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ARTICLE

Newer insights into the clinical profile of posterior lenticonus in children and its surgical, visual, refractive outcomes

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Abstract

Purpose To analyze the clinical profile of patients with posterior lenticonus and their surgical, visual, and refractive outcomes.

Results Retrospective interventional case series of 84 eyes of 63 patients with posterior lenticonus. The incidence of posterior lenticonus was 3.98% during a study period of 5 years. One-third of cases had bilateral posterior lenticonus. The mean age was 4.78 ± 4.28 years (unilateral cases were significantly older than bilateral, $P = 0.0001$). Males were 54%. Mean axial length and keratometry were 21.49 mm and 44.88 D, respectively. Eyes with the bilateral disease were significantly shorter (axial length, $P = 0.0012$) and smaller (horizontal corneal diameter, $P < 0.0001$) compared to those with unilateral disease. While 88% were pseudophakic; 12% were aphakic. The posterior capsular defect was noted intraoperatively in 44%. Sixty-eight percent of eyes had a pre-operative diagnosis of posterior lenticonus, 32% were diagnosed intraoperatively. The mean follow-up period was 1.3 years. Best-corrected visual acuity (BCVA) at 6 months was fair to poor in two-third of patients (median 20/100). The mean ± SD visual acuity (LogMAR) and spherical equivalence for unilateral and bilateral cases were 0.70 ± 0.27, 0.67 ± 0.26D ($p = 0.57$) and 2.04 ± 2.74, 5.15 ± 3.73D ($p = 0.0001$), respectively. Visual outcomes were better in children who are aged 2 years or more ($P = 0.0056$). Eight percent needed a second surgery.

Conclusion We report a higher prevalence of bilateral posterior lenticonus in this cohort. The clinical profile of bilateral disease differs from unilateral disease. The diagnosis is not always clinical. In the bag, intra-ocular lens (IOL) implantation is possible in the majority. The visual outcomes remain fair to poor, possibly due to late presentation and the presence of dense refractory amblyopia.

Synopsis The manuscript consists of the largest series of posterior lenticonus to date. It provides the prevalence of posterior lenticonus along with characteristics difference between unilateral and bilateral cases of posterior lenticonus. Newer insights in terms of diagnostics, pre-operative pick-up rate, how to improve, visual and refractive outcomes of unilateral and bilateral cases are described.

Introduction

Posterior lenticonus is a congenital anomaly of the lens characterized by a localized, round to oval, well-circumscribed protrusion of the posterior lens capsule and cortex [1]. Various theories have been proposed to describe the possible pathogenesis; like traction on posterior lenticonus on the posterior capsule (PC) with hyaloid artery [2] and aberrant hypoplasia of posterior lens fibers [3, 4]. The wider accepted theory states that the posterior lenticonus develops by herniation of cortical lens fibers and PC into the vitreous at an area of PC weakness during fetal development [5]. The presumed cause for the development of cataracts in the posterior lenticonus is mechanical due to posterior bowing of capsule and progressive degeneration of lens fibers [6]. The opacity usually starts at the posterior pole and may progress rapidly to involve the whole lens [7].

The existing literature states that the prevalence of unilateral posterior lenticonus is higher than bilateral cases and the majority of cases are sporadic [8–10]. Bilateral posterior

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lenticonus has been described to be associated with X-linked inherited disorders like Lowe syndrome or in cases of other metabolic syndromes [11, 12]. Unilateral posterior lenticonus has been reported to be inherited by autosomal dominant [13, 14] and rarely by autosomal recessive trait [15] and has shown an association with various ocular anomalies like microcornea [16], Duane syndrome [17], and anterior lenticonus [18].

Mistr and associates reported outcomes of posterior polar and posterior lenticonus cataracts (32 eyes) [10]. Visual outcomes of 25 cases at 4 weeks showed acuity better than 20/40 in 68% of eyes. A recent study by Lee and associates reported 47 eyes of 43 patients with posterior lenticonus-related cataracts [9]. The mean visual outcomes were 0.37 ± 0.57 LogMAR in total cataracts and 0.56 ± 0.50 in posterior polar cataracts (PPCs) (borderline significance, $p = 0.05$). Neither of the above studies reported the refractive outcomes. The aim of this study was to analyze the clinical profile of patients presenting with posterior lenticonus and their surgical, visual, and refractive outcomes and to identify the characteristic difference between the unilateral and bilateral cases.

Methods

We conducted the study at LV Prasad Eye Institute, Hyderabad in South India. The study adhered to the declaration of Helsinki and was approved by the Institutional Ethics Committee. We retrospectively reviewed the electronic medical records of patients with the international classification of diagnosis (ICD) code for posterior lenticonus and PPC from 2014 to 2019. We screened the surgical records of these patients and included the confirmed cases (either preoperatively or intraoperatively) of posterior lenticonus for final analysis. We confirmed the diagnosis pre-operatively either by slit-lamp biomicroscopy or immersion B scan (in total cataract cases with PC bulge or rupture of the PC with cortical matter prolapsed in anterior vitreous); or intra-operatively by noting a typical cone-shaped protrusion of the PC or sharply bordered PC defect with or without positive fish-tail sign (lenticular cortex hanging in vitreous cavity after PC dehiscence).

