Clinical profile and surgical outcomes in children with posterior lenticonus

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Abstract:
AIMS: The purpose of this study is to describe the clinical profile of children presenting with posterior lenticonus. We also report on visual outcomes following surgery in these patients.

SUBJECTS AND METHODS: Medical records of patients with a diagnosis of posterior lenticonus between January 2000 and December 2016 were reviewed. Data collected included demographic details, preoperative, intraoperative details like type of intraocular lens (IOL) and surgery, and amblyopia therapy. Only the patients with a follow-up of at least 6 months from the time of presentation were included in the analysis.

RESULTS: Forty-eight eyes with posterior lenticonus with at least 6 months of follow-up were studied. Twelve (25%) eyes had a preoperative posterior capsule (PC) dehiscence as seen on slit-lamp biomicroscopy or on ultrasonography. The mean presenting visual acuity in all eyes was 1.034 ± 0.56 logarithm of the minimum angle of resolution (logMAR) units. The mean final visual acuity in these patients was 0.57 ± 0.5 logMAR units. Postoperative visual acuity was found to have a borderline positive correlation with the presence of preoperative strabismus ($P = 0.049$). Younger age at presentation ($P = 0.533$) or the presence of preoperative PC dehiscence ($P = 0.735$) did not influence final visual acuity.

CONCLUSION: Visual acuity improves following surgery with IOL implantation in children with posterior lenticonus and cataract. With the availability of foldable lenses and improved surgical techniques, it is possible to place the IOL in bag despite a preexisting posterior capsular dehiscence. Children with posterior lenticonus who present with strabismus are likely to have poorer visual outcomes.

Keywords: Posterior lenticonus, Posterior subcapsular cataract, cataract surgery

Introduction

Posterior lenticonus is an uncommon lenticular entity which is frequently unilateral. It is a localized protrusion of the posterior capsule (PC) and cortical material of the lens. It was first described by Meyer in 1881.\textsuperscript{[1]} Earliest indication of posterior lenticonus can be seen ophthalmoscopically as an “oil droplet” in the central red reflex. It produces the pathognomonic scissoring movement during retinoscopy. The axial refraction is often markedly myopic, whereas the refractive error peripheral to the lenticus is often hyperopic. The retinoscopic reflex in patients with posterior lenticonus is often distorted, making preoperative optical correction of refractive errors difficult.\textsuperscript{[2]} A new syndrome having bilateral microcornea, posterior megalolenticonus, persistent fetal vasculature, and chorioretinal colobomas (MPCC) has also been described.\textsuperscript{[3]} Several theories have been proposed to explain the development of posterior lenticonus. These include subepithelial capsular hyperplasia, embryologic hyaloid artery traction,\textsuperscript{[4]} inherent weakness of the PC wall, and excessive strain of accommodation, resulting
in posterior herniation of the lens.\textsuperscript{[5]} Franceschetti and Rickli noted an overgrowth or aberrant hypertrophy of the posterior lens cortex.\textsuperscript{[2]} This overgrowth of lens fibers forces the displacement of a thinned, defective PC backwards. Posterior protrusion of the lens is very often associated with development of a cataract. Typically, the opacity of the lens arises from the posterior pole and involves the lens cortex and subcapsular area; however, involvement of the fetal nucleus has also been reported.\textsuperscript{[6]} Lens opacities associated with posterior lenticonus are not always localized within the posterior pole, and total cataracts have also been associated with posterior lenticonus.\textsuperscript{[7]} An ultrasonography or ultrasound biomicroscopy done in these patients may pick a localized protrusion in the posterior lens capsule and may be an ancillary tool in the diagnosis in these cases [Figure 1].

To the best of our knowledge, no study addresses the factors affecting the visual prognosis in patients with posterior lenticonus [Figure 2].

**Subjects and Methods**

Institutional review board clearance was obtained for this retrospective study. Medical records of patients with a diagnosis of posterior lenticonus between January 2000 and December 2016 were retrospectively reviewed. Data collected included demographic details, vision, anterior and posterior segment findings, intraoperative details for the patients who underwent surgery, and the visual outcomes. Visual acuity was assessed by Snellen’s chart or LEA symbols chart at 3 m in cooperative children. Central, steady, maintain method was used to assess vision in infants and uncooperative children. The diagnosis of posterior lenticonus was made clinically on slit-lamp examination. In those with a total cataract, diagnosis was confirmed on an ultrasound B-scan which shows posterior protrusion of lens. We excluded patients with a follow-up of <6 months from the date of presentation.

Statistical analysis was performed using SPSS for Windows software (version 12.0, SPSS, Inc. Chicago, Illinois, USA). Groups were compared using Chi-square/ Fisher’s exact test for categorical variables, means with the t-test. Significance was considered at $P < 0.05$.

**Surgical technique**

After routine cleaning and draping the patient, a superior clear corneal or scleral tunnel incision was made. Anterior capsulorhexis was done using an utrata forceps or a microcapsular forceps followed by automated irrigation/aspiration of the cortical material. Posterior capsulotomy, with anterior vitrectomy, was done wherever required using automated instrumentation.

