history or features of trauma, and without systemic or syndromic associations, presenting to the Department of Ophthalmology, Holy Family Hospital, Rawalpindi between 1st January, 2012 to 30th September, 2012 were included in this prospective, interventional study.

Results: The most common morphological type of isolated congenital cataract found in our study was lamellar cataract in 12 eyes (26.1%), and total white cataract also in 12 eyes (26.1%), followed by isolated blue dot cataract in 3 eyes (6.5%). Mixed morphologies were found in 13 (28.2%) eyes. Pre-operative visual acuity was better than 6/18 in 13 (28.3%) eyes, less than 6/18 in 15 (32.6%) eyes, and unrecordable in 18 (39.1%) eyes. Best corrected visual outcome was significantly improved, with a visual acuity achieved better than 6/18 in 25 (54.3%) eyes, less than 6/18 in 5 (10.9%) eyes and unrecordable in 16 (34.8%) eyes. (p= 0.000) The minimum follow up was 3 months and maximum follow up was 15 months.

Conclusions: Isolated lamellar and total white cataracts are the common morphologies of congenital cataract found in our study. Good visual outcome can be achieved with early surgical intervention and appropriate visual rehabilitation.

Congenital cataracts account for 1 out of every 2000 live births,[1] and are quite common, causing 10% of all preventable visual loss in children globally.[2] Pediatric cataracts are responsible for more than 1 million childhood blindness in Asia.[3] Visual loss is mainly due to stimulus deprivation amblyopia, strabismus and nystagmus which are proportionately related to the size, location and density of the opacity, especially if bilateral.[4,5] Several different classification systems exist including morphology, etiology, presence of specific metabolic disorders, associated ocular anomalies or systemic findings.[1]

Compared to adults, decision for surgery is more difficult as subjective visual assessment in children cannot be obtained, and surgeons rely largely on the morphology and location of the cataract and behavior of the child. Surgery needs to be undertaken within the first three months of life as indicated by experimental and clinical research,[5] as early detection and management is directly related to the visual outcome.

Controversy[6] still remains as regards to the age at which an IOL can be safely implanted inside the eye. Aphakia management poses a significant problem and needs spectacles or contact lenses. Success; however is directly related to parental compliance and child cooperation. Results of pediatric cataract surgery are based not only on the anatomic success but the postoperative maintenance of a clear visual axis, and aggressive management of pre-existing amblyopia and its prevention.

We embarked on this study, to observe different morphologies of the congenital cataracts which presented to us, and to manage them surgically, with appropriate visual rehabilitation, and to assess the visual outcome after management.

MATERIAL AND METHODS

A total of 46 eyes of 28 patients presenting to Ophthalmology Department, Holy Family Hospital, Rawalpindi from to 1st January, 2012 till 30th September, 2012 who were diagnosed as congenital cataracts on the basis of morphology (any age), and were operated during this period, were included in this study. Exclusion criteria included trauma, uveitis, glaucoma, anterior segment abnormalities, fundus abnormalities and systemic or syndromic associations. A detailed history and physical examination was done, along with visual acuity assessment, tonometry, slit lamp examination, retinoscopy, ophthalmoscopy, B-scan ultrasonography, keratometry and Intraocular lens (IOL) power assessment by SRK-II formula where necessary. The pupils were dilated with cyclopentolate 1% or phenylephrine 10%. All patients were treated with lens aspiration with anterior capsulorhexis via the limbal approach. Primary posterior capsulotomy with anterior vitrectomy was done only in selected cases due to absence of an AC maintainer in our hospital. Primary IOL implantation was done in children above two years of age. All cases were treated with topical steroid-antibiotics for at least 6 weeks. Cycloplegics or systemic steroids were needed in severe postoperative inflammation. The patients were followed up at 1st postoperative day, then 1st postoperative week, then monthly for at least 3 months. Thereafter, follow up was variable, with the range between 3 months to 15 months. Visual acuity was done with Snellen chart in adults, the picture Snellen chart in co-operative children, and fixation was noted in smaller children. Data was analyzed using SPSS version 16. Frequencies and percentages of age, gender, cataract morphology, and complications were noted. Pre and post-operative visual outcome was assessed and Chi square test was applied, with a p value less than 0.05 being considered significant.