The demographic details and pre-operative data included age at presentation, age at surgery, gender, presenting complaint, laterality of disease, best-corrected visual acuity (BCVA), anterior segment findings, and ancillary tests/investigations like Ultrasound A-scan, B scan, UBM, AS-OCT, TORCH profile, and other serological workups, if required. We utilized age-appropriate vision charts for documenting visual acuity. In infants where teller acuity chart (TAC) vision was not recordable, we noted the visual behavior as central, steady, and/or maintained (CSM). We graded BCVA as excellent (CSM; 20/25 better), good (CS, 20/50 to 20/25), fair (central unsteady, 20/100–20/50), or poor (eccentric fixation, 20/100 or worse). We did an anterior segment examination using slit-lamp biomicroscopy (conventional or handheld) in co-operative children and noted the lens status. In patients who had a dense cataract obscuring the view of the fundus, we performed an ultrasound B scan to rule out the presence of retinal detachment, persistent fetal vasculature, intra-ocular calcification, or a mass. We also did an immersion B scan to check for posterior capsular defects, if any. While we recorded horizontal corneal diameter (HCD), axial length (AL), and keratometry measurements in all patients; we could obtain anterior chamber depth (ACD) and lens thickness (LT) values for only those patients who underwent optical A-scan biometry. We photographically documented the posterior lenticonus and related cataracts in obvious cases using slit-lamp photographs or high magnification DSLR photographs. We also documented ultrasound biomicroscopy (UBM) and/ or Scheimpflug images in few patients.

All eyes with significant cataracts suspected to have deprivational amblyopia underwent surgery. We performed manual anterior capsulorhexis using a 26G needle and micro-rhexis forceps (Indo German Surgical Corporation, Mumbai, India) in all except one in which we performed a precision pulse capsulotomy (Zepto), Mynosys (Fremont, California). We avoided hydro dissection in all cases and aspirated the lens matter beginning from the periphery and aspirating the central most part at the end. In younger patients less than 8–10 years of age, we performed an additional posterior capsulotomy and limited anterior vitrectomy. We also performed a limited anterior vitrectomy if there was a pre-existing PC defect. We decided on the implantation of an IOL based on HCD (>10.5 mm) and AL (>16.5 mm). In patients with adequate capsular bag support and pre-existing small, eccentric PC defect, we performed in the bag implantation of single piece or multi-piece acrylic hydrophobic lens (AcrySof IOL Model SA60AT; Alcon, Novartis) based on the surgeon's preference. In those with inadequate capsular bag support, we implanted a multi-piece acrylic hydrophobic lens (AcrySof IOL Model MN60AC; Alcon, Novartis) in the sulcus. Occasionally, we also considered a polymethyl methacrylate (PMMA) IOL implantation in older patients. We used the SRK-II formula for the final IOL power calculation and decided on post-operative target refraction based on Enyedi guidelines [19]. We gave appropriate refractive correction to all patients within two weeks of surgery and prescribed aphakic glasses or contact lenses (preferably in unilateral cases) in patients who were left aphakic. We administered part-time occlusion therapy in those who had amblyopia, as early as 1-week in unilateral cases and 1 month in bilateral cases. Post-operatively we followed up all the patient's initial 3 months

for 1-year, and six-monthly thereafter. We performed the examination under anesthesia for younger children who were not co-operative for refraction, intraocular pressure (IOP) measurement, and detailed examination in an outpatient setting. We performed secondary IOL implantation (in the bag or sulcus) in aphakic eyes, once the child was above 2 years of age with adequate ocular measurements.

We analyzed the collected data using Microsoft Excel 2019 version 16.0.6742.2048. We compared preoperative parameters of unilateral and bilateral cases using mean and standard deviations. In unilateral cases, we compared the mean and standard deviations of AL of the affected versus the contralateral eye. We compared visual outcomes for unilateral versus bilateral cases and 2 years or less versus more than 2 years of age using an unpaired *t*-test. We also ran a linear regression analysis to understand the age-wise trend of visual outcomes. For the purposes of statistical analysis, we converted visual acuities to LogMAR. For analysis of refractive outcomes, we excluded aphakic cases to avoid skewing of results. We compared the mean and standard deviation of spherical equivalence of pseudophakic unilateral and bilateral cases. We defined the statistical significance with a cut-off of *P* value less than or equal to 0.05.

