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Congenital Cataract: Morphology And Management

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Abstract: ***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 92 eyes of 46 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, M.L.B. Medical college, Jhansi UttarPradesh between 1st February, 2017 to 31January, 2018 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 26 eyes (28.3%), and total white cataract in 22 eyes (23.9%), followed by isolated blue dot cataract in 7 eyes (7.6%). Mixed morphologies were found in 25 (27.2%) eyes. Pre-operative visual acuity was better than 6/18 in 22 (23.9%) eyes, less than 6/18 in 36 (39.2%) eyes, and unrecordable in 34 (36.9%) eyes. Best corrected visual outcome was significantly improved, with a visual acuity achieved better than 6/18 in 60 (65.2%) eyes, less than 6/18 in 10 (10.9%) eyes and unrecordable in 22 (23.9%) 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.*

Keywords: *Congenital cataract,morphology, surgical management, visual rehabilitation.*

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I. Introduction

Congenital cataracts and other disorders of visualdeprivation can damage the developing visual systemof a child, and permanently reduce central andperipheral vision. Therefore early diagnosis and surgery,appropriate refractive error correction, amblyopiatherapy and longterm follow‐ up are essential.[1]It is well known that recovery of normal visualfunction after cataract surgery is more probable inadults as compared to children due to impairment ofthe developing visual system.[2,3]The prevalence of congenital cataract has beenreported from 1 to 15 per 10,000 children worldwide,whereas it ranges from 1 to 3 per 10,000 births indeveloping countries.[4] The number of blind childrendue to congenital cataracts globally and in developingcountries are 200,000 and 133,000, respectively.[5]Red reflex examination at birth is an easy methodto screen for congenital cataracts leading to earlydiagnosis and timely surgery.[6] The sensitivity andspecificity of red reflex examination with no pupildilation immediately after birth have been reported as85 and 38.50%, respectively for detection of all typesof congenital ocular diseases,[7]According to the literature, the best visual outcomesmay be achieved when surgery is performed during thefirst 6 weeks of age in unilateral cases and within 5 to8 weeks of age in bilateral cases.[10] Furthermore, age atsurgery and time interval between affected fellow eyesin developed countries have been reported less thanone year and one week, respectively.[11,12,13,14] However, in aecent study by the authors of the current perspective,these figures were 3 years and 3 months,[1] respectively.Amblyopia is the major cause of visual impairment following congenital cataract surgery.[10,11] It was the reason for decreased vision in 76% of cases in the studyby Ledoux et al[16].Strabismus following congenital cataract has beenreported from 13% to 86% in the literature,[[15,16]. Ocular deviation inthese children represents unequal reduced VA in the both eyes due to unilateral or asymmetric bilateral lensopacities.Nondesirable surgical outcomes have been reportedeven in subjects with early cataract surgery, wearingappropriate glasses, contact lenses or implantation ofintraocular lenses (IOL), which has been due to the lackof long‐ term followup (at least up to the age of 10 years)stressing the fundamental role of followup visits formanagement of possible complicationsinchildrenwithcongenitalcataract**.**

II. Material And Methods

A total of 46 eyes of 28 patients presenting to Ophthalmology Department M.L.B. Medical Collage Jhansi up from to 1st February, 2017 till 31th January, 2018 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 medical college. 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. 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.

III. Results

A total of 92 eyes of 46 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 28 (60.8%) females and 18 (39.2%) males. Unilateral cataracts were seen in 4 (8.7%) patients only with bilateral involvement in 42 (91.3%) patients. Consanguinity was present in 26 (56.5%) patients. Morphologically, isolated lamellar cataract with riders was the most common type found in 26 eyes (28.3%), along with total white cataract, also in 22 eyes (23.9%), followed by isolated blue dot cataract in 7 eyes (7.6%), isolated nuclear, sutural and PSC (posterior subcapsular cataract) in 4 (4.3%) eyes each. A combination of different morphologies were found in 25 (27.2%) eyes, with combined blue dot and sutural in 8 (8.7%) eyes, blue dot and PSC in 5 (5.5%) eyes, nuclear and PSC in 6 (6.5%) eyes, coronary and PSC in 4 (4.3%) eyes and combined lamellar and sutural cataract in 2 (2.2%) eye (Table 1)

Table 1: Morphology of Congenital Cataract[17]

Morphology of Cataract	Frequency n (%)
Lamellar	26 (28.3)
Total white	22 (23.9)
Blue dot + Sutural	8 (8.7)
Blue dot	7 (7.6)
Nuclear + PSC	6 (6.5)
Blue dot + PSC	5 (5.5)
Nuclear	4 (4.3)
Sutural	4 (4.3)
PSC	4 (4.3)
Coronary + PSC	4 (4.3)
Lamellar + Sutural	2 (2.2)

