OUTCOME OF TREATMENT FOR BILATERAL CONGENITAL CATARACTS*

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INTRODUCTION

THE VISUAL RESULTS OF CATARACT SURGERY IN CHILDREN HAVE GENERally been poorer than in adults.[1-3] The difference is due in part to the various types of amblyopia that develop in children with cataracts, the association of nystagmus with cataracts of early onset, and the presence of other ocular abnormalities that adversely affect vision in eyes with developmental lens opacities. Following reintroduction of the aspiration technique for cataract removal by Scheie in 1960,[4] the surgical procedures for removal of the lens in childhood have been improved[5,6] and earlier surgery for congenital cataracts has been encouraged.[7-9] A current assessment of the outcome of surgery for congenital cataracts therefore seemed to be in order, and this paper addresses our experience with a group of children with bilateral congenital cataracts of diverse etiology.

PATIENTS AND METHODS

Fifty-one patients with bilateral congenital cataracts who had surgery by the authors at Boston Children's Hospital between 1971 and 1990 and whose postoperative visual acuity could be determined by recognition acuity tests were studied retrospectively. The cataracts were considered congenital if they were identified within the first 6 months (24 patients), were dominantly inherited in families with a history of congenital cataracts (19 patients); or were lamellar in configuration, suggesting an early developmental origin (19 patients). These categories were not mutually exclusive. At the time of cataract surgery, patient age ranged from 1 month to 22 years, depending on the time of presentation, the size and density of the cataracts, and the visual function of the patient as determined by visual behavior or measurement of visual acuity. Removal of

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lens material was accomplished by aspiration after incision of the anterior lens capsule or by a combination of aspiration and suction-cutting. In the earlier cases of the series, the posterior capsule was left intact at the time of aspiration and was later incised with a discission knife if it opacified. In more recent cases the posterior capsule was often removed with the suction-cutting instrument at the time of initial surgery. In a few patients an opacified posterior capsule was opened with a neodymium-yttrium aluminum garnet (Nd:YAG) laser. In most eyes, either a peripheral or a full iridectomy was performed as part of the initial surgery, the choice depending on how well the pupil dilated preoperatively. Aphakic correction was accomplished with spectacles or contact lenses. The patients were followed until 3 to 30 years of age (mean, 10.3 years; median, 8.5 years) for measurement of final visual acuity. Records were reviewed to identify patients with nystagmus, strabismus, amblyopia, and glaucoma. Nystagmus was recognized by clinical inspection, and strabismus was defined as overt ocular misalignment on cover test. Strabismic or deprivational amblyopia was considered to be present when that diagnosis was made by the examining physician. Since patching of the dominant eye was frequently carried out, the final visual acuities do not necessarily reflect the presence of amblyopia at some earlier point in time. Glaucoma was considered to be present when the ocular pressure was over 22 mm mercury on repeated measurements. The condition of the anterior-chamber angle was assessed by Koeppe gonioscopy.

RESULTS

Patients with congenital lamellar opacities had the best visual outcome from cataract surgery, and surgery improved their vision even if it was performed as late as the second decade (Table I). The average postoperative visual acuity in the better eye of 19 patients with lamellar cataracts was 20/38, and it improved to 20/30 if three patients who had nystagmus were excluded. Surgery for lamellar cataracts was performed from 7 months of age to 20 years, the average age at operation being 9.7 years. Deprivational amblyopia did not appear to occur, even in the older patients with bilateral lamellar or other partial cataracts unless the lens opacities were asymmetrical in the two eyes. The visual acuity in the better eye of all patients whose surgery was performed after 3 years of age improved to 20/40 or better, except in the previously mentioned patients with nystagmus and high myopia (Fig 1).

The visual outcome was less good in patients with more extensive cataracts (Table I), whose surgery was usually performed in the first year

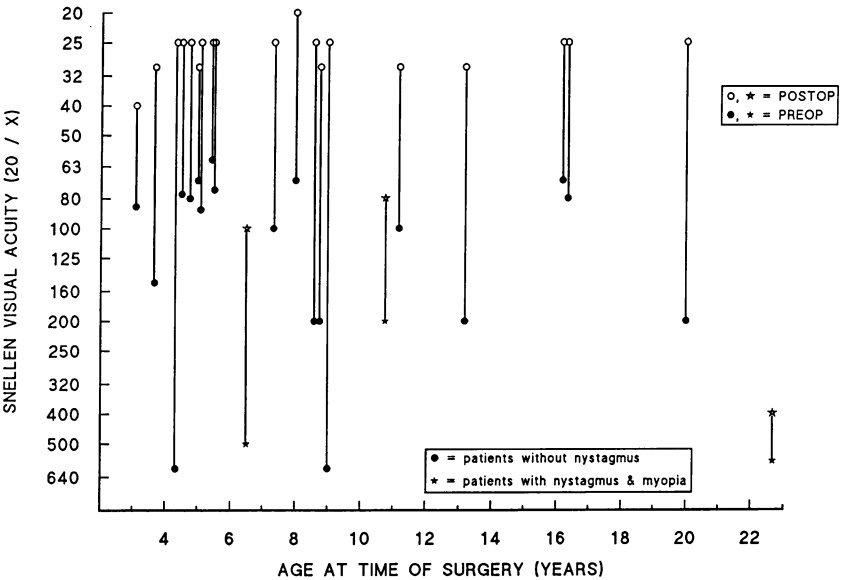

FIGURE 1
Preoperative and postoperative visual acuity in better eye of 22 patients plotted against age at time of surgery in that eye.