Results

In the period from 2014 to 2019, we performed 2286 pediatric lens aspirations ± IOL implantations, of which 176 were traumatic cataracts. So, we operated on 2110 non-traumatic pediatric cataracts during this period, of which 84 (3.98%) eyes had posterior lenticonus. These 84 eyes of 63 patients were included in the final analysis. The results for demographic details are shown in Table 1. Age of presentation and age at surgery was similar in all patients.

Table 1 Demographic details.

Number of Pediatric cataract cases (Jan 2014 to Dec 2019)	2286	
Traumatic cataract cases	176	
Posterior lenticonus cases	84	
% of Posterior lenticonus in the cohort	3.98%	
Total number	Patients/eyes	63/84
Laterality ($n = 84$)	Unilateral cases	43 (66%)
	Bilateral cases	21 (33%)
Gender ($n = 63$)	Male	34 (54%)
	Female	29 (46%)
Age	Minimum	2 months
	Maximum	15 years
	Mean ± SD	4.78 ± 4.28 years

Unilateral cases were significantly older than bilateral cases ($P = 0.0001$). Leukocoria was the most common presenting compliant $n = 42$ (50%), followed by decreased or blurred vision or poor visual behavior $n = 20$ (24%). Eight eyes (9.5%) had no complaints and were diagnosed on either routine examination, screening or showed a posterior lenticonus as an incidental finding. One patient had glare and photosensitivity as a presenting complaint. Pre-operative BCVA was poor in 80 (95%) eyes; whereas two eyes each had BCVA graded as fair and good, respectively. Figure 1 shows the lens morphology on slit-lamp examination.

Familial or cataracts in siblings were present in two (3%) patients (both bilateral). The first one was a 2-year-old male patient with a family history of pre-senile cataract in a father, the details of which were not available, and he was pseudophakic in that eye. The other patient was a 5-year-old girl whose elder sister (age 7 years) had a history of bilateral congenital cataracts, operated elsewhere.

Various other ocular associations in this cohort included sensory strabismus [$n = 27$ (43%), exotropia in 19 (30%); esotropia in 8 (13%)], nystagmus in 2 (3%), dissociated vertical deviation, Duane retraction syndrome and microcornea in 1 (1.6%) each. Exotropia was more commonly seen in unilateral cases (68.5%); whereas esotropia was more common in bilateral cases (62.5%). DRS and DVD were noted in a unilateral case; whereas both cases exhibiting nystagmus were bilateral.

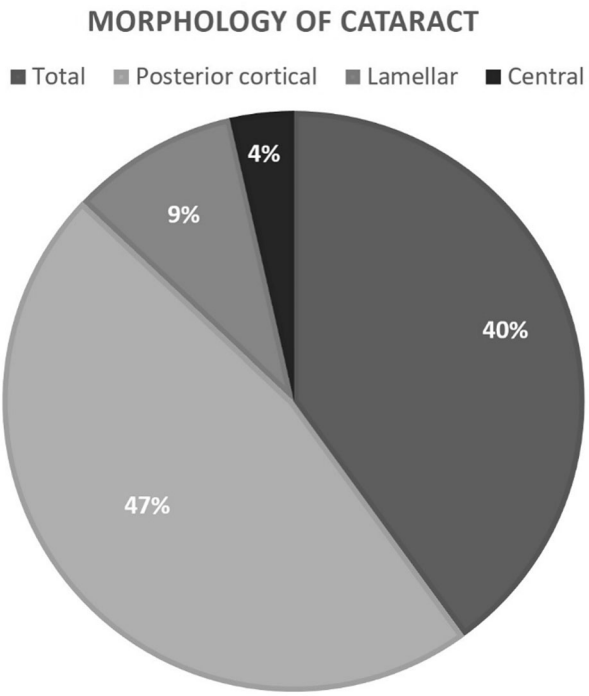

Fig. 1 Morphology of cataract. Pie diagram showing anterior segment findings on slit-lamp examination posterior lenticonus cataract.

Ultrasound B scan was performed in 62 eyes, of which 27 (44%) showed PC bulge and 2 (3%) scans showed evidence of PC rupture with the presence of cortex in the vitreous. The sensitivity of immersion B scan to diagnose posterior lenticonus was 47%. Confirmation of diagnosis of posterior lenticonus, either by slit lamp examination or by B scan ultrasonography was possible in 57 (68%) eyes, of which 33 (39.3%) were unilateral eyes and 24 (28.6%) were bilateral.

