

INTRAOcular LENS IMPLANTATION IN PEDIATRIC EYES WITH POSTERIOR LENTIGLOBUS

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ABSTRACT

Purpose: To report the outcome of intraocular lens (IOL) implantation in pediatric eyes with posterior lentiglobus.

Methods: Retrospective chart review of pediatric cataract surgery cases with primary IOL implantation.

Results: Of 553 eyes that received IOL implantation by the lead author, 364 eyes were identified as having been implanted with an IOL at the time of surgery for a nontraumatic cataract. Nineteen (5.2%) of 364 eyes were diagnosed with posterior lentiglobus. The average age at surgery was 5.8 ± 3.8 years (range, 0.6 to 13.9) years. All eyes had monocular cataract. Gender and ethnic distribution were as follows: male-female ratio, 12:7, and African American–Caucasian ratio, 4:15. A preexisting posterior capsule tear was noted at surgery in eight eyes, six of which presented with leukocoria from white cataracts. Site of IOL implantation was as follows: in-the-bag in 14 and sulcus in five, with optic capture into the anterior and posterior capsulotomy in four of those five. The average postoperative follow-up was 3.7 ± 3.0 years. Median postoperative visual acuity was 20/30, and postoperative strabismus was noted in five eyes. Secondary surgery was required in one eye for visual axis opacification and one eye for lysis of synechiae (diagnosed with postoperative spike in intraocular pressure). No other intraoperative or postoperative complications were observed.

Conclusion: Posterior lentiglobus causes a progressive unilateral cataract. Spontaneous rupture can result in leukocoria from a total white cataract. IOL implantation with posterior lentiglobus provided a satisfactory outcome in children.

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INTRODUCTION

Posterior lentiglobus is the name given to a progressive, well-circumscribed globular bulging of the posterior capsule of the lens. Many published articles refer to this anomaly as “lenticonus.”¹⁻⁸ However, as pointed out by Elschnig in 1895 (cited in Duke-Elder²), the term *lentiglobus* is anatomically more accurate because the bulging is globular and not conical. Historically, after Becker’s histological description of this entity in the rabbit in 1883, Meyer reported it in the human eye in 1888 (cited in Duke-Elder²).

In 1930, lentiglobus was reported to occur in 1 in 100,000 people (cited in Cheng et al⁹). Most cases (95%) are unilateral,^{4,10} and there is little published evidence that unilateral lentiglobus is a familial condition. In contrast, bilateral posterior lentiglobus is more likely to be inherited as an X-linked¹¹⁻¹³ or an autosomal dominant trait.^{8,11,14} It usually presents as the only ocular anomaly; however, it has rarely been reported in association with microcornea,¹⁴ Duane’s syndrome,^{4,15} and anterior lenticonus.^{1,16,17}

Posterior lentiglobus may rarely be noted at birth¹⁸ and may progress as early as the first month of life.¹⁹ However, more commonly it remains mild in the early years of life and may remain undiagnosed until later in childhood. Generally, it is detected between 3 and 15 years of age. Crouch and Parks⁴ and Cheng and associates⁹ reported that the average age at first diagnosis was 3 years 5 months and 5 years 7 months, respectively. Hosal and associates²⁰ noted a median age at diagnosis of posterior lentiglobus of 15 months (range, birth to 78 months). Early findings of posterior lentiglobus appear ophthalmoscopically as an “oil droplet” in the central red reflex. The axial refraction is often markedly myopic, whereas the refractive error peripheral to the lentiglobus is often hyperopic. The retinoscopic reflex is often distorted, making preoperative optical correction of refractive errors difficult. If the posterior cortex becomes densely cataractous, accurate identification of the opacity by biomicroscopy may be impossible preoperatively. If a very opaque cataract exists, ultrasound can aid in making the diagnosis.²¹ At the time of lens aspiration, bulging of the posterior capsule helps confirm the diagnosis of lentiglobus.

The changes in the lens develop as a small central or paracentral abnormality in the posterior lens capsule (Figure 1). The area of defect exhibits progressive bowing under the stress of intralenticular pressure or zonular traction. The subcapsular lamellae become disorganized and opaque. This opacification may progress rapidly over a span of several weeks. Thus, the presumed cause of the cataract is largely mechanical (cited in Amaya et al²²). As the capsule bows posteriorly, there is progressive distortion of the lens fibers themselves and cataract formation, which can be rapid (Figure 2).