of life. Nystagmus was associated with the reduced visual acuity in many of these patients. Unilateral reduction of vision was often due to strabismic or deprivational amblyopia. It was, in fact, difficult to say whether the strabismus or the lens opacity had been the primary cause of the amblyopia. The visual acuity in the better eye of 29 patients whose surgery was performed within the first 15 months of life is presented in Fig 2. The average postoperative visual acuity of those without nystagmus (n = 13) was 20/45 and of those with nystagmus (n = 16) was 20/80. Within the latter group earlier surgery did not seem to prevent the development of nystagmus. The onset of the nystagmus was difficult to establish exactly by chart review, but it was often first noted in the records between 2 and 4 months of age. Patients with nystagmus more often had corneas with horizontal diameters less than 9.5 mm, poorly dilating pupils, and no preoperative view of the fundus by indirect ophthalmoscopy than did those without nystagmus (Table II). Within individual families with dominantly inherited cataracts, affected family members tended to be uniform with respect to presence or absence of nystagmus (Table III).

Strabismus was recognized in 13 of the 51 patients preoperatively, but it was present in 35 patients postoperatively. Ocular misalignment was

TABLE I: CATARACT TYPE, SURGICAL PROCEDURES, VISUAL ACUITY, AND

PATIENT NO.*	CATARACT TYPE	SURGICAL PROCEDURES†		LAST POSTOPERATIVE VISUAL ACUITY‡			
		OD	OS	OD	OS	AGE (YR)	TEST
1	Dom inher complete	AC(0-1)	AC (0-2)	20/100	20/300	3	Allen
2	Dom inher complete	AC(0-1), D(0-4) D(0-5), D(1-3)	A(0-2), D(0-4) D(0-5)	20/400	20/100	8	Snellen
3	Idiopathic crystalline	A(0-2), C(1-0)	AC(0-1), C(3-8)	20/100	20/400	6	Snellen
4	Dom inher complete	AC(0-2)	AC(0-2)	20/85	20/85	7	E's
5	Idiopathic nuclear	A(0-2)	A(0-3), D(1-0) C(8-2)	20/30	20/80	9	Snellen
6	Dom inher complete	AC(0-5)	AC(0-2)	20/85	20/125	8	E's
7	Dom inher complete	AC(0-2), L(4-2)	AC(0-3)	20/85	20/65	4	Allen
8	Idiopathic complete	AC(0-3)	AC(0-3)	20/70	20/50	8	Snellen
9	Dom inher complete	AC(0-3)	AC(0-4), C(0-5)	20/85	20/125	7	E's
10	Congenital rubella	A(0-5), D(1-6) D(2-11), D(0-4)	A(0-3), D(0-7) D(2-11)	20/200	20/200	11	Snellen
11	Idiopathic complete	A(0-3), D(0-4)	A(0-6)	NLP	20/70	14	Snellen
12	Dom inher crystalline	AD(0-5)	A(0-3), D(0-5)	20/65	20/65	6	E's
13	Idiopathic lamellar	A(11-2)	A(0-3), D(1-3)	20/300	20/1000	11	Snellen
14	Idiopathic complete	AC(0-4)	AC(0-4)	20/40	20/40	6	HOTV
15	Dom inher crystalline	AD(0-5)	AD(0-4)	20/65	20/85	5	E's
16	Dom inher complete	A(0-5), D(2-1)	A(0-4), D(0-5)	20/100	20/80	12	Snellen
17	Dom inher nuclear	A(0-5), D(0-6) L(6-0), C(6-0)	AD(0-6)	20/100	20/40	6	HOTV
18	Congenital rubella	A(0-6), D(1-0) D(2-3)	A(0-5), D(1-0)	20/60	20/60	15	Allen
19	Dom inher crystalline	A(0-7), D(2-1)	A(0-5), D(0-10)	20/70	20/40	12	Snellen
20	Idiopathic complete	A(0-6)	A(0-5)	20/65	20/80	5	HOTV
21	Idiopathic lamellar	A(0-6), D(0-7)	AD(0-7)	20/40	20/30	7	E's
22	Idiopathic complete	A(0-6), D(0-7)	AD(0-7)	20/85	20/60	5	Allen
23	Dom inher crystalline	A(0-9), D(0-11)	A(0-8), D(0-9)	20/30	20/30	5	E's
24	Idiopathic lamellar	A(1-0)	A(0-8)	20/30	20/200	6	Snellen
25	Idiopathic lamellar	A(0-10)	A(0-9)	20/70	20/50	14	Snellen
26	Dom inher punctate	AD(0-10)	AD(1-0)	20/85	20/60	8	E's
27	Idiopathic lamellar	A(1-0), D(1-1)	A(1-1), D(1-3)	20/50	20/40	11	Snellen
28	Idiopathic lamellar	AC(1-6)	AC(1-2)	20/40	20/40	6	Snellen