The mean AL was 21.49 mm (range 18–26.85 mm). The mean ± SD AL in unilateral and bilateral cases was 22.16 ± 1.78 mm and 20.83 ± 1.86 mm, respectively ($p = 0.0012$). We compared the mean ± SD ALs of the affected eye (22.13 ± 1.77 mm) versus the contralateral eye (21.71 ± 1.33 mm) and found no statistically significant difference ($p = 0.22$). The mean keratometry value was 44.88 D (range 38.98–56.77 D). The mean ± SD keratometry values for unilateral (44.63 ± 1.94 D) versus bilateral (45.15 ± 3.18D) cases showed no significant difference ($p = 0.38$). The mean HCD was 11.47 mm (range 9–13.75 mm) and showed significantly lower values for bilateral cases (11.13 ± 0.72) versus unilateral cases (11.83 ± 0.76 mm, $p \leq 0.0001$). ACD was recorded in 38 patients and had a mean of 3.86 mm (range 2.76–4.7 mm). The mean LT recorded in 24 patients was 3.62 mm (range 2.57–4.58 mm). Pre-operative photographic documentation of cataracts was done in 43 (51%) eyes.

While 74 (88%) were pseudophakic (59 (70%) IOL in the bag, 15 (18%) IOL in the sulcus); 10 (12%) were left aphakic. Out of ten cases of surgical aphakia in the primary setting, nine eyes were of bilateral cases and only one of a unilateral case. The posterior capsular defect was noted intra-operatively in 37 (44%) eyes. Posterior capsulotomy and anterior vitrectomy were performed in 78 (93%) eyes. All patients were implanted with foldable acrylic lenses but one with PMMA. Age-appropriate under correction for target refraction was planned in 53 (63%) eyes. Intraoperative "fishtail sign" was observed in 5 (6%) eyes. In one case, optic capture was noted on the first postoperative day and the patient was taken for IOL repositioning. Another patient required a pars plana vitrectomy for cortical matter removal from the vitreous.

The mean follow-up period was 1.3 years (range: 1 month to 13 years). On a postoperative day 1, BCVA was fair to good in 12 (14%) and poor in 72 (86%) eyes. Occlusion therapy was started in 36 (58%) patients after a postoperative period of 1 month and prescribed suitable refractive correction. Of those who were prescribed patching therapy 83% were unilateral cases. At 6 months postoperative visit 59 patients were examined (Fig. 2); of whom BCVA was improved to excellent in one (2%), good to fair in 29 (45%) eyes whereas 29 (49%) eyes had a poor visual outcome. We also looked for age-wise visual outcomes by linear regression analysis (Fig. 3) ($Y = 0.7558 - 0.02324X$, $R2 = 0.1231$, $p = 0.0065$) and found better visual outcomes in older patients. On unpaired t-test, the mean visual outcomes (LogMAR) for patients of 2 years of age or less were worse (0.77 ± 0.24) than that of more than 2 years of age (0.59 ± 0.23, $p = 0.0056$). The mean ± SD visual acuities converted to LogMAR were 0.70 ± 0.27 and 0.67 ± 0.26, in unilateral and bilateral cases, respectively ($P = 0.57$). The mean ± SD spherical equivalence was 2.04 ± 2.74 and 5.15 ± 3.73 for unilateral and bilateral cases respectively ($P = 0.0001$) (Table 2).

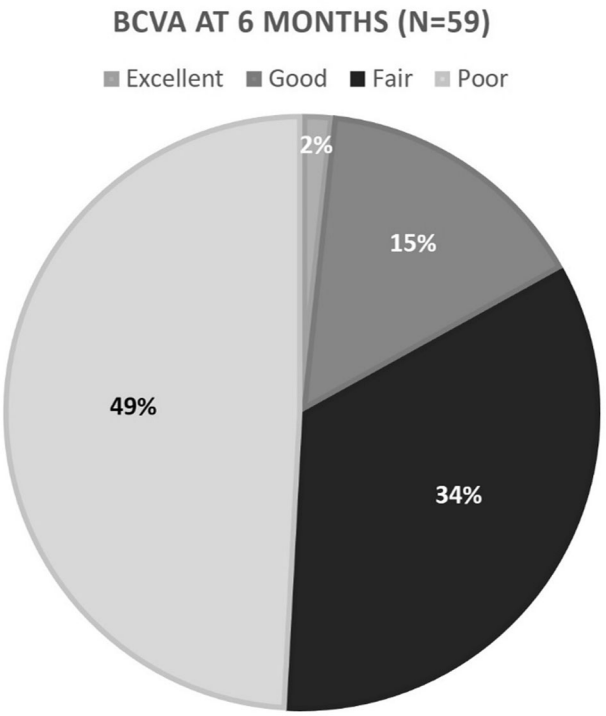

Fig. 2 Visual outcomes at 6 months. Pie diagram showing visual outcomes at 6 months after surgery for posterior lenticonus cataract.

The incidence of raised IOP was 4.7% on the first postoperative day and increased by 1% in a duration of 1 month. All these patients were managed using topical anti-glaucoma medications and did not need any additional intervention. Seven (8.2%) patients needed a second surgery in the form of membranectomy ($n = 2$, 2.3%), secondary IOL implantation ($n = 3$, 3.5%), pars plana vitrectomy ($n = 1$, 1.17% each), and IOL repositioning $n = 1$ (1.17%).