Although a relatively rare entity, posterior lentiglobus has been called the most common form of unilateral, developmental cataract in a normal-sized eye.^{4,10} The outcome of intraocular lens (IOL) implantation in 19 eyes with posterior lentiglobus is reported here.

METHODS

This project received an exempt status from the Institutional Review Board of the Medical University of South Carolina. A retrospective search of our pediatric cataract database was performed. Eyes with the diagnosis of posterior lentiglobus were identified. Preoperative data were collected, including age at surgery, gender, ethnicity, laterality of the cataract, visual acuity, axial length,

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Bold type indicates **ROS** member.

keratometry value, corneal diameter, and IOL power and type. Intraoperative data, including posterior capsule rupture and site of fixation of an IOL, were also collected, as were postoperative data consisting of date of the last examination, visual acuity, and any complications.



FIGURE 1

Unilateral posterior lentiglobus in the eye of a 5-year-old boy. The paracentral area of bulging posterior capsule is beginning to show cataractous opacification in the base of the capsular ectasia.

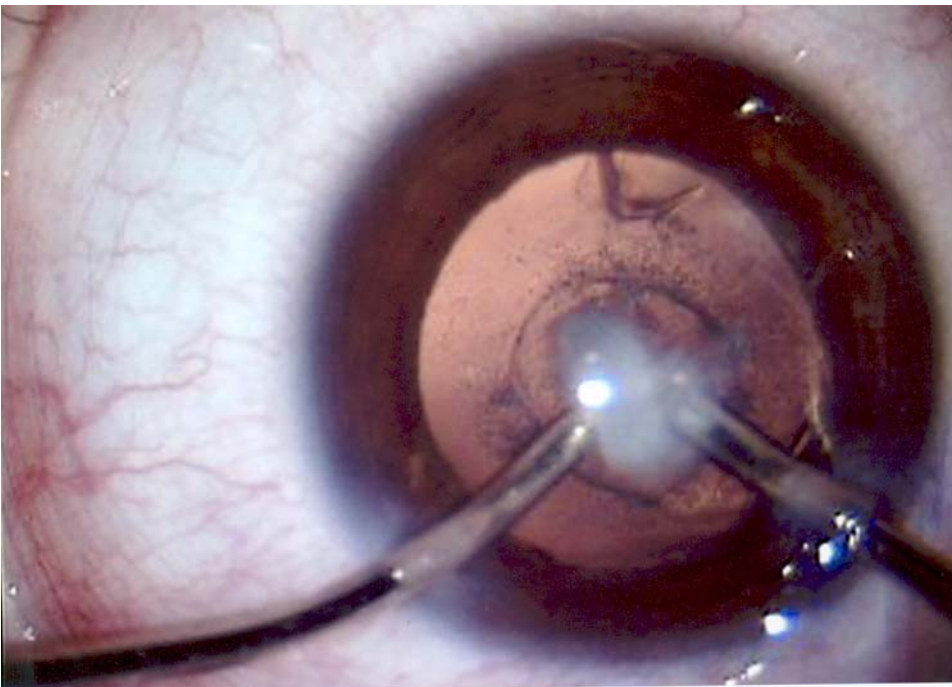


FIGURE 2

Intraoperative photograph showing the removal of a white cataract associated with posterior lentiglobus. The sharply demarcated globular defect in the capsule can be seen posterior to the surgical instruments. The “fishtail” sign (white dots of cortical material in the vitreous that fishtail back and forth on eye movement) was present, indicating a preexisting posterior capsule rupture.

RESULTS

Of 553 eyes that received IOL implantation by the lead author, 364 eyes were identified as receiving primary IOL implantation in nontraumatic eyes. Nineteen (5.2%) of 364 eyes were diagnosed with posterior lentiglobus. All 19 eyes in this series had monocular cataract. This represents 19 (14.1%) of 135 nontraumatic monocular cataracts. The Table describes the results of the individual eyes in detail. Average age at surgery was 5.8 ± 3.8 years (range, 0.6 to 13.9). Gender and ethnic distribution were as follows: male-female ratio, 12:7, and African American–Caucasian ratio, 4:15. Preoperative visual acuity was recorded as “no fixation” in five eyes. Central and steady but not maintained fixation was noted in two eyes. One eye each was noted to have light perception only, counting

