

PERSISTENT HYPERPLASTIC PRIMARY VITREOUS: DIAGNOSIS, TREATMENT AND RESULTS*

BY *Zane F. Pollard, MD*

INTRODUCTION

Persistent hyperplastic primary vitreous, known as PHPV, poses a challenge to the ophthalmologist because of the difficulty in obtaining a good visual result from treatment. Following posterior lenticonus, PHPV is the second most common cause of acquired cataract during the first year of life. In some cases, a cataract associated with PHPV may be present at birth. The purpose of this study is to differentiate patients with PHPV who obtain a good visual result from those who cannot expect visual improvement from treatment. My experience has shown that only those patients with anterior segment involvement in PHPV eyes obtained good visual results from surgery. The body of material presented will demonstrate to the ophthalmologist that surgery of the posterior segment of the eye in PHPV patients is essentially an exercise in futility if the intention is to provide a good visual result. The extensive case studies, the largest series reported in the English literature to date, will provide the ophthalmologist with significant data useful in communicating to parents whether they can realistically expect their child to have a satisfactory visual outcome.

During my residency in ophthalmology (1970-1973), the standard treatment of PHPV involved making the diagnosis in the first few months of the patient's life and then performing follow-up examinations every 3 months with the patient under anesthesia. When secondary and uncontrollable glaucoma set in, the eye was enucleated. The enucleation usually took place during the second or third year of life. In 1966, Raskind¹ reported on the enucleation of a 3-week-old infant's eye containing PHPV. The child's condition had rapidly progressed from a normal-looking external eye at birth to a cloudy cornea and secondary glaucoma 3 weeks later. This is the earliest case in the literature of PHPV requiring an enucleation. Karr and Scott² reported in 1986 on 25 patients with PHPV who were followed without surgery. Two of their patients, aged 6 and 3, required enucleation for complications related to secondary glaucoma. In a large percentage of patients, the natural history of untreated PHPV is characterized by recurrent intraocular hemorrhage and secondary glauco-

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ma. This process eventually leads to enucleation or phthisis.

During the past 22 years, I have followed 83 patients with extensive involvement of at least 1 eye with PHPV. Two patients had bilateral disease. Intraocular surgery was performed on 80 patients with the following objectives: (1) to try to save the eye by preventing the onset of secondary glaucoma, (2) to give the patient a black pupil, which would be cosmetically more acceptable than the white pupil of a cataract and/or PHPV membrane, and (3) to salvage vision and treat amblyopia in those cases in which there was a reasonable chance of success because of a clear media present postoperatively. After appropriate intraocular surgery and amblyopia therapy, strabismus surgery was performed on those patients who had at least 14 prism diopters of horizontal misalignment or 10 diopters of vertical misalignment.

This study analyzes the various presentations of PHPV and evaluates the differing outcomes after treatment. Information gleaned from this series of patients will enable the physician to inform the families of children with PHPV about reasonable expectations from surgery. A distinction will be made between eyes that can be salvaged from enucleation and those that can undergo visual rehabilitation. After 10 years of practice, it had become apparent to me that most children with PHPV had a poor visual outcome. A small percentage, however, did obtain good visual results in the 20/20 to 20/200 range. I will discuss preoperative findings as well as operative and long-term visual results in this extensive series of 83 consecutive patients with PHPV.

EMBRYOLOGY

PHPV has also been called persistent tunica vasculosa lentis, persistent posterior fetal fibrovascular sheath of the lens, congenital retinal septum, and ablatio falciformis congenita.³ The last two names include mention of a retinal fold, which is often seen in the posterior variety of PHPV. PHPV is the result of the anomalous development of the primary vitreous as it persists into the time of formation of the secondary vitreous, being associated with hyperplasia of the mesodermal elements contained in the primary vitreous and hyaloid artery system. The anterior extension became known as the tunica vasculosa lentis, so that eventually the entity was recognized as having two parts, an anterior and a posterior PHPV. In the early 10-mm embryo stage, vascularized mesoderm has entered the developing vitreous space, and the lens becomes separated from the primary or hyaloidean vitreous by a layer of vascularized membranes. The secondary, or definitive, vitreous formation occurs during the 13- to 70-mm embryo stage. By the time of the 70-mm embryo stage, the primary vitreous has ceased to grow and has, in fact, atrophied along with the remnants of the

hyaloid vascular system. At this point, these two primitive structures occupy a constricted zone in the middle of the secondary vitreous.⁴ This remnant of the primary vitreous is attached posteriorly around the optic disc and extends anteriorly through the secondary vitreous, where it is referred to as the canal of Cloquet. It then extends anteriorly to form the retrolental space of Erggelet. At the 170-mm stage, one can see well-formed fibrils, which run from the ciliary epithelium to the lens. These fibrils become the zonules, also known as the tertiary vitreous. The most anterior part of the secondary vitreous retains its connection with the retina at the area of the ora serrata known as the vitreous base.

The tunica vasculosa lentis, which is the anterior extension of the hyaloidean vitreous, is composed of a layer of vascular channels surrounding the lens.⁵ It originates from three areas: (1) the hyaloid artery, (2) the vasa hyaloidea propria, which is derived from the vessels that pass into the peripheral parts of the vitreous and enter the vascular coats of the lens at its equator, and (3) the anterior ciliary vessels, which originate from the major arterial circle of the iris. The anterior part of this structure is supplied mainly by the ciliary system, while the posterior part is supplied primarily by the hyaloid artery and its branches. By the eighth month of gestation, the anterior part usually shows a complete regression following the course of the posterior part, which generally has atrophied by the seventh fetal month. The fibrovascular sheath and the hyaloid artery compose the two aspects of the posterior system. Both are portions of the primary vitreous, and both can persist with the remnants of the primary vitreous. When the anterior vascularized capsule of the lens is present, that portion which has not atrophied can be used to judge the gestational age of premature infants. Hittner and associates⁶ showed that when the vascularized membrane covers the entire anterior surface of the lens, the gestational age is believed to be 27 to 28 weeks. When three fourths of the anterior capsule is covered with the vascular membrane, the gestational age is judged to be 29 to 30 weeks. When the vascularized membrane covers one half of the anterior lens surface, gestational age is measured at 31 to 32 weeks. When the membrane occupies only the peripheral one fourth of the anterior lens capsule, the gestational age is believed to be 33 to 34 weeks. This method of evaluating gestational age at birth correlates closely with the more formal Dubowitz clinical assessment of gestational age. Following dilation of a newborn's pupil, the remnant of the anterior vascular capsule of the lens is studied with a direct ophthalmoscope.

OTHER INVESTIGATORS' EXPERIENCE WITH PHPV AND SYNDROMES ASSOCIATED WITH PHPV

Much of what we know about PHPV has come from the pathologic studies of cases in which patients have been treated with enucleation.