Discussion

To the best of our knowledge, this is the largest reported series of posterior lenticonus-related cataracts. E-supplement summarizes the review of published literature on posterior lenticonus in children.

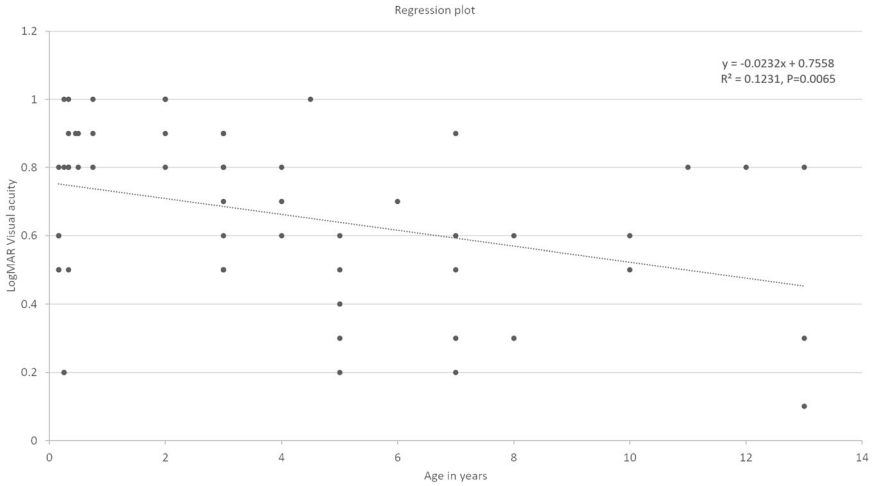

Fig. 3 Linear regression plot. Age-wise visual outcomes by linear regression analysis.

Table 2 Comparison of the clinical and surgical profile of unilateral versus bilateral posterior lenticonus.

Clinical profile		Unilateral (42 eyes of 42 cases)	Bilateral (42 eyes of 21 cases)	Total/*P* value
Age (Mean ± SD) years		6.58 ± 4.27	2.99 ± 3.54	$P = 0.0001$
Strabismus (n = cases)	Exotropia	13/19 (68.5%)	6/19 (31.5%)	19/63 (30%)
	Esotropia	3/8 (37.5%)	5/8 (62.5%)	08/63 (13%)
	DVD	1 (100%)	0	1/63 (1.6%)
	DRS type 1	1 (100%)	0	1/63 (1.6%)
Nystagmus		0	2 (100%)	2/63 (3%)
Axial length	Mean ± SD	22.16 ± 1.78	20.83 ± 1.86	$P = 0.0012$
HCD	Mean ± SD	11.83 ± 0.76	11.13 ± 0.72	$P < 0.0001$
Keratometry	Mean ± SD	44.63 ± 1.94	45.14 ± 3.18	$P = 0.38$
Pre-operative confirmation of diagnosis (n = eyes)		33/84 (39.3%)	24/84 (28.6%)	57/84 (68%)
Surgical profile				
Intraoperative confirmation of the diagnosis (n = eyes)		9/84 (11%)	18/84 (21.5%)	27/84 (32%)
IOL (n = eyes)	Single Piece (SA60AT)	31/84 (37%)	22/84 (26%)	53/84 (63%)
	Multi-piece (SA60MC)	9/84 (11%)	11/84 (13%)	20/84 (24%)
	PMMA	1/84 (1%)	0	1/84 (1%)
	Total (%) of IOL implantation in the primary setting	41/42 (98%)	33/42 (79%)	74/84 (88%)
	Aphakia is a primary setting	1/84 (1.6%)	9/84 (10.7%)	10/84 (12%)
Visual outcomes LogMAR (Mean ± SD)	Mean ± SD	0.70 ± 0.27	0.67 ± 0.26	$P = 0.57$
Refractive outcomes Spherical equivalence (Mean ± SD)	Mean ± SD	2.04 ± 2.74	5.15 ± 3.73	$P = 0.0001$
Second surgery details ($n = 7$)		$n = 4$ Membranectomy: 1 IOL repositioning: 1 Pars plana vitrectomy: 1 Secondary IOL: 1	$n = 3$ Membranectomy: 1 Secondary IOL: 2	

PL posterior lenticonus, *SD* standard deviation, *DVD* dissociated vertical deviation, *DRS* Duane retraction syndrome, *HCD* horizontal corneal diameter, *IOL* intraocular lens.

