Congenital cataract extraction with primary aphakia and secondary intraocular lens implantation in the posterior chamber

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PURPOSE. *To evaluate visual outcomes, eye movement abnormalities, and postoperative complications after secondary posterior chamber intraocular lens (IOL) implantation in aphakic children who had initial LAC correction.*
METHODS. *A retrospective study was carried out on 53 patients (94 eyes). Following aphakic surgery and LAC correction the patients received secondary IOL implantation in the posterior chamber. Special attention was paid to factors that may have influenced their visual outcomes, such as eye movement abnormalities, cataract unilaterality, and cataract density.*
RESULTS. *No association was found between age at surgery and the onset of strabismus (p=0.611) or with visual acuity (p=0.086). However, unilaterality and total cataract density were found to have a negative association with poor vision (p<0.001). Strabismus and nystagmus were found to have a statistically significant negative association with visual acuity (p=0.002). Posterior capsule opacification occurred in six eyes of five patients following cataract extraction; IOL dislocation occurred in four eyes after IOL intraocular implantation, and secondary glaucoma occurred in one eye.*
CONCLUSIONS. *IOL implantation before 12 months of age may be useful in unilateral cataract; in bilateral cataract, simultaneous surgical aphakia, LAC correction, and then IOL implantation at 2.5–3 years of age, together with anti-amblyopic therapy when strabismus or asymmetric cataract density occur, are efficient methods to obtain visual recovery. Extraocular muscle surgery may be required to correct strabismus. (Eur J Ophthalmol 2008; 18: 903-9)*

KEY WORDS. *Cataract, Aphakia, IOL, Intraocular lens implantation*

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INTRODUCTION

Indications and surgical techniques for cataract extraction are firmly established; however, aphakia correction, choice of surgical procedure, and the optimum age for surgery are controversial (1, 2).
Some studies, in fact, report visual recovery by means of intraocular lens implantation (IOL) in patients with unilateral cataract under 12 months of age (3-5) whereas other studies report no significant difference between the use of contact lens (CL) and IOL, and recommend using first CL and then IOL after reaching 12 months of age in unilateral cataract, and after 2–2.5 years of age in bilateral cataract (2, 6). The present study reports the results of patients corrected by CL after cataract extraction surgery and who subsequently received secondary IOL at 2.5–3 years of age, with regard to visual outcome, eye movement abnormalities, and postoperative complications.

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METHODS

A retrospective study was carried out on 53 patients with congenital cataract, 20 female (38%) and 33 male (62%), all of whom presented to the Paediatric Ophthalmology division of the Ophthalmology Department of the University Federico II of Naples between 1990 and 2005. A total of 41 patients presented cataract in both eyes and 12 had unilateral cataract, making a total of 94 affected eyes. The mean age at diagnosis was 3.5 months (0–12 months). All patients operated for congenital cataract in this period were included in the study (cataracts secondary to trauma or pathologies such as uveitis were excluded from the study).

Five patients also presented microphthalmos. Cataract extraction was carried out by the following surgical procedure: corneal or sclerocorneal incision, viscoelastic substance in the anterior chamber, anterior capsulorhexis, manual or automized extracapsular lens extraction (ECCE), posterior capsulorhexis or posterior capsulotomy, broad central anterior vitrectomy, aspiration of the viscoelastic substance, and incision closure with10-0 nylon suture. In preparation for surgery, antibiotics were injected into the anterior chamber as endophthalmitis prophylaxis and dexamethasone and further antibiotics were injected subconjunctivally during and after surgery.

Following surgery, the operated eyes were treated with topical antibiotics, corticosteroids, and mydriatic and cycloplegic agents. In 34 patients (68 eyes) who presented bilateral cataract, simultaneous surgery was carried out after ensuring the absence of ocular infections (such as blepharitis, conjunctivitis, and dacryocystitis) to minimize the risk of endophthalmitis. In order to reduce the risk of contamination from the first eye operated, the second eye was operated using a second sterile surgical set and a complete change of surgical gown and gloves.

