ORIGINAL ARTICLES – Clinical science

Long term follow up of primary trabeculectomy for infantile glaucoma

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Abstract

***Background*—The treatment for infantile glaucoma is surgical. Treatment options include goniotomy, trabeculotomy, combined trabeculotomy-trabeculectomy, and trabeculectomy.**

***Methods*—Patients who had a follow up of 5 years or longer after primary trabeculectomy were examined to determine the long term stability in infantile glaucoma.**

***Results*—In eyes with primary infantile glaucoma 92.3% achieved control of their glaucoma with a single trabeculectomy; 100% achieved control with two trabeculectomies; 85.7% of eyes with secondary infantile glaucoma achieved control with a single trabeculectomy. There were no serious complications experienced in either group.**

***Conclusion*—Primary trabeculectomy is a safe and successful operation for infantile glaucoma.**

(*Br J Ophthalmol* 1996;**80**:499–502)

The treatment for infantile glaucoma is surgical. It eliminates the resistance to aqueous outflow created by structural abnormalities in the anterior chamber angle.

This is achieved by internal drainage procedures such as goniotomy, or external drainage procedures such as trabeculotomy or trabeculectomy. Goniotomy provides good results[1–6]; however, it requires a clear cornea (which may be achieved by removing the corneal epithelium in most cases when corneal oedema is present) and a surgeon experienced in this technique. Trabeculotomy also requires a surgeon experienced in the technique, and even then the canal is not found in 11–15% of cases.[7 8]

Trabeculectomy is an easier procedure and more recently has been shown to have good results in infantile glaucoma.[9–12]

In this study we performed primary trabeculectomy in all cases, and present the long term outcome. This is a partial continuation of the report by Burke *et al.*[9] In their series, they reported on the effectiveness of trabeculectomy as a primary procedure including patients from two centres. In this study, only patients who underwent a primary trabeculectomy performed in the Children's Hospital, Temple Street, were included.

Materials and methods

Patients who had a follow up of 5 years or longer after primary trabeculectomy for infantile glaucoma were included in the study. In total there were 20 eyes from 13 patients. The follow up period ranged from 5 years 1 month to 14 years 1 month (mean 7.89 years).

An examination under anaesthetic was performed to confirm the diagnosis of infantile glaucoma when patients presented with the symptoms of epiphora, photophobia, and blepharospasm or the signs of buphthalmos and corneal haze. Anaesthesia was performed with thiopentone induction and maintained with a combination of oxygen, nitrous oxide, and isoflurane.

During anaesthesia, examination included measuring horizontal corneal diameters with Castroviejo calipers, documenting the presence or absence of corneal oedema and Haab striae, measuring intraocular pressures with a Perkins hand held applanation tonometer, and optic disc evaluation by direct and indirect ophthalmoscopy to determine the presence or absence of cupping (a cup-disc ratio of 0.3 or higher was considered suspicious). Associated ocular or extraocular anomalies were noted to differentiate primary from secondary infantile glaucoma.

A trabeculectomy was performed by converting to tracheal intubation using pancuronium, and anaesthesia was maintained by intermittent positive pressure ventilation and isoflurane.

The trabeculectomy was performed by one of two authors (RB or MO'K) using the following technique: a 6–7 mm limbal based conjunctival flap, a 5 × 5 mm lamellar scleral flap, a 1 × 3 mm trabeculectomy with controlled entry into the anterior chamber, and a peripheral iridectomy. The lamellar scleral flap was closed with 8/0 virgin silk (RB) or 10/0 nylon sutures (MO'K). The conjunctiva was closed with a continuous 8/0 plain collagen or Vicryl suture (RB) or 10/0 nylon (MO'K).

Postoperative follow up was based on serial examinations under anaesthetic measuring horizontal corneal diameter and intraocular pressures and optic disc evaluation. Surgical complications were also noted. Examinations

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Accepted for publication
19 January 1996

Table 1 Preoperative details of group 1

Patient no	Age at diagnosis	Eye	Corneal diameter (mm)	IOP (mm Hg)	Symptom	Corneal oedema	Disc cupping
1	4 Months	R	13.5	26	+	+	+
		L	13.5	26	+	+	+
2	5 Months	R	12.0	28	+	+	+
		L	12.5	24	+	+	+
3	5 Years	L	13.0	46	+	+	+
4	18 Months	L	13.5	16	+	+	+
5	4 Months	R	12.5	24	–	+	–
		L	12.5	16	–	+	+
6	11 Months	L	13.5	22	–	–	+
7	4 Months	R	12.0	22	+	+	+
		L	12.5	22	+	+	+
8	5 Months	R	13.5	22	–	+	–
		L	13.5	22	–	+	–

under anaesthesia were carried out at 1 month, 3 months, 6 months, 1 year, 18 months, 2 years, and yearly thereafter until the children were able to be examined in the outpatient department.