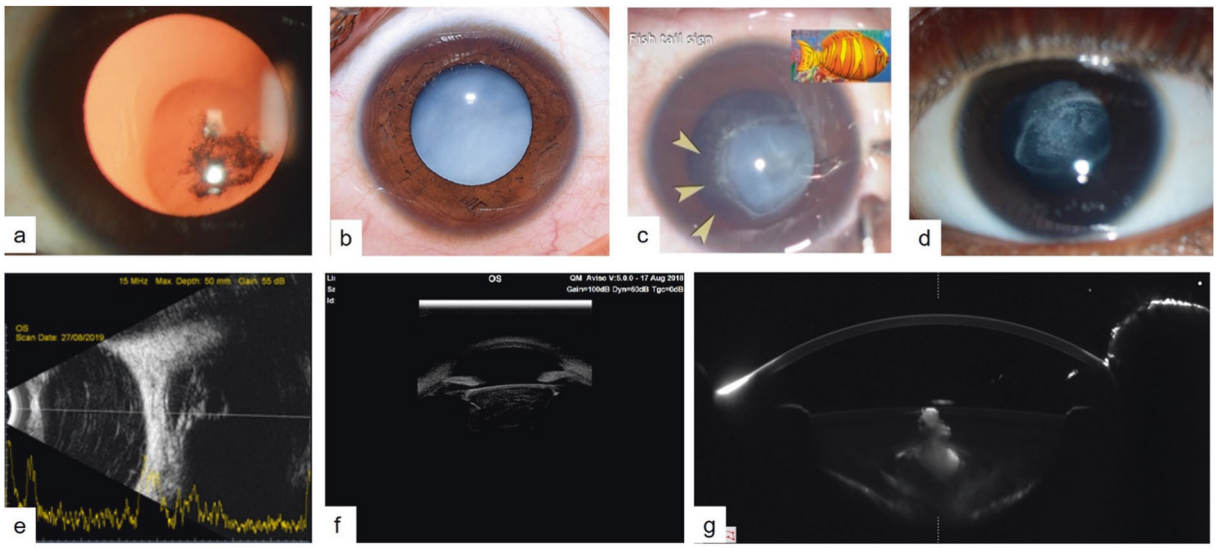

Fig. 4 Collage of various clinical morphologies, signs, and imaging modalities in posterior lenticonus. a Oil droplet reflex is seen on slit lamp with retro illumination associated with posterior cortical opacity. **b** Slit lamp photo of total cataract presenting as leukocoria. **c** Photo showing the posterior capsular defect and white dots in anterior vitreous—fishtail sign. **d** A photo showing the advanced case of posterior lenticonus with intact posterior capsule and surrounding posterior cortical lenticular opacity—atoll sign. **e** Ultrasound immersion B scan image showing bulging of the posterior capsule with no posterior capsule defect. **f** Ultrasound biomicroscopy (UBM) image of posterior lenticonus. **g** Scheimpflug image showing posterior lenticonus and defect in the posterior capsule.

We noted no sex predilection and found that the left and right eyes were equally affected in both unilateral and bilateral cases. Earlier literature [8, 9] has reported the prevalence of bilateral posterior lenticonus to be zero to ten percent. Contrary to this finding, we found a much higher prevalence of bilateral posterior lenticonus. One-third of the cases in our study cohort had bilateral posterior lenticonus. This gave us an opportunity to study and compare the clinical, surgical, and outcomes profile of unilateral versus bilateral posterior lenticonus; which is novel to the existing literature. Most of the cases of bilateral cataracts had simultaneous presentation and had leukocoria in both eyes, except for two cases in which other eye opacity was limited to the posterior cortex (Fig. 4a) and was detected on examination. Table 2 summarises the comparison of profiles of unilateral versus bilateral posterior lenticonus.

In most cases of posterior lenticonus, the age at diagnosis lies between three and 7 years [9, 20]. In our study cohort, leukocoria was noted first by the parents and was the most common presenting complaint in more than half of the eyes. Leukocoria is usually observed if there is dense lenticular opacity. This is consistent with total cataracts being the common anterior segment findings on slit-lamp examination in our study cohort (Fig. 4b).

The characteristic early findings of posterior lenticonus without cataractous changes appear ophthalmoscopically as an "oil droplet" in the central red reflex [7]. This is due to the axial refraction, which is often markedly myopic, whereas the refractive error peripheral to the cone is often hyperopic, a combination of which makes refractive error correction difficult leading to amblyopia. About one-third of the eyes showed an "oil droplet" reflex on a slit lamp and biomicroscopic examination (Fig. 3a). A lenticular cortex hanging in the vitreous cavity after posterior capsular dehiscence is described in the literature and is known as a "fishtail" sign [21]. The fishtail sign can be observed on slit-lamp examination or intraoperatively (Fig. 4c) [22]. An "Atoll" sign is also described in advanced cases of posterior lenticonus with intact PC on slit-lamp examination (Fig. 4d) [23].

Keratometry and AL in the posterior lenticonus are usually within the normal range [24]. We noticed that the eyes with bilateral posterior lenticonus were significantly shorter (AL, $p = 0.0012$) and smaller (HCD, $p \leq 0.0001$) compared to unilateral disease, which can be explained by the significantly younger age of presentation of bilateral cases Whereas, in unilateral cases, the AL of the affected eye did not show any significant difference compared to contralateral eye, signifying there is no significant axial myopia induced by the presence of posterior lenticonus.