OTHER FEATURES OF 51 PATIENTS WITH BILATERAL CONGENITAL CATARACTS

NYSTAGMUS	STRABISMUS PREOP	STRABISMUS POSTOP	AMBLYOPIA	OPEN-ANGLE GLAUCOMA	COMMENTS
+	–	ET	+	+OS	
+	–	ET	+	–	
+	–	ET	+	–	Angle-closure glaucoma OS
+	–	XT	–	0	
Occl	–	XT	+	+OU	
+	–	ET	+	+OU	
+	–	ET	+	0	
+	–	ET	–	–	
+	–	–	–	+OU	
+	ET	ET	–	+OS	
+	–	XT	NA	–	Angle-closure glaucoma OD
–	–	XT	–	0	
–	ET	ET	+	–	Asymmetrical cataracts
+	–	ET	–	0	
Occl	–	ET	+	–	
+	–	ET	+	–	
–	–	ET	+	0	
+	–	XT	–	+OD	
–	–	XT	+	–	
Occl	–	ET	–	0	
–	–	–	–	–	
+	E(T)	ET	+	–	
–	–	–	–	0	
–	–	XT	+	+OS	Asymmetrical cataracts
Trans	–	ET	+	–	
–	E(T)	E(T)	–	0	
–	ET	ET	–	0	
–	–	ET	–	0	

TABLE I: CATARACT TYPE, SURGICAL PROCEDURES, VISUAL ACUITY, AND

PATIENT NO.*	CATARACT TYPE	SURGICAL PROCEDURES†		LAST POSTOPERATIVE VISUAL ACUITY‡			
		OD	OS	OD	OS	AGE (YR)	TEST
29	Idiopathic lamellar	A(1-10)	A(1-3), D(7-5)	20/50	20/70	11	Snellen
30	Dom inher complete	AC(1-3)	AC(1-5)	20/60	20/85	9	E's
31	Idiopathic lamellar	A(3-3), D(4-5)	A(3-1)	20/70	20/40	5	E's
32	Dom inher crystalline	A(3-8), D(4-6) D(8-10)	A(3-7), D(4-6)	20/30	20/30	16	Snellen
33	Idiopathic lamellar	AC(4-3)	AC(4-4)	20/25	20/25	7	Snellen
34	Idiopathic lamellar	AC(4-6)	AC(4-4)	20/25	20/60	6	HOTV
35	Idiopathic lamellar	A(4-9), D(8-11)	A(4-11), L(9-3)	20/25	20/25	11	Snellen
36	Dom inher nuclear	AC(5-1)	AC(5-0)	20/25	20/25	7	HOTV
37	Idiopathic axial	AD(5-0)	AC(5-3)	20/30	20/30	11	Snellen
38	Hypocalcemic lamellar	A(9-0), L(12-0)	A(5-2), D(7-0)	20/25	20/200	9	Snellen
39	Dom inher lamellar	AD(5-3)	AD(5-5)	20/200	20/25	12	Snellen
40	Idiopathic lamellar	A(5-6), D(5-8)	A(5-8), D(6-0)	20/25	20/25	7	Snellen
41	Hypoglycemic lamellar	A(6-6), D(7-5)	A(6-5), D(6-6)	20/100	20/400	19	Snellen
42	Dom inher cortical	AD(7-4)	AD(7-6)	20/25	20/25	8	Snellen
43	Dom inher crystalline	A(8-0), D(8-3)	A(7-9), D(8-0)	20/20	20/20	9	Snellen
44	Idiopathic crystalline	A(8-7), L(9-11)	A(8-9)	20/25	20/25	10	Snellen
45	Idiopathic lamellar	A(8-9), D(8-10)	A(9-6), D(11-6)	20/30	20/40	12	Snellen
46	Idiopathic lamellar	A(10-6)	AC(10-9)	20/100	20/80	14	Snellen
47	Idiopathic punctate	A(13-9), D(13-9) L(18-0)	A(13-5), L(16-3)	20/30	20/40	22	Snellen
48	Idiopathic lamellar	A(15-1), D(17-11)	A(16-4), D(17-11)	20/40	20/25	24	Snellen
49	Idiopathic axial	A(16-6)	A(16-2), L(17-0)	20/30	20/25	18	Snellen
50	Idiopathic lamellar	A(20-0)	A(20-0)	20/30	20/25	20	Snellen
51	Idiopathic punctate	A(22-8), D(28-9)	A(22-0), D(24-0)	20/400	20/500	30	E's

+, present; −, absent; 0, data not available; ET, esotropia; XT, exotropia; (T), intermittent tropia; Occl, occlusional nystagmus; Trans, transiently present; NA, not applicable; OD, right eye; OS, left eye; OU, both eyes.

*Patients have been numbered according to the timing of their cataract surgery, from earliest to latest.