Intractable glaucoma and phthisis bulbi are the main reasons for enucleation of eyes containing PHPV (Figs 1 and 2). Reese,⁷ in his Jackson Memorial Lecture of 1955, pointed out that untreated cases can lead to phthisis bulbi. Haddad and associates⁸ wrote on an extensive study of enucleated eyes in 62 cases, stating that intractable glaucoma and phthisis bulbi are the main reasons for enucleation in eyes containing PHPV. All the infants in their study were full-term at birth and had no family history of PHPV. Fifty-eight of their patients were white, and 4 were black. In 51 patients, the abnormality was noted at birth or within the first few weeks of life. The disease was noted in the second or third decade of life in 10 patients and at age 71 in 1 patient. The histologic specimens of their patients have taught us much of what we know about PHPV and its natural history. The ciliary processes are elongated and pulled into the retro-
lental membrane. The peripheral retina can also be drawn into the retro-
lental membrane. As a result of these findings, some surgeons routinely approach PHPV via an anterior limbal approach so as to avoid the peripheral retina, which might be drawn into the membrane. This anterior limbal approach would also avoid a peripheral retina that might be inserting into the ciliary body. Two thirds of the patients in the study of Haddad and associates⁸ had microphthalmic eyes. These eyes were mildly microph-

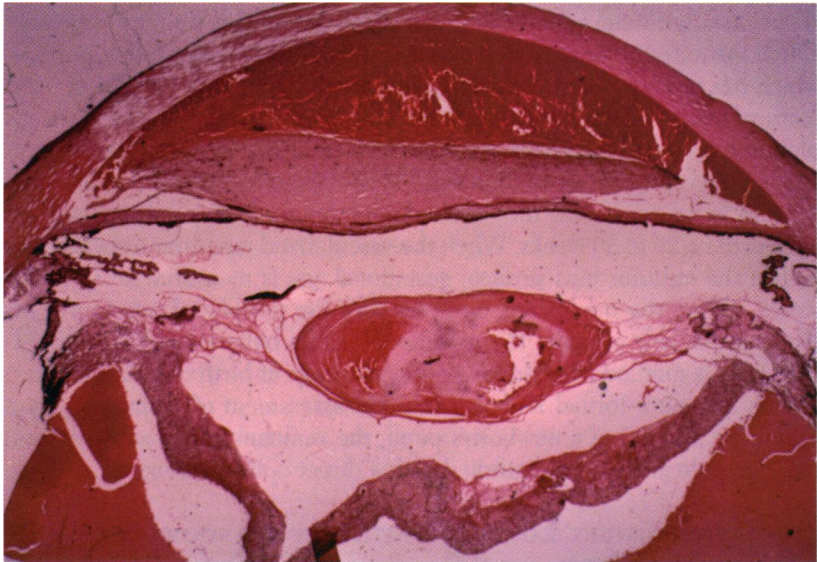


FIGURE 1

Pathologic specimen showing anterior chamber to be filled with blood in eye enucleated for intractable glaucoma secondary to recurrent hyphema in patient with PHPV.

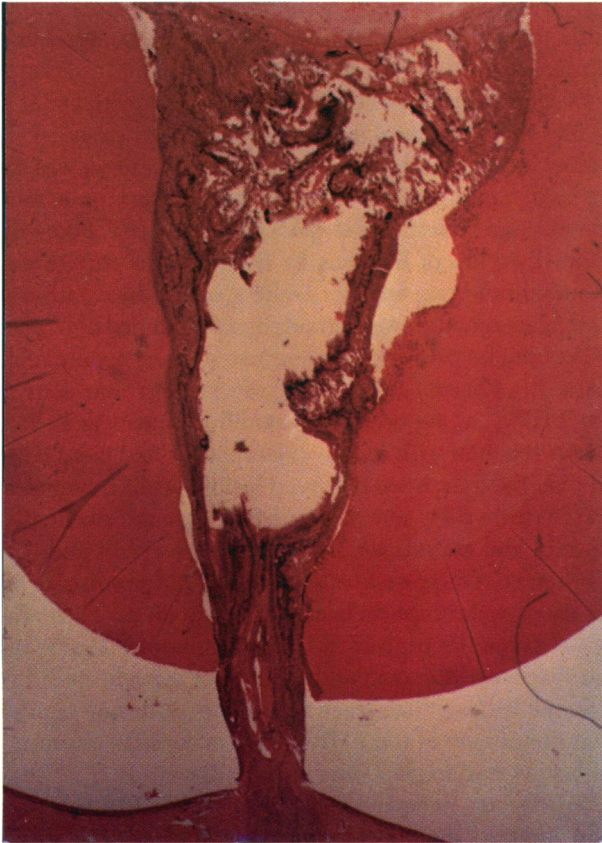


FIGURE 2

Same eye as in Fig 1 showing PHPV stalk going from lens to optic nerve.

thalmic in 32 of 55 unilateral cases. With the exception of 1 case, all patients with bilateral PHPV had extremely small eyes. In 13% of their patients' eyes, the PHPV occurred in a normal-sized eye; the remainder occurred in buphthalmic eyes. Of course, we do not know if the buphthalmic eyes were originally microphthalmic or normal in size prior to the onset of glaucoma. This same study reported that the patients with bilateral PHPV usually had severely microphthalmic eyes and showed other ophthalmic and systemic abnormalities that accounted for an early death in those infants with bilateral PHPV. The study reported bilaterality in 11%, whereas in my series to be discussed later, only 2.4% were bilateral. Eighty-four percent of my cases involved eyes that were microphthalmic,

and only 3 cases involved severely microphthalmic eyes with a corneal diameter of 7 mm or less. Two of my patients with bilateral disease were entirely normal except for their eyes.

In 56% of their cases, the retina was totally detached, and it abnormally inserted into the retrolental membrane in 30% of cases. A dehiscence of the posterior capsule with various stages of absorption was observed in 50% of patients' eyes. This accounts for a cataract or a membranous cataract, which can be observed in patients with PHPV. Some of the lenses were clear. Invasion of the lens by fibrovascular structures from the retrolental membrane can lead to swelling of the lens with a secondary angle-closure glaucoma or even intralenticular hemorrhage. Total absorption of the lens can be present. The lens can even be totally replaced with adipose tissue.⁹ Phacoanaphylactoid reaction to the lens has been observed in PHPV as reported by Caudill and associates.¹⁰ Extensive granulomatous reaction with foamy macrophages and giant cells containing cortical material have been observed histologically in enucleated specimens of PHPV from their series. Neutrophils around swollen lens fibers, epithelioid cells, and granulation tissue with eosinophils and lymphocytes have been observed. One case involved a 1-day-old premature infant, suggesting that the capsular rupture had occurred in utero. The definitive reason for posterior capsular rupture in PHPV is unknown. Some believe that a congenital absence or thinness of the posterior capsule contributes to the rupture. Some believe that traction on the posterior capsule by the fibrovascular membrane of the PHPV leads to capsular rupture.

Several factors lead to glaucoma in patients with PHPV. An immature angle may be present. Recurrent hemorrhage or uveitis can contribute to glaucoma. In addition, the development of an intumescent lens secondary to posterior capsule rupture with phacoanaphylactoid reaction contributes to the inflammation and glaucoma. Also, the intumescent lens can lead to a pupillary block and secondary angle-closure glaucoma. The association of a cataract in a microphthalmic eye can also be seen in the congenital rubella syndrome and in patients with congenital toxoplasmosis. Nine cases in Haddad's series contained a falciform retinal fold.