RESULTS

A total of 46 eyes of 28 patients ranging from 3 months to 25 years, with a mean age of 9.6 ± 8.1 years, were included in this study. There were 16 (57.1%) females and 12 (42.8%) males. Unilateral cataracts were seen in 3 (10.7%) patients only with bilateral involvement in 25 (89.2%) patients. Consanguinity was present in 16 (57.1%) patients. Morphologically, isolated lamellar cataract with riders was the most common type found in 12 eyes (26.1%), along with total white cataract, also in 12 eyes (26.1%), followed by isolated blue dot cataract in 3 eyes (6.5%), isolated nuclear, sutural and PSCO (posterior subcapsular cataract) in 2 (4.3%) eyes each. A combination of different morphologies were found in 13 (28.2%) eyes, with combined blue dot and sutural in 4 (8.7%) eyes, blue dot and PSCO in 3 (6.5%) eyes, nuclear and PSCO in 3 (6.5%) eyes, coronary and PSCO in 2 (4.3%) eyes and combined lamellar and sutural cataract in 1 (2.2%) eye (Table 1) (Fig. 1).

Table 1: Morphology of Congenital Cataract

Morphology of Cataract	Frequency n (%)
Lamellar	12 (26.1)
Total white	12 (26.1)
Blue dot + Sutural	4 (8.7)
Blue dot	3 (6.5)
Nuclear + PSCO	3 (6.5)
Blue dot + PSCO	3 (6.5)
Nuclear	2 (4.3)
Sutural	2 (4.3)
PSCO	2 (4.3)
Coronary + PSCO	2 (4.3)
Lamellar + Sutural	1 (2.2)

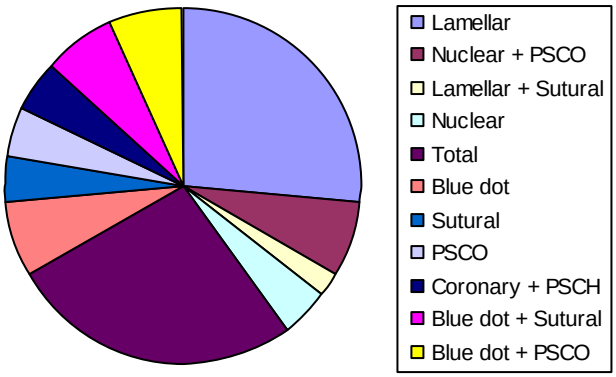

Fig. 1: Pie Chart of Congenital Cataract Morphology

Lens aspiration with Intraocular lens (IOL) implantation was done in 31 (67.4%) eyes, Lens aspiration with anterior capsulotomy alone, was performed in 13 (28.3%) eyes, and Lensectomy with posterior capsulotomy and anterior vitrectomy was done in only 2 (4.3%) eyes due to lack of an AC maintainer. IOL implantation was done in children above 2 years of age. Aphakic and uncooperative children required a secondary procedure for posterior capsular opacification with surgical capsulotomy alone or surgical capsulotomy with a secondary IOL later. Cooperative children and adults were treated with Nd-YAG laser capsulotomies. Visual rehabilitation was done in all patients, either with aphakic spectacles in children less than 2 years and residual refractive error was corrected with appropriate spectacles. Patching was advised to the parents in case of children.

At presentation, visual acuity ranged from light perception to 6/12, with only 13 (28.3%) eyes with visual acuity of 6/18 or better, 15 (32.6%) eyes had vision less than 6/18, and 18 (39.1%) eyes had unrecordable vision. The postoperative best corrected visual outcome was significantly improved (p= 0.000) ranging from unrecordable to 6/6, with 24 (52.2%) eyes having visual acuity of 6/18 or better (Table 2). 3 patients were lost to follow up at 3 months.