fingers, and hand movements. The remaining nine eyes had preoperative Snellen visual acuity ranging from 20/50 to 20/300 with a median of 20/100. Preoperative strabismus was noted in two eyes. Preoperative axial length and corneal diameter were 22.6 ± 1.5 mm and 11.9 ± 0.5 mm, respectively. Posterior capsule rupture was noted in eight eyes, six of which had resulted in leukocoria from a white cataract preoperatively. In four of these eyes, the preoperative A-scan tracing was highly suggestive of preoperative rupture. Site of IOL implantation was in-the-bag in 14 and sulcus in five, with optic capture into the anterior and posterior capsulotomy in four of those five. For postoperative results, only eyes with a minimum of 1 month follow-up ($n = 17$) were analyzed. Average follow-up was 3.7 ± 3.0 years ($n = 17$). Median postoperative visual acuity was 20/30, whereas postoperative strabismus was noted in five eyes. Secondary surgery was required in one eye for visual axis opacification and one eye for lysis of synechiae (diagnosed with a temporary spike in postoperative intraocular pressure). No other intraoperative or postoperative complications were observed.

DISCUSSION

Posterior lentiglobus increases the intraoperative challenges of cataract surgery in children because of the thin, floppy, and, at times, ruptured posterior capsule. Many theories have been proposed to explain the pathogenesis of posterior lentiglobus. Khalil and Saheb⁵ reported that the condition most likely resulted from herniation of cortical lens fibers and the posterior capsule into the vitreous at an area of weakened posterior capsule from fetal development. Franceschetti and Rickli (cited in Duke-Elder²) noted an overgrowth or aberrant hypertrophy of the posterior lens cortex. They theorized that this overgrowth of lens fibers forces the backward displacement of a thin and defective posterior capsule. A theory of overgrowth of posterior lens fibers, producing a phakoma of the lens, has also been suggested by Pergens (cited in Duke-Elder²). Lisch (cited in Duke-Elder²) further speculated that a disturbance in the tunica vasculosa lentis accounts for the anomaly. Kilty and Hiles²³ reported posterior lentiglobus in association with a persistent hyaloid artery remnant. The association of posterior lentiglobus with a persistent hyaloid artery remnant was originally described by Butler (cited in Duke-Elder²). Only one eye in the series reported here had a persistent hyaloid artery remnant. Marsh (cited in Duke-Elder²) suggested that highly plastic epithelium of a young child's lens can herniate posteriorly with the strain of accommodation. Mann (cited in Duke-Elder²) suggested a congenital defect, which arises within the lens sometime after the sixth month of gestation, because the lentiglobus fibers do not involve the Y-sutures or the embryonal nucleus. Crouch and Parks⁴ suggested that the inherent weakness of the diaphanous portion of the posterior lens capsule contributes to disruption of the normal lamellar arrangement of the lens fibers, resulting in cataractous changes. Cheng and colleagues⁹ noted that the cataract formation is due to disruption of the subcapsular cortical lens fibers in the area of the weakened, defective posterior capsule and that cortical overgrowth produces a progression of the lens opacification over the posterior lentiglobus. Trauma has also been reported to cause a lentiglobus-like defect.²⁴ Concussion trauma with subsequent rise in IOP in a predisposed eye (an eye with early posterior lentiglobus) may exaggerate the process of developing posterior lentiglobus and cataract. Ocular trauma may also merely bring the patient to an ophthalmologist, when detection of a previously undiagnosed lentiglobus may occur.

As is evident from the preceding literature review, the etiology of posterior lentiglobus remains somewhat of a mystery. In the series reported here, the area of capsule bulging varied in size from 2 mm to 7 mm and varied in location from central to paracentral. When not in the center, a nasal paracentral location was often noted. However, in only one case was a hyaloid artery remnant seen at the tip of the capsular bulge. Eyes with persistent fetal vasculature (PFV) (formally called persistence and hyperplasia of the primary vitreous) do not, characteristically, have any bulging or weakness of the posterior capsule. Therefore, posterior lentiglobus does not fit into the spectrum of PFV, because vessel remnants are usually not present and microphthalmia is absent. What causes the posterior capsule abnormality and the subsequent progressive bulging of the capsule that leads to cataract formation and, at times, spontaneous capsule rupture? Why is it nearly always unilateral, and why is it not associated with microphthalmia or anterior segment dysgenesis? These questions remain unanswered.