Intraocular lens (IOL) was implanted in bag wherever possible. IOL was implanted in ciliary sulcus in the absence of adequate posterior capsular support. Patients younger than 12 months of age did not have IOL implantation during primary surgery secondary IOL implantation was done later. A single-piece acrylic foldable IOL (Alcon [TX, USA, SA60AT]) was used for in-the-bag implantation while a 3-piece acrylic foldable IOL [AMOIL, USA] was used for sulcus fixation [Figure 3]. Postoperative target refraction was based on the patient’s age at the time of surgery. IOL power was calculated using...
the SRK II/SRK T formula. Topical steroid drops were prescribed in a dose tapering over 6–8 weeks along with cycloplegic drops. After cataract extraction, glasses were prescribed with a bifocal add for near work.

Amblyopia therapy was instituted wherever applicable.

Results

Electronic medical records of 180 patients with a diagnosis of posterior lenticonus were recovered and reviewed. Of these, 48 eyes met the inclusion criteria of a follow-up of 6 months or more and were included in the study. The mean age at presentation of these children was 7.2 ± 5.9 years. Most frequent complaint at presentation was decrease in vision (68.8%) followed by leukocoria (18.8%), and strabismus (12.5%). One patient (2.8%) each had a morning glory disc, nystagmus, and congenital nasolacrimal duct obstruction. Six (12.5%) eyes had a clear lens with only a posterior capsular bulge, and additional 6 (12.5%) eyes had visually insignificant cataract. These patients were treated conservatively with glasses and amblyopia therapy. Thirty-six (75%) eyes had visually significant cataract requiring surgery. Twelve (25%) eyes had a preoperative PC dehiscence seen on slit-lamp examination or on ultrasonography. Of the 36 patients who underwent surgery, IOL was implanted in the bag in 30 (83.3%) patients. In remaining 6 (16.7%) patients, IOL was implanted in the sulcus as a secondary procedure. In all 12 patients with preoperative PC dehiscence, we were able to place the lens in the bag.

The mean presenting visual acuity in all eyes was 1.034 ± 0.56 logarithm of the minimum angle of resolution (logMAR) units. The mean final visual acuity in these patients was 0.57 ± 0.5 logMAR units. Poor vision, defined as a visual acuity of <6/60, was found in 6 (12.5%) patients. Thirty-six (75%) of these patients required amblyopia therapy in the form of part-time occlusion and 10 (20.83%) patients underwent strabismus surgery in the follow-up period. Postoperative visual acuity was found to have a borderline positive correlation with the presence of preoperative strabismus ($P = 0.049$). There was no statistical correlation between postoperative visual acuity and younger age at presentation ($P = 0.533$) or the presence or absence of preexisting PC dehiscence ($P = 0.735$).

Discussion

Posterior lenticonus is a relatively uncommon entity with limited literature available on visual outcomes in these patients. Conservatively, patients with clear lens or those with visually insignificant cataracts can benefit using mydriatic drops along with appropriate correction of refractive error and amblyopia therapy.

Bradford et al.[8] have reported good results with occlusion in patients with partial cataracts and poor results in those with posterior lenticonus. Cheng et al.[1] found an improvement in visual acuity in 41% of the patients with posterior lenticonus using nonsurgical approach. Although amblyopia is frequently associated with posterior lenticonus, it is well known that there is a generally favorable outcome in terms of visual acuity following the cataract surgery and treatment of amblyopia.[9] Prognostic factors determining visual outcomes in posterior lenticonus patients are largely unknown. Age at diagnosis was studied as a factor determining visual outcome, and different studies have given equivocal results.[10] We also examined the age at presentation as an individual factor determining final visual acuity. However, we did not find any statistically significant correlation between the age at presentation and final visual acuity ($P = 0.533$).

Preoperative posterior capsular dehiscence has been reported as negatively correlated with the final visual outcome principally due to inability to implant IOL in the bag in these eyes. Ciliary sulcus implantation has been associated with multiple long-term complications like rubbing of haptics against the posterior iris surface, leading to pigment dispersion, chronic hyphema, and microhyphemas.[11] However, a study by Lee et al.[12] found that a preexisting PC defect had marginally significant better visual outcomes than patients with posterior polar opacity. The hypothesis suggested was that there is a rapid progression of cataract in eyes with posterior capsular dehiscence leading to an early diagnosis and intervention and thus better visual outcome. We did not find any statistically significant difference in visual outcomes in children with preexisting dehiscence and those without a dehiscence. In contrast to report by Lee et al., we were able to implant IOL in the bag in all children with preexisting posterior capsular dehiscence. We believe that this difference was because of availability of a foldable acrylic lens which is easier to implant in bag despite a preexisting posterior capsular rent compared to the series reported by Lee et al. where several lenses including rigid PMMA lenses were used.

Preoperative strabismus has not been studied as an additional factor affecting the final visual outcome in patients with posterior lenticonus.[11] We found a small but significant positive correlation between the visual outcome and the presence of preoperative strabismus ($P = 0.049$). The presence of strabismus in these cases, perhaps, is an indicator of long-standing nature of problem as well as a clear fixation preference for unaffected eye. We believe that parents should be counseled about guarded prognosis in children who present with lenticonus and strabismus.
Conclusion

Visual acuity improves following surgery with IOL implantation in children with posterior lenticonus and cataract. It is possible to safely place an IOL in the bag, despite pre-existing posterior capsular deficiency due to improved surgical techniques and foldable lenses. Children with posterior lenticonus who present with strabismus are likely to have poorer visual outcomes.

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Conflicts of interest
There are no conflicts of interest.

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