Table 2: Visual outcome of surgery

Visual Acuity	Frequency n (%)
Better than 6/18	24 (52.2)
Less than 6/18	6 (13.0)
Unrecordable	12 (26.1)
Missing	4 (8.7)

Early complications included severe inflammation in 22 (47.8%) eyes, mild inflammation in 13 (28.3%) eyes and striate keratitis in 10 (21.7%) eyes. These were managed appropriately with topical antibiotic-steroid combinations, cycloplegics and systemic steroids. Late complications included Posterior capsular opacification (PCO) in 40 (86.9%) eyes, retinal detachment in 2 (4.3%) cases, pseudophakic glaucoma in 1 (2.2%) case, and persistent uveitis leading to phthisis bulbi in 1 (2.2%) case. PCO was managed by surgical capsulotomies in children less than 4 years and older patients were treated with Nd-YAG laser capsulotomy. The patients are still on follow up and are part of a larger study.

DISCUSSION

Congenital cataract is a term used to define lenticular opacities at birth. Infantile cataract encompasses all lens opacities that develop within the first year of birth. The terms are used interchangeably due to some of these opacities being missed at birth only to be discovered later in life by ophthalmologists. They vary in severity from being non-progressive and visually insignificant to causing profound visual impairment.[1]

Bilateral congenital cataract accounts for 15%[7] of blindness in children worldwide. Idiopathic[2,7] cataracts are the most common. Underlying and associated causes of congenital cataract vary worldwide. Isolated hereditary cataracts account for 25% of cases, the most common being autosomal dominant, then autosomal recessive or X-linked.[2,8] Down, Patau, Edward, Turner and Cri du chat syndromes along with systemic diseases like galactosemia, Lowe, Fabry, Alport, Dystrophia myotonica, hypoglycemia, hypoparathyroidism and Marfan syndrome are frequent associations. Maternal infections like rubella, toxoplasma, cytomegalovirus, herpes simplex and varicella (TORCH) may be causative.[1,2,4,8,9]

Morphologically cataracts may be classified into fibre-based and non-fibre based. These include anterior or posterior polar cataracts, lamellar (round, grey shell surrounding a clear nucleus), nuclear or cataracta centralis pulverulenta, sutural or stellate, floriform (flower – shaped), coralliform (coral-shaped), blue dot (punctate cerulean cataract), coronary (supranuclear), subcapsular, total white, disciform, oil-droplet, spear and membranous cataracts. Lamellar cataract is the commonest.[1,2,4,8,9,10]

In our study, isolated lamellar and isolated total white cataract were the most common, but combined patterns accounted for the largest number of eyes. Other studies have shown lamellar,[10] nuclear[11] and total white[12] cataracts to be the commonest.

Visual loss in congenital cataract is predominantly caused by amblyopia, which arises in a number of ways[7]: stimulus – deprivation; competitive inhibition between the two eyes due to unilateral or asymmetrical bilateral cataract; improper aphakia management; or stimulus deprivation secondary to

posterior capsular opacification. Thus amblyopia reversal, treatment and prevention have profound long term implications on the patient.

In unilateral cataract, clinical observational studies have revealed that surgery by six to eight weeks[7] has a better visual outcome as compared to later intervention. This may also be the "critical period" for bilateral disease. Optimal timing for surgery is difficult to establish due to the association of aphakic glaucoma with very early surgery. Some have suggested that early IOL implantation may protect against this complication.[7,13]

Despite significant improvements in surgical, optical and visual rehabilitation techniques, an optimal surgical approach is yet to be established. Several techniques are available like lensectomy, anterior vitrectomy and/or combined with primary posterior capsulotomy. Two main approaches exist for pediatric cataract removal: the limbal approach and the pars plana approach, the latter being considered the most versatile[4]. The anterior chamber maintainer (ACM) is considered vital for pediatric cataract surgery. Anterior capsulorhexis, either manually or with a vitrectomy probe, along with elective posterior capsulectomy and deep anterior vitrectomy has been considered for infants under 2 years of age; above 2 years, this is considered optional.[1,2,4,7,9,13] The pars plana approach is indicated mainly for infants less than 2 years of age, particularly with bilateral cataracts. Simultaneous surgery reduces the risk of relative amblyopia which may occur even when few days apart.[4]