Lens aspiration with Intraocular lens (IOL) implantation was done in 70 (76.0%) eyes, Lens aspiration with anterior capsulotomy alone, was performed in 20 (21.7%) eyes, and Lensectomy with posterior capsulotomy and anterior vitrectomy was done in only 2 (2.2%) 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 22 (23.9%) eyes with visual acuity of 6/18 or better, 36 (39.2%) eyes had vision less than 6/18, and 34 (36.9%) eyes had unrecordable vision. The postoperative best corrected visual outcome was significantly improved (p= 0.000) ranging from unrecordable to 6/6, with 60 (65.2%) eyes having visual acuityof 6/18 or better (Table 2). 2 patients were lost to follow up at 3 months

Table 2: Visual outcome of Surgery[17]

Visual Acuity	Frequency n (%)

Better than 6/18	60 (65.2)
Less than 6/18	10 (10.9)
Unrecordable	20 (21.7)
Missing	2 (2.2)

Early complications included severe inflammation in 45 (48.9%) eyes, mild inflammation in 23 (25.0%) eyes and striate keratitis in 19 (20.6%) eyes. These were managed appropriately with topical antibiotic-steroid combinations, cycloplegics and systemic steroids. Late complications included Posterior capsular opacification (PCO) in 82 (89.2%) eyes, retinal detachment in 4 (4.3%) cases, pseudophakic glaucoma in 2 (2.2%) case, and persistent uveitis leading to phthisis bulbi in 2 (2.2%) case. PCO was managed by surgical capsulotomies in children less than 4 yearsand older patients were treated with Nd-YAG laser capsulotomy. The patients are still on follow up and are part of a large study.

IV. 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. 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 cataractacentralispulverulenta, 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.[18,19,21,25,26,27,].Visual assessment should be performed using patternsof fixation and supplemented when possible bypreferential looking charts, or pattern visual evokedpotentials. Measurement of corneal diameter,intraocularpressure, pupillary reflexes, ultrasonographyand indirect ophthalmoscopy should be carried out.(Table 3 & Table 4)

Table 3: Examination Protocol in paediatrics Cataract

History
1. Duration
2. F/H of Congenital Cataract
3. Visual Status: Ambulation in familiar andunfamiliar surroundings
4. Behavioural Pattern and School Performance

Birth History
1. History and Degree of consanguinity
2. H/O maternal infection in 1st Trimester
3. Gestational Age & Birth Weight
4. Birth trauma
5. Supplemental O2 therapy in Perinatal period
6. Developmental Milestones.

Table 4

Ocular Examination
1. Visual Acquity and Fixation Pattern
2. Refraction
3. Cover – Uncover test (Hirschberg's)
4. Note Nystagmus if any

5.5. SLIT LAMP EXAMINATION
- Associated Congenital Anomalies of iris, lens
- Type of Cataract
- Iridodonesis / Phacodonesis

6. Tension applanation if possible
7. Fundus examination if possible
8. B.Scan USG if there is no fundus view.

V. Timing Of Surgery

In unilateral cataract, clinical observational studies have revealed that surgery by six to eight weeks[24] 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.[24]Cataract surgerybefore 4 weeks of age appears to increase the risk ofsecondary glaucoma, whereas waiting beyond 8 weeksof age compromises visual outcome.If the cataract is incomplete at birth, close

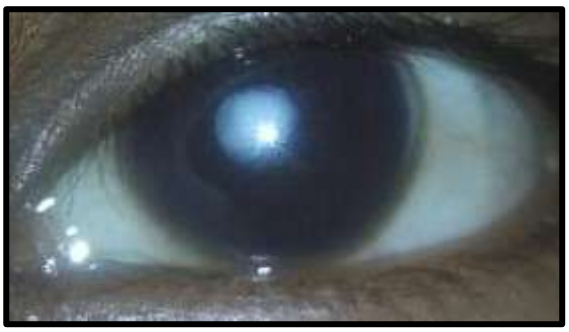
followup isadvised. Evidence of squint or nystagmus is an indicationfor immediate intervention. If the child has unilateralpartial cataract, occlusion therapy should be considered.Counselling of the parents is very important and shouldbe overstressed. It is important to make the parentsunderstand that the treatment of the child starts onlyafter surgery. The necessity for regular follow up, need toenforce the constant wearing of glasses, or contact lensdespite IOL implantation and the requirement of occlusiontherapy after surgery should be emphasized duringcounselling.Pre operative examination under shortanaesthesia with fully dilated pupils is mandatorybefore surgery. Examination under the operatingmicroscope or hand held slit lamp biomicroscope isperformed to assess the type and degree of cataract.The examinations performed under anesthesia include

1. Tonometry to rule out any associated glaucoma,
2. Measurement of corneal diameter
3. PosteriorSegment evaluation with an indirect ophthalmoscopewhenever fundus view is possible
4. Performing aB.Scan Ultrasonography in situations where there isno fundus view
5. Keratometry with a hand heldkeratometer and
6. A.Scan biometry for IOL powercalculation.

VI. Surgical Technique In Children

[28]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 peadiatric cataract removal: the limbal approach and the pars plana approach, the latter being considered the most versatile[21]. The anterior chamber maintainer (ACM) is considered vital for peadiatric 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[18,19,21,24,26].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[21].

