

The Development of Aphakic Glaucoma Following Lensectomy in Congenital Cataract in a NSW Children's Hospital

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ABSTRACT

Congenital cataract affects children and their vision from an early age and as such early diagnosis and treatment is vital. Following surgical lensectomy, children with congenital cataract will either have an intraocular lens inserted, be fitted with an aphakic contact lens or be prescribed aphakic spectacles. One possible complication of lensectomy in

these children is aphakic glaucoma. The aim of this study was to ascertain the prevalence of aphakic glaucoma in children with congenital cataract following lensectomy. A retrospective review of children presenting to the eye clinic at The Children's Hospital at Westmead NSW with congenital cataracts between 2008 and 2010 was performed.

Keywords: Congenital cataract, aphakic glaucoma

INTRODUCTION

Congenital cataract is one of the leading causes of stimulus deprivation amblyopia in infants¹ with an estimated 200,000 children blind from cataracts worldwide.² A congenital cataract is defined as any opacity of the lens that is identified within the first six months of life.^{3,4} Congenital cataracts can present as either bilateral or unilateral and early detection and treatment is vital in achieving adequate visual outcomes. Congenital cataracts have a devastating impact on visual development, and as a result this condition is a priority of the global Vision 2020 initiative.⁵

Treatment of congenital cataract requires early lensectomy to provide a clear optical pathway as well as optimal refractive correction. Congenital cataracts block sensory input to the retina and therefore cause significant stimulus deprivation amblyopia. This creates the need for a management plan that includes both amblyopia treatment and refractive correction. In relation to the refractive correction there are three options including intraocular lens (IOL) implant, contact lens wear or the prescription of spectacles. At The Children's Hospital at Westmead, the preferred refractive correction for aphakia following congenital cataract surgery is contact lenses until the child is older, at which time an IOL will be implanted. Once refractive correction is in place, amblyopia management

is commenced. The duration of occlusion therapy is determined by the amount of amblyopia and age of the child.

Postoperative complications following lensectomy include aphakic glaucoma, posterior capsule opacification, vitreous haemorrhage, inflammation and retinal detachment.⁶ Aphakic glaucoma is the most common long-term complication of congenital cataract surgery⁷ and refers to glaucoma that occurs after congenital cataract surgery. Unlike the name suggests, it can be present in the absence of aphakia when an IOL is inserted. The incidence of aphakic glaucoma varies and has been reported as low as 5% and as high as 41%.⁸⁻¹¹ A reason for the variability in the reported incidence is the duration of follow-up post lensectomy. A longer follow-up period may be associated with a higher incidence of aphakic glaucoma.⁸ A number of risk factors have been implicated, namely microcornea, early surgery and poorly dilated pupils which increase surgical manipulation, inciting more inflammation.⁷

The most common type of glaucoma following congenital cataract surgery is open-angle glaucoma.¹² Acute angle-closure aphakic glaucoma occurs less frequently and usually presents in the early postoperative period, generally within the first six months.¹³

The aim of this study was to ascertain the prevalence of aphakic glaucoma in children with congenital cataract following lensectomy.

METHOD

A retrospective analysis of the medical records of patients with a diagnosis of congenital cataract seen in the eye clinic

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at The Children's Hospital at Westmead over a twenty-four month period between 2008 and 2010 was performed. Ethics approval was granted by the institution for this study.

The data retrieved from medical records included gender, affected eye, age at lensectomy, type of aphakic correction, onset of aphakic glaucoma post lensectomy, intraocular pressure (IOP) measurement prior to medical intervention and treatment options for aphakic glaucoma.

For the purposes of this study, congenital cataract was defined as an opacity of the lens identified in an infant within the first six months of life with surgical intervention prior to twelve months of life. Aphakic glaucoma was defined as having persistent raised IOP of over 22 mmHg requiring medical intervention, without associated cause such as systemic disease or topical steroid use.

RESULTS

A total of 57 patient files with a diagnosis of congenital cataract were reviewed. Of these, nine were excluded from the study as the age at lensectomy was greater than 12 months. In total, 48 patients were included in the study; 25 (52%) were male and 23 (48%) were female.

Of the 48 patients, 69 eyes had a congenital cataract. Unilateral cataracts were slightly more prevalent at 56% (n=27) as compared to 44% (n=21) bilateral cataracts.

Age at Lensectomy

The age at which the lensectomy was performed ranged from 10 days to 12 months with the mean age being 3 months (SD±2.92) (Figure 1).

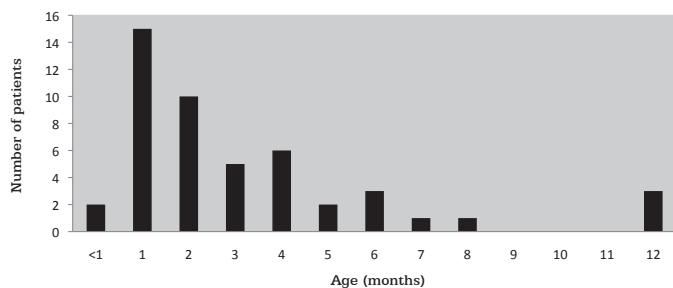


Figure 1. Age of patients at the time of lensectomy.