OTHER FEATURES OF 51 PATIENTS WITH BILATERAL CONGENITAL CATARACTS CONT'D

NYSTAGMUS	STRABISMUS PREOP	STRABISMUS POSTOP	AMBLYOPIA	OPEN-ANGLE GLAUCOMA	COMMENTS
+	ET	ET	+	−	
+	−	ET	−	+OD	
−	ET	ET	+	0	
−	−	−	−	−	
−	−	−	−	−	
−	−	ET	+	−	Asymmetrical cataracts
−	−	−	−	−	
−	−	−	−	0	
−	−	−	−	−	
−	ET	ET	+	−	Asymmetrical cataracts
−	ET	ET	+	0	Asymmetrical cataracts
−	−	−	−	0	
+	ET	ET	+	0	Asymmetrical cataracts
−	−	−	−	0	
−	−	−	−	−	
−	−	−	−	−	
−	−	−	−	−	High myopia, retinal detachment OU
+	ET	ET	−	−	High myopia
−	−	−	−	−	
−	XT	XT	+	−	
−	−	−	−	0	
−	XT	XT	−	0	High myopia
+	−	−	−	0	High myopia

†Surgical procedures: A, aspiration of lens; D, discission of posterior capsule or pupillary membrane; C, cutting of posterior capsule or pupillary membrane with suction cutter; L, laser capsulotomy; (1-2), 1 yr 2 mo of age.

‡Visual acuities have been converted from their original format to Snellen notation at 20 ft. Visual acuity tests: Snellen, chart numbers or letters; E's, isolated E symbols; HOTV, these letters presented singly on hand-held cards; Allen, Allen card pictures presented singly on hand cards.

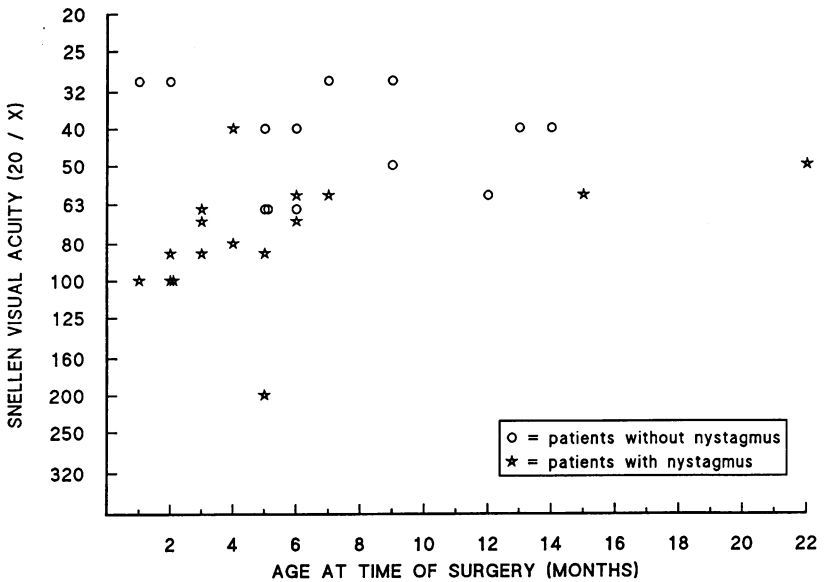

FIGURE 2
Postoperative visual acuity in better eye of 29 patients plotted against age at time of surgery in that eye.

TABLE II: OTHER OCULAR FINDINGS IN PATIENTS WITH COMPLETE CONGENITAL CATARACTS

	PATIENTS WITH NYSTAGMUS	PATIENTS WITHOUT NYSTAGMUS
Horizontal corneal diameter < 9.5 mm	8	1
Pupils dilating to 4 mm or less	12	1
No preoperative fundus view	15	3
Total no. of patients	16	13

TABLE III: DISTRIBUTION OF NYSTAGMUS AMONG AFFECTED MEMBERS OF FAMILIES WITH DOMINANTLY INHERITED CONGENITAL CATARACTS

	AFFECTED MEMBERS WITH NYSTAGMUS (PATIENT NO.)	AFFECTED MEMBERS WITHOUT NYSTAGMUS (PATIENT NO.)
Family 1	0	5 (12,15,19,23,32)
Family 2	1 (7 variable)	2 (17,36)
Family 3	2 (2,16)	0
Family 4	2 (6,9)	0

almost universal in patients whose surgery was performed in the first 2 years, occurring in 26 of 29 patients. It was also present in 8 of 22 patients whose surgery was performed after 2 years. Twenty-six patients had esotropia, nine were exotropic. More important from the visual standpoint was the presence of amblyopia in 21 of the 35 patients with strabismus. In some patients the amblyopia appeared to be primarily strabismic, but in six patients whose lens opacities were noted to be asymmetric preoperatively (patients 13, 24, 34, 38, 39, 41), deprivational amblyopia was probably the primary condition and the strabismus secondary.

Glaucoma developed in ten patients postoperatively. Two patients had angle-closure glaucoma in one eye, each with a small corneal diameter (7.5 and 8.0 mm), in spite of a large-sector iridectomy at the time of cataract extraction. One eye lost light perception, the other retains useful vision but has required trabeculectomy and several cyclocryotherapy treatments for pressure control. Open-angle glaucoma has developed in 11 eyes of eight patients, all of whom had surgery before 15 months of age. Seven of these eight patients are known to have had normal ocular pressures at the time of surgery. In no patient whose cataract surgery was performed after 3 years of age is glaucoma known to have developed. In the patients with open-angle glaucoma, the elevation of pressure was found at an average age of 8.4 years (range, 5 to 15 years). Gonioscopy has revealed open anterior chamber angles in all cases. Since 19 of the 51 patients have not yet had postoperative pressures recorded, the prevalence of glaucoma may be higher than our present estimate of 15% for the whole group and 27% for those operated before 15 months of age.

