Indications for vitrectomy in congenital retinoschisis

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SUMMARY Two patients with congenital retinoschisis developed rhegmatogenous retinal detachments with either a tractional component or associated vitreous haemorrhage. The second patient also had a large, raised schisis cavity under vitreous traction, which obscured visualisation of the macula. Vitrectomy is indicated for some patients with congenital retinoschisis.

Congenital retinoschisis, a relatively rare ocular disorder¹ first described by Haas² in 1898, has also been called congenital vascular veils,³ cystic disease of the retina in children,⁴ juvenile retinoschisis,⁵ and inherited retinal detachment.⁶ Usually bilateral,⁷ the disorder is inherited as an X-linked Mendelian recessive trait affecting males,⁸⁻¹⁰ though a few sporadic cases have been reported in females.¹¹

Retinoschisis has a predilection for the infero-temporal quadrant⁷ and rarely extends to the ora serrata. Large blood vessels frequently course through the inner retinal wall, and other vascular elements may traverse the schisis cavity.¹² Retinal holes in the inner layer are common and usually appear in multiples.¹³

Other ocular changes reported in retinoschisis include neovascular glaucoma,¹⁴ optic atrophy,^{5,15} and diminished visual acuity¹⁶ secondary to foveal dystrophy. Conway and Welsh¹² described the presence of a haemorrhagic retinal cyst surrounded by exudate extending into the macula.

Retinal detachment is a serious complication associated with congenital retinoschisis.^{5,7,13,17,18} Surgical treatment in the past has involved conventional scleral buckling procedures. The use of vitrectomy to remove vitreous haemorrhage and to repair a retinal detachment with a tractional component in patients with X-linked retinoschisis is, to the best of our knowledge, described for the first time here.

Case reports

CASE 1

An 11-year-old boy, who was diagnosed as having X-

linked retinoschisis at age 5, was referred to the University of Illinois Eye and Ear Infirmary with a retinal detachment in his right eye. He was the product of a normal pregnancy and had no congenital anomalies. Two older brothers also had X-linked retinoschisis. An ocular examination of his mother and three of his sisters was unremarkable. No consanguinity existed between the parents. When the patient was 4 years old, an electroretinogram (ERG) (performed with the patient under anaesthesia) showed an extinguished B wave. The boy's two brothers had similar ERG changes.

Ocular examination revealed a corrected vision of counting fingers at 6 feet (1.8 m) in the right eye and 5/200 in the left eye. External examination revealed 8° of left esotropia. Biomicroscopy was unremarkable. Ophthalmoscopy of the left eye revealed a large grey intravitreal membrane floating in the inferior vitreous between the inferior arcade and equator. The macula was distorted, and inferiorly a retinoschisis cavity was observed. A fundus examination of the right eye disclosed cystic macular changes. A full-thickness retinal detachment (Fig. 1) that involved the macula and most of the inferior retina extended from the 4 to 8 o'clock meridians. A small schisis cavity was noted in the inferior retina near the equator. Another area of retinoschisis was present inferior to the temporal arcades. A vitreous veil exerted traction on a retinal fold along the infero-temporal arcade. A partial-thickness retinal tear was also present below the retinal fold.

During surgery the partial-thickness retinal tear inferior to the macula was noted to be under considerable vitreous traction. The vitreous band was transected with a bipolar¹⁹ cautery attached to the fiberoptic membrane dissector and vitrophage to allow the underlying retina, including the partial-

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Fig. 1 Case 1. Composite fundus photograph showing flat detached retina inferiorly and in the macula.

thickness retinal tear, to settle. The inferior peripheral retina settled during the completion of the vitrectomy. Peripheral transscleral cryopexy was applied to the eye for 360°.

Postoperatively the retina remained attached (Fig. 2). At last examination seven months after surgery the best corrected visual acuity was 20/200.

CASE 2

A 2-year-old black boy had a red left eye that was diagnosed as viral conjunctivitis. On follow-up examination three weeks later the child was found to have a preretinal haemorrhage in the right eye and a possible retinal detachment in the left eye.

He was living in a foster home with his 3-year-old brother and no family history was available. Ocular examination of his brother was unremarkable.

Five weeks after his initial presentation the boy was admitted to hospital for an examination under anaesthesia. The right eye showed stellate cystoid changes in the macula and extensive retinoschisis in the inferior and temporal periphery, extending centrally with abrupt termination at the posterior pole. The left eye had a highly elevated bullous

schisis cavity inferiorly overhanging and obscuring visualisation of the macula (Fig. 3). A second area of schisis was present in the superior temporal area.

The patient was again examined under anaesthesia six weeks later and underwent drainage of schisis fluid, vitrectomy to relieve traction on the inferior schisis cavity, and placement of an encircling silicone band in the left eye. Stellate cystoid changes were observed in the left macula after the schisis cavities were flattened.