Twenty-seven patients (56%) underwent lensectomy at less than three months of age. Of the patients with unilateral cataracts, 13 (48%) underwent lensectomy before three months of age. Of the bilateral patients 14 (67%) underwent lensectomy before three months of age. All bilateral cases underwent a bilateral lensectomy (Table 1).

Treatment of Aphakia

Patients undergoing a lensectomy at The Children's Hospital at Westmead had either an IOL implanted at the time of the lensectomy or were prescribed contact lenses or spectacles

Age (months)	Unilateral (N=27)	Bilateral (N=21)
0	1 (3.7%)	1 (4.8%)
1	10 (37.0%)	5 (23.7%)
2	2 (7.4%)	8 (38.1%)
3	4 (14.8%)	1 (4.8%)
4	3 (11.2%)	3 (14.2%)
5	1 (3.7%)	1 (4.8%)
6	2 (7.4%)	1 (4.8%)
7	1 (3.7%)	0
8	1 (3.7%)	0
9	0	0
10	0	0
11	0	0
12	2 (7.4%)	1 (4.8%)

postoperatively. Contact lenses for the correction of aphakia were prescribed in thirty-six patients (75%) and as such was the most prevalent management in the cohort. Eight patients (17%) underwent a primary IOL implant and four (8%) were prescribed glasses (Figure 2).

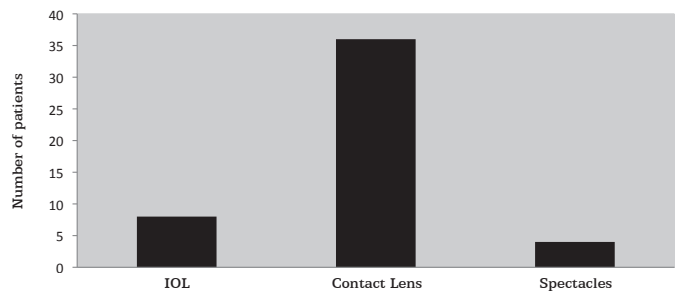


Figure 2. Type of primary correction of aphakia post lensectomy.

Aphakic Glaucoma

Fifteen eyes (22%) were diagnosed with glaucoma post lensectomy. This involved eleven patients, four of whom were bilateral cases.

The time of onset of glaucoma following lensectomy ranged from 14 days to 49 months with the average being 23.7 months (SD±19.8). Five of the sixteen eyes (31%) were diagnosed with acute angle-closure aphakic glaucoma and eleven (69%) were diagnosed with open-angle aphakic glaucoma. At the time of diagnosis IOP measurements ranged from 22 mmHg to 40 mmHg with an average IOP of 29 mmHg (SD±6.2) (Figure 3).

The age of the patients at the time of lensectomy who developed glaucoma was reviewed. Lensectomy in these patients was performed between 10 days and 5 months of age with an average age of 2 months (SD±2.41) (Figure 4).

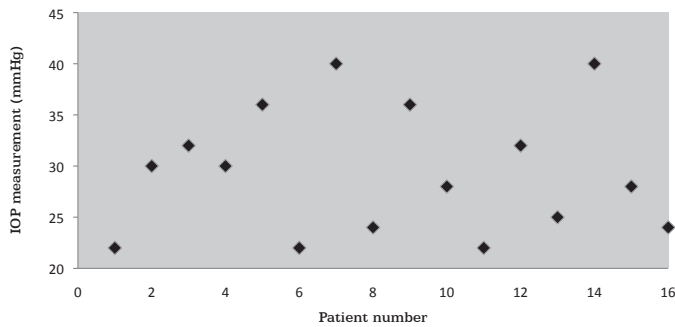


Figure 3. Intraocular pressure measurement in affected eyes at the time of diagnosis of glaucoma.

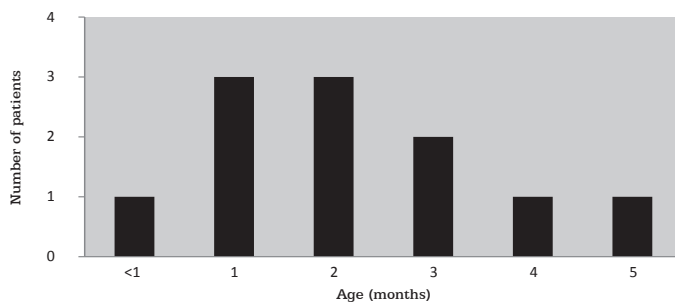


Figure 4. Age at time of lensectomy in those who went on to develop aphakic glaucoma.