One patient in the present series has had bilateral retinal detachments. He had idiopathic lamellar cataracts and high myopia in both eyes. His cataracts were aspirated at 8 and 9 years of age, with subsequent discission of the posterior capsules. The retinal detachments occurred at 13 and 16 years of age, and his vision after vitrectomy and scleral buckle procedures has fallen to 20/400 in one eye and counting fingers in the other.

DISCUSSION

A retrospective study of the outcome of surgery for congenital cataracts has several limitations. Although we believe that all patients included in the study had congenital lens opacities, not all patients were seen and followed by us from the time of birth. In particular, some patients with lamellar cataracts were not seen by us until they were several years old. On the other hand, their lens opacities were characteristic of congenital lamellar cataracts, a type of cataract we have not found to be acquired postnatally in otherwise healthy children. The surgery performed in our

series of patients was not identical in all cases. For instance, the posterior capsule was handled differently at different times during the study period. Additionally, the timing of surgery was not dictated by an established protocol, but was determined by the age at the time of referral and by the visual status of individual patients. Finally, some observations that in retrospect would have been useful for analysis were missing from our records.

We feel, nevertheless, that some useful observations can be made on the basis of this review of patients. There seem to be two general categories of patients with congenital cataracts. One is characterized by extensive lens opacities and early, obvious reduction of vision. These patients, who come to cataract surgery in the first year of life, often have smaller-than-normal corneal diameters, poorly dilating pupils, and a vulnerability to delayed postoperative open-angle glaucoma. The other category includes patients with partial, often lamellar lens opacities, corneas of normal size, and a remarkably good visual prognosis. Slightly over half of the patients in the first category develop nystagmus at 2 to 4 months of age, and the nystagmus is accompanied by a reduction of visual acuity in spite of a good anatomic result from surgery. Early surgery, within the time frame indicated in Table I and Fig 2, did not appear to abort the development of nystagmus, although some investigators have suggested that this might be so based on their experience with small numbers of patients.[7,8] It is possible that even earlier surgery than we have done, undertaken in the first few weeks of life, would have a more favorable influence on the development of nystagmus. On the other hand, the nystagmus may be a manifestation of a more general ophthalmic disorder that would not be influenced by the timing of surgery. Further evaluation of this question will require a randomized controlled study, since patient selection could influence the prevalence of nystagmus in any small series of patients, especially if all patients with congenital cataracts, regardless of type, were subjected to early surgery. Some concern has been raised about the possibility of a higher complication rate following cataract surgery performed in the first 2 months of life, an important concern, but also one based on retrospective study of patients not randomized with regard to the timing of surgery.[10]

Whereas it is well known that deprivational amblyopia is difficult to surmount in patients with unilateral congenital cataracts,[11,12] it is not a major factor in patients with bilateral partial or lamellar opacities, as long as their lens opacities are symmetrical. The visual prognosis in this group of patients, whose surgery is usually performed after 3 years of age, at a time when increasing visual needs begin to exceed the limits imposed by

the lens opacities, is excellent. The only patients in this second general category who fell short of this high expectation were a few who also had the unfortunate combination of nystagmus and high myopia.

Treatment of strabismic amblyopia following bilateral congenital cataract surgery is useful, although the ocular misalignment is sometimes hard to identify, and the amblyopia may be profound by the time it is recognized. Deprivational amblyopia due to asymmetry of cataracts from the outset is very difficult to reverse, similar to the situation in patients with monocular congenital cataracts.[11,12] An early start of treatment would seem to be the only hope of success in these asymmetrical cases.

The cause of delayed open-angle glaucoma following congenital cataract surgery is not yet well understood. It has been postulated that the trabecular meshwork is less porous because of early postoperative inflammation.[13,14] Another possibility is that there is an underlying developmental abnormality of the anterior chamber angle predisposing to the glaucoma and related to the developmentally small corneas and poorly dilating pupils that are seen in association with the more severe congenital cataracts. The 27% incidence of delayed open-angle glaucoma in patients who have had surgery in the first 15 months is probably an underestimate, since we do not yet have postoperative pressure measurements in all patients and the glaucoma develops late. Certainly ocular pressures should be monitored regularly after early congenital cataract surgery. The incidence of glaucoma following later surgery for congenital lamellar opacities is probably lower but is still undefined.

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DISCUSSION

DR ALBERT W. BIGLAN. This report summarizes the experience of two skilled pediatric ophthalmologists who have used contemporary surgical methods to treat 51 children with bilateral congenital cataracts. This retrospective study was conducted over a 19-year interval. The respectable mean follow-up of 10.3 years permits the authors to report reliable recognition visual acuity values, and it also provides us with a realistic estimation of "long-term" complications.

Inclusion criteria used to select patients who had congenital cataracts is undisputable. However, the selection of patients who were able to have their visual acuity determined by recognition tests does not give us information on patients who were unable to perform subjective testing and were excluded.

