Retinal detachment in congenital glaucoma

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SUMMARY This paper reports the occurrence of retinal detachment in 18 patients suffering from congenital glaucoma. Difficulty in establishing control of the glaucoma and the presence of high myopia were common findings. The poor results of detachment surgery were related to opacities in the media preventing evaluation of the retinal details and the development of massive preretinal retraction.

Detachment of the retina complicating congenital glaucoma was first described by Axenfeld,¹ who considered that the detachment was often the determining factor in the production of blindness in this condition. Knapp² also reported this development in an 11-year-old child with advanced anterior segment changes of congenital glaucoma; he considered that degenerative changes within the vitreous were implicit in the development of retinal detachment.

Cases of congenital glaucoma complicated by retinal detachment have also been mentioned within surveys of juvenile detachment.³⁻⁵ In these series a total of 11 cases of congenital glaucoma were encountered among 672 cases of retinal detachment within the first 2 decades of life.

This report concerns 18 patients with retinal detachment associated with congenital glaucoma. The clinical characteristics of these detachments and the salient features of the associated glaucoma are described together with the results of retinal detachment surgery. The incidence of retinal detachment in a closely studied series of patients with simple congenital glaucoma is also reported.

Patients and methods

We reviewed the clinical records of 189 patients with congenital glaucoma involving 290 eyes who were treated at Moorfields Eye Hospital between 1960 and 1979. Thirteen eyes of 11 patients suffered a retinal detachment. In addition 7 patients presented during this same period with retinal detachment in association with congenital glaucoma which had been managed elsewhere.

The glaucoma was classified as either simple or complicated. Simple congenital glaucoma refers to

cases which do not have any other developmental anomaly, either ocular or systemic, and show typical gonioscopic appearances.⁶ Complicated congenital glaucoma refers to cases in which glaucoma is related to other developmental defects of the eye with or without extraocular abnormalities.

A detailed profile of each patient was constructed from the clinical records, and particular attention was directed to the following factors: age of onset of glaucoma; family history of glaucoma or retinal detachment; glaucoma surgery undertaken and the control of intraocular pressure achieved; the maximum corneal diameter recorded and the refractive error; the age at which the patient developed retinal detachment, the type and extent of the detachment, and the nature of retinal breaks and other retinal abnormalities encountered; the retinal status of the fellow eye; retinal surgery undertaken and the results achieved.

Results

Retinal detachment occurred in 21 eyes of 18 patients with congenital glaucoma of varying aetiology. There were 13 males and 5 females. The details of the glaucoma and the principal features of the retinal detachments are presented in Table 1.

Congenital glaucoma was classified as simple in 16 and complicated in 2 of our patients. The age of onset of glaucoma ranged from 3 weeks to 12 months and was bilateral in 16 and unilateral in 2 cases. Glaucoma surgery was undertaken in 18 eyes of 16 patients, including filtering procedures in 10 eyes. Surgery was not performed on 1 eye with simple glaucoma which had undergone spontaneous arrest and both eyes of a patient with bilateral Sturge-Weber syndrome. Satisfactory control of the glaucoma was achieved in 10 eyes of 11 patients.

The interval between the onset of glaucoma and

Table 1 Clinical details of congenital glaucoma and retinal detachment in 18 patients