The secondary lens implantation procedure was as follows: corneal incision, viscoelastic substance in the anterior chamber, 360° anular synechiotomy, anterior vitrectomy when required, posterior chamber IOL, and 10-0 nylon suture. The choice of dioptric power was governed by echographic values, keratometry, the refraction of the healthy eye in unilateral cases, familiarity of refractive deficits, and surgical age; all patients were in fact 2.5–3 years of age, so we chose to hypocorrect in the order of 5–10% with respect to optimum IOL values.

First IOL implanted were Pharmacia Heparin Surface Modified AC IOL Model 722C; after a few years Acrysof MA60 BM (6 mm diameter and 12.5 mm long) were used; in the later years Hoya AF-1 lens (6 mm diameter and 13 mm long) were used. Mean lens power was 24.5 D (range 20–30 D).

Anterior vitrectomy was also carried out with IOL implantation.

The patients underwent a complete ophthalmologic examination both before and after surgery; special attention was paid to the evaluation of ocular alignment by the alternate cover test, Krimsky test, and also the Hirschberg test when necessary.

Postoperative visual acuity (VA) was evaluated by Albini's E and by Snellen's test in children over 3 years of age; refraction was evaluated by skiascopy. Cataract density was estimated by the degree of obstruction of the ocular fundus caused by the opacity (red reflex vision) during direct and indirect ophthalmoscopy.

All cataracts that impeded the vision of the red reflex at ocular fundus examination were considered total. Patients with strabismus or asymmetric bilateral cataract were treated with occlusive, antiamblyopic therapy. Seventeen patients were subsequently operated for strabismus; in these cases a deviation of less than 10 prism diopters was considered a success.

Mean follow-up was 65 months (range 46–108 months).

A statistical analysis of the influence of the following factors on VA was carried out: age at surgery, presence of strabismus, unilaterality/bilaterality, and cataract density. The 94 operated eyes were divided into three classes according to age: 0–4 months (35 eyes, 37%); >4 months and <12 months (45 eyes, 48%); and ≥12 months (14 eyes, 15%). Statistical analysis was carried out using chi-square and Fisher exact test. Values below 0.05 were considered statistically significant.

RESULTS

Sixty-five of the cataracts operated were total and 29 partial of which 13 were dense zonular, 12 posterior polar, and 4 posterior lenticonus. Mean age at cataract extraction was 7 months (2–24 months). Age at surgery was not found to be associated with strabismus onset ($p=0.611$) or with VA ($p=0.086$) (Tabs. I, II).

Cataract unilaterality, however, was found to be associated negatively with poor vision ($p<0.001$). As many as 10 eyes (83.5%) out of the 12 patients with unilateral cataract did not obtain >20/200 VA, while one eye (8.5%)

obtained 20/100 VA and one eye 20/40 VA. On the other hand, only 12 eyes of the patients with bilateral cataract (82 eyes) presented VA ≤20/200 whereas 36 eyes (44%) obtained VA >20/200 and 20/40 and 34 eyes (41%) obtained VA 20/40 (Tab. III).

Total cataract density was also shown to have a negative association with poor vision (p<0.01) only 12% of the eyes operated for total cataract versus 96% of partial cataract eyes obtained VA ≥20/40 (Tab. IV).

Amblyopia occurred in 92% of monolateral cataract patients (11 out of 12) and 36% of bilateral cataract patients (15 out of 41). Twenty-one patients (40%) presented strabismus before cataract extraction surgery of which 15 were esotropic and 6 exotropic. Nystagmus was present

TABLE I - VISUAL ACUITY IN 41 PATIENTS WITH BILATERAL CONGENITAL CATARACT

Visual acuity	No. of eyes (n=82 eyes)	%	Age at cataract extraction, mo			p value*
			≤4	4–12	≥12	
≤20/200	12	15	5/30 (17%)	3/42 (7%)	4/10 (40%)	0.086
20/200–20/40	36	44	14/30 (47%)	17/42 (40%)	5/10 (50%)	0.086
≥20/40	34	41	11/30 (36%)	22/42 (52%)	1/10 (10%)	0.086

