SURGICAL TREATMENT OUTCOMES OF CONGENITAL AND JUVENILE CATARACTS

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SUMMARY

Purpose: Evaluation of visual outcome after lens aspiration with or without intraocular lens implantation for isolated congenital and juvenile cataract in children aged 6 years and younger.
Material and Methods: Retrospective review of 48 children with isolated congenital and juvenile cataract who were surgically treated between January 1993 and December 2002 and had a minimal follow-up of 12 months.
Results: In the group of children with unilateral cataract, 33% (3 out of 9 children) of aphakic children and 45.5% (5 out of 11 children) of pseudophakic children attained a final best corrected visual acuity of 20/200 and 20/60 respectively. In the group of children with bilateral involvement, 35% (6 out of 17 children) of aphakic children have a final best corrected visual acuity of 20/30 and 63.7% (7 out of 11 children) of pseudophakic children have a final best corrected visual acuity of 20/25 or more.
Conclusion: The results of this study emphasize the need for early surgery and good organisation of postsurgical care in patients with pediatric cataract to optimise visual outcome. Furthermore patients with isolated unilateral congenital cataract surgically treated at an average age of 15 months without primary lens implantation and with variable and low compliance have suboptimal results. The effect of early surgery with primary lens implantation on the long term visual outcome in pediatric cataract needs to be further evaluated.

SAMENVATTING

Doel: Evaluatie van de visuele resultaten na lensaspiratie met en zonder primaire intra-oculaire lensimplantatie voor geïsoleerd congenitaal en juveniel cataract in kinderen zes jaar en jonger.
Materiaal en methoden: Retrospectieve studie van 48 kinderen met geïsoleerd congenitaal en juveniel cataract die heelkundig behandeld werden tussen Januari 1993 en December 2002.
Resultaten: In de groep patiëntjes met unilateraal cataract bereikten 33% (3 van de 9 patiëntjes) van de afake kinderen en 45.5% (5 van de 11 patiëntjes) van de pseudofake kinderen een best gecorrigeerde visuele acuiteit van respectievelijk 20/200 en 20/60. De groep patiëntjes met bilateraal cataract bereikten een best gecorrigeerde visus van 20/30 in 35% (6 van de 17 kinderen) van de afake kinderen en een best gecorrigeerde visus van 20/25 of beter in 63.7% (7 van de 11 kinderen) van de pseudophake kinderen.
Besluit: De resultaten van deze studie beklemtonen het belang van vroege interventie en goede organisatie van de postoperatieve opvolging in patiëntjes met pediatrisch cataract ter optimalisering van het visuele resultaat. We zien dat patiëntjes met geïsoleerd unilateraal congenitaal cataract heelkundig behandeld op een gemiddelde leeftijd van 15 maanden zonder primaire lens implantatie en gepaard gaand met een variabele en lage compliantie een suboptimaal visueel resultaat hebben. Het lange termijneffect van vroege heelkunde met primaire intraoculaire lensimplantatie op het visuele resultaat dient verder geëvalueerd te worden.

RÉSUMÉ

But: Evaluer les résultats fonctionnels de la chirurgie de la cataracte avec et sans implant de cristallin artificiel chez l'enfant de moins de 6 ans.
Matériel et Méthodes: Analyse rétrospective de 48 enfants avec une cataracte isolée congénitale et ju-

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vénile, qui ont été traités de janvier 1993 à décembre 2002.
Résultats: Dans le groupe d' enfants avec une cataracte unilatérale, 33% (3 des 9 patients) des enfants aphaques et 45,5% (5 des 11 patients) des enfants avec un implant, développaient une acuité visuelle corrigée respectivement de 20/200 et 20/60. Le groupe d' enfants avec cataracte bilatérale avait une vision corrigée de 20/30 pour 35% (6/17 enfants) des enfants aphaques, et une vision corrigée de 20/25 ou mieux pour 63,7% (7/11 enfants) des enfants pseudophaques.
Conclusion: Cette étude permet de rappeler l'importance d'une intervention précoce et d'une bonne organisation des soins postopératoires des enfants avec cataracte pédiatrique, afin d'optimaliser le résultat visuel. Les enfants avec cataracte congénitale isolée unilatérale opérés vers l'âge de 15 mois, sans implantation primaire d'un cristallin artificiel, et avec compliance variable, ont un résultat visuel suboptimal. Le résultat fonctionnel à long terme d'une opération précoce avec implantation dans la chambre postérieure, demande une évaluation ultérieure.