Treatment of Aphakic Glaucoma

Treatment methods for glaucoma varied throughout the period of follow-up for each patient. Treatment included both medical and surgical intervention. Medical options included prostaglandin analogs, beta-blockers, carbonic anhydrase inhibitors and combined medications. All glaucomatous eyes were initially treated with medical intervention. Nine eyes (56%) went on to have surgical intervention when IOP could not be controlled by topical treatment. Surgical interventions included Baerveldt drainage tube in seven eyes (44%), cyclodiode laser in one eye and anterior vitrectomy and peripheral iridotomy in one eye. Six eyes (40%) achieved adequately controlled IOP measurements on medical intervention alone.

DISCUSSION

Management of congenital cataracts remains a challenge for health professionals. The timing of surgery, choice of refractive correction and postoperative complications make congenital cataracts a complex disease to manage.

Although advanced techniques of cataract surgery have improved the outcome for these patients,³ the treatment of dense stimulus deprivation amblyopia and postoperative complications remains complex. The risk of developing aphakic glaucoma increases when surgery is performed in the first year of life. However, there is no clinically significant evidence to suggest that delaying lensectomy

within the first year of life reduces the risk.¹⁴ In this group of children 56% underwent lensectomy before three months of age. This corresponds with the literature that suggests early lensectomy at six weeks of age is recommended to ensure a clear visual pathway for the best visual outcome.¹⁵

Whilst aphakic glaucoma is the most common long-term complication of congenital cataract surgery,⁷ the diagnosis of glaucoma may be more difficult in children after congenital cataract surgery. This may be due to these eyes lacking the classic signs of congenital glaucoma, such as buphthalmos, epiphora and blepharospasm. Typically, patients with aphakic glaucoma are asymptomatic.^{9,12} Close monitoring of IOP in children with congenital cataract after lensectomy is required to ensure early diagnosis and treatment.

Aphakic glaucoma requires careful treatment and surveillance as it is associated with a poor visual prognosis.¹⁶ Aphakic glaucoma can develop in the early postoperative period (acute angle-closure glaucoma) or more commonly, many years after cataract surgery (open-angle glaucoma). It is therefore imperative that patients who have had a lensectomy for congenital cataract are followed up long-term. Research that has had a long period of follow-up has reported a higher incidence of aphakic glaucoma.⁸ The period of follow-up in our research was only two years and the incidence of aphakic glaucoma found was 23%. Therefore, it can be hypothesised that with a longer follow-up the incidence of aphakic glaucoma may be higher.

It is significant to note that eight patients in our study received an IOL implant at the time of the lensectomy, and only one of these patients developed aphakic glaucoma. This patient received medical intervention for a postoperative period of two months, at which time the IOP stabilised and medical therapy was withdrawn. This may indicate a more acute, inflammatory increase in IOP rather than true aphakic glaucoma. This finding corresponds to the literature reporting that an IOL may be protective against aphakic glaucoma.¹⁵ Despite the indication that early IOL implant reduces the risk of developing aphakic glaucoma, it is not always the best treatment choice when features such as microphthalmos and persistent hyperplastic primary vitreous are present.

It is noteworthy that treatment of aphakic glaucoma differs from that of other paediatric glaucomas.¹⁷ While the first line of treatment for paediatric glaucoma is surgery,¹⁸ the first line of treatment for aphakic glaucoma is medical management.⁷ Medical treatment should be the first choice of treatment for patients with aphakic glaucoma, followed by surgical intervention if appropriate IOP control cannot be gained. In our study nine of the sixteen eyes with aphakic glaucoma went on to require surgical intervention, with 44% of affected eyes receiving a drainage tube. It has been reported that drainage implant surgery may be more likely to succeed in controlling IOP than other surgical techniques in patients with aphakic glaucoma.¹²

CONCLUSION

Congenital cataracts require early detection and intervention to enable the best patient care and visual outcomes. Early screening can be achieved by the red reflex check on all newborns prior to discharge from hospital. Upon diagnosis of a congenital cataract, immediate assessment with an ophthalmologist is recommended with lensectomy being performed within the first three months of life.

Aphakic glaucoma is a common and serious postoperative complication in congenital cataract patients following lensectomy. Patients who have undergone lensectomy for congenital cataract must be followed up by an eye healthcare professional for the rest of their life, as aphakic glaucoma can present at any time after lensectomy.

Understanding the risk factors of aphakic glaucoma is important in the management of the disease. In our study it was found that late-onset open-angle aphakic glaucoma is more common than acute angle-closure aphakic glaucoma. The incidence of aphakic glaucoma was higher in patients who were left aphakic following lensectomy and in those who had a lensectomy in the first three months of life. These risk factors must be considered when determining the timing of surgical intervention for congenital cataract. Medical management is often the first line of treatment for aphakic glaucoma, however in some cases this is not sufficient to control the glaucoma and surgical intervention is required. This highlights the importance of regular monitoring of all patients who have undergone lensectomy for congenital cataract.

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