Doctors Robb and Petersen have divided their study group into two categories: those patients with complete (extensive) cataracts and those with partial cataracts. This separation is logical because the visual prognosis and outcome, in this author's experience, differs between these two groups.

Doctors Robb and Petersen have found that eyes with complete cataracts were frequently associated with anatomic and functional anomalies such as decreased corneal diameters, pupils that dilated poorly, and visual deprivation nystagmus. Children with partial cataracts had eyes that were anatomically normal and lacked signs of visual deprivation.

The authors should be commended on reporting recognition visual acuity values. Reporting the grating visual acuity would probably include more patients in their study, but it would underestimate the prevalence of amblyopia.

The authors report the patient's functional visual acuity by reporting vision in the eye with the best visual acuity in each child. Although this provides us with a "real world" functioning visual acuity, patients with bilateral cataracts are actually given two chances to achieve this level of acuity. In Table I, the authors do provide us with data for the fellow eyes. In some cases, the visual acuity in the fellow eye is disappointing and this should not be overlooked. Some fellow eyes have decreased vision due to amblyopia and some of these eyes are undoubtedly undergoing treatment. Others have decreased vision due to complications such as glaucoma and retinal detachment.

The authors clearly demonstrate that children with partial cataracts can achieve levels of visual acuity that will permit them to compete in school, drive automobiles, and have gainful employment as they get older. On the other hand,

children who had eyes with complete cataracts may not do as well.

I attempted to see if we are making progress in the treatment of children with bilateral congenital cataracts. I reviewed visual acuity results obtained in children with partial and complete cataracts who were treated prior to the introduction of the vitreous suction-cutting instruments (*Ophthalmology* 1979; 86:1586-1598). These results were compared with the visual acuity results obtained in this study and two earlier studies in which complete or partial congenital cataracts were removed early in life with a vitreous suction-cutting instrument (Am J Ophthalmol 1982; 93:615-621; 94:441-449). In this study, I selected eyes from the author's Table I that had cataracts that were complete or partial (lamellar) and these are listed in Table I.

TABLE I: VISUAL ACUITY RESULTS AFTER CATARACT SURGERY FOR BILATERAL CONGENITAL CATARACTS

		VISUAL ACUITY	
		< 20/60	> 20/200
Francois 1979			
Complete cataract	(100 eyes)	25%	50%
Partial cataract	(167 eyes)	70%	17%
Gelbart, et al 1982			
Combined	(48 eyes)	60%	27%
Parks 1982			
Lamellar (partial)	(32 eyes)	100%	
Robb and Petersen 1992*			
Complete	(26 eyes)	19%	11%
Lamellar (partial)	(38 eyes)	73%	18%

*Eyes selected from Table I.

I was initially disappointed to see that the percent of eyes that attained visual acuity "better than 20/60" had not changed very much, but when I reviewed the data on individual eyes, I was encouraged to see that most eyes in this category had recognition visual acuities better than 20/30, and this does reflect progress. We have also made progress in reducing the number of eyes that become legally blind (visual acuity 20/200 or less). This trend has also been true in our community. The Western Pennsylvania School for Blind Children, has had a decrease in the number of children attending school because of visual loss due to congenital cataracts (Table II). The progress we have made is due in part to realization that it is important to perform cataract surgery early in life. Other factors include the introduction of new surgical instruments and techniques and our improved ability to manage complications such as glaucoma and retinal detachment.

TABLE II: CHILDREN ENROLLED IN THE WESTERN PENNSYLVANIA SCHOOL FOR BLIND CHILDREN

SCHOOL YEAR	TOTAL ENROLLMENT	STUDENTS WITH CONGENITAL CATARACTS
1962-63	199	16
1971-72	194	23
1981-82	183	12
1991-92	180	2

In this report, 68% of patients with bilateral congenital cataracts developed strabismus. Children with complete cataracts had an even higher prevalence, especially if cataract surgery was performed early in life. The high prevalence of strabismus should not be surprising because of the frequency that anisometropia, aniseikonia, and visual deprivation in this population.

Glaucoma developed in at least 20% of patients with bilateral congenital cataracts. What is of more concern is that glaucoma developed more frequently in patients who had their cataract surgery performed before 15 months of life. The authors estimate that the prevalence may even be higher, since some patients still have not had formal measurement of their intraocular pressures. Simon and associates (*Ophthalmology* 1991; 98:670-674) followed 34 eyes with congenital cataracts for 5 years and found that 24% of these eyes developed glaucoma. Keech and co-workers (*Am J Ophthalmol* 1989; 108:136-141) noted that glaucoma developed in 11% of the eyes they treated, and Chrousos and Parks (*Ophthalmology* 1984; 91:1238-1241) found "chronic glaucoma" occurred in 6.1% of the eyes in their study.

Glaucoma after cataract surgery in children is a serious complication, and it is very difficult to treat. Glaucoma may be associated with the underlying condition (rubella), or it may be related to complications of the surgical procedure or, as the authors suggest, there may be a coexisting defect in the facility of outflow of aqueous associated with cataracts.

