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Primary trabeculectomy in congenital glaucoma

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SUMMARY The reported success rates in the treatment of congenital glaucoma with goniotomy, trabeculotomy, and trabeculectomy suggest that trabeculectomy should be performed if the other procedures fail. We propose that the decision to perform primary trabeculectomy in primary and secondary congenital glaucoma reduces the effect which the many variable findings in surgical anatomy may have on the outcome of other procedures. This is a retrospective study of the results of primary trabeculectomy in 21 consecutive eyes of 15 patients with congenital glaucoma. Eighteen of 13 patients' eyes were controlled after a single trabeculectomy and remained controlled after a mean follow-up of 3·9 years (range 1·5 to 6·7 years). The role of primary trabeculectomy in congenital glaucoma merits further consideration.

Congenital glaucoma is a major cause of visual handicap in children and is responsible in some series for 2·5 to 10% of all children registered blind.[1] Its pathogenesis is disputed[2] but may be a variable embryological defect in the development of the anterior chamber[3] leading to glaucoma of all degrees of severity. The management of congenital glaucoma is primarily surgical, with goniotomy or trabeculotomy being the accepted procedures of choice.[4–7] While this treatment is effective, a delay in control of intraocular pressure is not uncommon, as patients regularly require more than one operation to achieve satisfactory intraocular pressure control.[2 7]

Some surgeons consider the results of primary trabeculectomy in children to be less satisfactory[1 8]: hence its general use as a secondary procedure when goniotomy or trabeculotomy are contraindicated or have failed. The purpose of this study was to evaluate the effectiveness of trabeculectomy as the primary procedure in 15 consecutive cases (21 eyes) of congenital glaucoma.

Patients and methods

Fifteen consecutive cases (21 eyes) of childhood glaucoma seen consecutively between 1980 and 1985 at our unit were studied. All observations were recorded by either one or both authors. The follow-up period after trabeculectomy ranged from 18 months to 6·7 years, mean 3·9 years.

The ocular family history, age of onset, and presenting signs and symptoms were collated. Corneal diameters, the cup-disc ratio, gonioscopy findings, refractive errors, the intraocular pressure, visual acuity, and ocular motility were recorded where possible before and after treatment. Visual acuities in patients too young for assessment of numerical values were extrapolated from oculomotor fixation patterns.[9] Anisometropia was defined as a spherical difference of $\geq$2·0 dioptres and/or a cylindrical difference of $\geq$1·50 dioptres. Horizontal corneal diameters were measured under general anaesthesia by means of Castroviejo calipers, and values greater than 10·5 mm at birth or 11·5 mm at 1 year were considered suspicious. Cup-disc ratios were evaluated by direct and indirect ophthalmoscopy where possible, and ratios of 0·3 or higher were considered suspicious. Cycloplegic refractions were performed and ocular motility was assessed by an orthoptist. Intraocular pressures were measured under general anaesthesia by means of a Perkins hand-held applanation tonometer and with the slit-lamp applanation tonometer when children were old enough to tolerate this technique. Intraocular pressures of $\leq$16 mmHg in the first year and <18 mmHg up to 3 years of age under general anaesthesia were considered normal[10 11] in the absence of other signs of overt disease or of disease progression.

All suspicious cases were initially examined under anaesthesia (EUA) and trabeculectomy done when

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Table 1 *Summary of age, operative procedures, intraocular pressure, and corneal diameter in group 1*

