Management of persistent hyperplastic primary vitreous by pars plana vitrectomy

Gholam A. Peyman, Donald R. Sanders, and Krishan C. Nagpal

From the Department of Ophthalmology, University of Illinois Eye and Ear Infirmary, Chicago, Illinois

In the past the surgical treatment of persistent hyperplastic primary vitreous (PHPV) has been mainly carried out via an anterior segment approach. With the newly developed vitreous cutting and aspirating instruments, pars plana vitrectomy may be a workable surgical alternative (Smith and Maumenee, 1974; Michels, Machemer, and Mueller-Jensen, 1975; Peyman and Sanders, 1975).

We describe two cases of PHPV handled by pars plana surgery.

Patients

Case 1

A 3-month-old Black boy was noted to have leucocoria in the right eye at two months of age. Prenatal and developmental histories were unremarkable. There was no family history of ocular abnormalities. Physical examination showed only an easily reducible right inguinal hernia.

The child’s left eye was normal, with corneal diameter of 11.5 x 11.5 mm. He had a deviation in the right eye with a constant 24 degrees of right esotropia. The right eye was normal externally with a corneal diameter of 11 x 11 mm. Intraocular pressure by Schiötz tonometry was 24 mmHg in the left eye and 21 mmHg in the right. Gonioscopy showed normal angles in both eyes. A posterior polar cataract in the right eye spared the periphery of the lens, through which was visible a large fibrovascular stalk (Fig. 1) with prominent vessels emanating from the optic disc. Surrounding the base of the stalk was a localized central tractional retinal detachment. There was no evidence of elongated ciliary processes at the periphery of the lens.

Fig. 1. Preoperative appearance of patient's right eye demonstrating cataract and intravitreal fibrovascular stalk (arrow)
Surgical technique

Under general anaesthesia with four 4-0 silk bridle sutures under the rectus muscles, a no. 15 Bard-Parker blade cut a 3.5 mm long limbus-parallel sclerotomy down to the choroid, 2.5 mm posterior to the corneoscleral limbus in the inferotemporal quadrant. The ends of a 5-0 Dexon mattress suture, placed through the sclerotomy lips, were tied loosely. Toothed forceps grasping the sclerotomy lips stabilized the eye while a radiofrequency diathermy probe (Peyman and Dodich, 1972), inserted through the sclerotomy, coagulated the large vessels of the intravitreal stalk without complication. A no. 52S Beaver blade inserted into the equator of the lens created a track through the sclerotomy for the vitrophage for the vitrectomy instrument. The lens fragmenter unit (Peyman, Huamonte, and Goldberg, 1975) of the vitrophage broke up the lens, which was removed together with the posterior polar cataractous plaque and the anterior portion of the fibrovascular stalk, with the wide-angle cutting unit of the vitrophage (Peyman, 1976). An anterior vitrectomy was performed with the same unit. The vitrophage was removed, and the 5-0 Dexon mattress suture was tied and reinforced centrally with an interrupted suture of 5-0 Dexon. Sutures of 5-0 catgut closed the peritomy, and the eye received atropine and an antibiotic corticosteroid preparation topically.

Two weeks after operation the eye was quiet, and only a slight vitreous haze clouded the media (Fig. 2). The stump of the fibrovascular stalk appeared retracted, and the tractional retinal detachment was somewhat flattened. Ocular examination has remained unchanged after being followed-up for six months.

CASE 2

A 4-month-old White boy was noted to have leukocoria in the left eye at approximately six weeks of age. Prenatal and developmental histories were non-contributory. At three months of age the child was noted to have, in the left eye, a small cornea, shallow anterior chamber, small cataractous lens, and a white retrolental mass with elongated ciliary processes nasally. Intraocular pressure (Schiotz) was 17 mmHg and the fundus could not be visualized. X-ray films of the orbit revealed no intraocular calcifications, and ultrasonography revealed a slightly microphthalmic eye with a thickening at the posterior lens capsule. The patient was then referred to us for treatment.

The child's right eye was normal with corneal diameters of 11 ± 1.5 mm. There was unsteady fixation in the left eye with a variable deviation. The left eye appeared microphthalmic with corneal diameters of 10 ± 0.25 mm. Intraocular pressure (Schiotz) was 17 mmHg (4/5). A distinct dense pupillary membrane extended from collarette to collarette; in addition, an intact tunica vasculosa lentis and a retrolental mass extended into the lens (Fig. 3). A red reflex could be elicited temporally around the retrolental mass. The operative procedure was identical to that in Case 1 except that the intravitreal stalk was not diathermized. There were no operative or postoperative complications and on the first postoperative day the conjunctiva was mildly hyperaemic, the cornea was clear, and retinal fundus detail could be seen with a hand-light. At two weeks postoperatively only mild conjunctival hyperaemia was noted (Fig. 4), and at examination three months postoperatively there was no evidence of inflammation, the ocular media were clear, and the retina was normal (Fig. 5).

Discussion

Most surgical approaches to the treatment of PHPV have been via an anterior segment approach (Collins, 1968; Wolfe, 1954; Reese, 1955; Acers and Costen, 1967; Gass, 1970; Smith and

FIG. 2 Appearance of eye 10 days postoperatively demonstrating clear pupillary opening with no evidence of ocular inflammation

FIG. 3 Preoperative view demonstrating cataractous lens
Maumenee, 1974); either a discission or excision of the retrolenticular tissue through a large corneoscleral limbal incision followed an aspiration procedure. Nevertheless, this large incision provides poor operative exposure for removal of retropupillary tissue.

Michels and Ryan (1975) reported two cases of PHPV among 100 vitrectomy cases although the operative technique and postoperative results were not specified. Michels and others (1975) have said that they prefer to use an incision at the limbus rather than at the pars plana in these microphthalmic eyes. We advocate early surgical intervention in these cases before complications such as bleeding into the lens or vitreous or traction on the ciliary body occurs or becomes worse. With development of the cataract, a lens-induced flat anterior chamber and secondary glaucoma may develop. Although the visual prognosis in these cases is poor because of macular aplasia, tractional detachments and deprivation amblyopia, removal of the anteroposterior traction caused by the hyperplastic primary vitreous allows the eye to grow and to achieve reasonable cosmesis.

**Summary**

Two children with persistent hyperplastic primary vitreous (PHPV) underwent vitrectomy and lens-ectomy via the pars plana to remove the fibrovascular stalk. Postoperatively the eyes were quiet, only a slight vitreous haze obscured the fundus view in the immediate postoperative period, and the stumps of the stalks retracted. Early surgical treatment of PHPV may prevent later serious complications.

**References**


COLLINS, E. T. (1908) J. Amer. med. Ass., 54, 1051


——, and DODICH, N. A. (1972) Ibid., 3, 32