Some degree of persistence of the hyaloid vascular system can be present in 3% of all full-term infants. Up to 95% of premature babies can have a remnant of the hyaloid system.¹¹

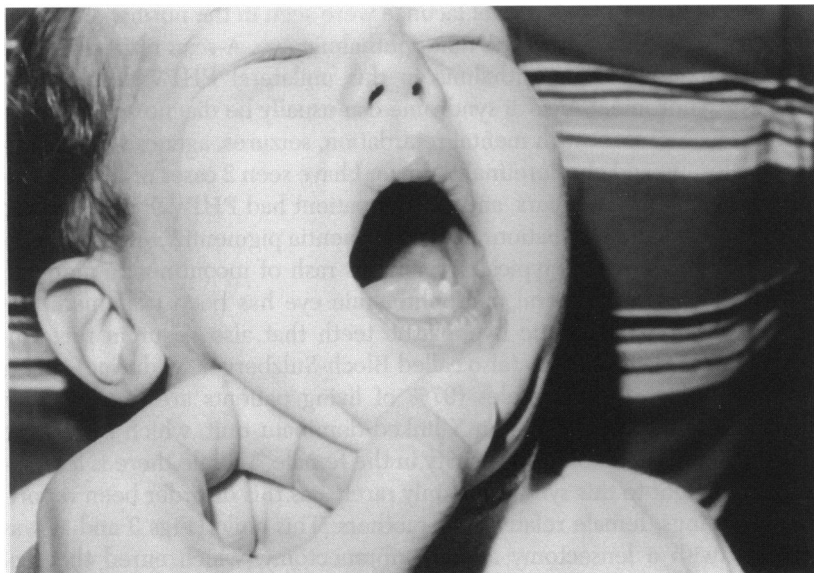
In one of Haddad's cases, a medulloepithelioma of the ciliary body was present in a 2½-year-old boy in association with PHPV. Another patient, a 1½-year-old boy, had 1 eye that had been microphthalmic since birth. It contained a retinoblastoma. Retinoblastoma uniformly presents in a normal-sized eye. The occurrence of a retinoblastoma and PHPV in a microphthalmic eye is a rare event.¹² Morgan and McLean¹³ reported on a 4-week-old girl who underwent an enucleation for retinoblastoma in her

left eye. The horizontal corneal diameter was 11.0 mm in the left eye. The right eye had a corneal diameter of 10.0 mm and underwent a lensectomy and anterior vitrectomy for an anterior PHPV. The child's mother had bilateral retinoblastoma, and 2 of her siblings also had retinoblastoma. Hara and colleagues¹⁴ reported on enucleating the eye of an 8-month-old boy because a needle biopsy showed clusters of blastic cells erroneously believed to be retinoblastoma. The eye contained PHPV and retinal dysplasia with rosettes. The eye was microphthalmic. An adenoma of the pigmented ciliary epithelium associated with PHPV in an enucleated eye from a 3-year-old girl has been reported.¹⁵ Shields and associates¹⁶ reported on a case that had the features of PHPV with leukokoria, shallow anterior chamber, cataract, and retrolental fibrovascular tissue. The eye was finally enucleated and found to contain a malignant teratoid medulloepithelioma of the ciliary body and not a PHPV. None of the patients in my series had an intraocular tumor. None of my patients had a family history of PHPV. Wang and Phillips¹⁷ reported the occurrence of PHPV in fraternal twins.

Haddad and associates⁸ reported systemic abnormalities in their bilateral cases only. Cleft lip and palate, polydactyly, and microcephaly were the main systemic abnormalities. The two bilateral cases in my series were otherwise normal. PHPV has been reported in Aicardi syndrome, in which the typical chorioretinal lacunae were seen in the normal-sized eye, with PHPV presenting in the microphthalmic eye. A-scan ultrasound was used to confirm microphthalmia in this unilateral PHPV occurring in Aicardi syndrome.¹⁸ Aicardi syndrome can usually be diagnosed in female infants who present with mental retardation, seizures, agenesis of the corpus callosum, and chorioretinal lacunae. I have seen 2 cases of Aicardi syndrome in the past 20 years, and neither patient had PHPV. PHPV has also been reported in association with incontinentia pigmenti.¹⁹ An 11-month-old girl (Fig 3) with a typical serpiginous rash of incontinentia pigmenti and PHPV in a unilateral microphthalmic eye has been examined. She showed the characteristic hypoplastic teeth that also occur in this syndrome (Fig 4). This entity (also called Bloch-Sulzberger syndrome) occurs almost exclusively in females (97% of living patients are female); it is believed to be inherited as an X-linked dominant trait, which is lethal in the male, with variable expressivity in the female.²⁰ While there is a familial component to this syndrome, only rarely has this disorder been reported in siblings, female relatives, or mothers. This child (Figs 3 and 4) was treated with a lensectomy and membranectomy, which cured the secondary angle-closure glaucoma. No visual rehabilitation was possible because of extensive involvement of the posterior segment and mental retardation. The fellow eye remains normal after a 7-year follow-up. Wald and colleagues²¹ reported on retinal detachment in 6 eyes of 4 patients

**FIGURE 3**

Child with serpiginous rash and incontinentia pigmenti.

**FIGURE 4**

Hypoplastic teeth in same child as in Fig 3 with incontinentia pigmenti.

with incontinentia pigmenti. They reported extensive preretinal and vitreous fibrous organization leading to the tractional detachment. They did not report PHPV in their patients. Goldberg and Custis²² reported on a very large series of 13 patients with incontinentia pigmenti. They had patients with preretinal fibrosis, retinal detachment, fibrovascular proliferation, and vitreous hemorrhage. They did not report PHPV. Zweifach²³ reported retinal dysplasia with incontinentia pigmenti but not PHPV. However, in the reported case, the patient had a pipelike stalk emanating from the optic nerve to the lens. This might have been PHPV. Scuderi and coworkers²⁴ reported PHPV in association with retina dysplasia. They found dysplastic retina with rosettes in association with the fibrovascular membrane of PHPV. Baloglou²⁵ also reported a case of PHPV in a 5-month-old girl in association with a dysplastic retina. Offret and associates²⁶ reported dysplastic retina in association with PHPV.

There have been reports of PHPV in association with morning glory syndrome. Characteristically, the morning glory disc is enlarged with an excavated disc containing a tuft of glial tissue in its center. Numerous retinal vessels leave the disc from its edge and course onto the retina. Peripapillary chorioretinitic atrophic changes can often be seen. Some have suggested that this morning glory disc may be a variant of persistent hyperplastic primary vitreous. Brown and associates²⁷ reported 2 cases of PHPV in association with morning glory disk, one in a 37-year-old man and 1 in a 5-year-old boy. Cennamo and colleagues²⁸ reported on another case of morning glory syndrome with PHPV and two lens colobomas in a unilaterally involved eye. The eye was of normal size with A-scan biometry. They believed that morning glory syndrome, optic pit, and optic nerve coloboma were variants of the same disease process and that PHPV was a condition in which morning glory syndrome was part of the same entity.

Teske and Trese²⁹ reported on a 10-week-old infant with PHPV in a microphthalmic eye and changes of retinopathy of prematurity in the other eye. The child was not premature but had been exposed to regular use of cocaine during the entire 9 months of pregnancy. The investigators believed it was unlikely that the PHPV was related to the use of cocaine, but they expressed interest that the association was present and that PHPV was associated with changes of retinopathy in its fellow eye.

Norrie's disease has many findings in common with PHPV. Norrie's is a rare X-linked recessive disease also known as oculoacousticocerebral degeneration. It presents a triad of retinal malformation or dysplasia, deafness, and mental retardation. A grey-white mass behind the lens has been described and is thought to be retinal dysplasia. Rosette formation is seen as well as retinal detachment and retinal folds. PHPV and Norrie's disease usually have microphthalmia and cataracts, retrolental masses, and secondary glaucoma. LaRussa and Wesson³⁰ reported a case of Norrie's dis-

ease in which surgery was performed to remove the lens, after which a totally detached retina attached to a fibrous retrolental stalk was noted. This patient had PHPV. There is some controversy whether retinal dysplasia reported in other cases of Norrie's disease is an entity different from PHPV. There are enough similarities in the two conditions to cause confusion when making the diagnosis.