*Chi-square test

TABLE II - VISUAL ACUITY IN 12 PATIENTS WITH UNILATERAL CATARACT

Visual acuity	No. of eyes (n=12 eyes)	%	Age at cataract extraction, mo			p value*
			≤4	4–12	≥12	
≤ 20/200	10	83	2/2 (100%)	5/5 (100%)	3/5 (60%)	0.086
20/200–20/40	1	8.5			1/5 (20%)	0.086
≥ 20/40	1	8.5			1/5 (20%)	0.086

*Chi-square test

TABLE III - OCULAR ALIGNMENT BEFORE AND AFTER CATARACT EXTRACTION

Ocular alignment	Patients before surgery	% Of total patients	Patients after surgery	% Of total patients
Strabismus	21/53	40	38/53	72
Esotropia	15/53	28.5	26/53	49
Exotropia	6/53	11.5	12/53	23
Hypertropia	0/53	0	2/53	4
DVD	0/53	0	4/53	8
Nystagmus	11/53	21	11/53	21

DVD = Dissociated vertical deviation

TABLE IV - THE INFLUENCE OF CATARACT DENSITY ON VISUAL ACUITY IN 94 EYES

Visual acuity	Eyes with total cataract (n=65)	Eyes with partial cataract (n=29)	p value*
≤20/200	22/65 (34%)	0 (0%)	<0.01
20/200–20/40	35/65 (54%)	2 (7%)	<0.01
≥20/40	8/65 (12%)	27(93%)	<0.01

*Chi-square test

in 11 patients (21%) and was associated with strabismus in 8. Strabismus was observed in 38 patients (72%) after surgery: 26 had esotropia and 12 exotropia (Tab. V). Furthermore, two patients presented hypertrophy associated with esotropia, and four patients had DVD associated with nystagmus and esotropia.

The presence of eye movement disorders such as strabismus or nystagmus was found to be a statistically significant negative factor for visual acuity (p=0.002) (Tab. VI).

Table VII reports refractive results after secondary IOL implantation. In some patients the refraction remained stable, in others there was a slight tendency towards myopia; only one patient required further surgery for severe myopia.

The opacification of the posterior capsule appeared as a complication in 6 eyes (7%) of 5 patients following cataract extraction surgery. These eyes were reoperated. Dislocation of the IOL was seen in 4 eyes (4%) of 4 patients following secondary IOL implantation; this required repositioning in 2 patients; secondary glaucoma occurred in 1 eye (1%) of a patient with microphthalmos.

Satisfactory results were obtained in all 17 patients undergoing strabismus surgery: 13 patients became orthotropic and residual deviation remained in 4 cases (Tab. VIII).

DISCUSSION

Despite numerous articles regarding visual recovery of aphakic children after congenital cataract surgery the subject is controversial. There are different schools of thought with regard to the choice of surgical procedure, the optimum age for surgery in order to minimize the risk of amblyopia, and the best method to achieve visual recovery. Some studies recommend the use of IOL in unilateral cataract patients even prior to the age of 12 months (3, 4). Lambert et al (5) studied a group of 25 children operated in the first months of life for unilateral cataract extraction, comparing eyes corrected with CL with eyes corrected with IOL. Although better visual outcomes were observed in the children corrected with IOL, they required a greater percentage of second surgeries (83% IOL versus 23% CL). Autrata et al (7), working on a group of 41 children who had undergone unilateral congenital cataract extraction surgery in the early months of life, report better visual acuity results, lower strabismus incidence, and better binocular vision in the IOL corrected patients compared to the CL corrected patients but a greater percentage of complications requiring repeat surgery. On the other hand, differences in visual acuities after correction with IOL or CL have not been reported elsewhere (8, 9). All the patients in our study were corrected with CL after cataract extraction and subsequently, at the age of 2.5–3 years they were corrected by IOL implantation. Satisfactory results were obtained in the bilateral cataract patients; however, despite antiamblyopic therapy, disappointing results were obtained in the unilateral cataract patients. One patient only, who had initially presented a partial cataract which later became total (operated for ECCE at 24 months and IOL at 36 months), obtained a visual acuity of 20/40. Amblyopia was observed in 49% of pa-