KEY WORDS

congenital cataract - juvenile cataract - surgery - outcome

MOTS-CLÉS

cataracte congénitale - cataracte juvénile - chirurgie - résultats fonctionnels

INTRODUCTION

Congenital cataract is a common cause of poor vision in infants. Improved surgical methods and the insight in the importance of early detection, allowing early treatment as well as appropriate optical correction and amblyopia treatment, have improved visual outcome of childhood cataracts. In spite of this knowledge, the treatment of childhood cataract still remains a challenge. Even with early and intensive treatment, the visual outcome of unilateral cataract can be disappointing. Bilateral dense cataracts attain better visual results if operated before the age of eight weeks (9,10). Less severe bilateral lens opacities may have better outcome if intervention is delayed. We retrospectively reviewed infants and children with unilateral or bilateral cataracts without associated ocular or systemic disease, who underwent surgery before the age of 6 years between 1993 and 2002 at the University Hospital Leuven, Belgium. Visual outcome and complications are reported.

METHODS

We reviewed the records of all children aged 6 years and younger who underwent cataract surgery between January 1993 and December 2002 at the University Hospital Leuven, Belgium. One hundred and seven records were reviewed. Children with associated ocular anomalies, systemic or mental disorders, less than 12 months follow-up and absence of follow-up at our department were excluded (Table 1). Forty-eight patients were eligible for inclusion in this study.

Table 1: *Exclusion criteria*

	No patients		*No patients*
Retinoblastoma	2	Aniridia	1
Uveitis with secondary cataract	3	Microphthalmia	2
PHPV	13	Disorders of the eye as a whole	1
Traumatic cataract	4	(microphthalmia, cataract, glaucoma)	
Marfan syndrome	4	Psychomotor retardation	3
Cockayne syndrome	2	of unknown origin	
Conradi syndrome	1	Epilepsy	2
Rubella syndrome	1	Secondary lens implantation	3
Dubowitz syndrome	1	Follow-up less than 12 months	7
Down syndrome	1	Surgery in other Hospital	2
Hallerman-Streiff-François syndrome	1	Others	5

Four subgroups are considered. Unilateral cataract was present in 20 patients. Eleven infants were treated with primary IOL implantation (unilateral pseudophakia, group P1), 9 infants were primary corrected with contact lenses (unilateral aphakia, group A1). Bilateral cataracts were present in 28 patients. Eleven children were treated with primary IOL implantation (bilateral pseudophakia, group P2), 17 were primary corrected with contact lenses (bilateral aphakia, group A2).

