Long-term visual results after pars plicata lensectomy-vitrectomy for congenital cataracts

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SUMMARY We performed a pars plicata lensectomy-vitrectomy on 32 patients (47 eyes) with congenital cataracts. Ocular abnormalities, mainly nystagmus, strabismus, and microphthalmia, were present in 29 patients. No complications occurred intraoperatively or postoperatively in 39 eyes with up to 8½ years' follow-up (average 2.2 years). The pars plicata approach is a good surgical technique for the management of congenital cataracts.

Congenital and early developmental cataracts have resulted in a large percentage of cases of childhood blindness.^{1,2} The surgical management of such eyes has been debated in the literature for over 50 years. Early techniques such as discission, linear extraction, and intracapsular extraction were plagued by a persistently high rate of surgical complications.³⁻⁷ More recently the discission and aspiration technique for congenital cataracts has been advocated.⁸⁻¹¹ However, the incidence of complications with this technique, including vitreous loss, secondary glaucoma, retinal detachment, and, in particular, secondary membrane formation, remains quite high. Despite the use of phacoemulsification instruments, postoperative complications often necessitated additional procedures during the critical period of visual development.¹²⁻¹⁴

A major innovation in the management of congenital cataracts has been the application of automated vitrectomy instruments.^{15,16} Both translimbal and transciliary body approaches are in use.¹⁷⁻¹⁹ While the anterior segment approach through the limbus may effectively produce an optically clear visual axis, the procedure is complicated by a high incidence of cystoid macular oedema and by vitreous incarceration in the limbal incision.^{20,21} We have performed pars plicata lensectomy-vitrectomy in congenital cataracts for over 12 years with good surgical results and a low incidence of postoperative cystoid macular oedema.²²⁻²⁶ This report describes the long-term visual outcome of infants and juveniles

who have had congenital and developmental cataracts managed with this approach.

Material and methods

Thirty-two patients (47 eyes) were referred to the vitreous service of the University of Illinois Eye and Ear Infirmary between 1974 and 1986. Six patients between the ages of 5 weeks and 18 years had a unilateral cataract. In only one was an autosomal dominant mode of inheritance suggested by family history; no aetiology was identified for the remaining five patients.

Twenty-seven patients between the ages of 3 weeks and 10 years had bilateral cataracts. Bilateral lensectomy-vitrectomy was performed in 15, while 10 of the remaining 11 patients underwent another procedure on the contralateral eye. In five patients the congenital cataracts were attributed to maternal infection with rubella. All five also had microphthalmia. Three of these patients had valvular heart defects, deafness, and mental retardation as well. Three patients had dominantly inherited congenital cataracts. In addition one patient had Down's syndrome, another had mental retardation and a seizure disorder, and a third was diagnosed with Peters' anomaly. Among the other patients one was born with raised titres to cytomegalovirus; one had a congenital herpesvirus infection resulting in hydrocephalus, severe mental retardation, and cataracts; and one had microphthalmia with cataracts and multiple craniofacial anomalies.

Before surgery, whenever possible, all eyes were

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examined for corneal size, intraocular pressure, clarity of the media, and visual acuity. Muscle balance and the presence of nystagmus were noted. None had undergone previous surgery. All procedures were performed with the patients under general anaesthesia.

SURGICAL PROCEDURE

The eye was maximally dilated preoperatively. After surgical preparation and sterile draping 4–0 black silk sutures were used to retract the eyelids. The conjunctiva was opened through a limbal peritomy, and bridle sutures were placed beneath the rectus muscles. In infants a 3.5 mm long sclerotomy was made 2.0 to 2.5 mm behind and parallel to the limbus with a no. 67 Bard-Parker blade. In older children the sclerotomy was made 3 mm from the limbus.