There has been appropriate concern that the use of a suction-cutting device to remove a cataract and posterior lens capsule may increase the risk for retinal detachment. The advantage this technique offers is that the posterior lens capsule is almost completely removed and this reduces the need for surgery to treat an opacified capsule (*Ophthalmology* 1983; 90:344-345). In this study, a retinal detachment developed in only two eyes in the 51 patients. This incidence is similar to the 1.5% reported by Keech and co-workers (*Am J Ophthalmol* 1989; 108:136-141) and Chrousos and Parks (*Ophthalmology* 1984; 91:1238-1241). At this time, the use of a suction-cutting device appears to be a safe and effective method for removing cataracts in young children.

What can we take home from this presentation? First, we should note that over the last 2 decades, there has been an improvement in the visual outcome for children with bilateral congenital cataracts.

Second, the work of von Noorden (*Invest Ophthalmol* 1973; 17:721-731) and others has shown that it is necessary to perform surgery and provide optical correction early in life because there is a critical period for visual development. In support of this, Rodgers and associates (*Arch Ophthalmol* 1981; 99:999-1003) have found that children who were treated before 8 weeks of life had better visual acuity when compared with children who had their cataracts removed after the second month of life. It is unsettling that the Robb and Petersen study and the study reported by Keech and co-workers (*Am J Ophthalmol* 1989; 108:136-141), both show an increase in complications when cataracts are removed before 8 weeks of age. This may be due to anomalies associated with the cataracts in these patients or to risk factors yet to be defined.

Third, performing the cataract surgery very early in life does not ensure a good result. Robb and Petersen have demonstrated that even with early treatment, eyes still retain visual deprivation nystagmus.

What, then, is the optimal time for treatment of patients with congenital cataracts? In light of the preceding, I still feel that children with bilateral complete congenital cataracts who have absent red reflexes should have their cataracts removed within the first 2 months of life. These eyes should receive prompt correction of their aphakic refractive error, and this is usually accomplished with contact lenses.

In summary, treatment of a child with a cataract requires that the ophthalmologist have a thorough understanding of development of the visual system. The rehabilitation of these eyes requires commitment of the ophthalmologist and the parents to provide continued follow up to diagnose and to manage complications such as amblyopia, strabismus, and glaucoma. Adhering to these principles, children will, for the most part, enjoy excellent vision and become productive members of our society.

Dr David S. Walton. It has been my place in addition to seeing a few patients with Marfan disease to have the opportunity to see some children with glaucoma following cataract extraction. I commend Doctor Robb on his paper. As he pointed out, secondary glaucoma has replaced retinal detachment as the principal post-surgery threat when children are operated on for cataracts. When these patients present with glaucoma, one often sees a picture like this with obstruction of the pupil with residual lens tissue. They can look like this with the lens tissue not looking so prominent but rather with distorted pupils due to the binding down of the iris to the residual lens tissue. I have seen in my practice approximately 75 such cases. On gonioscopy, significant abnormalities are present. When one does preoperative, precataract gonioscopy, generally the filtration angles are normal. In this patient, with persistent hyperplastic primary vitreous one would suspect that perhaps preoperative gonioscopy might have been abnormal. What one usually sees when gonioscopy is done after surgery are striking abnormalities consisting of pigmentation in the angle and poor insertion of iris-like tissue over the meshwork. It is difficult to see with lower magnification, but it can be a very

striking abnormality contrasting with the beautiful angle of a young child with very little pigmentation.

I think the causes of glaucoma is important and we need more opportunity to look at those eyes pathologically that have this problem. Also, eyes that did not have glaucoma but have had childhood cataract surgery should be studied exhaustively. The cause of glaucoma is not clear, but in those patients who have had near perfect angles before surgery and which subsequently develop glaucoma and have abnormal angles such as I am shown, one is certainly suspect about the surgery and the postoperative reaction to the surgery and residual lens tissue. possibly the surgical technique of operating in the posterior chamber to facilitate the removal of as much lens material as possible will be helpful in lessening the risk of this glaucoma. I would very much like to hear Doctor Robb's comments about this.

I would like to underscore once again that glaucoma following this surgery is a serious problem affecting especially young children operated for cataracts in the first year of life. For their families and the children alike, it is a significant tragedy.

DR JOHN T. FLYNN. I would like to join in congratulating Doctors Robb and Petersen on an excellent piece of work in their retrospective study. This should serve as a spur to the pediatric community to get our act together and get some good case control studies going in this entity and finally to from case control studies to treatment clinical trials. My one question to Doctor Robb is with regard to the so-called partial or lamellar cataracts. Before the verbal age, what are his indications for surgery in that group of patients and then after the verbal age (of about 3), when presumably you start to get measurable recognition acuities, what are his indications for surgery in that group of patients?