The protocol followed at the University Hospital Leuven for children presenting with cataract is described below. Preoperatively all patients underwent a full ophthalmological assessment including visual acuity, eye motility and position, slit lamp examination, dilated fundoscopy, retinoscopy, keratometric reading and A-scan ultrasound determination of axial length. To assess vision we used age appropriate tests. Correction was worn during the testing. In some preverbal patients, monocular and binocular fixation behaviour was assessed and a Snellen visual acuity was assigned to each fixation pattern as described by Bradford et al (5). The pupils were dilated with cyclopentolate 0,5%, fundoscopy and streak retinoscopy were performed. In children with asymmetric cataracts, a trial of occlusion was recommended to eliminate the possibility of superimposed amblyopia. If vision was difficult to assess, we reassessed the child after a short period. In small and very uncooperative children, ocular examination under general anaesthesia was performed. Surgery was planned if the lens opacities were severe enough to interfere significantly with visual development. Babies or children with unilateral cataracts were not referred to the paediatrician; all the bilateral cases without a positive family history were referred to the paediatric department for a complete work-up. Informed consent was obtained for all patients from the parents after discussion of the risks, benefits and postoperative care.
Children in group A2 (bilateral aphakia) underwent surgery for the second eye within 1 week (mean 3.9 days) without occlusion and without optical correction of the first eye during the interval between the operation on the first and on the second eye. Contact lenses were inserted at the time of the second operation. In group P2 (bilateral pseudophakia) the average time between surgery of the first eye with the poorer vision and the second eye was 3.2 months (2-7months).
Intraocular lens power was calculated with the SRK II formula. In unilateral cataract the IOL power was aimed at emmetropia in the early postoperative period for immediate visual rehabilitation. In bilateral cases we tried to obtain a slight hypermetropia in accordance to age and expected eye growth. In most patients we used heparin coated PMMA lens (Pharmacia, 811C or 812C), more recently we use the single and three piece acrylic foldable IOL (Alcon Laboratories Inc., SA60AT or MA30BA).
Surgery was performed by one surgeon (BF), experienced in both surgical procedures. General anaesthesia was used in all patients. The pupils were dilated with cyclopentolate 0.5% and phenylephrine 5% instillated three times starting one hour before surgery.
Because of the variability of the eye position under anaesthesia a superior bridle suture is beneficial. In children with bilateral congenital cataracts who needed surgery in the first 12 weeks of life, a standard lensectomy and anterior vitrectomy without IOL was performed. Children with unilateral cataract under the age of one were also left aphakic. Two corneal stab incisions were made at the 2 and 10 o'clock position. After filling the anterior chamber with Healon® (10 mg/ml, Pharmacia Ophthalmics, Uppsala, Sweden), the anterior capsule was opened. If possible a continuous curvilinear capsulorhexis with forceps was performed; otherwise a can-opener technique was used. The lens material was aspirated using a bimanual irrigation-aspiration technique. With a vitrectomy tip the posterior capsule was opened centrally and an anterior vitrectomy was performed. In all cases a peripheral iridectomy was made at the twelve o'clock position. Corneal incisions were closed with a Nylon 10.0 suture. Optical correction of the aphakia with contact lenses (Silsoft super plus, Bausch & Lomb) was standard.
When the decision to implant an IOL was made, a partial thickness 3.2 mm limbal tunnel was performed. A manual anterior continuous capsulorhexis was performed using forceps. In white cataracts trypan blue staining was used to visualise the capsule. An irrigation and aspiration hand piece was used to aspirate the lens

material. The posterior capsule was left intact in specific situations (older children, earlier cases, higher vitreous pressure) or a posterior rhexis with or without an anterior vitrectomy was done. The capsular bag was filled with Healon® and the IOL was placed in the capsular bag. The tunnel was closed watertight with Nylon 10.0 sutures.

At the end of the procedure depot steroid (Celestone Chronodose®) was injected subconjunctivally. An antibiotic ointment and one drop of Atropine 1% was placed, the eye was patched and a shield was placed over the eye. Postoperatively all children received a combination steroid- antibiotic eye drop four to six times daily for two weeks after which it was tapered slowly tailored to the patient's needs. Topical atropine sulphate 0.5% eye drops were used once or twice daily for two weeks. Follow-up visits were planned at day one, day four, day eight and then every two weeks until week 6 followed by controls at 4 monthly intervals. Intraocular pressures were measured every 3-6 months using Goldmann or Perkins tonometry. Amblyopia treatment was initiated immediately after surgery following the Great Ormond Street occlusion protocol for unilateral and bilateral cataract (6). The refractive correction was prescribed 6 weeks after surgery. Bifocals or multifocals were adapted depending on the case.

Table 2: *Visual acuity and compliance with occlusion therapy in children after lensectomy for unilateral cataract (***group A1**).

Patient **n=9**	*eye*	*Present. (months)*	*Surgery (months)*	*aetiology*	*preop VA*	*postop VA*	*Follow-up*	*Occlusion*
MB	OS	2	3	C	PL	20/50	65	Good
DWC	OD	2	3	C	PL	<20/400	15	Very poor
TT	OD	4	4	C	PL	<20/400	42	None
CJ	OS	5	5	C	PL	20/200	48	Good
HL	OD	8	9	C	PI	<20/400	72	Good
VS	OD	11	13	C	PL	<20/200	63	None
VJ	OD	21	22	J	PL	20/100	61	Good
MJ	OS	19 days	29	C	PL	<20/400	55	None
LY	OD	3	44	C	PL	<20/400	31	Very poor

C: congenital, J: juvenile, PL: perception of light, VA: visual acuity

Table 3: *Visual acuity and compliance with occlusion therapy in children after IOL implantation for unilateral cataract (***group P1)**.