The ciliary body was exposed at the sclerotomy site and gently diathermised. A 5–0 polyglactin 910 (Vicryl) or 7–0 polyfilament (Supramid) mattress suture was passed through the sclerotomy and tied with a loose double-throw knot. A 52-s Beaver blade was inserted into the lens with a slicing motion and removed for insertion of a wide-angle Peyman vitrophage.²⁷ If the blade did not easily penetrate the centre of the lens (rare in children), a Peyman lens fragmentor was used to pulverise the lens nucleus for about 60 seconds.²⁸ The wide-angle cutter was placed in the eye, the knot was tightened, and the lens material was cut and aspirated. The lens capsule was not removed until the entire nucleus and cortex were aspirated. The lens material behind the iris was sucked into the vitrophage and brought into the pupillary space to be cut and removed, followed by the anterior and posterior capsule.

If the pupil did not dilate, a sector iridectomy was performed with the vitrophage. An anterior vitrectomy was preceded by a deep vitrectomy confined to the central core down to the optic disc. A modified Goldmann posterior pole lens and an operating microscope were used for direct visualisation. The vitrophage was then removed, and the scleral mattress suture was tightened until the sclerotomy was watertight. If the eye was soft, intraocular pressure was re-established with an injection of normal saline solution through the sclerotomy site into the vitreous chamber with a 30-gauge needle. The conjunctiva was closed with 6–0 plain catgut, and atropine and Maxitrol (neomycin, polymixin B, and dexamethasone) drops were instilled, and patch and shield were placed.

Results

Our follow-up has ranged from one week after discharge to 11 years (average 2.2 years). Nineteen of

our patients were seen for follow-up at our institution for less than two years. However, the median visual acuity of those receiving visual rehabilitation for at least two years was 20/100. The patients' best corrected visual acuities were recorded preoperatively and at the time of the last visit.

No surgical or postoperative complications occurred in 39 eyes. Two patients had transient corneal oedema postoperatively that resolved spontaneously over several days. One patient had transient corneal oedema and intravitreal bleeding from the iris. The child with Down's syndrome had bilateral cystoid macular oedema noted by fluorescein angiography four months postoperatively, which cleared by 32 months. One case of increased intraocular pressure and iritis in the immediate postoperative period responded to acetazolamide and steroid drops. Three months postoperatively this patient suffered a retinal detachment that was treated successfully with a scleral buckle and cryotherapy. One eye had a retinal dialysis intraoperatively. Another patient had a retinal detachment two weeks postoperatively; both conditions were treated successfully. No other retinal detachments were observed up to 8½ years after pars plicata lensectomy-vitrectomy (Table 1).

Of the 10 patients whose contralateral eye had a procedure other than the pars plicata lensectomy-vitrectomy, five had secondary membrane formation. Discission and aspiration in one such case resulted in increased intraocular pressure, choroidal atrophy, and fibrous ingrowth of the cornea; postoperative visual acuity was light perception. The right eye, which underwent a pars plicata lensectomy-vitrectomy, suffered no complications; postoperative visual acuity was 20/60.

None of the patients whose contralateral eye had an alternative procedure had a better final visual acuity in that eye than in the eye that had pars plicata lensectomy-vitrectomy (Table 2).

Results of the long-term outcome in our monocular cases are hindered by a lack of follow-up information. However, the children with acquired monocular cataracts who were operated on at ages 4, 18, and 10 years, respectively, showed an improvement of 5 and 6 lines on the Snellen chart in two cases and from counting fingers to 20/50 in the third. There was one case of a choroidal detachment postoperatively that resolved after six weeks (Table 3).

All but four patients with bilateral congenital cataracts had nystagmus, strabismus, or both. Two (patients 14 and 23) had a visual acuity of 20/200 preoperatively. One (patient 13) with central, steady, and maintained vision preoperatively was operated on after 3 years of age. Severe mental retardation in the fourth patient (patient 8) made accurate visual assessment difficult.