While no genetic abnormality has been assigned to PHPV, there have been reports of cases in the same family. Menchini and associates³¹ reported on PHPV in 2 brothers, one 33 years old and the other 25. One had anterior PHPV in 1 eye only, and the other had the posterior variety. Wang and Phillips¹⁷ also reported PHPV in nonidentical twins. Burke and O'Keefe³² reported a case of PHPV in a buphthalmic eye with a pressure of 50. The contralateral eye had megalocornea. The horizontal corneal diameter in the involved eye of that 7-month-old child measured 15 mm, and the horizontal corneal diameter measured 13 mm in the uninvolved eye. Simple megalocornea with no other pathology was present in 6 siblings.³² Lin and associates³³ reported on persistent hyperplastic primary vitreous seen in a woman whose involved eye was enucleated at age 5 because of intractable glaucoma. Her 15-month-old son had PHPV in one microphthalmic eye. Alward and colleagues³⁴ reported on a 4-day-old infant with bilateral PHPV and bilateral congenital glaucoma. Both corneas were cloudy, both anterior chambers were deep, and both corneas measured 10.5 mm horizontally. The infant was full-term and had no other abnormalities. Bilateral trabeculotomies were performed for the glaucoma, after which the patient underwent bilateral lensectomy and vitrectomy at separate surgical settings. Awan and Humayun³⁵ reported on two adults, aged 62 and 71, both of whom had unilateral PHPV in an eye with poor vision. The uninvolved eye of each patient had open-angle glaucoma. In 1 of the patients, anomalous blood vessels were observed along the entire circumference of the anterior chamber angle in the eye without the PHPV.

Spaulding³⁶ reported on the enucleation of a blind and painful eye in a 71-year-old man. Histopathologic examination showed a persistent hyperplastic primary vitreous. This is one of the oldest patients in the literature with PHPV.

PHPV has also been seen in combination with microphthalmia and a communicating orbital cyst. This unilateral involvement also contained a hypoplastic optic nerve and a macular coloboma through which there was the intraocular extension of the orbital cyst.³⁷

Meisels and Goldberg³⁸ reported on 5 patients with a persistent anastomotic blood vessel between the anterior and posterior tunica vasculosa lentis as a manifestation of PHPV. The vessel was radially oriented on the anterior surface of the iris. It traveled around the pupillary margin to anas-

tomose with a retrolental fibrovascular PHPV. This prominent radial iris vessel can suggest the presence of PHPV. I have seen this anomaly twice in association with PHPV.

CLASSIFICATION OF PHPV

In the past 40 years, 2 clinical forms of PHPV have come to be recognized. Reese⁷ extensively discussed the anterior form, which was called *persistent tunica vasculosa lentis* and mainly affected the anterior segment. It is now called *anterior hyperplastic primary vitreous*. The second type of presentation has been called *posterior hyperplastic primary vitreous* and has been classified as a separate entity from its anterior cousin by Pruitt and Schepens.³⁹ They discussed posterior hyperplastic primary vitreous as a separate entity, giving credit to Michaelson,⁴⁰ who also agreed that the posterior form was an entity unto itself and that PHPV should be clinically divided into an anterior and a posterior form. They stressed overlap, stating that anterior and posterior forms could occur in the same patient. Pruett stressed the retinal findings in discussing 30 patients. Twenty-five of the patients had unilateral involvement and 5 were affected bilaterally. Microcornea was found in 46% of his patients. The involved eye had a horizontal corneal diameter that measured 1.0 to 1.5 mm less than its fellow eye. No axial lengths were recorded in the paper. Since I have been performing A-scans on my patients with PHPV during the past 8 years, I concur that the presence of microcornea in an eye with PHPV usually means that microphthalmia is also present. I have performed surgery on 1 patient with PHPV whose horizontal corneal diameter measured 9.0 mm in the involved eye and 10.5 mm in the uninvolved eye. The axial length was 20.22 mm in the involved eye and 20.18 mm in the uninvolved eye. All other patients in whom I have measured the axial length with A-scan ultrasound have shown at least 0.65 mm less measurement in the involved eye. At birth, the horizontal corneal diameter in a normal infant measures up to 10.5 mm.⁴¹ A measurement of more than 12.00 mm in the first year of life is highly suggestive of glaucoma. The mean axial length in a newborn is 16.8 mm.⁴² A retinal fold associated with radial vitreoretinal traction was observed in 70% of Pruitt's patients. Vitreous membranes were observed in 88%, and a retinal detachment occurred in 45% of the patients. A pigmented or dragged macula was sometimes involved with a retinal fold in 70% of the cases. A clear lens was observed in 89% of the cases, vitreous hemorrhage in 3%, and open-angle glaucoma in 3%. Other abnormalities reported with posterior hyperplastic primary vitreous were hypoplastic optic nerve and a failure of the retinal vessels to extend anterior to the equator. As one can see from all of these posterior abnormalities, it was not surprising that most of Pruett's patients had less than 20/200 vision. The

few patients with good visual acuity had minimal involvement. He reported 1 patient with 20/20 vision, but this patient had a normal-sized eye. The only manifestation of PHPV was a membrane of mesodermal elements anterior to the disk extending into the vitreous cavity. The macula was undisturbed leading to a 20/20 eye. This most likely represented an enlarged Bergmeister's papilla. Retinal dialysis and full-thickness macular hole have been seen in cases of purely posterior hyperplastic primary vitreous.⁴³ Rubinstein⁴⁴ reported on 14 cases of posterior PHPV. One patient had 6/9 vision, 6 were 6/18 to 6/60, and the rest were counting fingers to light perception. Preretinal glial nodules, which are felt to be hamartomatous astrocytomas, can be seen as a posterior finding in PHPV.^{45, 46} In my series, which will be discussed in this paper, 21 cases were of the purely anterior type, 10 were purely posterior, and 52 were a combination of anterior and posterior PHPV (Table I).

Since Reese began talking about anterior PHPV and Pruitt and Schepens began to clarify posterior PHPV, I believe that prognostically it is most important to categorize our patients into either posterior PHPV, anterior PHPV, or a combination of posterior and anterior PHPV. Later, as the presentation of the case histories are discussed, it will become apparent that the patients with the purely anterior form have the most reasonable chance for a successful visual outcome from the treatment of their PHPV. The purely posterior cases and those with a combination of anterior and posterior PHPV uniformly have poor visual results, but those with glaucoma or potential glaucoma (having a very shallow anterior chamber) can be treated in order to save the eye from the effects of elevated intraocular pressure.

FINDINGS IN POSTERIOR PHPV

Posterior PHPV has as its hallmark a retinal fold, which can be found in as many as 70% of the patients. The eye is usually microphthalmic but can be of normal size. Leukokoria is present if the PHPV membrane is large enough, dense enough, and forward in the vitreous. Usually the lens is clear but can become cataractous with time if the vessels from the membrane grow forward enough to enter the lens through the posterior capsule. The lens can swell and cause a secondary angle-closure glaucoma, or the lens can be resorbed, leaving a membranous cataract.⁴⁷ Vitreous mem-

TABLE I. PRESENTATIONS OF PERSISTENT HYPERPLASTIC PRIMARY VITREOUS

Purely anterior presentation	21 patients (25.3%)
Purely posterior presentation	10 patients (12.0%)
Combination of anterior and posterior presentation	52 patients (62.7%)

branes are commonly found, and a stalk going from the membrane to the optic nerve head may be seen. If only a hyaloid artery is present (Fig 5), then there is no visual obscuration. If just a Bergmeister's papilla remains, then these patients also will have no visual loss. Patients with posterior PHPV who have been described as having good vision usually have mild involvement with a remnant of the hyaloid artery and/or a somewhat enlarged Bergmeister's papilla with no other posterior pole abnormalities. If the vessel invades the lens, a cataract will form (Fig 6). Often, with posterior PHPV, the patient will have poor vision because there is a tractional retinal detachment of the posterior pole. This may be associated with an abnormal macula. The macula may be hypoplastic with hypopigmentation, there may be a pigment maculopathy, or the macula may be abnormal because it was involved with a retinal fold or tractional detachment of the posterior pole. A hypoplastic or dysplastic optic nerve head may be present. All of these posterior polar abnormalities can be additive in making the odds against the patient with PHPV too great to permit visual rehabilitation. Because of poor vision, strabismus can be present. While glaucoma is usually not a finding in the purely posterior type, I have seen one patient with a purely posterior PHPV develop open-angle glaucoma with a normal angle on gonioscopy. The anterior chamber depth was normal in that patient. The findings in posterior PHPV are summarized in Table II.