Patient **n=11**	*eye*	*Present. (months)*	*Surgery (months)*	*Type of IOL*	*aetiology*	*preop VA*	*postop*	*Follow up (months)*	*Occlusion*
JN	OS	7	8	H	C	PL	20/30	20	Good
JT	OS	9	10	S	C	PI	20/200	40	Poor
HK	OS	31	25	A	J	20/250	20/200	14	Good
VJ	OS	12	44	H	C	20/400	<20/200	54	Good
FA	OS	42	46	H	J	<20/400	20/100	26	Good
VPR	OS	52	52	H	J	20/200	20/30	18	Good
MoA	OD	51	53	H	J	20/200	20/50	29	Good
DSR	OS	55	57	A	J	20/80	20/60	14	Poor
DM	OD	62	63	A	J	CF's	20/200	19	Good
DuM	OD	55	67	A	J	20/250	20/100	14	Good
MA	OS	7	68	H	C	20/80	20/25	23	Good

H: heparin coated PMMA IOL (Pharmacia & Upjohn, 811C or 812C); A: Acrysof IOL (Alcon Laboratories Inc, SA60AT or MA30BA), S: sulcus IOL 720C Pharmacia & Upjohn. VA: visual acuity, J: juvenile, C: congenital

Table 4: *Visual acuity after bilateral lensectomy for bilateral cataract (***group A2**).

Patient **n=17**	*eye*	*Present. (months)*	*surgery (months)*	*aetiology*	*preop VA*	*postop VA*	*Follow up (months)*	*Occlusion*
VM	OD	6 days	2	C	PL	20/30	106	Good
	OS		2	C	PL	20/30	106	
SoN	OD	16 days	2	C	PL	20/100	100	Poor
	OS		2	C	PL	20/30	100	
SS	OD	15 days	2	C	FF	20/50	72	Good
	OS		2	C	FF	20/50	72	
SN	OD	9 days	2	C	PL	20/100	21	None
	OS		2	C	PL	20/100	21	
DuK	OD	1	2,5	C	searching	20/25	103	
	OS		2,5	C	searching	20/25	103	
AB	OD	2,5	2,5	C	PL	<20/400	62	None
	OS		2,5	C	PL	20/120	62	
DK	OD	1	2,5	C	PL	20/100	39	Good
	OS		2,5	C	PL	20/100	39	
AL	OD	3 weeks	3	C	PL	20/30	100	Poor
	OS		3	C	PL	20/200	100	
VdkA	OD	2,5	3	C	searching	20/100	78	Poor
	OS		3	C	searching	20/60	78	
DRM	OD	3	3	C	FF	20/80	65	Good
	OS		3	C	FF	20/80	65	
GD	OD	3	3	C	PL	20/200	24	
	OS		3	C	PL	20/200	24	
TS	OD	4	4	C	PL	20/400	98	Poor
	OS		4	C	PL	20/80	98	
AJ	OD	4	5	C	PL	20/60	114	None
	OS		5	C	PL	20/60	114	
VBA	OD	5	5	C	ODSign	20/50	52	None
	OS		5	C	ODSign	20/50	52	
BJ	OD	1.5	6	C	PL	20/30	69	Good
	OS		6	C	PL	20/50	69	
AM	OD	11	14	C	PL	20/25	31	Good
	OS		14	C	PL	20/25	31	
NA	OD	13	19	C	PL	20/50	60	None
	OS		19	C	PL	20/200	60	

C: congenital, PL: perception of light, FF: fixing and following, ODsign: oculodigital sign, VA: visual acuity

RESULTS

Forty-eight patients (76 eyes) with a minimum of 12 months follow-up were included in the study. Twenty-eight patients (58.3%) had bilateral and 20 patients (41.7%) had unilateral lens opacities. In the group with bilateral lens opacities, cataracts were congenital in 17 patients. All of them were treated by lensectomy without consecutive IOL implantation. The remaining 11 children had bilateral juvenile cataract and were implanted primarily with a posterior chamber IOL. In the unilateral cataract group, 9 eyes underwent a lensectomy, 8 of those were congenital in origin, one juvenile. A posterior chamber IOL was implanted in the remaining 11 eyes, 4 of those were congenital lens opacities, 7 juvenile. Cataracts presenting and diagnosed after the age of 1 year are specified as juvenile cataracts.

VISUAL OUTCOME

Unilateral cases (n=20)

The mean age at surgery was 15 months (range 3-44 months) for the unilateral aphakic group and 45 months (range 8-68 months) for the unilateral pseudophakic group. Mean follow-up period was 67.5 months (range 18-98 months). The visual acuity attained following cataract surgery for group A1 and group P1 is shown in Table 2 and 3.