Table 1 Bilateral congenital cataracts treated with pars plicata lensectomy-vitreotomy in both eyes

Patient no	Eye	Procedure*	Age at surgery	Aetiology	Follow-up	Visual acuity†			Comments
						Preop.	Follow-up	Complications	
1	OD	PPLV	44mo	Rubella	18mo	No view	CNSNM	None	Pendular nystagmus
	OS	PPLV	44mo			No view	CNSNM	None	
2	OD	PPLV	7wk	Rubella	12mo	No view	CNSM	None	Fine pendular nystagmus
	OS	PPLV	10wk			No view	CNSM	Transient corneal oedema	
3	OD	PPLV	4.5yr	Rubella	1wk	No view	LP	None	Pendular nystagmus
	OS	PPLV	4.5yr			No view	LP	None	
4	OD	PPLV	6mo	Autosomal dominant	6yr	No view	20/60	Transient corneal oedema	Occlusion nystagmus
	OS	PPLV	8mo			No view	20/60	None	
5	OD	PPLV	13wk	Idiopathic	4mo	No view	CNSM	None	Left estropia, high myopia
	OS	PPLV	16wk			No view	CNSM	None	
6	OD	PPLV	13wk	Idiopathic	4mo	No view	CNSM	Retinal dialysis	Pendular nystagmus
	OS	PPLV	16wk			No view	CNSM	None	
7	OD	PPLV	11mo	Idiopathic	5mo	No view	CNSM	None	Fine pendular nystagmus
	OS	PPLV	9mo			No view	CNSM	None	
8	OD	PPLV	12wk	Herpesvirus, retardation	2yr	No view	CSM	None	Straight without nystagmus
	OS	PPLV	9wk			No view	CSM	None	
9	OD	PPLV	10mo	Autosomal dominant	1yr	No view	CSM	None	Left esotropia
	OS	PPLV	8mo			No view	CSM	Retinal detachment	
10	OD	PPLV	14wk	Idiopathic	6mo	No view	CNSM	None	Pendular nystagmus
	OS	PPLV	16wk			No view	CNSM	None	
11	OD	PPLV	9wk	Cytomegalovirus	4.5yr	No view	20/60	None	Fine pendular nystagmus
	OS	PPLV	10wk			No view	20/60	None	
12	OD	PPLV	8yr	Retardation, seizures	3yr	20/400	20/100	None	Right esotropia
	OS	PPLV	10yr			20/400	20/100	None	
13	OD	PPLV	37mo	Idiopathic	2.5yr	CSM	20/100	None	Alternating exotropia
	OS	PPLV	40mo			CSM	20/100	None	
14	OD	PPLV	5yr	Idiopathic	1yr	20/100	20/30	None	Straight without nystagmus
	OS	PPLV	4.5yr			20/200	20/40	None	
15	OD	PPLV	8yr	Idiopathic	3yr	HM	20/200	None	Left exotropia
	OS	PPLV	9yr			HM	HM	Retinal detachment, iritis	

*PPLV indicates pars plicata lensectomy-vitreotomy.

†Preop. indicates preoperative; CNSMM, central, but neither steady nor maintained fixation; CNSM, central, not steady, but maintained fixation; LP, light perception; CSM, central, steady, maintained fixation; HM, hand motion.

Discussion

The pars plicata approach is an advantage to the pars plana procedure in children.²⁸ An incision made in the pars plana of young children may increase the risk of retinal detachment. Because the pars plicata is further anterior from the vitreous base and ora serrata, the risk of retinal detachment is lessened. The pars plicata approach also has many advantages over the translimbal approach. The pars plicata is in the direct axis of the lens, allowing for more complete removal of lenticular material. In contrast to the limbal approach, the cornea is not manipulated during surgery. Deep visualisation of the fundus is possible with a modified Goldmann lens, and lens fragments in the vitreous can be removed, thus preventing persistent inflammation postoperatively. An extensive vitrectomy also prevents pupillary block glaucoma resulting from a vitreous prolapse into the anterior chamber. Lastly, the vitreous is removed without trauma to the iris, which has been a suspected cause of cystoid macula oedema.²⁹

The visual outcome after surgery for congenital cataracts also depends on the age of the patient at surgery, the density of the lens opacity, the period over which the cataracts developed, the presence of nystagmus or other ocular abnormalities, and the postoperative management. In our population a large percentage of cataracts in infants and young children was not discovered until many months after birth. Only six (19%) of our patients had cataracts identified and operated on before 3 months of age. Recent studies by Gelbart and associates³⁰ and by Rogers and coworkers³¹ emphasise the need for surgery to be performed in the first eight weeks of life to achieve optimal visual rehabilitation of infants with dense bilateral cataracts.

