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CLINICAL STUDY

Bilateral uneven cataracts in children: amblyopia management by sequential intraocular lens implantation

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Abstract

Purpose **To report on amblyopia management of children with bilateral uneven cataracts treated by sequential intraocular lens implantation.**

Methods **Children with bilateral uneven cataracts who needed bilateral cataract surgery were prospectively enrolled in Oxford Eye Center, Johannesburg, South Africa and Southwest Eye Hospital, Chong Qing, China. In the same patient, the amblyopic eye with the denser cataract underwent primary intraocular lens implantation, whereas the better eye was temporarily left aphakic as an alternative to patching. A secondary intraocular lens implantation was performed in the aphakic eye when best-corrected visual acuity in the amblyopic eye attained its best potential.**

Results **Thirteen children were included in this non-comparative study. Average age at surgery was 3.02 ± 1.87 years with an average follow-up period of 9.35 ± 5.23 years. In the amblyopic eyes, 10 out of 13 (77%) had less than 20/120 best-corrected visual acuity before amblyopia treatment. The optical penalization of the dominant eye (temporary aphakia) lasted on average 8.38 ± 4.05 weeks. The best-corrected visual acuity of the amblyopic eye improved to 20/50 or better in six eyes (46%), and ranged from 20/60 to 20/200 in five eyes (38%); in remaining two eyes, the best-corrected visual acuity stayed below 20/200. Best-corrected visual acuity was restored to 20/30 or better following intraocular lens implantations in 12 of the dominant eyes (92%).**

Conclusions **Optical penalization by temporary aphakia of the dominant eye is a convenient means for treating amblyopia in children with bilateral uneven cataracts.**

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Keywords: amblyopia; cataract; optical penalization; aphakia

Introduction

Bilateral developmental cataracts in early childhood are not always equal and may be of uneven density. Children with bilateral uneven cataracts (BUCs) are generally toddlers with developmental cataracts that often present with light perception in the dense cataract eye and navigational vision or better in the fellow eye. Consequently, the eye with the denser cataract becomes amblyopic because of the rivalry that exists due to the disparity between the perceptions from the two eyes. Deprivation amblyopia caused by unilateral congenital cataract has the most potential to cause severe amblyopia.[1–6] Children with BUCs often present with amblyopia that is similar to the unilateral congenital cataract amblyopia. As history is not always available, the depth of amblyopia in the denser cataract eye is unknown until it is assessed postoperatively. The earlier the cataract has existed in the worse eye, the deeper the amblyopia will be.[1–6] Young children with BUCs are less amenable to occlusion therapy than infants with unilateral congenital cataracts because of their age and the stress to parent–child relationships.[7] Furthermore, amblyopia therapy by patching in toddlers with BUCs is

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problematic because of the lens opacities in the less affected, dominant eye. An alternative to patching is to penalize the dominant eye.[8,9,10] To maximize amblyopia treatment, it is possible to consider operating on both eyes and initially only implanting an intraocular lens (IOL) in the weaker eye, leaving the dominant eye temporarily aphakic as a means of optical penalization. Secondary IOL implantation in the dominant eye is deferred for a variable period of time dependent on when the best-corrected visual acuity (BCVA) in the weaker eye shows no further improvement during testing.

To date, the literature has reported on the management of unilateral and bilateral congenital cataracts.[11–21] However, BUCs in children that need bilateral surgery are rare, and the specific issue of amblyopia management in these cases has not yet been reported. Our report addresses this issue and we propose a strategy for the management of amblyopia in children with BUCs.

Materials and methods

From October 1990 to March 2007, children younger than 7 years of age with BUCs were prospectively enrolled in the study. Paediatric cataracts associated with microcorneas (corneas less than 9.5 mm in diameter), microphthalmia (axial length shorter than 18 mm), uveitis, or trauma were excluded. The cataracts were considered surgically operable when a red reflex was non-existent through an undilated pupil in non-verbal children, or BCVA was $\leqslant$20/80 in verbal children. Informed consent was obtained from the parents or the guardians for cataract surgery, and sequential IOL implantation after the positive and negative aspects of this approach, as well as alternative methods for amblyopia treatment have been explained.