TABLE V - VISUAL ACUITY IN RELATION TO LATERALITY

Visual acuity	Unilateral cataract	Bilateral cataract	p value*
≥20/40	1/12 (8.5%)	34/82 (41%)	<0.001
20/200–20/40	1/12 (8.5%)	36/82 (44%)	<0.001
≤20/200	10/12 (83%)	12/82 (15%)	<0.001

*Chi square test, Fisher exact test

TABLE VI - RELATIONSHIP BETWEEN STRABISMUS AND VISUAL ACUITY: DATA OBTAINED IN 53 PATIENTS

Visual acuity	Strabismus (38/53)	No strabismus (15/53)	p value*
≥20/40	25%	50%	0.002
20/200–20/40	36%	42.5%	0.002
≤20/200	39%	7.5%	0.002

*Chi-square test

TABLE VII - REFRACTIVE DATA AFTER INTRAOCULAR LENS IMPLANTATION

No.	Sex	Type	Age, mo	Refraction 1 year after	Refraction 2 years after	Refraction 3 years after	Final refraction	Follow-up, mo
1	M	U	30	OS +1.25	OS 1.25	OS +0.75	OS +0.75	56
2	F	B	36	OD –0.50 OS –1	OD –1 OS –2	OD –2 OS –2.5	OD –3 OS –4	67
3	M	U	36	OD +1	OD + 0.75	OD +0.50	OD +0.50	46
4	M	B	33	OD –2 OS –2.5	OD –2.5 OS –3	OD –3.5 OS –4.25	OD –3.5 OS –4.25	57
5	M	U	34	OD +0.75	OD +O.50	OD +0.50	OD +0.50	76
6	F	U	33	OD –1	OD –2	OD –3	OD –4	65
7	M	B	36	OD +0.50 OS +0.50	OD –0.50 OS–0.5	OD –0.5 OS–0.5	OD –0.5 OS–0.5	71
8	M	B	35	OD –1.5 OS –2.75	OD –1.5 OS –3.25	OD –2.25 OS –3.75	OD –2.25 OS –3.75	54
9	M	U	31	OS +1	OS +1	OS +0.75	OS +0.75	49
10	M	U	33	OD +1.50	OD +1.25	OD + 1	OD +0.75	58
11	F	U	34	OS –1	OS –2	OS –2	OS –2.5	71
12	M	B	35	OD +3 OS +3	OD +2 OS +2	OD +1 OS +1.50	OD –2.75 OS –1.5	72
13	M	B	32	OD +1.25 OS +1	OD +0.75 OS +1	OD +0.75 OS +1	OD +0.75 OS +1	88
14	M	B	31	OD +1 OS +1	OD E OS E	OD E OS E	OD E OS E	97
15	M	B	33	OD +4.50 OS +4.50	OD +4.25 OS +4.5	OD +3.50 OS +3.25	OD +3 OS +2.50	67
16	F	U	32	OS +1	OS +1	OS +0.75	OS +0.75	68
17	F	B	30	OD –0.25 OS–0.25	OD –0.50 OS–0.50	OD –0.50 OS–0.50	OD –0.50 OS–0.50	71
18	M	U	30	OD +1.50	OD +2	OD +1	OD +0.5	86
19	M	B	36	OD –1.25 OS –0.5	OD –1.75 OS –1	OD –2 OS –1.50	OD –3.50 OS –3.25	108
20	M	B	33	OD –3.50 OS –2	OD –4.50 OS –3	OD –4.50 OS –3	OD –5 OS –3.25	67
21	M	B	30	OD–1.5 OS –3.5	OD–1.5 OS –3.5	OD –2 OS –4	OD –2.5 OS –4.5	55
22	F	U	31	OD +1	OD +0.75	OD +0.75	OD +0.75	64
23	F	B	32	OD –1 OS–0.75	OD –2 OS –1.25	OD –2.50 OS –1.25	OD –3.25 OS –1.50	65