Although we could not determine the time over which the lens completely opacified, 12 eyes (67%) with no view of the fundus preoperatively had a visual acuity no better than central fixation by age 2½ years (Table 4). Conversely, eight eyes (53%) with incomplete cataracts and at least central fixation preoperatively had a preoperative visual acuity of 20/60

Table 2 Bilateral congenital cataracts in which one eye had pars plicata lensectomy-vitrectomy and the contralateral eye another procedure

Patient no	Eye	Procedure*	Age at surgery	Aetiology	Follow-up	Visual acuity†			Comments
						Preop.	Follow-up	Complications	
16	OD	PPLV	6 mo	Idiopathic	2 yr	No view	CNSM	None	Fine pendular nystagmus
	OS	D & A	5 mo			No view	NCNSNM	Posterior capsular plaque	Intermittent left exotropia
17	OD	PPLV	31 wk	Down's syndrome	6 yr	No view	CNSNM	Cystoid macular oedema	Nystagmus
	OS	D & A	32 wk			No view	CNSNM	Cystoid macular oedema	Right esotropia
18	OS	PPLV	3 wk	Rubella	3 yr	No view	20/100	None	Fine pendular nystagmus
	OD	I & A	4 wk			No view	20/100	None	
19	OS	PPLV	1 yr	Multiple anomalies	1 wk	No view	Clear view	None	Pendular nystagmus
	OD	I & A	9 mo			No view	Clear view	Secondary membrane	
20	OD	PPLV	12 mo	Peters' anomaly	16 mo	No view	CNSNM	None	Coarse pendular nystagmus
	OS	Sector	15 mo			No view	CNSNM	None	
21	OD	PPLV	4.5 yr	Rubella	1 wk	LP	LP	Transient corneal oedema	Pendular nystagmus
	OS	None				LP	LP	Bleeding iris	Right esotropia
22	OD	PPLV	4 yr	Autosomal dominant	11 yr	No view	20/60	None	Left esotropia
	OS	D & A	8 mo			No view	LP	Glaucoma, fibrous ingrowth	
23	OS	PPLV	9 yr	Idiopathic	7 yr	20/200	20/40	None	High myopia, straight
	OD	D & A	7 yr			20/200	20/60	Secondary membrane	without nystagmus
24	OD	PPLV	5 yr	Idiopathic	None	20/80	20/40	None	Left exotropia
	OS	Phaco	5 yr			20/400	20/200	None	
25	OD	PPLV	1 yr	Idiopathic	8-5 yr	CSM	20/100	None	Pendular nystagmus
	OS	Phaco	1 yr			CSM	20/400	Secondary membrane	Left exotropia
26	OS	PPLV	22 mo	Idiopathic	5 yr	CSM	20/30	None	
	OD	D & A	16 mo			CSM	20/40	Secondary membrane	Right exotropia

*PPLV indicates pars plicata lensectomy-vitrectomy; D & A, discission and aspiration; I & A, irrigation and aspiration; Phaco, phacoemulsification.

†Preop. indicates preoperative; CNSM, central, not steady, but maintained fixation; NCNSNM, neither central, steady, nor maintained fixation; CNSNM, central but neither steady nor maintained fixation; LP, light perception; CSM, central, steady, maintained fixation; HM, hand motion.