The preoperative assessment included a history of the patient and his or her family, VA measurements adapted to the patient's age (preferential looking chart in non-verbal children and Sheridan and Gardiner charts or Snellen chart in verbal children), slit-lamp examination, dilated fundus examination, keratometry, and A- and B-scan ultrasound studies. In young uncooperative patients, some of the preoperative measurements were performed under general anaesthesia before surgery. The IOL power was calculated by the SRK II formula, and aimed at an undercorrection of 20 or 10% in children younger or older than 2 years, respectively.[22,23]

Paediatric cataract surgery was performed in the same manner for all patients by two surgeons (Yu and Dahan) who had several occasions to work together in paediatric cataract surgery sessions worldwide. The two eyes were operated simultaneously on the same day using different surgical sets for each eye. The basic method of paediatric cataract surgery did not change during the period of the study.[15] An anterior chamber maintainer was used during all the operations. Using a limbal approach, a manual or mechanized anterior capsulotomy (according to feasibility) was performed and the cataract was aspirated manually using a cortex aspirating cannulae; a vitrectomy apparatus was used for the posterior capsulotomy and anterior vitrectomy leaving a peripheral capsular annulus. A posterior chamber IOL (one piece PMMA with encircling 'C' loops, overall diameter $\leqslant$12 mm) was implanted in the bag or sulcus in the amblyopic eye (according to the feasibility during the surgery), whereas the fellow eye was temporarily left aphakic. Postoperatively, both eyes were refracted and the BCVA assessed by an experienced paediatric orthoptist. For non-verbal children, preferential looking chart was used. For verbal children, Sheridan and Gardiner cards and/or Snellen charts were used. The residual refractive error was corrected by spectacles in the amblyopic eye, whereas the less affected dominant eye was temporarily left uncorrected as a mean of optical penalization. The parents or guardians were instructed to give mental support and special care to the child during the optical penalization period. The BCVA of both eyes was monitored weekly by the orthoptist. The duration of the optical penalization was individually tailored by the orthoptist in consultation with the surgeon according to the difference in BCVA between the two eyes. The secondary IOL implantation was always performed in the sulcus using the same type of IOL as for the fellow eye. The secondary implantation was scheduled when it was felt that no further improvement in the BCVA could be achieved for the amblyopic eye. Whenever necessary, conventional occlusion amblyopia therapy was carried out after the second IOL implantation until the child reached 12 years of age. Although most authors advise to stop amblyopia therapy by 6–8 years of age, we advised the parents to carry on with occlusion for half an hour a day between 8 and 12 years of age to prevent regression.[24]

Results

Thirteen children (9 boys, 4 girls) with BUCs were included in this non-comparative prospective study. Patient nos. 1–12 were operated by Dahan in Johannesburg, South Africa, and patient no. 13 was operated by Yu in Chong Qing, China. Average age at the time of surgery was 3.0 ± 1.9 years, and the average duration of the follow-up testing lasted 9.4 ± 5.2 years (see Table 1). The preoperative, early postoperative, and the last visit BCVA of the amblyopic eyes, and the duration of the penalization period are listed in Table 2. Only one of the patients had strabismus. Patient no. 13 presented with a variable exotropia of the weaker eye. In the amblyopic eyes, 10 out of 13 (77%) had a BCVA that

Table 1 Summary table of patient demographics, age at the time of surgery, and follow-up period

Patient	*Sex*	*Age at surgery (years)*	*Amblyopic eye*	*Length of follow-up (years)*
1	M	6	R	17
2	M	0.75	L	15
3	M	3	L	15
4	M	2.5	R	14
5	F	0.75	R	13
6	M	1.5	R	12
7	F	2.25	L	11
8	M	5	L	2
9	F	4	L	8
10	F	1.5	R	8
11	M	3	L	4
12	M	6	L	2
13	M	3.5	R	0.5

F = female; L = left eye; M = male; R = right eye.

Table 2 Changes to the best-corrected visual acuity for the weaker eye over the period of treatment for amblyopia

Patient	*Treatment duration (weeks)*	*Best-corrected visual acuity*		
		Preop	*Early postop*	*Last visit*
1	3	20/600	20/60	20/20
2	12	LP	20/20000	20/60
3	3	20/200	20/120	20/25
4	2	20/200	20/120	20/20
5	12	LP	20/20000	20/120
6	8	LP	20/20000	20/40
7[a]	12	LP	20/400	20/200
8	14	LP	20/20000	20/4000
9	8	LP	20/1200	20/50
10	12	LP	20/200	20/50
11	10	LP	20/1200	20/200
12	8	LP	20/20000	20/4000
13[b]	5	20/1200	20/600	20/200

Early postop = within a week after surgery; LP = light perception; Preop = before surgery.
[a]Patient needed an IOL exchange at the age of 7 because of excessive myopization (>7 D).
[b]Patient presented with a variable, exotropia, of the non-dominant eye.