24	M	B	30	OD –0.75 OS –1.50	OD –0.75 OS –1.50	OD –0.50 OS –1.5	OD –2.25 OS –3.5	89
25	M	B	30	OD +1.50 OS +1.25	OD +1.25 OS +1	OD +1 OS +1	OD E OS E	49
26	M	B	34	OD+0.50 OS +0.50	OD E OS E	OD –0.50 OS –1	OD–0.75 OS–1	52
27	M	B	36	OD +0.50 OS +1	OD E OS +O.75	OD –1 OS +O.25	OD –2.5 OS –1.5	59
28	M	B	32	OD +1.50 OS +1	OD +1 OS +0.75	OD +0.50 OS +O.25	OD +0.25 OS E	63
29	M	B	31	OD E OS–0.50	OD E OS E	OD E OS E	OD E OS E	72
30	M	B	33	OD +1.50 OS +1.25	OD +1.50 OS +1.25	OD +1 OS +1.25	OD +1 OS +1	71
31	F	B	34	OD +0.75 OS +1	OD + 0.50 OS +0.50	OD E OS E	OD E OS E	59
32	F	B	35	OD +0.50 OS +0.25	OD E OS E	OD –0.50 OS –0.75	OD –0.5 OS –0.75	98
33	M	B	30	OD +2 OS +1.75	OD +1.75 OS +1.50	OD +1.50 OS +1.25	OD +1 OS +1	52
34	F	B	33	OD +0.50 OS +0.50	OD +0.50 OS +O.50	OD E OS E	OD E OS E	48
35	F	B	32	OD +1.50 OS +1.25	OD +150 OS +1.25	OD +1 OS +1.25	OD +1 OS +1.25	65
36	M	B	34	OD +1 OS +1	OD +1 OS +1	OD +0.50 OS +0.25	OD E OS E	65
37	M	B	36	OD +1.50 OS +1.25	OD +1.25 OS +1	OD +0.75 OS +1	OD –0.50 OS +0.25	61
38	F	B	33	OD –0.25 OS –0.25	OD –0.25 OS –0.25	OD–0.50 OS –0.50	OD –075 OS –0.75	58
39	M	B	35	OD +1 OS +0.75	OD +0.75 OS +0.50	OD –0.25 OS –0.25	OD –0.50 OS –0.75	55
40	F	B	30	OD +0.50 OS +0.50	OD E OS E	OD –0.25 OS –0.25	OD –0.25 OS –0.25	77
41	M	B	33	OD +1 OS +1	OD +1.50 OS+1.50	OD +2 OS +2	OD +2 OS +2.50	81
42	F	B	34	OD +2 OS +2	OD +1.5 OS +2	OD +1 OS +1.50	OD +1 OS +0.75	68
43	F	B	34	OD +1 OS +1	OD +1 OS +0.50	OD +0.50 OS +0.50	OD E OS E	49
44	M	B	32	OD +0.5 OS +0.5	OD +0.50 OS +0.5	OD E OS E	OD E OS E	51
45	M	B	30	OD –0.50 OS –0.5	OD –1 OS –0.5	OD –1.5 OS –1	OD –2 OS –1.50	52
46	M	B	36	OD –0.25 OS –0.50	OD –0.50 OS –0.50	OD –0.75 OS –0.75	OD –1 OS –1.25	60
47	M	U	34	OS –1.50	OS –2.25	OS –2.50	OS –5.25	72
48	F	B	35	OD +0.75 OS +1	OD +0.50 OS +0.50	OD –0.50 OS –0.50	OD –0.50 OS –0.50	62
49	F	B	32	OD +0.50 OS +1	OD +0.50 OS +1	OD E OS E	OD E OS E	56
50	F	U	32	OS +1.25	OS +1	OS +0.75	OS + 0.50	48
51	F	B	33	OD +1 OS +1	OD +0.5 OS +0.5	OD +0.25 OS –0.25	OD –0.5 OS –0.75	65
52	M	B	36	OD +2 OS +1.75	OD +1.50 OS +1	OD +1.50 OS +1	OD +0.75 OS + 0.50	54
53	F	B	36	OD +0.50 OS +0.75	OD E OS +0.50	OD –1 OS –0.75	OD –1.75 OS –1	50