Criteria for successful outcome included resolution of corneal oedema, stabilisation or reduction in horizontal corneal diameter, reversal of disc cupping, and an intraocular pressure measurement of 18 mm Hg or less.

Results

A total of 20 eyes from 13 patients underwent primary trabeculectomies for infantile glaucoma. Thirteen eyes had primary infantile glaucoma (group 1) and seven eyes had secondary infantile glaucoma (group 2)—one patient with Rieger's anomaly, two patients with Sturge-Weber syndrome, one patient with Lowe's syndrome, and one patient with Rubenstein-Taybi syndrome who had bilateral glaucoma.

The preoperative details of patients with primary infantile glaucoma are shown in Table 1 (group 1), and the details of patients with secondary infantile glaucoma are shown in Table 2 (group 2).

In group 1, the age at diagnosis ranged from 4 months to 5 years (mean 13.9 months). In group 2, the age at diagnosis ranged from 2 months to 5 years (mean 21.2 months).

In group 1, five out of the eight patients had symptoms of photophobia, epiphora, and blepharospasm. All five of these patients (eight eyes) also had corneal oedema, Haab striae, optic disc cupping, and horizontal corneal diameters of greater than 12 mm (range 12–13.5 mm: mean 12.8 mm). Of the remaining five eyes, four had corneal oedema, Haab striae, and corneal diameters of greater than 12.5 mm (range 12.5–13.5 mm: mean 13 mm). Only one of these eyes was noted to have significant cupping. The remaining eye had a corneal diameter of 13.5 mm and optic disc cupping. Eleven of the 13 eyes had pressures of 22 mm Hg or greater (range 22–46 mm Hg: mean 25.8 mm Hg). The remaining two eyes had pressure of 16 mm Hg, but corneal diameters of 12.5 mm and 13.5 mm and both had corneal oedema, Haab striae, and optic disc cupping.

Table 2 Preoperative details of group 2

Patient no	Diagnosis	Age at diagnosis	Eye	Corneal diameter (mm)	IOP (mm Hg)	Symptom	Corneal oedema	Disc cupping
1	SW	3 Years	R	13.0	26	–	–	–
2	RT	3 Months	R	12.5	26	–	+	–
			L	12.0	26	–	+	–
3	SW	5 Years	R	12.5	26	–	–	+
4	Rieger's	2 Months	L	9.0	13	–	+	+
5	Lowe's	3 Months	R	12.75	24	+	+	+
			L	13.0	22	+	+	+

SW=Sturge-Weber syndrome; RT = Rubenstein-Taybi syndrome.

Table 3 Postoperative details of group 1

Patient no	Eye	Follow up (months)	Reversal of oedema	Reversal of cupping	IOP control	No of procedures
1	R	119	+	+	+	1
	L	119	+	+	+	1
2	R	83	+	+	+	1
	L	84	+	+	+	1
3	L	93	+	+	–	2
4	L	61	+	+	+	1
5	R	63	+	N/A	+	1
	L	63	+	+	+	1
6	L	169	N/A	+	+	1
7	R	74	+	+	+	1
	L	74	+	+	+	1
8	R	91	+	N/A	+	1
	L	92	+	N/A	+	1

N/A=not applicable, because this sign was not present postoperatively.