FINDINGS IN ANTERIOR PHPV

The patients with the purely anterior variety of PHPV are the ones who have the best chance for a successful visual rehabilitation. In this entity the posterior pole is totally normal with no evidence of a retinal fold. There is no abnormality of the optic nerve or macula. These patients characteristically present with leukokoria very early in life. The white pupil is often a result of the retrolental membrane (Fig 7), or it may also be a result of the effects of the PHPV membrane and a cataractous lens, which occurs when the posterior pole of the lens is invaded by the fibrovascular membrane of the PHPV. The ciliary processes can be quite elongated, as they are drawn into the fibrovascular membrane (Fig 8). The ciliary processes can appear to be of normal size or only mildly elongated if they are pulled only into the periphery of the membrane (Fig 9). Most ophthalmologists feel that the elongated ciliary processes are pathognomonic of PHPV, but Olsen and Moller⁴⁸ have pointed out that this can be seen in any process of retrolental shrinkage. The eye is usually microphthalmic, but this can be subtle, as seen in Fig 10 with a unilateral PHPV.

If the fibrovascular membrane invades the lens, an intralenticular hemorrhage⁴⁹⁻⁵¹ can be produced (Fig 11). Sometimes this can be accompanied by a dilated iris vessel (Fig 12). Some of the membranes have extensive

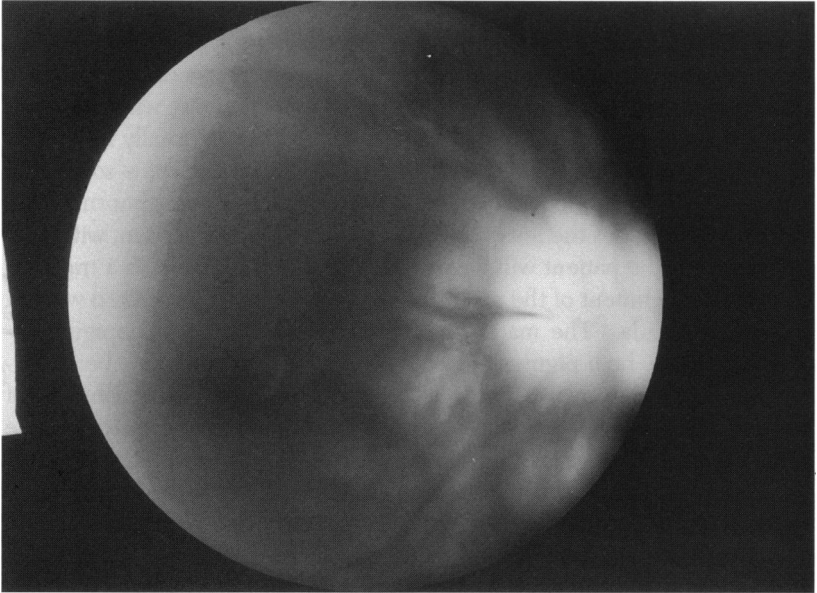


FIGURE 5

Case showing only a hyaloid artery.

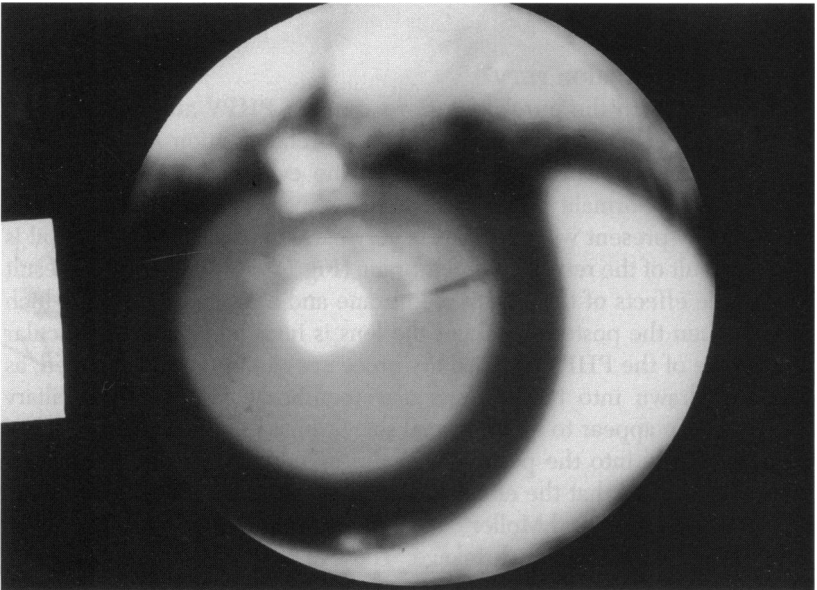


FIGURE 6

Hyaloid artery with mild cataract in eye with 20/40 acuity. Hyaloid artery has invaded posterior aspect of lens.

TABLE II. CHARACTERISTIC FINDINGS IN PURELY POSTERIOR PHPV

Leukokoria
Microphthalmia
Retinal fold
Tractional retinal detachment of posterior pole
Hypoplastic optic nerve
Dysplastic optic nerve
Vitreous membranes and stalk
Pigment maculopathy
Hypoplastic macula
Clear lens
Strabismus

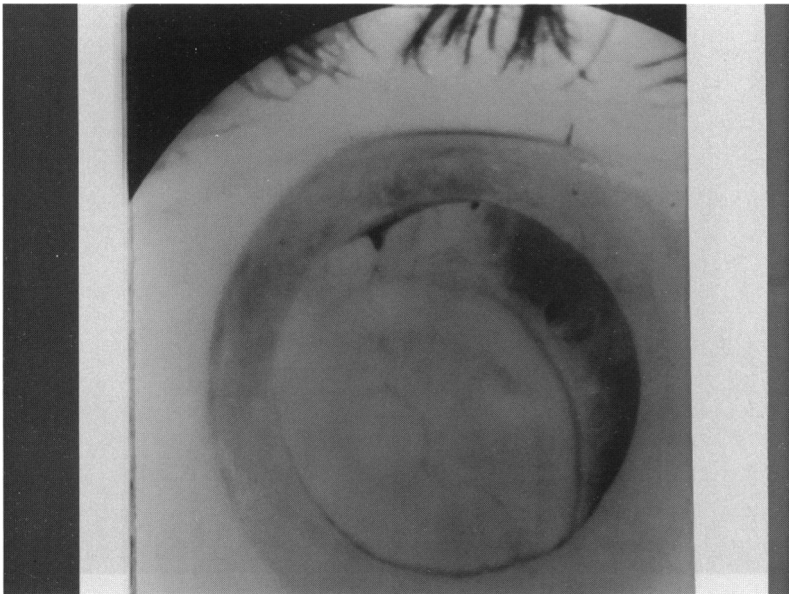


FIGURE 7

Leukokoria due to PHPV membrane with clear lens.