He was readmitted for an examination under anaesthesia 10 months later. Examination of the left eye revealed a reaccumulation of fluid in the inferior and temporal schisis cavity, which again covered the macula. Three months later the child underwent surgery in the left eye, which involved drainage of fluid from both schisis cavities, and then scattered transscleral cryocoagulation was applied to the now flattened inferior and temporal schisis cavity. Twenty months after operation the retina remained attached (Fig. 4).

Nineteen months after the initial admission to hospital a vitreous haemorrhage (Fig. 5) and an inferior retinal detachment were discovered in the

Fig. 2 Case 1. Fundus photograph after vitrectomy and retinal detachment repair.

right eye during routine examination. After a vitrectomy to remove blood from the vitreous cavity two large full-thickness retinal tears were observed. Subretinal fluid and fluid in the large schisis cavity

was drained, and the two retinal tears were treated with cryopexy and an encircling silicone band, and a 5 mm radial silicone sponge was placed behind both retinal tears.

Fig. 3 Case 2. Left eye fundus photograph showing huge schisis cavity covering the macular area. Arrow points to optic disc.

Fig. 4 Case 2. Fundus photograph after vitrectomy, retinal detachment repair, and cryocoagulation. Note attached retina and typical schisis changes of the macula; arrows indicate cryocoagulation scar.

Fig. 5 Case 2. Fundus photograph of right eye showing vitreous haemorrhage covering a detached inferior retina.

Fig. 6 Case 2. Same eye as in Fig. 5 after vitrectomy, retinal detachment repair, and buckling procedure.

Postoperatively a cataract developed in the right eye, which required a lensectomy a year later. At the time of the cataract surgery the retina and schisis cavities were noted to be flat (Fig. 6). Measurement of visual acuity was not possible because of the patient's youth.

Discussion

Congenital retinoschisis may be progressive or stationary or may spontaneously regress.¹³ The clinical spectrum of ocular changes varies considerably among affected individuals. Deutman²⁰ states that macular retinoschisis occurs in all affected patients. Foveal retinoschisis in one report was present in 98% of involved individuals.²¹ According to Deutman²⁰ in almost half the affected eyes macular lesions may represent the only sign of this disorder. The macular changes in older people may appear atrophic and pigmented.²²⁻²⁴ Peripheral retinoschisis, characterized by intraretinal splitting involving the nerve fibre layer,²⁵ occurs in approximately 50% of eyes.²⁶ The inner layer is often immobile and elevated in a concave configuration. An absolute scotoma is present corresponding to the area of retinoschisis.²⁵

Two severe vision-threatening complications of congenital retinoschisis are retinal detachment and vitreous haemorrhage.¹² Retinal detachments are caused by a combination of inner and outer layer holes in areas of retinoschisis, or retinal tears created by vitreous traction in abnormal retina.¹³ Vitreous haemorrhage may result from vitreous traction on

retinal vessels coursing through the elevated inner retinal layer of a schisis cavity.

A variety of vitreous pathology has been observed in eyes with congenital retinoschisis. All 15 patients in a series reported by Lisch²⁶ showed varying degrees of vitreous liquefaction. Vitreous strands were found in 50% of patients by Bec and associates.²⁷ Kraushar *et al.*¹³ reported a series of 40 patients (77 eyes) with congenital retinoschisis. Retinal detachment developed in 22% of these affected eyes. Associated with retinal detachment were pathological vitreous changes, including vitreous membranes attached to and in some instances elevating the equatorial retina. Vitreous traction present in the posterior pole resulted in pseudopapilloedema (13% of eyes) and ectopic macula (6% of eyes). Vitreous haemorrhage occurred in 40% of eyes. The investigators believed that in some instances vitreous haemorrhage was caused by traction on retinal blood vessels. Other less common signs of vitreous traction were tenting of the inner retinal layer (12%) or blood vessels (4%) and geographic areas of white without pressure (35%).

The retinas in both our patients and the inferior schisis cavity overhanging and obscuring visualisation of the macula were under vitreous traction. The retinas in both cases were successfully reattached following vitrectomy and scleral buckling. Vitrectomy in eyes with congenital retinoschisis is indicated when a significant tractional component is associated with either retinal detachment, elevation of a schisis cavity covering the macula, or vitreous haemorrhage originating from blood vessels located in the inner

retinal layer of a schisis cavity. The tractional changes may result from proliferative vitreoretinopathy²⁸ complicating a retinal detachment or vitreous pathology inherent in congenital retinoschisis. Scleral buckling does not treat the vitreous pathology in these eyes, though it may relieve to some extent the tractional component of the retinal detachment. Vitrectomy is necessary to remove vitreous haemorrhage and to eliminate vitreous traction on the vessels and the retina.

Although vitrectomy prevents progression of vitreous tractional changes, it will not eliminate reaccumulation of fluid in the schisis cavity, as was the case in our second patient.

Drainage of subretinal fluid combined with cryocoagulation of the suspected retinal holes creates a permanent chorioretinal scar, thereby preventing reaccumulation of fluid in the schisis cavity.

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