In group 2, only one of the patients had symptoms of photophobia, epiphora, and blepharospasm. This patient had Lowe's syndrome with bilateral corneal oedema, Haab striae, corneal diameters of 12.75 mm and 13 mm, pressures of 22 mm Hg and 24 mm Hg respectively, and optic disc cupping. Two patients with Sturge-Weber syndrome both had intraocular pressures of 26 mm Hg and corneal diameters of 13 mm and 12.5 mm associated with a port wine stain, in the absence of corneal oedema or Haab striae. Only one had cupping. The patient with Rubenstein-Taybi syndrome had bilateral corneal oedema and Haab striae, with intraocular pressures of 26 mm Hg bilaterally and corneal diameters of 12.5 mm and 12 mm in the right and left eyes respectively. The patient with Reiger's anomaly had a microphthalmic eye with a corneal diameter of only 9 mm. However, the presence of corneal oedema and significant cupping confirmed the diagnosis of glaucoma, even though the pressure was measured at only 13 mm Hg.

The postoperative details of groups 1 and 2 are shown in Tables 3 and 4 respectively.

Table 4 Postoperative details of group 2

Patient no	Eye	Follow up (months)	Reversal oedema	Reversal of cupping	IOP control	No of procedures
1	R	127	N/A	N/A	+	1
2	R	130	+	N/A	+	1
	L	129	+	N/A	+	1
3	R	107	N/A	+	+	1
4	L	66	–	*	–	1
5	R	75	+	+	+	1
	L	75	+	+	+	1

N/A=not applicable, because this sign was not present preoperatively. * denotes that cupping could not be assessed owing to corneal opacification.

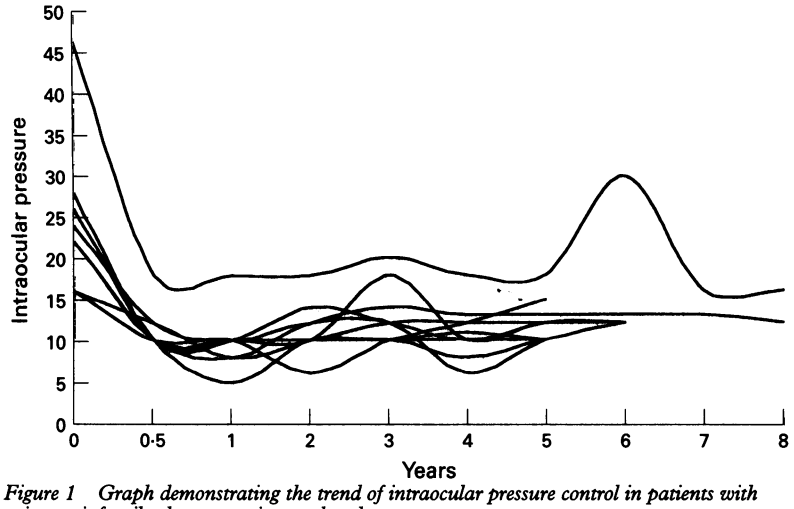

Figure 1 Graph demonstrating the trend of intraocular pressure control in patients with primary infantile glaucoma, since trabeculectomy.

In group 1, the symptoms of epiphora, photophobia, and blepharospasm resolved in all five patients who experienced it preoperatively. Corneal oedema resolved in all 12 eyes that it was present in preoperatively. Reversal of cupping was noted in all 10 eyes that had it preoperatively. An intraocular pressure of less than 18 mm Hg was maintained at all times postoperatively in 12 of the 13 eyes in this group. The other eye was controlled for 6 years, at which stage the intraocular pressure increased to 30 mm Hg and required a repeat trabeculectomy. This eye has maintained an intraocular pressure of less than 18 mm Hg since then (follow up 1 year 11 months).

The intraocular pressure measurements over the follow up period have been demonstrated in Figure 1. The control appears stable, with little variability in the measurements. Failure at 6 years is seen in a single patient. The overall success rate in group 1 was thus 92.3% with a single trabeculectomy, and 100% with two trabeculectomies.

In group 2, the symptoms of epiphora, photophobia, and blepharospasm resolved in the one patient who experienced them. Corneal oedema resolved in four of the five eyes that had it preoperatively. The remaining eye had chronic corneal oedema and as a result, developed an opaque cornea. Reversal of cupping occurred in three out of the four eyes that had it preoperatively. The remaining eye developed an opaque cornea and so the disc could not be evaluated. An intraocular pressure of less than 18 mm Hg was maintained at all times in six of the seven eyes in this group. The remaining eye had intermittent pressure rises. This was the microphthalmic eye with Rieger's anomaly, that developed an opaque cornea secondary to chronic corneal oedema. This eye lost perception of light but was comfortable and so no further procedures were undertaken. The overall success rate in group 2 was thus 85.7% with a single trabeculectomy.