was <20/120 before amblyopia treatment. The optical penalization of the less affected eye (temporary aphakia) lasted on average 8.4 ± 4.1 weeks. For six children (46%), the BCVA improved to 20/50 or better, whereas in five subjects (38%), the BCVA ranged from 20/60 to 20/200; in two remaining children, the BCVA remained below 20/200. The preoperative, early postoperative, and the last visit BCVA for the dominant eyes are listed in Table 3. All of the dominant eyes maintained a superior or equal BCVA when compared with the amblyopic eyes. There were no cases of reversal amblyopia in the better eyes during the optical penalization period. Following

Table 3 Changes to the best-corrected visual acuity for the dominant eye made temporarily aphakic over the period of amblyopia treatment

Patient	*Best-corrected visual acuity*		
	Preop	*Early postop*	*Last visit*
1	20/200	20/30	20/20
2	20/200	20/60	20/20
3	20/120	20/30	20/20
4	20/80	20/30	20/20
5	20/200	20/60	20/20
6	20/200	20/50	20/20
7	20/600	20/60	20/25
8	20/120	20/40	20/20
9	20/1200	20/120	20/30
10	20/400	20/80	20/20
11	20/200	20/60	20/30
12	20/400	20/50	20/30
13	20/400	20/100	20/60

Early postop = within a week after the first operation; Preop = before surgery.

the secondary IOL implantations in the better eyes, BCVA was restored to 20/30 or better in 12 eyes (92%).

None of the patients needed additional surgical intervention, such as secondary posterior capsulotomy/vitrectomy. There were no cases of IOL dislocations, optic capture or eccentric pupils, and none of the patients developed glaucoma or retinal detachment during the follow-up period. The lack of serious complications in our subjects was probably due to the fact that most of the patients were older than 1 year of age, and all the operations were performed by experienced paediatric cataract surgeons. One patient needed an IOL exchange at the age of 7 (5 years after the initial surgery) because of excessive myopization (>7 D) in spite of a 3 D undercorrection at the time of first IOL implantation.

Discussion

Congenital cataracts or cataracts that develop in early childhood account for 5–20% of treatable blindness in children worldwide.[18] Cataracts in childhood not only reduce visual perception but also interfere with normal visual development. Cataracts that cause visual deprivation can potentially cause the most severe degree of amblyopia.[8] Merely removing the cataract is usually insufficient to reverse amblyopia; therefore, effective postoperative amblyopia management is an integral part of restoring a child's vision.[1–3,5–21,25]

Although the patients were recruited during a period of over 17 years, our series is relatively small because of the rarity of the condition. Most of the cataracts in the paediatric age, present either as bilateral and equal, or as unilateral. Hence, a rich literature on the management of bilateral and unilateral paediatric cataracts exists.

Our study of 13 children (26 eyes) with BUCs suggests an alternative approach to the postoperative amblyopia management to that routinely used.[1–3,5–10,25] Optical penalization of the dominant eye was achieved by temporary aphakia, whereas the amblyopic eye was favoured by an IOL implantation. The uncorrected vision in the aphakic eyes was measured as less than counting fingers at one foot (20/4000) during the penalization period. This low vision in the better eye assured a definite advantage of the amblyopic eye over the better eye immediately after the first operation. Amblyopia treatment was effectively enforced continuously throughout the day without relying on compliance with patching. The stress associated with applying a patch over the better eye was avoided. The patients were conveniently left unpatched, whereas the BCVAs for both eyes were monitored by a paediatric orthoptist throughout the optical penalization period. None of the patients experienced reversal amblyopia during or after the penalization period.

In two patients, the BCVAs of the amblyopic eyes only improved marginally to 20/4000 because of deep preexisting amblyopias. We assume that in these two patients, the onset of the cataract in the weaker eye probably occurred during the first 6 months of life causing deep irreversible amblyopia.

Occlusion therapy combined with accurate optical correction is probably the best method to treat amblyopia following cataract surgery.[1–3,5–10,25] Good compliance in patching decreases the duration of required therapy and has been shown to be an important factor in visual rehabilitation,[1,8,9,10] Nevertheless, a patching regimen is difficult to maintain, especially when applied for several years in toddlers.[1,6] Long-term patching therapy is very demanding for the child and the family, and can cause extreme stress to the parents and parent–child relationships.[10] In addition, several authors have found that an overly intensive patching regime hinders the development of binocular function,[5] as well raising other concerns related to psychological and perceptual development of the child.[3,5,7] One limitation of our study is that we have not compared the optical penalization method with occlusion therapy to determine whether patching, and its associated problems, would be a more efficient method of improving VA from the amblyopic eye. As cases of BUCs that need bilateral cataract surgery are rare, establishing a control group was not feasible. In addition, we have not measured binocularity and stereopsis, although none of the patients except one presented with strabismus or developed it during the follow-up period.