In the unilateral aphakic group (group A1, n=9) only 3 eyes (33.3%) attained a visual acuity of 20/200 or more. Poor vision in this group was

Table 5: *Visual acuity after bilateral IOL implantation for bilateral cataract (***group P2**).

Patient **n=11**	*eye*	*Present. (months)*	*Surgery (months)*	*Type of IOL*	*aetiology*	*Preop VA*	*postop VA*	*Follow up (months)*
MM	OD	24	28	H	J	20/100	20/30	52
	OS		25	Phaco S	J	20/400	20/60	49
CE	OD	36	38	H	J	20/100	20/30	12
	OS		36	H	J	LP	20/30	12
VL	OD	39	40	H	J	<20/400	20/25	30
	OS		42	H	J	<20/400	20/25	28
OM	OD	43	45	H	J	20/100	20/25	42
	OS		43	H	J	LP	20/25	44
OJ	OD	17	46	H	J	20/60	20/20	12
	OS		44	H	J	20/60	20/20	14
GN	OD	57	48	H	J	20/200	20/20	50
	OS		50	H	J	20/100	20/20	46
CR	OD	51	62	A	J	20/100	20/30	21
	OS		59	H	J	20/400	20/30	24
VHM	OD	20	60	H	J	20/200	20/40	31
	OS		63	H	J	20/200	20/40	28
VOG	OD	62	63	H	J	20/100	20/20	61
	OS		67	H	J	20/100	20/20	57
BH	OD	45	66	H	J	20/100	20/30	32
	OS		69	A	J	20/80	20/25	29
LR	OD	48	72	H	J	20/60	20/20	30
	OS		65	H	J	20/100	20/20	37

H: heparin coated PMMA IOL (Pharmacia & Upjohn, 811C or 812C); A: Acrysof IOL (Alcon Laboratories Inc, SA60AT or MA30BA), Phaco S: PMMA IOL (PhysIOL SA).J: juvenile, VA: visual acuity

POSTOPERATIVE COMPLICATIONS AND OPHTHALMOLOGICAL FINDINGS AT FOLLOW UP

An overview of postoperative complications and findings during follow up is given in Table 6.

Table 6: *Postoperative complications and ophthalmological findings at follow up*

	PCO	*Glaucoma*	*Endophthalmitis*	*Strabismus*	*Nystagmus*
Group A1 (n = 9)	0	0	0	3 (33%)	1 (11%)
Group A2 (n = 34)	10 (29%)	2 (5.8%)	1 (3%)	10 (29%)	4 (12%)
Group P1 (n = 11)	3 (27%)	0	0	1 (9%)	1 (9%)
Group P2 (n = 22)	15 (68%)	0	1 (5%)	2 (9%)	0

PCO: posterior capsular opacification

in a part attributed to late presentation in 2 patients and low compliance with occlusion therapy in 5 (56%) of 9 children. In the unilateral pseudophakic group (group P1, n=11) 5 eyes (45.5%) attained a visual acuity of 20/60 or more. One child (9%) attained a visual acuity of less than 20/200 fifty-four months after surgery despite good follow-up and compliance to occlusion therapy.

Bilateral cases (n=28)

The mean age at surgery was 4.7 months (2-19 months) for the bilateral aphakic group and 52 months (25-72 months) for the bilateral pseudophakic group. Mean follow-up period was 82.1 months (range 23-140 months). The visual acuity attained for group A2 and group P2 is shown in Table 4 and 5.

In the bilateral aphakic group (group A2, n=17) the final best corrected visual acuity of the worse eye was better than 20/60 in 6 (35.3 %) patients, 6 (35.3%) children attained a visual acuity of 20/30 or better in the best eye. In the bilateral pseudophakic group (group P2, n=11) final best corrected visual acuity of the worse eye was better than 20/60 in 10 patients (90.9%), 7 (63,7%) children attained a visual acuity of 20/25 or more in the best eye.

Opacification of the posterior capsule (PCO) or anterior hyaloid face occurred in 57.6% of children treated with a posterior chamber IOL. Seventy one percent of children who underwent lens extraction with primary posterior capsulotomy without anterior vitrectomy developed PCO compared to 32% where an anterior vitrectomy was performed (Table 7). Ten patients required secondary YAG laser capsulotomy. In 5 of those patients reopacification of the capsulotomy occurred requiring surgical capsulotomy to clear the visual axis. Thirteen patients underwent a secondary surgical capsulotomy when the capsule was thought to be too thick or the child too uncooperative for YAG laser capsulotomy.