Case	Onset of glaucoma and classification	Glaucoma surgery	IOP control	Diameter cornea (mm)	Refractive error Dsph eq	V/A	Age at detachment	Presentation	Features of detachment	Surgery	Results	Associated findings
1, M L	5 Months simple	G × 3	Poor	15	-12.50	HMs	6 Yr	Hypotension	Total/no breaks	Inoperable	—	Cataract
		TR × 1 G × 3	Fair	15	-11.00	6/36	—	—	—	—	—	—
2, F L	4 Months simple	G × 3	Good	14	-16.00	6/60	—	—	—	—	—	Amblyopia
		G × 3	Good	14	-19.00	6/36	7 Yr	Visual loss	Temporal half round holes and multiple disinsertions 80°	1. Local explant 2. Encirclement 3. Pars plana vitrectomy	Failed MPR	—
3, M L	6 Months simple	G × 1	Poor	14.5	-7.00	6/60	8 Yr	Vitreous haemorrhage	Total/no breaks	Enucleation	—	Rubeosis iridis bullous keratopathy
		G × 1	Good	13	-9.50	6/9	—	—	—	—	—	—
4, M L	2 Months simple	G × 4 T × 1 C × 1	Poor	13.5	-2.50	6/60	6 Yr	Hypotension	Total/no breaks	Inoperable	—	Cataract/luxation
		G × 2 C × 1	Poor	13.5	-3.50	CFs	9 Yr	Hypotension	Total/no breaks	Inoperable	—	Cataract/luxation
5, M L	4 Months simple	G × 3	Fair	14	-15.00	3/60	2 Yr	Hypotension	Total/no breaks	Encirclement	Failed MPR	Cataract
		G × 4	Fair	13	-20.00	6/60	5 Yr	Hypotension	Total/360° giant tear	Inoperable	—	—
6, M L	6 Months simple	G × 3	Fair	16	-11.00	6/60	8 Yr	Hypotension	Total/no breaks	Inoperable	—	Cataract
		G × 1	Good	14	-9.00	6/12	—	—	—	—	—	—
7, M L	1 Month simple	G × 3	Poor	14.5	-4.00	—	8 Mth	Hypotension	Total/no breaks	Inoperable	—	Corneal scarring cataract
		G × 2 S × 1 CA × 1	Poor	14.5	Not known	PL	—	—	—	—	—	Bullous keratopathy
8, F L	10 Months simple	G × 3	Poor	13	-8.00	6/60	—	—	—	—	—	—
		S × 1 G × 2 S × 1 T × 1	Poor	13	-4.50	CFs	14 Yr	Postop finding	Total/no breaks choroidal detachment	Inoperable	—	—
9, F L	2 Months simple Normal	G × 3 T × 1	Fair	13.5	-9.00	6/60	14 Yr	Visual loss	Total/upper temporal U tear	Encirclement and local explant	Failed MPR	Amblyopia
		—	—	11.5	-0.50	6/6	—	—	—	—	—	—

G = Goniotomy. TR = Trephine. T = Trabeculectomy. C = Cyclocryotherapy. CA = Cycloanaemisation. S = Scheie's thermosclerostomy. I = Iridencleisis.

Table 1 Continued

Case	Onset of glaucoma and classification	Glaucoma surgery	IO P control	Diameter cornea (mm)	Refractive error Dsph eq	VA	Age at detachment	Presentation	Features of detachment	Surgery	Results	Associated findings
10, M	R 1 Month simple	G × 3	Fair	14.5	Not known	—	—	—	—	360° prophylactic cryotherapy	—	Corneal scarring Cystoid degeneration of ora serrata Lens subluxation
L		G × 2	Fair	13.5	-8.00	—	4 Yr	Visual loss	Total/270° giant dialysis	Encirclement, pars plana vitrectomy, silicone oil	Upper half reattachment	
11, M	R 3 Weeks simple	T × 1	Fair	13	Not known	—	3 Yr	Hypotension	Total/no breaks	1. Encirclement 2. Silicone oil	Success	Corneal scarring
L		T × 1	Poor	13.5	Not known	—	—	—	—	—	—	Bullous keratopathy
12, M	R 3 Months Axenfeld's syndrome	G × 4 S × 3 C × 1	Poor	14	-10.00	HMs	10 Yr	Hypotension	Inferior half single U tear	Local explant	Failed MPR	—
L		G × 2 T × 1	Poor	15	-2.50	6/60	—	—	—	—	—	—
13, F	R 2 Months bilateral Sturge-Weber syndrome	Nil	Good	13.5	+1.50	6/60	6 Yr	Vitreous haemorrhage	Total serous	Silicone oil	Failed I/O haem. Success	Choroidal haemangioma
L		Nil	Good	13.5	+1.50	6/60	6 Yr	Vitreous haemorrhage	Total serous	Silicone oil	—	Choroidal haemangioma
14, F	R Not known simple	Nil spont. arrest	Good	14	+11.50	CFs	24 Yr	Visual loss	Temporal half single U tear	Encirclement	Failed	Aphakia
L	Normal	—	—	11.5	+0.50	6/12	—	—	—	—	—	—
15, M	R 12 Months simple	I × 1	Good	13.5	-10.00	6/60	14 Yr	Visual loss	Temporal half 90° giant tear	1. Local explant 2. Silicone oil	Failed MPR	—
L		I × 1	Good	14	-8.00	6/60	—	—	—	—	—	Amblyopia
16, M	R 2 Months simple	I × 1 CD × 1 I × 1	Fair	14	-8.00	6/60	32 Yr	Visual loss	Total 180° giant dialysis	Scleral resection	Failed MPR	—
L			Good	13.5	-8.00	6/24	—	—	—	—	—	—
17, M	R 8 Months simple	G × 2	Good	13	-2.50	6/12	—	—	Flat U tear	Prophylactic cryotherapy	—	—
L		G × 1 S × 1	Fair	13.5	-10.00	CFs	8 Yr	Visual loss	Total/no breaks	Encirclement	Failed	Amblyopia
18, M	R 4 Months simple	I × 1	Good	13.5	+11.00	CFs	—	—	—	—	—	Aphakia
L		I × 1	Good	13	+5.00	6/36	33 Yr	Visual loss	Total, round hole UNQ	Encirclement	Success	—