Nineteen (5.22%) of 364 eyes were diagnosed with posterior lentiglobus in the series reported here. Parks and coworkers¹⁰ noted it in 33 (19.8%) of 167 eyes. In the current series, among monocular nontraumatic cataract, the prevalence was 19 of 135 (14.1%). Hosal and coworkers²⁰ noted it in 19 (34.5%) of 55 eyes with nontraumatic cataract in children before 8 years of age. In the current series, care was taken to distinguish posterior polar cataracts with no sign of a bulge in the posterior capsule from posterior lentiglobus. An unequivocal central or paracentral bulge in the posterior capsule was required for the diagnosis of posterior lentiglobus. Posterior polar cataracts in children often have a moth-eaten appearance to the posterior capsule, but no bulge is seen. This may explain the lower percentage of posterior lentiglobus in the current series as compared to the report of Parks and

TABLE. DETAILS OF 19 PATIENTS WITH POSTERIOR LENTIGLOBUS

ID	EYE	AGE (YR)	SEX	RACE	PREOP VISION	PREOP STRABISMUS	AXIAL LENGTH	K	CORNEAL DIAMETER	PREOPERATIVE DIAGNOSIS	TYPE IOL	SITE OF FIXATION OF IOL	POSTOP FOLLOW-UP (YR)	POSTOP COMPLICATION	POSTOP VISION	POSTOP STRABISMUS
1	OD	4.0	M	Cauc	20/100	ET	21.9	45.4	11.0	Posterior lentiglobus	Acrysof MA 60	Capsular bag	6.5	No	20/30	ET + HT
2	OS	13.7	M	Af-Am	20/80	No	ND	ND	ND	Posterior lentiglobus	Storz P359UV	Capsular bag	ND	ND	ND	NA
3	OS	9.4	M	Cauc	20/50	No	22.1	ND	ND	Posterior lentiglobus	Acrysof MA 60	Capsular bag	6.0	Postop IOP rise, synechiolysis	20/30	NA
4	OS	7.2	M	Af-Am	HM	No	22.3	46.3	11.8	Posterior lentiglobus, total cataract	AcrySof MA60AT	Optic capture	1.7	No	20/25	NA
5	OD	6.1	F	Cauc	20/160	No	22.4	42.8	12.5	Posterior lentiglobus	AcrySof SA60AT	Optic capture	4.2	No	20/25	NA
6	OD	1.1	M	Cauc	No fixation	No	22.1	43.5	11.5	Posterior lentiglobus	AcrySof SA60AT	Capsular bag	3.4	Removal of VAO	20/50	NA
7	OD	0.6	M	Cauc	CSUM	No	19.2	44.1	12.0	Posterior lentiglobus	AcrySof SA60AT	Capsular bag	4.6	No	20/40	ET
8	OD	7.6	F	Cauc	20/200	No	22.6	43.7	ND	Posterior lentiglobus	Storz P359UV	Sulcus	11.3	No	20/20	NA
9	OS	7.6	M	Af-Am	20/300	No	25.6	42.5	11.0	Posterior lentiglobus	AcrySof SA60AT	Capsular bag	1.6	No	20/250	NA
10	OS	1.4	M	Cauc	No fixation	No	22.5	44.6	12.5	Total cataract, PFV with posterior lentiglobus and vitreous anomaly	AcrySof SN60AT	Capsular bag	0.4	No	CSUM	XT
11	OS	7.4	F	Cauc	LP	No	23.5	ND	ND	Total cataract	AcrySof MA60AT	Optic capture	0.2	No	20/20	NA
12	OD	2.9	M	Cauc	No fixation	No	24.1	44.1	12.5	Total cataract, posterior lentiglobus	Acrysof MA 60	Capsular bag	4.3	No	20/25	NA
13	OD	6.4	F	Cauc	20/60	No	21.3	44.5	ND	Posterior lentiglobus	Acrysof MA 60	Capsular bag	5.2	No	20/25	NA
14	OD	5.4	M	Cauc	CF	No	21.7	41.8	12.0	Posterior lentiglobus	AcrySof SN 60	Capsular bag	0.4	No	20/200	NA
15	OS	13.9	M	Cauc	20/100	No	23.4	42.5	ND	Posterior lentiglobus	Storz P359UV	Capsular bag	0.6	No	20/25	NA
16	OS	3.3	F	Cauc	No fixation	No	22.6	40.8	ND	Posterior lentiglobus	Pharmacia 820A	Capsular bag	7.1	No	20/125	X(T)
17	OS	2.5	F	Cauc	CSUM	No	22.3	44.3	12.0	Posterior lentiglobus	AcrySof SA60AT	Capsular bag	0.2	No	20/30	X(T)
18	OD	3.2	M	Cauc	No fixation	No	22.5	43.5	12.0	Posterior lentiglobus	AcrySof SA30AL	Optic capture	3.1	No	20/20	NA
19	OS	6.9	F	Af-Am	20/300	ET	25.4	44.6	ND	Posterior lentiglobus	Acrysof MA 60	Capsular bag	ND	ND	ND	NA

Af-Am = African-American; Cauc = Caucasian; CF = counting fingers; CSUM = fixation that is central and steady, but unmaintained; ET = esotropia; HM = hand motion; K = keratometry; LP = light perception; ND = no data; PFV = persistent fetal vasculature; VAO = visual axis opacification; XT = exotropia; X(T) = intermittent exotropia.

coworkers,¹⁰ in which no cataract was designated as posterior polar.