In the immediate postoperative period four eyes developed a mild fibrinous uveitis that resolved rapidly and with topical steroids in three eyes. Two eyes developed a more serious inflammation: one eye (OJ, Group P2) developed endophthalmitis five days postoperatively. Topical, intravenous antibiotic therapy and subconjunctival injection of steroids, vancomycin and amukin were necessary to quieten the eye. The treatment was tapered over the next few weeks until the eye became quiet again. Two and a half months later the patient presented with posterior capsular opacification. A surgical capsulotomy was performed with recurrence of the endophthalmitis two months afterwards. We restarted the topical and systemic antibiotic therapy. The endophthalmitis resolved rapidly under this treatment. Another patient (GD, Group A2) developed an endophthalmitis 16 months after cataract surgery. An anterior chamber rinsing combined with a vitrectomy and intraocular antibiotics was performed. Postoperatively a subconjunctival injection with antibiotics and corticosteroids was given, followed by steroid antibiotics eye drops. Both cases were probably suture-related because they resolved under intensive treatment.

Table 7: *PCO after implantation of an IOL in the treatment of pediatric cataract*

	No PCO	PCO	Post-operative months*
No PPC (n = 7)	0	7 (100%)	12,3 (3-19)
PPC - AV (n = 7)	2 (29%)	5 (71%)	10,2 (1-27)
PPC+ AV (n = 19)	12 (63%)	7 (37%)	16,5 (3-29)

PCO: posterior capsular opacification, AV: anterior vitrectomy, PPC: primary posterior capsulorhexis
*: time in months to appearance of significant PCO after surgery

One patient with bilateral aphakia presented with a unilateral iris bombans 8 days postoperatively because of synechiae formation between the iris and the residual lens capsule. The child was treated with a secondary surgical iridectomy and postoperative topical Timoptol 0.5% eye drops. Two bilateral aphakic children developed glaucoma 3,5 and 6,5 years postoperatively and are treated with topical therapy. Cystoid macular oedema or retinal detachment has not been observed so far.

DISCUSSION

Childhood cataract has an estimated prevalence of 1.2 to 2.3 per 10 000 births. In case of bilateral cataracts 1/3 is inherited without systemic abnormalities. Other causes include metabolic disorders such as galactosemia, chromosomal abnormalities such as trisomy 21, intrauterine insults such as congenital Rubella. They can be part of a systemic syndrome such as Hallermann-Streiff-François syndrome and Lowe syndrome. On the other hand unilateral cataracts are not usually inherited or associated with systemic disease; PHPV and posterior lenticonus are frequent. They may occur with

Table 8: *Final monocular visual acuity*

Group	< 20/400	<20/200-20/400	<20/60-20/200	20/20-20/60
A 1 (n = 9)	5 (55.6%)	1 (11.1%)	2 (22.2%)	1 (11.1%)
P 1 (n = 11)	0	1 (9%)	5 (45.5%)	5 (45.5%)
A 2 (n = 34)	1 (2.9%)	1 (2.9%)	14 (41.2%)	18 (52.9%)
P 2 (n = 22)	0	0	0	22 (100%)

A1:unilateral aphakia; A2: bilateral aphakia; P1: unilateral pseudophakia; P2: bilateral pseudophakia

congenital infections, such as congenital rubella. The majority of cases however are idiopathic. Slit-lamp examination is mandatory to exclude asymmetrical bilateral cataracts (28). In all cases of childhood cataract examination of the parents is mandatory.
Our group consists of patients with heterogeneity in the type of cataract, differences in ages at surgery and variable compliance with therapy. Our results are probably representative for the real outcome of surgical treated childhood cataracts as a group (Table 8).