If the posterior cortex becomes densely cataractous, accurate identification of the opacity by biomicroscopy may be impossible preoperatively. In dense cataracts, ultrasonography with immersion scan (Fig. 4e) gives better diagnosis suggestive of increased thickness of the lens anteroposteriorly, localized posterior capsular defect with a bulge with or without capsular rupture, and cortical matter in anterior vitreous [25].

As stated earlier, pre-operative clinical diagnosis of posterior lenticonus was certain in only 57 (68%) cases; the

rest of 32% of cases were diagnosed to have posterior lenticonus intraoperatively. The PC defect is present more often in the eyes with total cataract as compared to posterior polar opacity, especially in this age group of the population, as shown in the comparative study by Lee et al. [9]. The posterior lenticonus has a dynamic course of progression as compared to PPC [26] and hence progresses faster to total white opacity. The PC defect in cases of posterior lenticonus is usually elliptical/oval [7]. Whereas, the defect in PPC is usually round and lacks vitreous degeneration [27]. These characteristics are useful to distinguish the two entities intraoperatively, in cases where there is a total cataract. The percentage of intraoperative confirmation of the diagnosis was more for bilateral cases compared to unilateral cases. Out of all eyes, half had inherent PC weakness without any defect. About 44% of eyes had posterior capsular defect; whereas 6% of eyes had a vitreous loss. This is an important finding which adds to the current knowledge of the clinical profile of posterior lenticonus that posterior lenticonus is not always a clinical diagnosis and many cases with dense opacities may have undetected underlying posterior lenticonus. The accuracy of technique for conducting the immersion B scan by the technical staff, could be the contributory reason for its low sensitivity (<50%) in diagnosing posterior lenticonus in this study cohort. Here the role of UBM (Fig. 4f) or anterior segment optical coherence tomography (AS-OCT) (Fig. 4e) to demonstrate the PC and cortical details more accurately, must be reiterated.

Almost all patients had significant cataracts at presentation and underwent cataract surgery soon after the presentation. Only one patient had a progressive opacity, which was observed for around 8 months before surgery was indicated. Hence, the overall age at surgery was similar to that of age at presentation.

Though the visual outcomes were similar for unilateral and bilateral cases, the age-wise trend showed that the older the patient, the better the visual outcomes. Also, patients more than 2 years of age had better visual outcomes. This may be explained by the presence of denser cataracts in the early age of life leading to denser amblyopia. We also looked into refractive outcomes of pseudophakic cases in this cohort after optical correction and amblyopia therapy. We found that refractive outcomes at the last follow-up visit were significantly better for unilateral cases compared to bilateral cases. This can be secondary to higher under correction planned for bilateral cases because of the younger age group.

Our study reported a low rate (2.35%) of intraoperative complications as mentioned in two of our patients needing IOL repositioning for optic capture and pars plana vitrectomy for cortex removal. There was also a low incidence of postoperative rise in IOP and all those who had raised IOP were managed successfully on a course of topical anti-glaucoma medications for a limited period.

In 1991 Cheng et al. [24] reported 41 eyes treated for posterior lenticonus. Their treatment consisted of surgical aspiration of the lens, implantation of an IOL, and amblyopia management. BCVA improved over a pre-operative visual acuity within the first six postoperative months after cataract extraction by two or more lines in 43% (15/35) of the patients tested. The BCVA observed during the entire follow-up period indicated that while 49% (19/39) of the eyes achieved postoperative visual acuity of 20/20 to 20/40, 10% (4/39) were at <20/200. Longer follow-up showed loss of visual acuity due to recurrent amblyopia.

Mistr et al. [18] reported outcomes of posterior polar and posterior lenticonus cataracts. The cohort had 32 eyes with unilateral posterior lenticonus. The mean age of surgery was 5.21 ± 3.83 years. Surgical options differed in approach (corneal, limbal/scleral tunnel), posterior capsulorhexis (pars-plicata/anterior approach), and IOL implantation (78% in the bag, 16% in the sulcus and piggyback IOL in 6%). Visual outcomes of 25 cases at 4 weeks showed acuity better than 20/40 in 68% of eyes. Strabismus was infrequent pre and post-operatively (12–16%).

Lee and associates ([9] in 2014 reported 47 eyes of 43 patients with posterior lenticonus-related cataracts. Patients younger than 24 months, were left aphakic in the primary setting and had delayed secondary IOL implantation (25.5%) after 24 months. In the bag implantation of IOL was possible in 68% of eyes; whereas 32% had IOL implantation in the sulcus. The mean visual outcomes were 0.37 ± 0.57 LogMAR in total cataract and 0.56 ± 0.50 in PPC (borderline significance, $p = 0.05$). Strabismus was present in about 60% of the cases. Neither of the above studies reported the refractive outcomes.