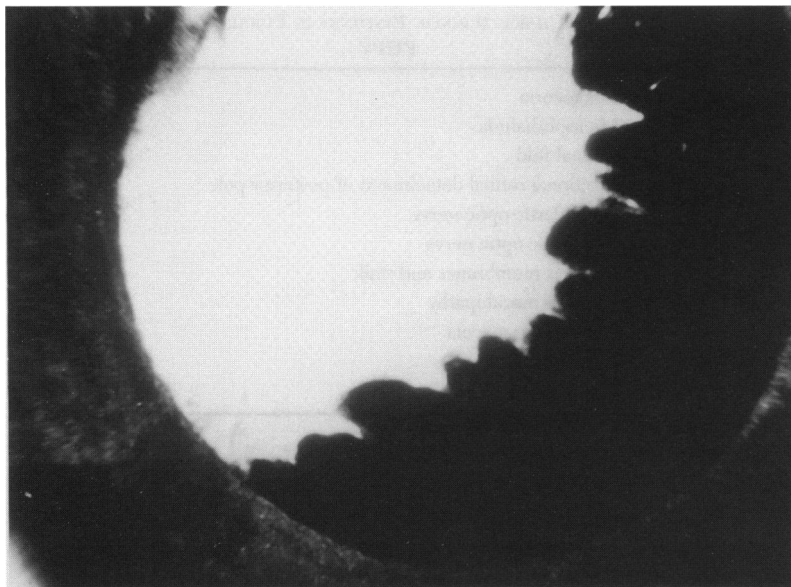


FIGURE 8

Eye with PHPV membrane containing elongated ciliary processes and clear lens.

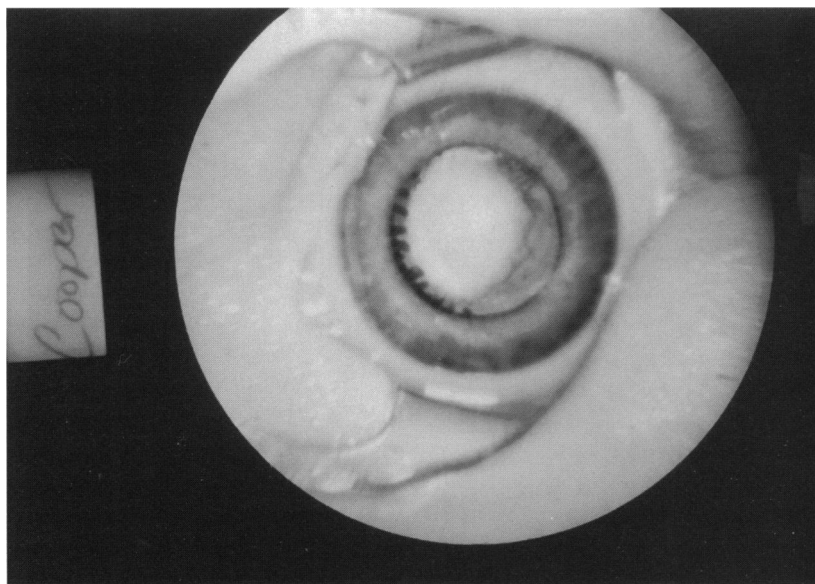
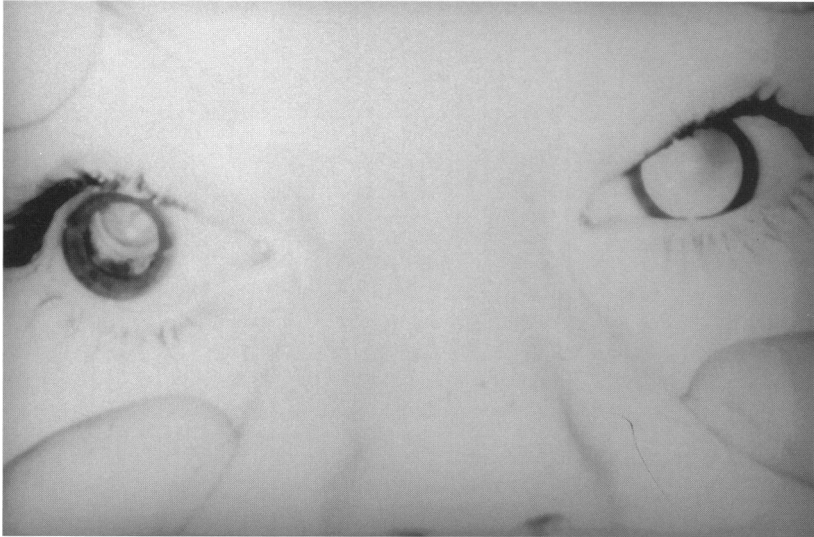


FIGURE 9

Eye with mildly elongated ciliary processes pulled into periphery of PHPV membrane. Cataractous lens is present.

**FIGURE 10**

Mildly microphthalmic eye with PHPV on right side.

development of vessels, but they do not always bleed, as indicated by the eye containing an extensive anterior PHPV membrane with numerous vessels that bleed neither preoperatively nor intraoperatively (Fig 13). These patients often present with a shallow anterior chamber (Fig 14), which may be the cause of a secondary angle-closure glaucoma. The retrolental membrane has invaded the lens and pushed the lens-iris diaphragm forward, causing a secondary angle-closure glaucoma. Extensive posterior synechiae, as well as peripheral anterior synechiae, may form as the chamber shallows.

Strabismus may be present at the initial presentation; or if the infant is too young to have developed strabismus, the family is told to expect its onset. Often these children are seen within a few days of birth when no strabismus is apparent. However, as their fixation matures and as amblyopia is always present, the eye usually becomes esotropic. Occasionally, the patient may develop a dissociative hypertropia in the involved eye. While strabismus is a common presentation, the two most common entities noted by the parents are microphthalmia and leukokoria. The findings in anterior PHPV are summarized in Table III.

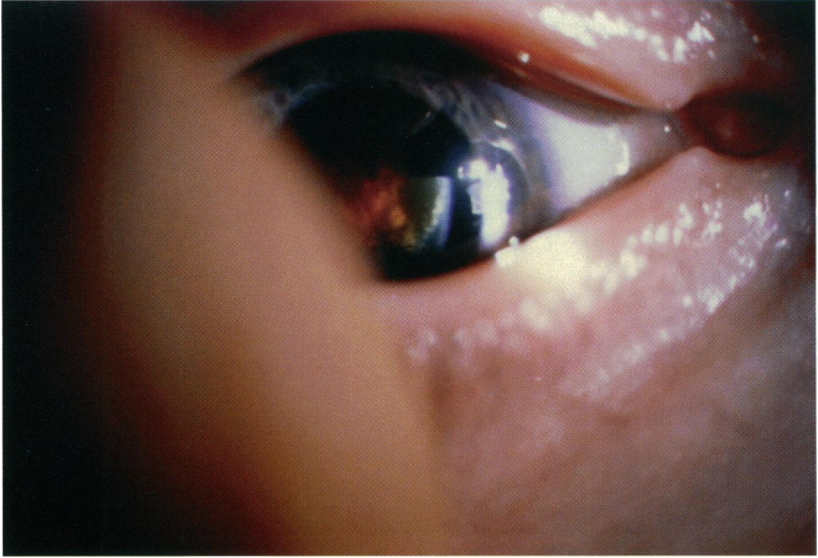


FIGURE 11
Intralenticular hemorrhage secondary to PHPV.