IOL implantation has been advocated in children two years[2] and above, due to problems arising due to IOL power, size, availability, material, refraction change and long term IOL safety.[6] However, many ophthalmologists now implant IOLs in younger age groups like one year with successful outcomes.[14-16] IOL power should be under corrected by 20% in children less than 2 years, and in children between 2 and 8 years, under corrected by 10%.[4,9] The postoperative residual refractive error is corrected with spectacles. Pediatric IOLs should be in the range of 10.5-12mm ideally[17]. Techniques of IOL placement include in-the-bag, ciliary sulcus or IOL optic placement behind the capsular bag.[18] Hydrophilic acrylic IOLs have fewer postoperative complications[15] as compared to rigid PMMA lenses. Heparin coated[7] PMMA IOLs reduce postoperative uveitis. In our study, we implanted either hydrophilic acrylic or rigid PMMA IOLs, with comparable results.

Pediatric eyes are especially prone to complications like fibrinous anterior uveitis, posterior capsular opacification, lens reproliferation (Soemmerring ring), secondary pupillary membranes, aphakic or pseudophakic glaucoma in 25% (often years later), endophthalmitis, retinal detachment (also late) and unpredictable final refraction.[1,2,4,6,7,8,9,13,19]

The visual outcome depends on cataract type, timing of intervention, quality of surgery, and above all, amblyopia management. Poor visual outcome with refractory amblyopia is associated with dense cataracts, unilateral cataracts, late presentation to the ophthalmologist, and poor compliance to occlusion therapy[20]. Bilateral cataracts have been associated with a lesser risk of refractory amblyopia. Dense, central, large and posterior cataracts lead to early amblyopia, and a subsequent poor visual outcome. Partial, less dense, anterior, and smaller cataracts even if detected late, can be managed effectively with a good visual outcome[21]. In our study, most lamellar cataracts although detected late, resulted in very good post-operative vision.

Limitations of our study were many. This is not a study on pediatric patients alone and to evaluate morphology, we included older patients as well. Lack of an ACM prevented us from managing children less than 2 years of age appropriately with a primary posterior capsulotomy and anterior vitrectomy and only irrigation and aspiration was done, which resulted in early PCO formation, necessitating surgical capsulotomies and increasing the number of surgical procedures for every patient.

Final visual outcome in children was poorer as compared to older patients, due to poor parental compliance with spectacles, patching and follow up. Appropriate management of congenital cataract in a developing country poses a lot of problems both for the doctors and the patients. Lack of essential equipment, together with illiteracy, poverty and irregular follow up affect tremendously the management of such cases. Late presentation of children to hospitals results in refractory amblyopia. Unaffordability of contact lenses, poor compliance with aphakic glasses and reluctance to patching all contribute to poor postoperative visual outcomes in aphakic children. Similarly in children who present later and are implanted IOLs, refractory amblyopia is difficult to reverse and owes mostly due to poor compliance of patching. However, partial cataracts even when detected later, when treated, yield good results with much patient and doctor satisfaction.

Early diagnosis and management along with parental advice and support is the key to successful visual rehabilitation. Strategies to screen and detect congenital cataract within the first three months of life are needed for early diagnosis and routine ocular examination[5] of neonates and young infants should be done routinely by ophthalmologists to prevent late detection and subsequent poor visual outcome.

CONCLUSION

Congenital cataract varies considerably in morphological appearance with the major types being lamellar, total white, combined pattern and blue dot. Early surgical management with aggressive postoperative rehabilitation and amblyopia therapy is essential for effective visual outcome. Visual outcome is better for partial, bilateral cataracts as compared to total white or unilateral cataracts.