Average age at surgery in the current series was 5.8 ± 3.8 years (range, 0.6 to 13.9). Parks and coworkers¹⁰ reported an average age at surgery of 3.37 ± 2.4 years. Cheng and colleagues⁹ reported an age range of 2 months to 11 years and 8 months (mean, 4.5 years). Hosal and colleagues²⁰ noted a median age at surgery of 49 months (range, 5 to 85 months). A preponderance in female patients has been reported in the literature (cited in Duke-Elder²). Parks and colleagues¹⁰ noted a female preponderance, whereas Cheng and coworkers⁹ noted no predilection for either sex. In the current series, more male than female patients were operated on for posterior lentiglobus (male-female ratio, 12:7). Median preoperative visual acuity was 20/300 in the current series, as opposed to 20/50 as described by Parks and coworkers.¹⁰

Even eyes with an intact posterior capsule are at higher risk of intraoperative rupture, and appropriate surgical maneuvering must be performed to avoid intraoperative rupture. In eyes with a preexisting rupture of the posterior capsule, the lentiglobus defect can be hidden behind a dense white central cataract, especially when viewed through an undilated pupil.²⁵ Preoperative evaluation under maximum dilatation helps to unveil the important diagnostic signs. Hydrodissection is contraindicated in cases of cataract with known or suspected posterior lentiglobus.²⁵ Hydrodissection leads to a sudden buildup of hydraulic pressure, which can cause uncontrolled tearing of the posterior capsule at the lentiglobus defect, sending lens material into the vitreous and threatening the capsular bag stability. With closed-chamber bimanual irrigation and aspiration, the incisions can be made to promote a tight fit around the instruments to maintain anterior chamber stability by reducing fluctuations of the iris-capsule diaphragm and minimizing stress on the lentiglobus defect during the surgery. It is occasionally possible to convert a preexisting capsule rupture into a posterior curvilinear continuous capsulorrhexis (PCCC) to support the IOL better and resist further tearing. However, sometimes the preexisting tear is very large, and it is not possible to convert it to a round PCCC of desired size. When operating on an eye with lentiglobus, it is important to take care not to extend the opening in the posterior capsule. Avoiding hydrodissection and using bimanual irrigation and aspiration help to avoid extending the posterior capsule. This also helps to achieve in-the-bag fixation of an IOL. In the current series, in-the-bag fixation was achieved in 14 eyes.

Median postoperative visual acuity in the current series was 20/30. Hosal and coworkers²⁰ noted that 42.1% of their patients (eight of 19 eyes) with monocular posterior lentiglobus cataracts attained visual acuity of 20/40 or better in the affected eye, 36.8% (seven of 19) attained 20/50 to 20/80, and 21.1% (four of 19) attained less than or equal to 20/100. The investigators further noted that 22.2% (four of 18) also attained binocularity and had 100 sec/arc or better stereoacuity. Cheng and coworkers⁹ noted that 19 eyes (49%) attained postoperative visual acuity in the 20/20 to 20/40 range, seven (18%) eyes attained 20/50 to 20/100, four (10%) eyes attained 20/200, and four (10%) eyes attained less than 20/200. Two young patients (5%) had central, steady, and maintained visual fixation reflexes, and three additional patients (8%) had central, steady, but not maintained fixation.

In conclusion, posterior lentiglobus commonly causes a monocular developmental cataract in the absence of microphthalmia. It may be undiagnosed early in life when the capsular bulge produces a highly myopic and somewhat distorted central lens reflex but no noticeable cataract. Later, it can present after spontaneous capsule rupture as a total cataract camouflaging the lentiglobus defect. A-scan ultrasound can help in the diagnosis of preoperative posterior capsule rupture in eyes with total cataract. Despite the congenital and mostly unilateral nature of posterior lentiglobus, the visual prognosis is often good, because the cataract characteristically develops slowly over time after visual fixation reflexes have formed. However, a delayed diagnosis of posterior lentiglobus may result in dense amblyopia from form vision deprivation, which may be resistant to occlusion therapy after surgery.