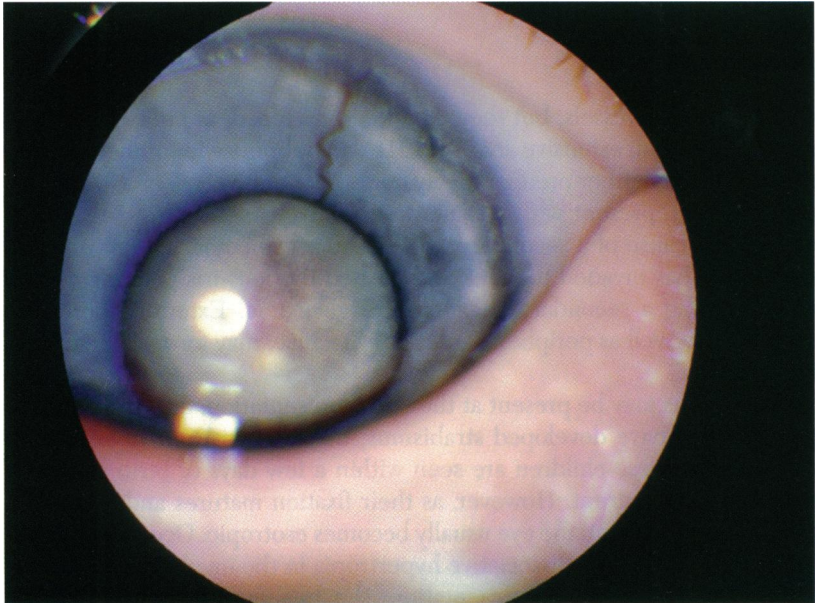


FIGURE 12
Intralenticular hemorrhage with dilated iris vessel in eye with PHPV.

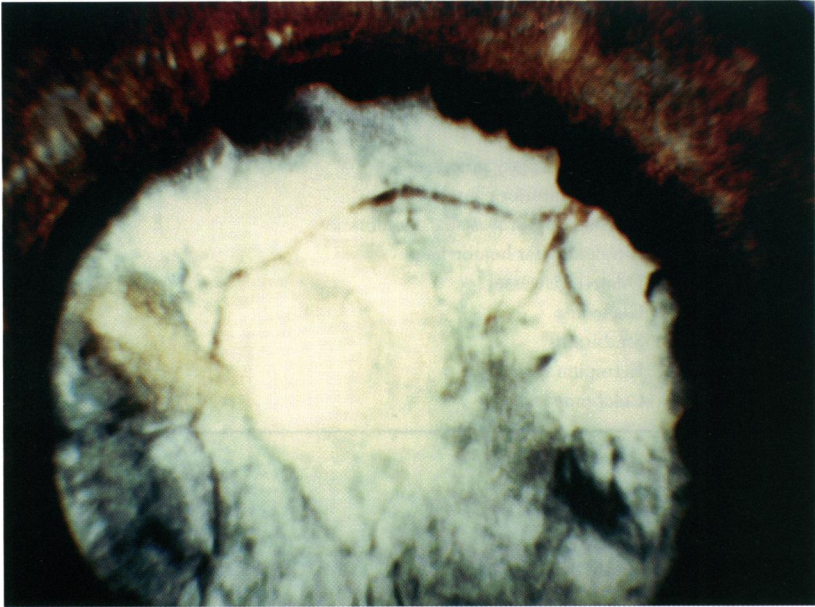


FIGURE 13

Extensive vessels in anterior PHPV membrane.

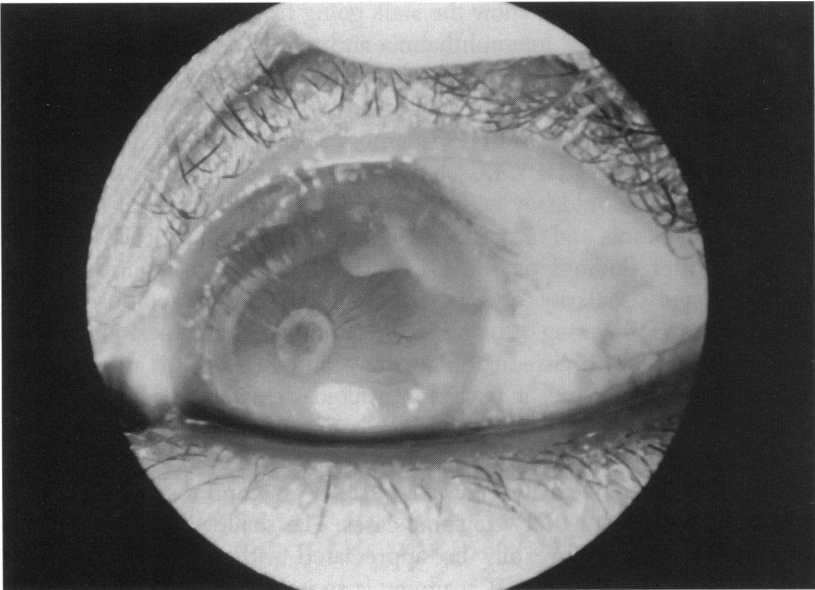


FIGURE 14

Patient who has PHPV with a nonexistent anterior chamber.

TABLE III. CHARACTERISTIC FINDINGS IN ANTERIOR PHPV

Microphthalmia
Leukokoria
Cataract
Elongated ciliary processes
Shallow anterior chamber
Retrolental fibrovascular membrane
Intralenticular hemorrhage
Dilated iris vessel
Glaucoma
Strabismus
Ectropion uvea
Coloboma iridis

RADIOLOGY: ULTRASOUND, COMPUTED TOMOGRAPHY, MAGNETIC RESONANCE IMAGING

In this age of radiologic explosion of new diagnostic machinery, ultrasound, computed tomography (CT), and magnetic resonance imaging (MRI) have come forward to aid in the diagnosis of PHPV. The early diagnosis of PHPV may be difficult when a totally cloudy cornea is present. B-scan ultrasonography can show the stalk going from the posterior pole to the lens, as well as the microphthalmia and retinal detachment that may be present (Fig 15).^{52, 53} Even high technology of color-flow doppler sonography has been used to show the vascular nature of PHPV lesions.⁵⁴ CT scanning also can show the PHPV membrane and has been useful in the diagnosis of bilateral PHPV. Bisset and Towbin⁵⁵ presented the CT examination of a 4-week-old boy with bilateral PHPV with bilateral microphthalmia, leukokoria, and intraocular masses. Mafee and associates⁵⁶ pointed out further findings on the CT study of PHPV. Postcontrast study showed hypervascularity of the vitreous. This can be confused with retinoblastoma. Layering of blood in the lateral decubitus position can be seen in patients with vitreous hemorrhage and PHPV. This retrohyaloid layered fluid represents a posterior hyaloid detachment. Hemorrhage within the vitreous gel does not layer, but if the posterior hyaloid is detached and there is posterior bleeding, then layering in the retrohyaloid space can be seen. A small optic nerve can be seen on the CT scan, and this is associated with PHPV in some cases. The shallowing of the anterior chamber can also readily be appreciated with CT scanning. The absence of calcification on CT scanning is an important point in differentiating PHPV from retinoblastoma, as calcification usually can be seen