In comparison with earlier studies [9, 19] (poor acuity in 85% and 68% eyes, respectively) the pre-operative BCVA was significantly poorer (95% eyes) in our study cohort. The proportion of cases with total cataracts was also significantly higher (39%); compared to earlier studies (19% [19] and 25.5% [9]). Though the mean age of presentation was similar, data on the period between the first appearance of leukocoria and presentation to the clinic is not available; which possibly can be more in view of the societal context of our population cohort. The percentage of cases with co-existent sensory strabismus or nystagmus is also higher in this cohort, which implies a longer duration of visual deprivation. Our results were consistent with earlier studies [9, 19, 25] for optical correction. IOL implantation was successful in the majority of the patients. Amblyopia therapy in the form of patching was instituted in 58% of cases.

Studies have reported the visual prognosis of posterior lenticonus after cataract extraction and amblyopia therapy in the form of patching to be favorable [24, 25]. In this

study cohort, appropriate optical correction and prompt amblyopia therapy were initiated postoperatively in the majority of the patients. The visual outcomes were fair to poor (20/50–20/100) in most. The possible reasons for this could be delayed presentation, poorer BCVA at the initial presentation compared to earlier studies, and the presence of dense refractory amblyopia.

We reviewed the existing literature on bilateral posterior lenticonus. The earliest report of bilateral posterior lenticonus dates back to 1950 by Fronimopoulos and associates [28] who reported the occurrence of bilateral anterior lenticonus with rudimentary posterior lenticonus in a 25-year-old farmer with no other significant ocular or systemic problem and no family history of a similar problem. In 1983 Pollard reported a series [14] of familial bilateral posterior lenticonus in three families suggesting autosomal recessive inheritance in one and autosomal dominant inheritance in two families. Schipper and associates [29] reported seven members from the same family with bilateral posterior lenticonus suggesting autosomal dominant inheritance pattern. Vivian and colleaugues. [13] found bilateral posterior lenticonus in a mother and her two sons and suggested that bilateral posterior lenticonus might be inherited as an X-linked trait. Later in 2000 Russell-Eggitt [12] reported 15 children from 13 pedigrees; 11 of 13 pedigrees were comparable with X-linked inheritance or autosomal dominant inheritance with variable expression. Various reports [30–33] have reported simultaneous occurrence of bilateral anterior and posterior lenticonus in patients with Alport syndrome. In 2018 Khokar and associates reported a rare association of bilateral posterior lenticonus and persistent fetal vasculature in a 10-year-old girl [34].

In our large cohort of posterior lenticonus, only two patients had a family history suggesting likely autosomal recessive and autosomal dominant inheritance patterns. There was no association with any renal or metabolic syndromes in any of our patients as reported earlier [11, 12]. One patient each had Duane retraction syndrome and microcornea as other ocular associations. No patients showed any evidence of persistent fetal vasculature. This possibly could answer the question of what investigations to consider preferably in suspected cases of posterior lenticonus. In limited-resource settings, an ocular investigation (UBM or AS-OCT) can be preferred over systemic workup.

Our study has few limitations. We could have performed the genetic analysis in cases of bilateral and familial posterior lenticonus but it was not possible due to patient profile; as most of them could not afford to undertake whole genome sequencing. UBM or AS-OCT may help assess the status of PC in cases with total cataract and may have better sensitivity than immersion ultrasound B scan in diagnosing posterior lenticonus. We deferred these additional imaging modalities in most cases due to economic reasons. Though all the surgeons who performed the surgeries were pediatric ophthalmologists who are long-term fellowship-trained in the field with experience of either 2 years or more, the involvement of more than one surgeon may have an impact on extrapolating surgical outcomes. Another limitation was the ability to analyze the visual outcomes at 6 months in two-third of patients, only, as the rest were either short of 6 months postoperative period or lost to follow-up.

To conclude, our study provides newer insights into the clinical profile of posterior lenticonus. Bilateral posterior lenticonus has a much higher prevalence than what has been described in the literature and the clinical profile of those with bilateral disease differs from that of unilateral disease. The diagnosis is not always clinical, and one must suspect posterior lenticonus in total cataracts. Cataract extraction and optical correction with IOL implantation is possible in the majority of patients; despite the presence of pre-existing PC defects, intraoperative complications are rare. Visual outcomes are fair to poor; possibly due to late presentation and the presence of dense refractory amblyopia.

Summary

What was known before

- Posterior lenticonus is a known entity in children all publications are about unilateral posterior lenticonus cases.

What this study adds

- Largest series of posterior lenticonus to date.
- Provides the prevalence of posterior lenticonus.
- Characteristics difference between unilateral and bilateral cases of posterior lenticonus.
- Newer insights in terms of diagnostics, pre-operative pick-up rate, how to improve, visual and refractive. outcomes of unilateral and bilateral cases.

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Compliance with ethical standards

Conflict of interest The authors declare no competing interests.

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