G = Goniotomy. TR = Trepine. T = Trabeculectomy. C = Cyclocryotherapy. CA = Cycloaenaeisation. S = Scheie's thermocyclosterotomy. I = Iridencleisis. UNQ = Upper nasal quadrant.

the development of retinal detachment ranged from 7 months to 33 years. The presenting features of retinal detachment included ocular hypotension with or without lens subluxation in 9 eyes, subjective visual loss in 8 eyes, vitreous haemorrhage in 3 eyes, and the observation of retinal detachment shortly after filtering surgery in 1 eye. A wide range of refractive errors was encountered. Of a total of 18 eyes in which the error could be measured 15 eyes were found to have myopia in excess of 2.50 dioptres and in 8 eyes to be 10 or more dioptres as expressed in spherical equivalents.

Retinal detachment was unilateral in 15 and bilateral in 3 cases. With the exception of serous detachments in both eyes of the patient with bilateral Sturge-Weber syndrome the detachment was considered to be rhegmatogenous in all cases. However, retinal breaks were identified in only 9 eyes and included u-tears in 4 eyes, round holes in the temporal periphery with multiple incomplete dialyses in 1 eye, giant pre-equatorial tears in 2 eyes, and giant dialyses in 2 eyes.

Retinal detachment surgery was undertaken in a total of 13 eyes of 12 patients. The limited potential for useful visual recovery together with the presence of significant corneal and lens opacities were the major factors precluding any attempt at surgical management in the remaining 8 eyes. Encirclement with or without additional silicone sponge explants was performed in 8 eyes and local procedures alone in 3 eyes. Silicone oil injection was used in 3 eyes with massive preretinal retraction. Pars plana vitrectomy for massive preretinal retraction was performed on 2 eyes.

In 3 eyes detachment surgery was successful as defined by retinal reattachment for 6 months or longer following surgery. In 2 of the 5 eyes in which silicone oil injection was used anatomical reattachment with visual improvement was achieved. The causes of failure of detachment surgery included severe intraocular haemorrhage in one eye and massive preretinal retraction in 8 eyes.

Cases classified as complicated congenital glaucoma included a patient with Axenfeld's syndrome which proved extremely resistant to all forms of attempted glaucoma control and the case of bilateral Sturge-Weber syndrome. The latter patient developed bilateral serous retinal detachments in association with diffuse choroidal angiomas (Fig. 1). The only treatment considered likely to achieve retinal reattachment was the use of silicone oil injection. In the left eye a successful result was achieved whilst surgery of the right eye was complicated by the development of severe intraocular haemorrhage.