Author's Affiliation

Dr. Sana Nadeem
Senior Registrar
Ophthalmology Department
Fauji Foundation Hospital, Rawalpindi

Dr. Muhammad Ayub
Senior Registrar
Ophthalmology Department
Holy Family Hospital, Rawalpindi

Dr. Humaira Fawad
Consultant Ophthalmologist
Ophthalmology Department
District Headquarters Hospital, Rawalpindi

REFERENCES

1. **Rosenfeld SI, Blecher MH, Bobrow JC, Bradford CA, Glasser D, Berestka JS.** Lens and Cataract. Section 11. Basic and Clinical Science Course. American Academy of Ophthalmology. San Francisco. 2005; 33-9.
2. **Simon JW, Buckley EG, Drack AV, Hutchinson AK, Plager DA, Rabb EL, Ruttum MS, Aaby AA.** Paediatric Ophthalmology and Strabismus. Section 6. Basic and Clinical Science Course. American Academy of Ophthalmology. San Francisco. 2005; 277-89.
3. World Health Organisation. Prevention of childhood blindness. Geneva: WHO; 1992.
4. **Yanoff M, Duker JS.** Ophthalmology. Second Edition. Mosby: St Louis. 2004; 279-379.
5. **Rahi JS, Dezateux C.** National cross sectional study of detection of congenital and infantile cataract in the United Kingdom: role of childhood screening and surveillance. The British Congenital Cataract Interest Group. BMJ. 1999; 318: 362-5.
6. **Javadi MA.** Pediatric cataract surgery. Editorial. J Ophthalmic Vis Res. 2009; 4: 199–200.
7. **Wormald R, Henshaw K, Smeeth L.** Evidence-Based Ophthalmology. London: BMJ Publishing group; 2008. www.books.google.com. 2012; 47-51.
8. **Kanski JJ, Bowling B.** Clinical Ophthalmology. A systematic approach. Seventh Edition. Elsevier: London. 2011; 298-304.
9. **Denniston AKO, Murray PI.** Oxford handbook of ophthalmology. Oxford University Press: Karachi. 2007; 235-627.
10. **Jain IS, Pillay P, Gangwar DN, Dhir SP, Kaul VK.** Congenital cataract: etiology and morphology. J Pediatr Ophthalmol Strabismus. 1983; 20: 238-42.
11. **Wilson ME, Trivedi RH, Morrison DG, Lambert SR, Buckley EG, Plager DA et al.** The Infant Aphakia Treatment Study: evaluation of cataract morphology in eyes with monocular cataracts. J AAPOS. 2011; 15: 421-6.
12. **Kaid Johar SR, Savalia NK, Vasavada AR, Gupta PD.** Epidemiology based etiological study of pediatric cataracts in Western India. Indian J Med Sci. 2004; 58: 115-21.
13. **Mazhar-ul-Hasan, Qidwai UA, Aziz-ur-Rehman, Bhatti N, Alvi RH.** Complication and visual outcome after pediatric cataract surgery with or without Intraocular lens implantation. Pak J Ophthalmol. 2011; 27: 30-4.
14. **Yorston D.** Intraocular lens implants in children. J Comm Eye Health. 2001; 14: 57-8.
15. **Rowe NA, Biswas S, Lloyd IC.** Primary IOL implantation in children: a risk analysis of foldable acrylic v PMMA lenses. Br J Ophthalmol. 2004; 88: 481-5.
16. **Flicroft DI, Knight-Nanan D, Bowell R, Lanigan B, O'Keefe M.** Intraocular lenses in children: changes in axial length, corneal curvature, and refraction. Br J Ophthalmol. 1999; 83: 265–9.
17. **Bluestein EC, Wilson ME, Wang XH, Rust PF, Apple DJ.** Dimensions of the pediatric crystalline lens: implications for intraocular lenses in children. J Pediatr Ophthalmol Strabismus. 1996; 33: 18-20.
18. **Gimbel HV, DeBroff BM.** Posterior capsulorhexis with optic capture: maintaining a clear visual axis after pediatric cataract surgery. J Cataract Refract Surg. 1994; 20: 658-64.
19. **Speeg-Schatz C.** Results and complications of surgery of congenital cataract. [Article in French] J Fr Ophthalmol. 2011; 34: 203-7.
20. **Harrad R.** Modulation of amblyopia therapy following early surgery for congenital cataracts. Br J Ophthalmol. 1995; 79: 793.
21. **Ondráček O, Lokaj M.** Visual outcome of congenital cataract surgery. Long term clinical results. Scripta Medica. 2003; 76: 95-102.