

Surgical outcomes after intraocular lens implantation for posterior lenticonus-related cataract according to preoperative lens status

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PURPOSE: To address the surgical outcomes of pediatric patients with cataracts associated with posterior lenticonus who required cataract extraction and intraocular lens (IOL) implantation according to preoperative lens status.

SETTING: Department of Ophthalmology, Seoul National University Children's Hospital, Seoul, South Korea.

DESIGN: Comparative case series.

METHODS: Patients who had cataract extraction and IOL implantation for posterior lenticonus were divided into 2 groups according to the preoperative lens status. Clinical features and visual outcomes in both groups were comparatively analyzed.

RESULTS: Forty-seven eyes of 43 patients were studied. Thirty-five eyes had lens opacities localized to the posterior pole, and 12 eyes presented with total opacity of the lens. Preexisting posterior capsule defect was identified intraoperatively in 11 eyes with total lens opacity. Eyes with preexisting posterior capsule defects more frequently required ciliary sulcus fixation of the IOL ($P=.01$). The mean follow-up after cataract extraction was 66.9 months \pm 35.9 (SD). The mean final corrected distance visual acuity of patients with total opacity (0.37 ± 0.57 logMAR) was better than that of patients with posterior polar opacity (0.56 ± 0.50 logMAR), with borderline significance ($P=.05$).

CONCLUSIONS: A preexisting posterior capsule defect, found most often in eyes that presented with total lens opacity, could be an obstacle to capsular bag fixation of the IOL. Posterior lenticonus patients with total lens opacity had marginally significantly better visual outcomes than patients with posterior polar opacity.

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Posterior lenticonus is a rare congenital lenticular anomaly¹ characterized by localized protrusion of the posterior lens capsule and cortex. Although there are many theories of what causes posterior lenticonus, including posterior polar traction of the lens by a hyaloid artery remnant,^{2,3} aberrant hyperplasia of posterior lens fibers,^{4,5} and weakness of the posterior lens capsule,^{6,7} the exact pathogenesis of posterior lenticonus remains unclear.

In patients with posterior lenticonus, the typical localized and oil droplet-shaped protrusion of the lens induces refractive errors of lenticular origin, such as myopia and a high degree of astigmatism. As the

disease progresses, lens opacification may occur at the conical protrusion⁶ and is one of the major causes of posterior lenticonus-related visual disturbance. 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.⁸ Lens opacities associated with posterior lenticonus are not always localized within the posterior pole, and total cataracts have also been associated with posterior lenticonus.⁹ In these patients, leukocoria may be a presenting symptom of posterior lenticonus.¹⁰

The diagnosis of posterior lenticonus is not as difficult if posterior conical protrusion of the lens is

apparent preoperatively. However, cortical cataracts or total lens opacity may accompany posterior lenticonus in some patients.¹⁰⁻¹² In patients with an atypical presentation, intraoperative findings are crucial for the diagnosis of posterior lenticonus. In fact, according to a case series that retrospectively studied Korean patients with posterior lenticonus-associated cataracts,¹³ in 15 (32.6%) of 46 eyes, posterior lenticonus was first diagnosed during cataract extraction. If a preexisting, sharply bordered defect of the posterior capsule involving the posterior pole of the lens is detected after aspiration of the lens cortex, the diagnosis can be posterior lenticonus.^{4,6,7}

Most cases of posterior lenticonus are unilateral,⁷ and in some cases the lens opacity may be present at birth.¹⁴ Although amblyopia is common among patients with posterior lenticonus, the visual prognosis of posterior lenticonus is favorable after cataract surgery and visual penalization.^{12,15} However, factors affecting the visual prognosis are largely uncertain in patients with posterior lenticonus. The aim of this study was to address surgical results after cataract surgery and posterior chamber intraocular lens (PC IOL) implantation for posterior lenticonus and to analyze the visual outcomes according to the preoperative lens status.

PATIENTS AND METHODS

A retrospective review of medical records at Seoul National University Children's Hospital from January 1999 to December 2010 was performed to identify patients who had cataract extraction and PC IOL implantation for posterior lenticonus complicated by cataract. Ethical approval was obtained from the Institutional Review Board, Seoul National University Hospital. Patients with a follow-up of less than 12 months after cataract extraction and whose Snellen corrected distance visual acuity (CDVA) could not be measured at the final follow-up examinations were excluded.

The diagnosis of posterior lenticonus was made based on preoperative and intraoperative findings. Posterior lenticonus was diagnosed preoperatively if localized conical protrusion of the posterior lens cortex and posterior capsule was observed on the preoperative slitlamp biomicroscopic examination. The intraoperative diagnosis of posterior

lenticonus was made when a typical cone-shaped protrusion of the posterior capsule or a sharply bordered defect of the posterior capsule involving the posterior pole of the lens was detected after uneventful aspiration of the lens cortex.

Data Collection

The following preoperative, intraoperative, and postoperative data were collected through a retrospective review of medical records. The preoperative data included sex, age at presentation, laterality of disease, family history of pediatric cataract, underlying disease, accompanying eye disease, presenting symptoms, preoperative horizontal ocular alignment, preoperative lens status, preoperative CDVA, and preoperative management (including penalization, pharmacologic dilation of pupil, and correction of refractive error). All patients were placed into groups according to the preoperative lens status. Patients who had lens opacity not localized to the posterior pole and who had no red reflex on dilated fundus examination were in the total opacity group. Others with lens opacity localized to the posterior pole were in the posterior polar opacity group.

Intraoperative data, such as age at cataract extraction, intraoperative status of the posterior lens capsule, and position and type of IOL, were also collected. The preoperative CDVA and final Snellen CDVA were converted to the logMAR scale for statistical analysis. The CDVA of patients who preoperatively were able to perceive light only was not converted to the logMAR scale.¹⁶

Surgical Technique and Postoperative Management

Some patients with unilateral small opacity localized to the posterior pole of the lens were initially treated with nonsurgical methods (occlusion therapy and/or pharmacological dilation of pupil). All these patients ultimately had cataract extraction due to unresponsiveness or poor compliance. All surgical procedures were performed by the same experienced pediatric ophthalmologist (Y.S.Y.).

For cataract extraction, a mechanical anterior capsulorhexis, irrigation/aspiration (I/A) of the lens, posterior capsulotomy, and anterior vitrectomy were performed through a superior scleral tunnel incision. In patients expected to have adequate posterior capsule support after cataract extraction, the PC IOL was fixated in the capsular bag. If the patient had an intact anterior capsule rim in the absence of sufficient posterior capsule support, the PC IOL was implanted in the ciliary sulcus. Because poly(methyl methacrylate) (PMMA) IOLs have been proven in pediatric eyes, a 1-piece PMMA IOL was used for in-the-bag and for sulcus fixation. Over a study period of 11 years, 3 types of 1-piece PMMA IOLs (811 B, Pharmacia & Upjohn, Inc.; LK55 A, Lucid Korea Co., Ltd.; and MZ60BD, Alcon Surgical, Inc.) were available at each timepoint. From 2008, foldable IOLs with acrylate optics were used. A 1-piece acrylate IOL (SA60AT, Alcon Surgical, Inc.) was used for in-the-bag placement, and a 3-piece acrylate IOL with PMMA haptics (MA60BM, Alcon Surgical, Inc.) was used for sulcus placement because it was expected that only the 3-piece acrylate IOL with PMMA haptics would provide adequate tension for stable sulcus fixation. [Figure 1](#) shows a diagram of the IOLs used in the study. The postoperative refraction was

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