IOL implantation has been advocated in children two years2 and above, due to problems arising due to IOL power, size, availability, material, refraction change and long term IOL safety.[23] However, many ophthalmologists now implant IOLs in younger age groups like one year with successful outcomes. 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%.[21,26] The postoperative residual refractive error is corrected with spectacles. Peadiatric IOLs should be in the range of 10.5-12mm ideally. Techniques of IOL placement include in-the-bag, ciliary sulcus or IOL optic placement behind the capsular bag. Hydrophilic acrylic IOLs have fewer postoperative complications as compared to rigid PMMA lenses. Heparin coated[24] PMMA IOLs reduce postoperative uveitis. In our study, we implanted either hydrophilic acrylic or rigid PMMA IOLs, with comparable results.



Preoperative congenital cataract postoperative congenital cataract

In infants with bilateral cataracts it is advantageous toperform surgery in both eyes at the same time, toprevent an amblyopia in the second eye.The lens can be approached through the limbus or parsplicata. Although temporal clear corneal incisions arefavoured in adults, it may not be a good choice inpeadiatric cataracts. Most peadiatric patients have withthe rule astigmatism and temporal incisions may inducefurther worsening of with the rule astigmatism. Hencea superior limbal or scleral tunnel incision is preferred.Using the limbal approach, a high viscosity ophthalmicviscoelastic material should be used to overcome thevitreous pressure and prevent the shallowing of the ant.chamber. If the pupil is small, flexible iris retractors canbe used to enlarge the pupil. Anterior Capsule stainingwithTrypan blue makes the anterior capsulorhexiseasier. If an IOL is implanted the anterior capsulorhexisshould be round, smaller than the optic and placed inthecenter. The capsule is thick and elastic in children,which makes it more difficult to perform a

manualcontinuouscapsulorhexis. The capsulorhexis openingtends to be larger than intended. The anteriorcapsulorhexis can be created preferably with a needleand forceps or it can also be created using a diathermy.Mechanisedcapsulotomy by a vitrector is easier toperform and is the third option for anterior capsulemanagement. The vitrector should be placed with itscutting port posteriorly in contact with the intactanterior capsule. The cutter should be turned on andsuction increased. Cutting rates of 150-300 cuts perminute and aspiration of 150-250 cc/min should beused for vitrectorhexis.Afterrhexis most surgeons perform a hydrodissectionto separate the lens capsule from the cortical material and to shear the epithelial cells away from the capsule.Hydrodissection has a shearing effect on lens epithelialcells and retards PCO.Forremoval of the cortical material, a phacoemulsificationhand piece, a vitrectomy tip, or an automated irrigationaspiration device can be used. It is usually possible toremove the nucleus and cortex with irrigation andaspiration and heparin can be used in irrigating solutionto minimize the inflammation after surgery. Phaco probeand ultrasound energy is sometimes needed in densecataracts. The aqualase liquefaction technique using awarmwaterstream would probably be helpful inremoving these dense cataracts. It is important to removeall the lens epithelial cells to prevent later pco.Since the intact pco rapidly inchildren and maintenance of a clear visual axis is necessaryto prevent amblyopia., a posterior capsulorhexis is preferred by most surgeons. Sometimes rhexis is impossible and avertical posterior capsulotomy with a needle mayuffice. If fibrotic parts are found in the posteriorcapsule, scissors can be used. If persistanthyaloid arteryis found adherent to posterior lens capsule, it shouldbe cut with scissors , and cautery is seldom indicated.The IOL should be placed in the bag rather than theciliary sulcus because of the complications like pupillarycapture and IOL decentration after sulcus fixation.It is debatable whether an anterior vitrectomy shouldbe performed at the primary surgery. Inflammatoryreaction in anterior vitreous is severe in children andcan result in fibrous membrane formation.Anteriorvitrectomy is necessary in children< 2 years of age along with a posterior capsulorhexisas they are subject to severe posterior capsularopacification and intense uveal inflammation. It maynot be necessary in children > 2 years or when you are implanting an IOL which has good biocompatibility withthe anterior vitreous face. It can be performed through the pars plana or through limbalincision upto a depthof 2 mm. This technique appears to be a good way ofpreventing the formation of after cataract.Another technique involves performing an opticcapture, where, the IOL is pressed through the posteriorcapsulorhexis while the haptics remain in the bag.The viscoelastic should be completely removed , andno vitreous should be in the anterior chamber. The sclerais soft and elastic in children and it is hard to achieveaself sealing incision in most cases. So the incision should be closed by sutures. Endophthalmitis is themost serious complication and prophylatic antibioticsare indicated in all cases.

VII. Correction Of Aphakiaiol Implantation

Today most children are implanted with an IOL during surgery and the criteriaof IOL implantation depend on the childs age and whether the cataract is unilateral or bilateral. It is perfectly safe and acceptable to perform primary implantation in a child older than one year.

VIII. Contact Lens

If no IOL is implanted, contact lensesare given as early as possible to prevent stimulusdeprivation amblyopia.Spectacles- In some children with bilateral aphakiaspectacles are better tolerated than contact lenses.

IX. 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.

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