Case no	Uni/ bilateral	Age (yr) at and type of surgery	Follow-up Postop. (yr)	Intraocular pressure (mmHg)						Corneal diameter preop. (mm)	
				Preop		1 month postop.		Now			
				R	L	R	L	R	L	R	L
1	B	0·4 R trab. 0·4 L trab.	2·2 2·2	26	26	5	5	10	10	13·5	13·5
2	B	0·7 R trab. 0·7 L trab.	6·7	25	25	10	9	8	5	12·5	12·5
3	B	0·3 R trab. 0·3 L trab.	2·5 2·5	21	24	<10	<10	<10	<10	12·5	13·0
4	U	0·6 L trab.	1·5	11	28	12	10	15	12	11·0	12·5
5	B	0·5 R trab. 0·5 L trab.	2·7 2·7	18	18	12	10	10	10	12·5	13·0
6	U	2·2 L goniot. 12·2 L trab.	5·4	18 12	36 32	16	15	16	17	12·5	15·0
7	U	0·5 R trab.	6·3	25	12	12	12	12	13	12·0	10·5
8	U	0·9 L trab.	6·4	9	22	9	10	15	13	11·5	13·5
9	U	1·2 L trab.	5·4	13	30	13	8	14	10	11	13
10	B	0·8 L trab. R trab. 1·0 L goniot.† R+L trab. R+L ccryo. 2·1 L ccryo.‡	2·4 1·1	20 13 17	20 18 19	13 13 	10 10 	 18	 17	14·5 15·0	14·5 15·0§

*Trab.=trabeculectomy. †Goniot.=goniotomy. ‡Ccryo.=cyclocryotherapy. §Corneal diameter now.

indicated. In bilateral cases the second eye was operated on at a second sitting, usually one week later. Serial EUAs were subsequently performed one- to four-monthy until the patient was old enough to tolerate a reliable outpatient assessment.

The anaesthesia was carried out by the same anaesthetist in the great majority of procedures. EUAs were performed with thiopentone induction and maintained with a combination of oxygen, nitrous oxide, and isofluorane. Trabeculectomy was performed following conversion to tracheal intubation using pancuronium and maintained by intermittent positive pressure ventilation and isofluorane.

One of us (RB) performed a trabeculectomy using a 6–7 mm limbal-based conjunctival flap followed by a 5 mm×5 mm shallow lamellar scleral flap. A 1mm×3mm trabeculectomy was performed with a slow controlled entry into the anterior chamber to prevent rapid loss of pressure, followed by a peripheral iridectomy. Viscoelastic material was used to deepen the anterior chamber in the last four cases. The lamellar scleral flap was resutured tightly with five 8-0 virgin silk sutures and the conjunctiva closed with a running 6-0 plain collagen or 6-0 Vicryl suture which included Tenon's capsule. In two of the early cases, where Schlemm's canal was identified at the anterior ends of the radial incisions, a trabeculotomy was also performed with the Harms double trabeculotomy probe.

The criteria for surgical success were stable or improved optic disc appearance, resolution of corneal oedema, lack of evidence of further corneal enlargement disproportionate to normal growth, stabilisation of refractive error, and average intraocular pressures (IOP) of 18 mmHg or less in the absence of other signs of decompensation.

Results

The 15 cases (10 males, five females) were divided into two groups. Group 1 had 10 patients (seven males, three females; 15 eyes) with primary infantile glaucoma and group 2 (glaucoma associated with congenital anomalies) consisted of five patients (six eyes), four with Sturge-Weber syndrome and one with Rubinstein-Taybi syndrome. The main presenting symptoms, listed by frequency, in group 1 were hazy cornea (seven cases), enlarged corneal diameter (five cases), photophobia (four cases), epiphora (three cases); whereas in group 2 three patients presented with an enlarged corneal diameter, one with a hazy cornea, and one case was referred for eye assessment because of Sturge-Weber syndrome.

In group 1 the intraocular pressure was normal in nine of 10 patients (13 of 15 eyes) on examination under anaesthesia one month after primary trabeculectomy (Table 1). This was associated with clearing of corneal oedema and reversal of optic disc cupping. From the outset none of these nine cases required any other form of intervention, after a mean follow-up of

Table 2 *Summary of age, operative procedures, intraocular pressure, and corneal diameter in group 2*

Case no	Uni/ bilateral	Age (yr) at and type of surgery	Follow-up Postop. (yr)	Intraocular pressure (mmHg)						Corneal diameter preop. (mm)	
				Preop		1 month postop.		Now			
				R	L	R	L	R	L	R	L
11	U	3·3 R trab.	2·9	24	10	13	12	13	14	13	11·5
12	U	4·1 L trab.	2·3	16	25	17	10	16	12	12	12·5
13	U	1·8 R trab/t-ot.*	5·7	27	11	14	8	12	10	13	11·5
14	B	0·4 R trab.	3·2	26	18	<10	<10	10	10	12·5	12·5
		L trab.	3·2								
15	U	7·4 L trab.	3·3	16	26	14	16			12·5	12·5
		8·3 L trab/t-ot.*		18	32	16	22				
		9·5 L trab revision		16	30	15	20				
		10·4 L Yag laser	0·5	16	29			16	21		

*Trabeculectomy/trabeculotomy.