Unilateral congenital cataracts pose the biggest challenge in terms of amblyopia treatment mainly because of the strong cortical rivalry induced by the perceptual difference that exists between the inputs from two eyes.[1–6,26] A retrospective study of unilateral congenital cataract by Lundvall and Kugelberg[6] found that only 6 out of 30 patients (20%) achieved VA of 20/200 or better in the cataract-extracted eye. A similar condition exists when bilateral cataracts develop unevenly in early childhood. The eye with the denser cataract normally becomes amblyopic because of the superior visual perception arising from the other eye. Although the degree of amblyopia caused by the dense cataract is uncertain until it has been assessed postoperatively, it is dependent on the age of the child at the onset of cataract formation and the length of visual deprivation. Although comparatively rare, young children with BUCs who need bilateral cataract surgery pose a dilemma in terms of the sequence and the timing of the surgery in both eyes. An alternative and conventional approach to our study would be to first operate the weaker eye only until the vision of the operated eye surpasses that of the fellow eye. This conventional approach was used by the authors before the prospective study on BUCs. Good results were achieved when the preoperative VA in the better eye was lower than the postoperative VA in the amblyopic eye. This approach seems, at first glance, more reasonable because it requires only one operation for each eye, although our method requires two separate operations for the better eye. In other words, the dominant eye is exposed twice to surgical risks. Furthermore, the conventional approach allows in-the-bag primary IOL implantations in both eyes. In our reported method, the secondary IOL implantation in the better eye is mostly limited to the sulcus because by the time of the second operation, the capsular leaflets are already sealed and difficult to separate. However, the preexisting amblyopia in the weaker eye might be so deep that the child might still prefer to use his non-operated dominant eye postoperatively, in which case occlusion therapy is mandatory. Our proposed method avoids patching, and provides an intensive and convenient way to penalize the dominant eye.

An alternative similar optical penalization consists of rendering both eyes aphakic and fitting the child with contact lenses. Amblyopia treatment in bilateral aphakia is conveniently achieved by leaving the contact lens out of the dominant eye for a period of time each day. This method requires excellent compliance and a dedicated setup for paediatric contact lenses fitting.

Bilateral paediatric cataract surgery and IOL implantation during the same session have been reported in the literature,[27] but there are no reports on selective IOL implantation for amblyopia management. Simultaneous bilateral IOL implantation might perpetuate the rivalry between the inputs from the two eyes unless the occlusion therapy is effective and

successful. Although patching is probably the most effective way to improve vision after paediatric cataract surgery, it is difficult to implement in children above 1 year. Most of the patients in our study were toddlers for whom good compliance with amblyopia treatment was uncertain. It is not uncommon for toddlers' parents to quit and abandon occlusion therapy especially in cases of deep and persistent amblyopia.[6,26] Our proposed method enforces an intensive period of optical penalization and ensures follow-up until the secondary implantation in the dominant eye is performed. A certain risk of dense amblyopia developing in the better eye exists in the event of failing to return to a timely follow-up during the penalization period. Fortunately, the parents are usually keen to cooperate and anxious to return for the secondary implantation in the better eye. Recent studies recommend moderate patching or penalization as an alternative to aggressive patching to encourage binocular development.[10] Although our study cannot claim that its method is superior to conventional patching (no control group), the suggested method offers a convenient way to reduce the vision in the better eye in an intensive and continuous manner.

BUCs are relatively rare, but the issue of postoperative amblyopia therapy is similar to unilateral congenital cataracts. Optical penalization by temporary aphakia considerably reduces the VA in the dominant eye, thus providing a valid alternative to patching and a convenient way to enforce an intensive amblyopia therapy. The children were left unpatched during which time the dominant eye's VA was markedly reduced temporarily. The amblyopic eye benefited from full optical correction and no suppression through the better eye throughout the penalization period. This study shows that the temporary aphakia is a practical and convenient way to enforce amblyopia treatment; however, its superiority over the conventional occlusion therapy remains to be proven by further comparative studies.

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