The amblyogenic factors in congenital unilateral cataracts are not only visual deprivation but also unequal competition between the two eyes. The latter becomes important once the visual deprivation is prolonged to 12 to 30 weeks of life (3). Attaining good visual results in unilateral cataract demands early surgery during the first two months, prompt aphakic correction, aggressive amblyopia treatment, and good compliance (2,4,12). None of the children with unilateral cataract in our series was operated before the age of 2 months because of presentation after that age in most of them. Furthermore, we had to cope with significant variations in age of surgery and compliance with therapy, as were Lundvall and Kugelberg (19) who reported similar results. Treating unilateral cataract is a difficult decision that has to be taken with full consent of the parents and in the best interest of the baby (27). The neonates diagnosed with bilateral cataracts are operated as soon as possible with less than one week time between the two eyes. The best corrected visual acuity in the worse eye was 20/60 or more in 35.3% in those infants at a mean age of 6.5 years (range 21 to 114 months). This result is comparable to other reports concerning children with bilateral congenital cataracts without co-morbidity (10,18,26). For juvenile bilateral cataracts visual outcome is more satisfactory. All those infants, who were primarily implanted with an IOL, attained a visual acuity of more than 20/60 in the worse eye, moreover 54,5% attained a visual acuity of 20/25 or more in the worse eye, similar to other reports (6,8,23). Primary IOL implantation in late bilateral cataracts showed a higher incidence of visual acuity better than 20/63 compared to aphakic contact lens correction (17). At our department extended wear contact lenses are used to correct aphakia in cases where no IOL is implanted. In cases of contact lens intolerance we change the contact lenses for aphakic spectacles, followed by bifocal glasses at the age of 3-3.5 year. Implanting IOL's in children aged two years and older is now widely accepted (28). Lambert and Lynn (16) reported that primary IOL implantation in cases of unilateral cataract surgery during the first six months of life results in an improved visual outcome but a higher rate of complications requiring reoperation compared with children treated with contact lenses. IOL implantation is now also accepted as a safe alternative for the treatment of bilateral cataracts under the age of 2 years (23). However we still have to wait to see the long-term effects of this management of bilateral congenital cataracts. In accordance with Joseph and Yair Morad (13) we would like to stress the fact that contact lens use should continue to receive serious consideration as treatment option for paediatric aphakia following cataract surgery, especially in children younger than 2 years old. In this group surgical complication rates following primary IOL implantation may be highest and refractive outcomes most unpredictable because of rapid ocular growth before this age (11).

At our department infants with unilateral cataract aged six months and older and children with bilateral cataracts aged two years and older are always primary implanted with an IOL. Today we increasingly use primary IOL implantation in younger age groups from 6 months onwards, depending on the axial length. There is a general agreement about the fact that primary posterior capsulotomy with anterior vitrectomy is necessary to prevent or delay opacification of the visual axis in children (14,31). In the adult eye the influence of the IOL on the occurrence of PCO is well recognized (7,22), with the newer Acrysof® foldable acrylic IOL (Alcon) resulting in a lower rate of PCO (29,30,33). Ram and associates (25) concluded that the management of the posterior capsule rather than IOL design and material influences the incidence of PCO after cataract surgery in children. Postoperative uveitis can be a harbinger of complications such as cystoid macular oedema, synechia formation, glaucoma, PCO and sometimes corneal decompensation. Endophthalmitis following intraocular surgery in children is not uncommon, a survey of over 500 paediatric ophthalmologists suggested an incidence of 7 per 10 000 cases (32). We had to cope with two cases of serious ocular inflammation; both probably suture-related.

Glaucoma is a well recognized sequela of congenital cataract surgery, but there is still a lack of adequate information regarding its presentation, risk factors and management. The reported prevalence of postoperative glaucoma varies between 6% and 26% of eyes among children operated before, as well as after 1 year of age (20,21,26). The incidence of primary open angle glaucoma seems to be lower in older children undergoing primary IOL implantation following cataract surgery (1) in contrast to infants undergoing similar surgery during the first 4 weeks of life (15,24,34).

CONCLUSION

The overall results of patients with pediatric cataract treated with primary IOL implantation are better compared to those left aphakic.

Bilateral childhood cataracts without associated ocular or systemic disease can attain functional visual results in case of early surgery with attention to the posterior capsule and anterior vitreous, immediate optical correction, intensive occlusion treatment and good compliance. A vision of 20/200 seems to be a realistic goal for the group of patients with unilateral congenital cataract, surgically treated without primary IOL implantation at an average age of 15 months, associated with variable and low compliance. Therefore, early surgery during the first two months, prompt aphakic correction, aggressive amblyopia treatment, and good compliance are necessary to attain good visual result in this group of patients. The decision to treat unilateral cataract is one that has to be taken with full consent of the parents and in the best interest of the baby.

Whether early surgery with primary IOL implantation and improvement of compliance strategy improves the results on long term needs to be further investigated.

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