U = Unilateral; OS = Left eye; B = Bilateral; OD = Right eye; E = Emmetropia

TABLE VIII - STRABISMUS SURGERY IN 17 PATIENTS AFTER CATARACT EXTRACTION

Patient	Age at cataract surgery	Sex	Ocular alignment before cataract extraction	Type of cataract	Ocular alignment after cataract extraction	Laterality of cataract	Angle of deviation (prism diopters) before strabismus surgery	Strabismus surgery	Angle of deviation (prism diopters) after strabismus surgery
1	5	M	ET	Total	ET	UN	20	MRc+LR	ORTH
2	4	M	ORTH	Total	XT	UN	20	MR+LRc	ORTH
3	3	M	ORTH	Total	ET+H	BIL	25	MRc+LR+IOc	ET
4	7	F	XT	Total	XT	BIL	30	LRc	ORTH
5	8	F	ET	Zonular	ET+N+H	BIL	20	MRc+LR+OIc	ET+N+H
6	7	M	ORTH	Total	XT+N	BIL	30	LR	XT+N
7	8	F	ORTH	Posterior subcapsular	ET	BIL	20	MRc+LR	ORTH
8	3	F	ET	Total	ET	UN	25	MRc+LR	ORTH
9	5	M	ET	Total	ET	UN	30	MRc+LR	ORTH
10	6	M	ET	Total	ET	UN	30	MRc+LR	ORTH
11	9	F	ET	Zonular	ET	UN	20	MRc+LR	ORTH
12	5	M	ET	Total	ET	UN	25	MRc+LR	ORTH
13	7	M	XT N	Total	XT+N+H	BIL	35	MRc+LR+IOc	XT+N+H
14	6	F	ORTH	Total	ET	BIL	30	MRc+LR	ORTH
15	7	F	ET	Zonular	ET	UN	25	MRc+LR	ORTH
16	4	M	ORTH	Total	ET	UN	30	MRc+LR	ORTH
17	4	M	XT N	Total	XT+N	UN	25	LRc	ORTH

ET = Esotropia; UN = Unilateral; MRc = Medial rectus recession; LR = Lateral rectus resection; ORTH = Orthotropia; XT = Exotropia; H = Hypertropia; BIL= Bilateral; Ioc = Inferior oblique recession; N = Nystagmus

tients (26 patients); amblyopia occurred in 92% of unilateral cataracts versus 36% of bilateral cataracts. Twenty-three out of 26 patients with amblyopia presented eye movement disorders. Furthermore, 12 of the 15 patients with bilateral cataract and amblyopia presented asymmetric density cataract.

It may be hypothesized that strabismus occurring in bilateral cataract arises from the difference in cataract density between the two eyes: in fact the total density was found to be associated with poor vision. Thus our study found that unilaterality, total cataract density, the presence of eye disorders such as strabismus and nystagmus had a negative influence on visual acuity. Opacification of the posterior chamber is one of the principal complications of cataract surgery and increases the risk of deprivation amblyopia (2, 10, 11). Some studies have observed a significant decrease in postcataract surgery opacification of the posterior capsule, employing capsulorhexis in association with anterior vitrectomy (1, 2). Posterior capsule opacification following cataract extraction occurred in 6 eyes (6%) of 5 patients in our study, necessitating surgical intervention adopting posterior capsulectomy and anterior vitrectomy or pars plana vitrectomy. Only one case of glaucoma in one eye (1%) occurred, in a patient with microphthalmos; this was treated medically. Zwaan et al (11) also observed 1% glaucoma incidence in eyes corrected with IOL. We are in agreement with other authors who claim that the use of IOL in the posterior chamber reduces the risk of postoperative glaucoma (1, 2, 11, 12).

We believe that in unilateral congenital cataract, IOL implantation under 12 months of age may be opportune, even though other authors have not obtained satisfactory results (13). In bilateral cataract, simultaneous surgical aphakia, LAC correction, and then IOL implantation at 2.5–3 years of age, together with antiamblyopic therapy when strabismus or asymmetric cataract density occurred, are efficient methods to obtain visual recovery. Finally, extraocular muscle surgery may be required to correct strabismus.

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