Table 3 Monocular congenital cataracts removed by pars plicata lensectomy-vitrectomy

Case no.	Age at surgery	Aetiology	Follow-up, mo	Visual acuity*			Comments
				Preop.	Follow-up	Complications	
1	4 yr	Idiopathic	14	20/400	20/60	None	Variable esotropia Left esotropia
2	5 wk	Idiopathic	4	No view	CNSNM	None	
3	6 mo	Idiopathic	8	No view	CSM	None	
4	10 yr	Idiopathic	1	Count fingers	20/50	None	
5	18 yr	Idiopathic	1	20/200	20/40	None	
6	5 wk	Autosomal dominant	2	No view	Clear view	Choroidal detachment	

*Preop. indicates preoperative; CNSNM, central but neither steady nor maintained fixation; CSM, central, steady, and maintained fixation.

Table 4 Age at surgery and visual results after pars plicata lensectomy-vitrectomy for total cataract (n=18)*

Final visual acuity	Age				%
	0-3 Mo	3-12 Mo	1-2 Yr	≥2 Yr	
Light perception	-	-	-	3	17
CSM fixation to 20/100	2	4	1	2	50
20/100-20/60	1	-	-	-	5
20/60	2	2	-	1	28

*Includes those patients with 'no view' preoperatively and follow-up past age 2½ years.

Table 5 Age at surgery and visual results after pars plicata lensectomy-vitrectomy for incomplete cataract (n=15)*

Final visual acuity	Age, yr				%
	0-1	1-2	2-4	>4	
Hand motion to 20/200				1	7
20/200-20/100				1	7
20/100-20/60		1	2	2	33
20/60-20/30		1		7	53

*Includes those patients with at least fixation capability preoperatively.

or better (Table 5). These results agree with the findings of François³² that 50% to 70% of cases with complete congenital cataracts had a postoperative visual acuity of less than 20/200, while 35% to 87% of cases with incomplete congenital cataracts had a final visual acuity of greater than 20/60.

Nystagmus, the most common ocular abnormality associated with bilateral visual deprivation amblyopia, occurred in 17 (65%) of our patients.^{4 5 30 32} None achieved a visual acuity of better than 20/60. Although the amplitude of the nystagmus decreased during follow-up in some cases, it never resolved completely. Gelbart and associates,³⁰ Rogers and associates,³¹ and Parks³³ noted the development of pendular nystagmus in infants with bilateral congenital cataracts by 12 to 13 weeks of age. This may represent the critical period at which irreversible amblyopia begins.^{34 35}

Strabismus occurred with bilateral congenital cataracts in 14 (54%) of our patients. Gelbart and colleagues³⁰ reported esotropia in 21 of 24 patients with congenital cataracts despite early surgery and a short interval between operations for each eye. A recent Russian study of children with bilateral aphakia demonstrated strabismus in 50 of 52 patients.³⁶ France and Frank³⁷ described a 58% incidence of esotropia preoperatively; an additional 21% were identified after lens aspiration, refractive correction, and patching therapy.

Our experience with monocular congenital cataracts removed by pars plicata lensectomy-vitreotomy has been limited. However, our three children with incomplete cataracts and no strabismus at the time of surgery regained useful vision, a finding consistent with that in the literature.³⁸ Recently, Beller and Hoyt have reported good visual results in eight of 11 infants who were operated on within the first six weeks of life, corrected frequently with extended-wear contact lenses, and treated with careful patching monitored by visual evoked potentials.³⁹

Despite advances in our understanding of amblyopia in animal models and in humans, the management of both monocular and binocular congenital cataracts remains a tremendous clinical challenge. We have performed pars plicata lensectomy-vitreotomy for congenital cataracts in 32 patients. All retained a large clear pupillary space for the follow-up period. As in other studies, ocular abnormalities, including nystagmus, strabismus, and microphthalmia, have contributed adversely to the visual outcome.^{1 4 5 17 30-32 37} Our long-term results are comparable to those found in the literature.^{1 9 10 13 30 32} Despite the risk of retinal detachment, the pars plicata approach is a good surgical technique for the management of congenital cataracts.

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