The retinal status of the fellow eye was assessed

Fig. 1 Central horizontal B-scan section of right eye showing retinal detachment and choroidal haemangioma (arrow).

wherever possible. In one eye a flat u-tear in temporal retina was identified for which prophylactic cryotherapy was undertaken. In a further case the fellow eye of a patient with a giant retinal dialysis showed marked cystoid degeneration of the ora serrata in all quadrants, which was treated by 360° cryotherapy.

Twelve eyes of 10 patients in this study belonged to a closely studied series of 158 cases of simple congenital glaucoma involving 246 eyes followed by the late A. Lister and one of the authors (N.S.C.R.). The results of surgery and the overall complications within this series have been the subject of a previous report.⁷ Over a follow-up period ranging from 2 to 19 years this represents an incidence of retinal detachment of 6.3%.

Discussion

Early reports of retinal detachment in congenital glaucoma concerned eyes severely compromised by the effects of uncontrolled glaucoma.^{1,2} An increased risk of retinal detachment was thought to accompany surgical intervention in cases of uncontrolled glaucoma.² However, there have been few reports of this complication in patients observed over a prolonged period following the introduction of more effective methods of glaucoma control.

Myopia is the most common refractive error in congenital glaucoma and is usually between 1 and 7 dioptres.⁸ The axial length and the degree of myopia are not constantly related, since the anticipated myopia is mitigated by changes in corneal curvature and posterior displacement of the lens. Myopic fundus changes in hydrophthalmia were described by Parsons⁸ and included retinal pigment atrophy and disc crescents. Myopia was an almost constant finding in our patients, exceeding 10 dioptres in 8 of the eyes. This excess of highly myopic eyes

implies significant accelerated axial growth, though we did not determine the axial length of the eyes.

With a single exception all patients received miotic therapy at some stage in their clinical course. No significant association between the use of miotics and the development of retinal detachment was recognised. None of the patients were known to have received irreversible cholinesterase inhibitors, which might be considered inappropriate in cases of advanced uncontrolled glaucoma with the attendant risk of retinal detachment.

The features of congenital glaucoma commonly encountered in our patients included bilateral involvement and the necessity for repeated surgical intervention. These patients are commonly found within the estimated 15% of all patients with simple congenital glaucoma who are not effectively controlled by goniotomy alone.⁷ Serious impairment of vision was often present before the onset of retinal detachment as a result of anterior segment changes and glaucomatous optic nerve damage. However, in several cases the fellow eye also had poor visual function attributed to severe corneal scarring, dense amblyopia, or glaucomatous optic atrophy.

In 19 of 21 eyes the retinal detachment was considered to be of rhegmatogenous origin, though retinal breaks were only identified in 9 eyes. The presence of corneal scarring or cataract precluded visualisation of retinal details in many cases. Of the retinal breaks that were identified, vitreoretinal traction played a predominant role in their origin. Giant retinal breaks were located at varying distances from the ora serrata but in all instances appeared to be pre-equatorial. Giant retinal breaks have been described in association with various developmental anomalies.^{5 9 10}

The results of detachment surgery in cases of congenital glaucoma are frequently disappointing in our experience. Of a total of 13 eyes considered likely to benefit from detachment surgery a successful outcome was achieved in only 3 eyes of 3 patients. The major factors contributing to these poor results were the difficulty in the evaluation of peripheral retinal details owing to opacities in the ocular media and the tendency to develop massive preretinal retraction. Evidence of the latter was often detected at the time of presentation, and postoperatively several eyes proceeded rapidly to an inoperable state. In 5 eyes the only surgical technique con-

sidered likely to achieve retinal reattachment was silicone oil injection, and a successful result, that is, more than half of the retina reattached accompanied by visual improvement, was achieved by this method in only 2 eyes.

Prolonged follow-up of patients suffering from congenital glaucoma has identified a variety of factors which may limit the visual outcome or prejudice continued visual function. These include amblyopia,¹¹ delayed recurrence of glaucoma, and late endothelial decompensation necessitating keratoplasty.¹² Retinal detachment is a further complication which may develop in buphthalmic eyes at varying intervals after the onset of glaucoma and would appear to carry an extremely poor prognosis.

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