4·1 years (range 1·5 to 6·7 years). One patient (case 10) required two trabeculectomies in each eye over four months followed by left goniotomy and later repeat cyclocryotherapy to both eyes. The intraocular pressure became normal for short periods in both eyes after each procedure but then gradually became decompensated. The intraocular pressure and other parameters remained stable one year after cyclocryotherapy. This case had severe disease at the initial operation (age 9 months) as shown by horizontal corneal diameter of 14·5 mm in each eye, gross corneal scarring, and gross distortion of limbal anatomy. An obvious Barkan's membrane was not seen in any of these 10 patients.

In group 2 the intraocular pressure was normal in four of five patients (five eyes) on examination under anaesthesia one month after primary trabeculectomy (Table 2). None of these eyes have since become decompensated after a mean follow-up of 3·5 years (range 2·3 to 5·7 years). One patient (case 15) with Sturge-Weber syndrome who was referred aged 7·3 years with left unilateral glaucoma proved difficult to control. This patient required four operations before satisfactory control was attained, which included a primary trabeculectomy, a trabeculectomy/ trabeculotomy, trabeculectomy revision, and YAG laser division[12] of the fibrous bleb wall.

No immediate or late surgical complications were noted among the 18 eyes controlled after a single trabeculectomy except for a small hyphaema in three eyes, which cleared completely within three days of surgery. In particular, no patients developed scleral ectasia, staphyloma, or vitreous loss.[8] Two patients (cases 7 and 12) developed slight superior pupil peaking secondary to localised peripheral anterior synechiae between the iris periphery and the posterior edge of the draining fistula; however, aqueous outflow was clinically unimpaired. This complication occurred spontaneously in one case and developed in the other (case 7) following blunt trauma associated with a small hyphaema. There was documented reversal of optic-disc cupping (total or partial) in 10 of 11 eyes in whom media opacities did not preclude a view of the optic disc prior to surgery, while the cup-disc ratio of the 11th eye (case 6) was unchanged. The corneal diameters and refractive error of all 18 eyes stabilised; none became increasingly myopic on follow-up.

There were two patients (cases 10 and 15, three eyes) whose intraocular pressures were initially controlled after surgery but subsequently decompensated (case 10 within two months, case 15 after nine months). The cup-disc ratio, intraocular pressure, and refraction were stable after a total of 12 procedures to three eyes.

The relative incidence of anisometropia, strabismus, and the final visual acuity was assessed for both groups of patients (Tables 3 and 4). In group 1 four of 10 patients had anisometropia, three of 10 patients had an exotropia, and 12 of 15 affected eyes had a visual acuity equivalent to 6/24 or better.

In group 2 three of five patients had anisometropia and three of five had strabismus (esotropia two cases, exotropia one case), while four of six affected eyes had a visual acuity equivalent to 6/24 or better. The visual acuity in one patient (case 12) was indeterminable owing to severe mental retardation.

Discussion

Of the 10 patients in group 1 nine (13 eyes) had control of IOP within one month of primary trabeculectomy and have not become decompensated after an average of 4·1 years (range 1·5 to 6·7 years). None of these nine patients suffered serious ocular complications, and all but one has a visual acuity equivalent to 6/24 or better, despite the presence of anisometropia in three, and strabismus in two cases

Table 3 *Summary of ocular findings in group 1*

Case no.	*Uni/bilateral*	*Strabismus*	*Anisometropia*	*Haab striae*	*Visual acuity*	
					R	*L*
1	B			++	6/12	6/12
2	B			++	CSM	CSM*
3	B			++	6/12	6/12
4	U		+		CSM	CSM
5	B	+‡			CSM	CSM
6	U		+		6/5	6/9
7	U	+	+	+	6/24	6/6
8	U				6/6	6/6
9	U	+	+	+	6/6	4/60
10	B			++	UC US UM	UC US UM†

*CSM=central, steady, maintained. †Uncentral, unsteady, unmaintained. ‡Exophoria for near, exotropia for distance.