REFERENCES

1. Batra DV. Bilateral anterior and posterior lenticonus. *Ophthalmologica* 1965;150:107-110.
2. Duke-Elder SS. Posterior lentiglobus (lenticonus). In: Duke-Elder SS, ed. *System of Ophthalmology. Vol 3. Normal and Abnormal Development. Part 2. Congenital Deformities*. St Louis: Mosby; 1964:700-706.
3. Howitt D, Hornblass A. Posterior lenticonus. *Am J Ophthalmol* 1968;66:1133-1136.
4. Crouch ER Jr, Parks MM. Management of posterior lenticonus complicated by unilateral cataract. *Am J Ophthalmol* 1978;85:503-508.
5. Khalil M, Saheb N. Posterior lenticonus. *Ophthalmology* 1984;91:1429-1430.
6. Simons BD, Flynn HW Jr. A pars plana approach for cataract surgery in posterior lenticonus. *Am J Ophthalmol* 1997;124:695-696.
7. Osher RH, Marques FF, Marques DM, Cionni RJ. Focal multistriae: postoperative finding in posterior lenticonus patients. *J Cataract Refract Surg* 2003;29:2029-2031.
8. Pollard ZF. Familial bilateral posterior lenticonus. *Arch Ophthalmol* 1983;101:1238-1240.
9. Cheng KP, Hiles DA, Biglan AW, Pettapiece MC. Management of posterior lenticonus. *J Pediatr Ophthalmol Strabismus* 1991;28:143-149.
10. Parks MM, Johnson DA, Reed GW. Long-term visual results and complications in children with aphakia. A function of cataract type. *Ophthalmology* 1993;100:826-840.
11. Gibbs ML, Jacobs M, Wilkie AO, Taylor D. Posterior lenticonus: clinical patterns and genetics. *J Pediatr Ophthalmol Strabismus* 1993;30:171-175.
12. Russell-Eggitt IM. Non-syndromic posterior lenticonus a cause of childhood cataract: evidence for X-linked inheritance. *Eye* 2000;14(Pt 6):861-863.
13. Vivian AJ, Lloyd C, Russell-Eggitt I, Taylor D. Familial posterior lenticonus. *Eye* 1995;9(Pt 1):119-123.
14. Bleik JH, Traboulsi EI, Maumenee IH. Familial posterior lenticonus and microcornea. *Arch Ophthalmol* 1992;110:1208.

15. Capobianco S, Magli A. Retraction syndrome with posterior lenticonus. *J Pediatr Ophthalmol Strabismus* 1980;17:96-100.
16. Vedantham V, Rajagopal J, Ratnagiri PK. Bilateral simultaneous anterior and posterior lenticonus in Alport's syndrome. *Indian J Ophthalmol* 2005;53:212-213.
17. Kapoor S. Familial anterior and posterior lenticonus. *Ophthalmologica* 1979;178:186-189.
18. Seidenberg K, Ludwig IH. A newborn with posterior lenticonus. *Am J Ophthalmol* 1993;115:543-544.
19. Mohny BG, Parks MM. Acquired posterior lentiglobus. *Am J Ophthalmol* 1995;120:123-124.
20. Hosal BM, Biglan AW, Elhan AH. High levels of binocular function are achievable after removal of monocular cataracts in children before 8 years of age. *Ophthalmology* 2000;107:1647-1655.
21. Fett D, Paez JH, Isenberg S. Infantile leukocoria caused by posterior lenticonus. *Ann Ophthalmol* 1984;16:679-680.
22. Amaya L, Taylor D, Russell-Eggitt I, et al. The morphology and natural history of childhood cataracts. *Surv Ophthalmol* 2003;48:125-144.
23. Kilty LA, Hiles DA. Unilateral posterior lenticonus with persistent hyaloid artery remnant. *Am J Ophthalmol* 1993;116:104-106.
24. Tipshus AF. Posterior traumatic lenticonus. *Arch Ophthalmol* 1969;82:548-549.
25. Vasavada AR, Praveen MR, Nath V, Dave K. Diagnosis and management of congenital cataract with preexisting posterior capsule defect. *J Cataract Refract Surg* 2004;30:403-408.