Other than hyphaema, there were no serious operative or postoperative complications noted in either group. Of note, there were no flat anterior chambers, no vitreous loss, no cataracts, no subluxed lenses, no retinal detachments, and no cases of endophthalmitis.

Discussion

Goniotomy was first described in 1893 by Carlos de Vincentiis.[13] It was not popularised until 1938 when Barkan redescribed it as an operation for infantile glaucoma.[14-16] Since then it has remained one of the operations of choice, with good results in a previously untreatable disease.[1-6] When performed with direct visualisation of the angle, successful control has been reported in various series from 81–100% with one or multiple procedures.[1-6] However, in up to 50% of cases there may be significant corneal oedema obscuring an adequate view of the angle structures. In this study, 85% of patients had corneal oedema and as a result many would have been unsuitable for goniotomy without epithelial debridement. Recurrence rates of up to 34%[17] may also necessitate the need for multiple procedures.

Trabeculotomy ab externo was described by Burian[18] and Smith[19] independently in 1960. It is reported to give similar results to goniotomy, with the advantage that corneal clarity is not necessary. It is thus a suitable operation for a larger percentage of patients. However, subsequent failure has resulted in a successful outcome in only 51% at 24 months.[8] This is thought to be due to overgrowth of the trabecular tissue covering the trabeculotomy site.

Trabeculectomy was introduced as an operation for glaucoma by Cairns in 1968.[20] However, it has not been favoured in infantile glaucoma following poor results by Beauchamps and Parks.[21] They reported a 50% success rate with a high rate of complications. In their series, all the eyes undergoing trabeculectomies had either undergone previously failed goniotomies or had complicated secondary glaucomas. More recent series have shown more promising results when trabeculectomy is used as a primary procedure,[8 11] without a high incidence of serious complications. In this study, the success rate was 92.3% in eyes with primary infantile glaucoma, and 85.7% in eyes with secondary infantile glaucoma with a single trabeculectomy. There were no serious complications and the control has been maintained over a period of at least 5 years (mean follow up: 7.89 years, range: 5 years 1 month to 14 years 1 month). This suggests the long term stability of control following trabeculectomy. It has previously been reported that the majority of failed trabeculectomies for infantile glaucoma occur in the first year after surgery.[8] In this study, there were two failed procedures. The first was a patient with primary infantile glaucoma who failed 6 years after his trabeculectomy. He underwent a second trabeculectomy, which has to date given him good control (follow up 1 year 11 months). The second was a patient with secondary infantile glaucoma associated with Rieger's anomaly and microphthalmia. She had intermittent intraocular pressure rises from 6 weeks after surgery, but underwent no further procedures because she developed an opaque cornea with iris adhesions.

The age of diagnosis has also been shown to be an important prognostic factor in infantile

glaucoma. A poor prognosis appears to be associated with patients presenting before 2 months or after 2 years of age.[22] In this study, seven out of eight patients with primary infantile glaucoma were diagnosed between 4 and 18 months. The other patient was diagnosed at the age of 5 years. This was the patient who developed late failure of trabeculectomy. This would support the poor prognosis associated with later presentation.

All patients in this study were white. This tends to be associated with a better prognosis for trabeculectomies of all age groups. This may also explain why the results in all previously published series have varied.

Elder[23] reported on combined trabeculotomy-trabeculectomy, demonstrating a success rate of 93.5% for the combined procedure over a 2 year period in primary infantile glaucoma. This compares similarly to 'trabeculectomy only' in our patients.

Infantile glaucoma has been considered as having a poor prognosis following trabeculectomy. This may have been due to the fact that trabeculectomy has been restricted to cases in which there has been multiple failed procedures. Russell-Eggitt stated that young age is a significant negative factor in bleb survival.[24] Miller reported that the risk of relapse in eyes having undergone trabeculectomy in childhood is significant especially if antiproliferative agents are not used.[12] The use of antimetabolites as an adjunct to trabeculectomy has improved the success rate of surgery in patients at high risk for failure. However, many serious complications have been described following their use.[25] Our results suggest that it is not necessary to use antimetabolites for primary trabeculectomies in infantile glaucoma.

The results of this study demonstrate that primary trabeculectomy is a successful operation for infantile glaucoma when performed in a tertiary referral unit.

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