(Table 3). Our good visual results can be attributed to the prompt optical correction of ametropia and anisometropia, and early control of intraocular pressure, as well as the vigorous treatment of potential ambylopia.[13]

It is generally acknowledged that the treatment of primary infantile glaucoma is surgical. The accepted procedures of choice are goniotomy provided there is a clear cornea and trabeculotomy ab externo for a hazy cornea.[1] It is generally considered that both procedures are comparable,[14] and success rates of 80% and over have been described.[4-7] However, surgical control with either technique warrants more than one procedure in a significant number of patients.[2 7] Trabeculectomy is not a first line procedure in primary infantile glaucoma, since it is claimed to be associated with a higher incidence of complications and a lower incidence of success in reducing the intraocular pressure to normal.[2 8] Beauchamp and Parks described an overall success of 50% in controlling IOP, which was reported as similar to previous experience.[8] Our surgical success rate of 87% after a single procedure is higher than most reported series on goniotomy or trabeculotomy. Rao and colleagues report a 75% success rate following primary trabeculectomy.[15] Our cases differed from those of Beauchamp and Parks in that all but one had trabeculectomy prior to 1·5 years old, whereas the patients in their series were generally older and hence more likely to have other ocular sequelae of buphthalmos, which may influence the outcome.[16]

There were five patients in group 2; four had Sturge-Weber syndrome and one had Rubinstein-Taybi syndrome. Of these patients four (five eyes) had a normal intraocular pressure within one month of trabeculectomy and have not become decompensated since then (mean follow-up 3·5 years; range 2·3 to 5·7 years). The visual acuity is better than or equivalent to 6/18 in these patients (case 12, indeterminable) despite the presence of anisometropia in two and strabismus in two (Table 4).

Glaucoma develops in 30% of patients with Sturge-Weber syndrome. It is unilateral in 90% of cases,[17] and the treatment is similar to that for other cases of congenital glaucoma except that goniotomy, the suggested primary surgical procedure, yields unsatisfactory results.[17] Our results with trabeculectomy were very encouraging and without complications, though numbers were small.

Apart from two patients (three eyes) in our series in whom trabeculectomy failed, complications were

Table 4 *Summary of ocular findings in group 2*

Case no.	*Systemic diagnosis*	*Uni/bilateral*	*Strabismus*	*Anisometropia*	*Haab striae*	*Visual acuity*	
						R	*L*
11	Sturge-Weber syndrome	U		+		6/9	6/6
12	Sturge-Weber syndrome	U	+		+	*	*
13	Sturge-Weber syndrome	U				CSM	CSM
14	Rubinstein-Taybi syndrome	B	+	+	+	6/18	6/6
15	Sturge-Weber syndrome	U	+	+		6/6	6/9

*Indeterminable (severe mental retardation).

minimal. We share the view of Anderson[14] that the response to surgery in a given case is largely governed by the underlying nature of the case. In case 10 the symptoms dated back to the first weeks of life, but diagnosis was delayed, and the child presented at 9 months with permanent corneal scarring and horizontal corneal diameters of 14·5 mm and grossly distorted limbal anatomy. Case 15 presented with Sturge-Weber syndrome and developed a prominent bleb in contrast to the majority of our cases, which had a diffuse or slightly elevated bleb. It has been proposed that prominent bleb formation may be associated with an increased risk of late failure of trabeculectomy.[18]

Though maligned, primary trabeculectomy reduces the effect which the many variables in the anatomy of the anterior segment have on the outcome of other surgical procedures.[19] Eighteen of 21 eyes were surgically controlled after one procedure and have not relapsed. The gloomy picture of paediatric trabeculectomy surgery resulting in inadequate control of intraocular pressure and poor visual outcome has been refuted by our experience. We believe that the role of trabeculectomy as the primary surgical procedure, particularly in cases with primary congenital glaucoma, accompanied by vigorous treatment of potential ambylogenic factors merits further consideration.

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