PEER DISCUSSION

DR EVELYN A. PAYSSE: Dr. Wilson has presented an excellent review on posterior lentiglobus including his experience with this entity at Storm Eye Institute. One of the major points he brings up is that even though posterior lentiglobus is a relatively uncommon type of pediatric cataract, it is the most common cause of non-traumatic unilateral cataract in children. The prevalence in his series was roughly 5% in all non-traumatic cataracts but 14% of his non-traumatic unilateral cataracts. Others report varying prevalence ranging between 20%-34%.¹⁻² Is it possible this condition is becoming less common?

Dr. Wilson discusses the many theories on the etiology of posterior lentiglobus. There are many. One theory proposes that there is a congenital weakness of the posterior capsule related to the presence of a persistent hyaloid artery. Another proposes a congenital weakness of the posterior capsule associated with increased intralenticular pressure, which in turn causes bulging of the subcapsular cortex into the defect and derangement of these fibers and secondary cataract. No one really knows why posterior lentiglobus occurs.

Dr. Wilson discussed the progression of this developmental cataract and how it's usually diagnosed between the ages of 3 and 5 years old with ages of surgery ranging widely from 3-15 years of age.²⁻⁴ He does not directly discuss the tempo of this progression but the implication is that it varies substantially as the range of age at surgery is very large in comparison to the age at diagnosis.

Dr. Wilson's postoperative visual acuities very impressively went from a mean preoperative acuity of 20/300 to a mean postoperative acuity of 20/30. Such good visual results despite these cataracts being unilateral in a pediatric population underscore the fact that posterior lentiglobus is typically slowly progressive, allowing for good visual development early on and the ability to wait on surgical intervention until a significant opacity occurs.

Dr. Wilson then gave us some technical tips and tricks to use when facing a patient with posterior lentiglobus in need of cataract surgery. He discussed the very important use of the A-scan in helping to diagnose this condition preoperatively so that we may adjust our surgical approach. Because of the weakness of the posterior capsule, hydrodissection should be avoided and the use of a tight I/A unit to maintain intraocular pressure during the case is critical.

To summarize, Dr. Wilson has presented an excellent review of posterior lentiglobus, his approach to surgery, and his excellent visual results. His results underscore the fact that we can and should wait for these cataracts to become visually significant before intervening and to carefully approach them surgically in order to avoid complications.

REFERENCES:

1. Parks MM, Johnson DA, Reed GW. Long-term visual results and complications in children with aphakia. A function of cataract type. *Ophthalmology* 1993;100:826-840.
2. Hosal BM, Biglan AW, Elhan AH. High levels of binocular function are achievable after removal of monocular cataracts in children before 8 years of age. *Ophthalmology* 2000;107:1647-1655.
3. Crouch ER Jr, Parks MM. Management of posterior lenticonus complicated by unilateral cataract. *Am J Ophthalmol* 1978;85:503-508.
4. Cheng KP, Hiles DA, Biglan AW, Pettapiece MC. Management of posterior lenticonus. *J Pediatr Ophthalmol Strabismus* 1991;28:143-149.

DR MALCOLM R. ING: I have talked to a number of pediatric cataract surgeons and it seems that whenever there is a violation of the capsule, they are selecting three-piece lenses versus the one-piece lens, which is becoming very popular in adult surgery. Their rationale was that the three-piece lens has fewer tendencies for dislocation and decentration, and may be a more stable lens. What is your impression?

DR ALBERT W. BIGLAN: In a report in 1991 we looked at 40 eyes, with about 50 percent having achieved 20/40 or better visual acuity. However, we had eyes with in the bag implantation, some with iris fixated lenses, and others with anterior chamber lenses and other eyes that were rehabilitated with contact lens. There was even an eye rehabilitated with an epikeratophakia graft. Your results update us with the results that should be expected with contemporary treatment. It is important to emphasize not to do hydrodissection

during surgery. This can result in an rupture of the posterior lens capsule and produce an admixture of the lens contents with vitreous. This can cause devastating problems.

DR ALLAN J. FLACH: What were the off-label things that you did as were listed in your presentation slide?

DR KENNETH W. WRIGHT: Are you concerned about doing an elective posterior capsulectomy and partial anterior vitrectomy considering the increased risk of late retinal detachment rather than electing a YAG procedure? I want to emphasize the importance of waiting in some of these cases because they are slowly progressive, you can often refract them and wait until they are older. My preference would be to put the intraocular lens in and do a YAG later when they are a little older.

DR J.BRONWYN BATEMAN: I am surprised that only 14 percent of the cases are unilateral. Were some of the other cases either an early form of this disease or perhaps not being picked up at the time of surgery? In the US it just seems like that should be a higher percentage since we've controlled the intrauterine infections so well.