DR MARSHALL M. PARKS. No doubt Doctor Robb and I see the same cataract patient pathology. However, as I view these patients, the main bulk of the pathology falls into two separate diseases, the lamellar cataract and the nuclear cataracts. Doctor Robb alluded to the lamellar cataracts, but I did not hear anything about the nuclear cataracts. The nuclear cataract involves the embryonic and fetal nucleus, present in the newborn with microcornea and a 3 mm axial opacity which is as amblyogenic as a total cataract. The lamellar cataracts are totally different, not present at birth, and since they are acquired I do not list them under the heading of congenital cataracts. Unless we initiate treatment of bilateral nuclear cataracts within the very first weeks of life, the fixation reflex in neither eye develops causing nystagmus to become apparent between 2 and 3 months of age. But the amblyopia process is going on all during those first 2 months of life prior to the appearance of the nystagmus at which time it has become irreversible. Therefore, to prevent irreversible amblyopia surgical clearing of the pupils and optical correction of the aphakia must be provided as soon as possible during the first 2 months of life. Doctor Robb's cataract patients with microcornea had nystagmus and yet many of our identical patients visually rehabilitated at 1 week or 2 months of age, including aphakic correction, don't have

nystagmus. The questions I pose are just how soon were his patients operated and provided aphakic optical correction, and with what degree of intensity were they followed within the first very few weeks of age? By the way, my incidence of aphakic glaucoma occurring in nuclear cataract patients within 5 years post-operatively is identical to Doctor Robb's experience. Also, our experience is identical for visual success even with late surgery in patients with lamellar cataracts.

DR W. RICHARD GREEN. In previous publications, the inability of the pupil dilate has been associated with rubella. I would like to ask Doctor Robb if there was an association of this feature with the total cataracts and with rubella.

DR GEORGE WEINSTEIN. I note that neither of the authors nor the primary discussant mentioned optical correction of the congenital cataract problem. Doctor Parks did touch upon this.

We live in an era of intraocular lens implantation. One might argue that this is an unsavory term if used with respect to the management of patients with congenital cataracts, but I must say that my personal experience in using lens implants in children has been very favorable. This is not with respect to patients in the earliest age group of onset, but rather for those who, as the authors described, developed their cataracts at some later time.

Of course, there is the problem of evolution of the eye, especially elongation of the eyeball and the related changes in the refractive error. However, if most of the refractive error can be corrected by a lens implant, the child has a much better chance at visual rehabilitation.

Of course, some concerns remain. The treatment of the posterior is in question. But I wondered if the authors would willing to address the question of the replacement of the lens with an intraocular lens implant, both the advantages and disadvantages of this.

DR RICHARD M. ROBB. I would like to thank all the commenters and especially Doctor Biglan for his thoughtful review of the manuscript. He did go back to review Doctor Francois' paper of 1979 on the results of surgery for congenital cataracts. One of the things that was interesting about that paper was that Doctor Francois had not operated on any patient prior to 6 months of age at that time. That stands out now because I think there is no question that surgery for congenital cataracts has been improved since the time that linear extractions or multiple decisions of the lens were the standard procedures. Congenital cataract surgery now can be done at an early age safely and effectively and cleanly.

Doctor Walton brings up the question of abnormalities of the anterior chamber angle in explanation of the acquired glaucoma. I respect Doctor Walton's statement about the findings on gonioscopy, and I am sure that in time he and others will help to clarify what the cause of this glaucoma is. The emphasis on glaucoma as a complication, I think, is perfectly appropriate. We have in the past worried more about retinal detachments but clearly open-angle glaucoma, which is a very

difficult complication to treat, has become a serious threat.

Doctor Flynn asked about indications for surgery and that would take more time than I have now to outline in detail. But in general it is a matter of assessing the visual performance of the patient Prior to the time of subjective visual acuity testing the assessment is made by behavioral means and by preferential looking testing. And after that by tests of visual acuity, they are familiar to all of you. I think it is always a question of balancing the child's visual behavior against the trade off, which is the requirement for aphakic correction. We commit a child to the use of an aphakic correction for an indefinite period of time once the cataracts are removed.

Doctor Parks' comments about lamellar vs nuclear cataracts are interesting and I recognize that he has divided his cases in that way. In reviewing our material it was not clear that this kind of a distinction was important to make. Doctor Parks is certainly representative of many pediatric ophthalmologists in bringing up the issue of early surgery and whether that will help to abort the development of nystagmus. We are not convinced at this point that that is so and I think we need some of what Doctor Flynn was alluding to: we need studies that are prospective studies in which the kinds of cataracts are well defined preoperatively so that we can tell whether or not the timing of surgery has a bearing on the development of nystagmus.

Doctor Green had a question about rubella. I believe there were two patients with rubella who were among the ones with poorly dilating pupils, but that does not account for all the patients who had poorly dilating pupils. Poor pupillary dilatation seems to be a distinctive feature of the early severe cataracts, even beyond those that are caused by rubella.

Finally, Doctor Weinstein had a question question about intraocular lenses. This is gradually being brought up among pediatric ophthalmologists, but I must say that the interest is filtering down from above rather than from below. That is, those of us who are considering the issue of very early surgery really haven't thought that it was wise to add the complications of an IOL to the surgery in the first or second month of life. I think that in the long run those of you who do IOL's and do them well, will probably begin to use them in the older children that we have to deal with. I would emphasize, however, that removing the posterior capsule at the time of surgery has been a major improvement in dealing with pediatric cataracts. It avoids the problem of secondary membranes which are almost universal if the posterior capsule is not removed at the time of initial surgery.

Thank you.