DR EDWIN. M. STONE: Could you comment on bilateral cases, specifically if there are known familial cases and whether there is a different incidence of this condition in different ethnic groups?

DR EDWARD L. RAAB: You mentioned the occasional necessity of operating on these children because of a refractive aberration, even when the lens opacity is not yet well developed. Would you agree that it is probably subject to the same judgmental determination as we use for a PSC, namely using about a 3 mm size as a threshold? Also, are all of these, in your experience, central in the posterior capsule? I have had one case that was markedly off to the side, and the opacity never involved the center of the lens until much later, and I was able to defer surgery for a long time.

DR THOMAS D. FRANCE: When you identify a lentiglobus early you can watch it develop and have a good opportunity of obtaining good vision without deprivation amblyopia. When you see a 3-year-old that has a white cataract, and the parents indicate that it appears new, what ways do you have to determine that you are not dealing with a long-standing congenital cataract that is going to end up with deprivation amblyopia, as opposed to this lentiglobus, where you are going to get a better visual result?

DR M. EDWARD WILSON: I had noted in the literature, in the Dr. Parks series and others series, that the incidence was higher. If you look at the Parks series, there was no posterior polar category. It may just be that I have to see a definitive weak bulging capsule to call it posterior lentiglobus. If it's a posterior polar cataract with a moth-eaten posterior capsule I do not call that posterior lentiglobus. I have to see a definitive bulge before I put it in this category. Some of those other series' did not even have any other categories, other than PSC so maybe it is just lumping or splitting. So, I am not sure whether it's decreasing or not. Also my series is just those that had surgery and, as has been mentioned, they can be slowly progressive and be eccentric. I did not include all of those that I diagnosed with posterior lentiglobus, but rather just those that went to surgery.

I use a three-piece AcrySof lens if the lens is going in the sulcus. If I've made a determination that I have enough capsule to place the lens securely in the capsular bag, then I use the one-piece lens. So, it's a judgment based on capsule stability. I do not put the single-piece lens in the sulcus but I do choose it when I put the lens in the capsular bag. If you put a three-piece lens in a bag that already has a rupture, the IOL unfolding might further split the posterior capsule. If I have an open posterior capsule a single-piece lens that can be delivered without any additional trauma to the capsule is an advantage.

Even though it is the standard of care, intraocular lenses in children are off label. We would like the FDA to change the labeling, just based on the literature that is already available, at least beyond infancy.

Dr. Wright asked about the elective vitrectomy versus the YAG. If the child's old enough, and if the capsule is stable and has a chance of staying clear, I would do a YAG laser. Long follow-up has shown that vitrectomy and posterior capsulectomy, when it is indicated in a child, has a lot less risk in a formed vitreous than in the adult vitreous. I agree that it should not be done if it is not necessary. In young children that will not sit for YAG, or when the posterior capsules are already abnormal, or there are these fluffy white dots in the vitreous that need to be cleaned up anyway, then I perform the posterior capsulectomy and partial anterior vitrectomy.

Dr. Bateman asked about the incidence of unilateral cases and yes, there were perhaps other cases in my series. Dr. Stone asked about bilateral cases. I did not have any bilateral cases. The ones that were reported in the literature were familial. That seems to be a disorder that looks the same as the unilateral variety, but it is a genetic condition and rare. Concerning ethnic group differences; we found that our ethnic composition on these patients was basically the same as the ethnic configuration of our clinic in general, so I did not see a predilection. We had more boys than girls whereas in the past literature, there were more girls than boys.

Dr. Raab asked about the judgment of when to remove one of these when it is really just a refractive problem. Early diagnosis, patching, and careful refractions are needed. The judgment about when to operate is very difficult, but unless you pick them up early, they will get refractive amblyopia before they get a cataract. I agree that 3 mm or more in the center is a good indication of when a cataract is visually significant, but I base the decision for surgery on best corrective visual acuity and a trial of patching. Those that are very eccentric may not need surgery.

Dr. France asked how I determine whether a white cataract in a 3 year-old child is a neglected congenital cataract or a ruptured posterior lentiglobus. I am not completely certain which it is until surgery but there are many clues that are reasonably reliable. A neglected congenital cataract will be more likely to have strabismus, microcornea, a well defined posterior capsule on A-scan ultrasound and a relatively normal lens thickness – also measured with ultrasound. The posterior lentiglobus that progresses slowly and then acutely ruptures, hydrates, and turns white will usually have a normal corneal diameter, straight eyes, and a thickened lens with a poorly defined posterior capsule on A-scan ultrasound.