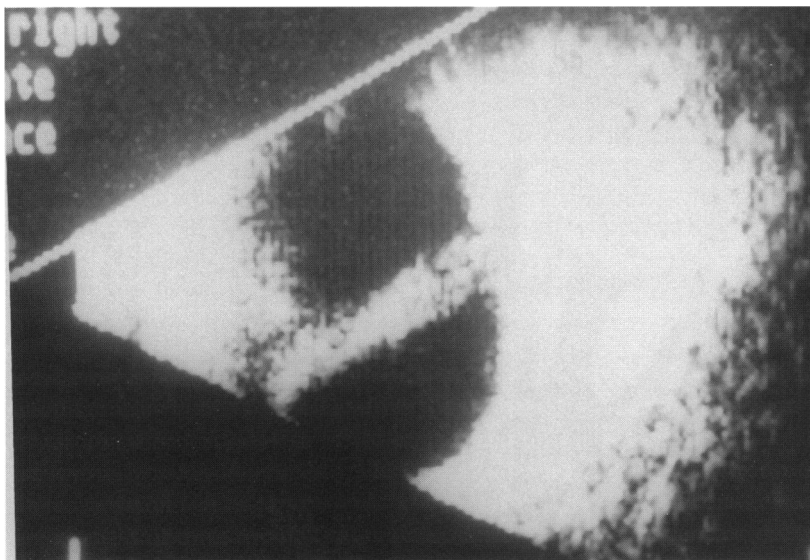


FIGURE 15

B-scan ultrasonography showing microphthalmic eye with PHPV extending from posterior pole anteriorly toward lens.

with retinoblastoma.⁵⁷ Calcification has been reported by Morris⁹ histopathologically in a case with PHPV, but this is quite rare. Mafee and Goldberg⁵⁸ showed the superiority of CT scanning over MRI for the detection of calcification in differentiating retinoblastoma from PHPV. They felt that MRI was superior to CT in differentiating noncalcified retinoblastoma from other disease entities, such as Coats' disease, *Toxocara* infection, retinal detachment, and PHPV. The morphology of these various lesions can be seen clearer with MRI. They discussed a case with bilateral PHPV in a child with Norrie's disease in which the lesions were quite clearly shown on MRI and CT scanning. I have studied a 3-month-old patient with a T1 MRI who had a PHPV in a microphthalmic left eye (Fig 16). A hyaloid artery can be seen running from the posterior pole to the posterior chamber. There is hemorrhage with layering of blood in the posterior vitreous. The anterior two thirds of the vitreous appears white, since it contains more serum than the posterior third, which has a dark appearance because of the presence of blood. No lenticular substance can be seen in the left eye, but only a membrane remains where the hyaloid artery ends anteriorly. A normal lens is seen in the right eye.



FIGURE 16

T1-weighted MRI showing axial view of microphthalmic left eye containing PHPV. Layering of blood in posterior third of vitreous can be seen with hyaloid artery extending from posterior pole to membrane in posterior chamber. (Courtesy Dr John Alarcon, Department of Radiology, Scottish Rite Children's Hospital, Atlanta.)

DIFFERENTIAL DIAGNOSIS OF PHPV

In making the diagnosis of PHPV, every physician must be primarily concerned with the possibility of retinoblastoma. Shields and associates⁵⁹ reported on several hundred consecutive patients referred to their service because of suspected retinoblastoma. Of this group, 58% did have retinoblastoma. In the remaining patients, the most commonly confused diagnoses were PHPV (28% of patients), Coats' disease (16%), and ocular toxocariasis (16%). Howard and Ellsworth⁶⁰ studied 500 consecutive patients referred with a diagnosis of retinoblastoma. In this group, 53% turned out to have some diagnosis other than retinoblastoma. Of 27 other diagnoses listed, the most common was PHPV, followed by retrolental fibroplasia, posterior cataract, coloboma of the optic disc or posterior pole, uveitis, and larval granulomatosis. Buyck and associates⁶¹ reported on 23 children under age 6 with leukokoria. Retinoblastoma and Coats' disease were seen in children only over 8 months of age. (I personally have seen retinoblastoma occurring bilaterally as early as 6 weeks of age and have enucleated an eye for Coats' disease as early as 5 months of age.) When

they highly considered Coats' disease, they recommended examining the subretinal fluid for the presence of fatty exudates, ghost cells, and cholesterol crystals, which were believed to be highly conclusive for a diagnosis of Coats' disease. Tasman⁶² studied 16 children with Coats' disease at the Wills Eye Hospital. The average age of patients with the disease was 10 years. The youngest patient in his series was 2 and the oldest was 16. Two patients had bilateral disease. The extreme subretinal exudation and the areas of telangiectasia separate Coats' disease from PHPV. Sherman and colleagues⁶³ reviewed the CT findings in 2 children who subsequently had 1 eye enucleated, as retinoblastoma could not be ruled out. Each child had unilateral Coats' disease. One child was a 31-month-old boy and the other was a 7½-month-old boy. CT scanning for each child showed increased density within the globe, which enhanced with contrast agent. The investigators believed that they could not differentiate Coats' disease from retinoblastoma on CT examination, primarily because no calcium was seen in either of these eyes. Elevated levels of aqueous lactic dehydrogenase (LDH) are believed by many physicians to be supportive of the diagnosis of retinoblastoma. Other conditions causing an elevated level of aqueous LDH are traumatic hyphema, absolute glaucoma, endophthalmitis, and intraocular cell necrosis. In one report, the aqueous:serum LDH ratio in Coats' disease was 3:1.⁶⁴ A report by Lifshitz and colleagues⁶⁵ presented one case of Coats' disease with a 6:1 ratio of aqueous:serum LDH.

Retinopathy of prematurity can be confused with PHPV, especially if the retinopathy has progressed to the point of a total retinal detachment with the vitreous becoming filled with membranes. Usually the bilaterality and the history of prematurity help in differentiating this entity from PHPV. Some of the cicatricial changes of retinopathy can be difficult to distinguish from PHPV, such as the tractional retinal folds going from the posterior pole to the periphery and the funnel detachment of the retina seen in the late stages of retinopathy of prematurity.^{66, 67}

Ocular toxocariasis, when it has progressed to produce a totally detached retina, can be difficult to differentiate from PHPV and/or retinoblastoma. There was hope at one point that an elevated ELISA for *Toxocara* would differentiate toxocariasis from retinoblastoma, but there are children with retinoblastoma and positive titers for *Toxocara*, meaning that they have been exposed to *Toxocara* in addition to harboring retinoblastoma.⁶⁸ The greatest number of children with ocular toxocariasis are in the 4- to 8-year-old age-group, obviously much older than those with PHPV. This is because children need to be mobile and either crawling or walking in areas contaminated with the feces of infected dogs in order to contract the disease. When the child puts a stick or blade of grass into his or her mouth, and that object has been contaminated with eggs containing the larva of *Toxocara*, then the possibility exists for contracting the disease.

A retinal fold can present in a patient with ocular toxocariasis (Fig 17). It is easy to see how this fold could be confused with a retinal fold occurring in PHPV. Ocular toxocariasis can also present with a huge glial mass coming off of the optic nerve extending into the vitreous (Fig 18). These patients often are referred with a diagnosis of PHPV. Table IV lists the entities confused with PHPV.

TREATMENT OF PHPV

In 1955⁶⁹ Reese referred to PHPV, saying that “he had not been able to discover a single recognizable case in an adult.” He believed that if left untreated, most eyes would develop spontaneous hemorrhage, secondary glaucoma, corneal opacification, and retinal detachment or phthisis. The glaucoma is usually a secondary angle-closure glaucoma associated with swelling of the lens after rupture of the posterior capsule by the invading fibrovascular PHPV. Pupillary block can also be involved in the etiology of the glaucoma. Peripheral anterior synechiae, anterior displacement of the lens-iris diaphragm from the retrolental membrane, retinal detachment, and seclusio pupillae can contribute to the etiology of the glaucoma. While it is true that most untreated cases will eventually need enucleation, there

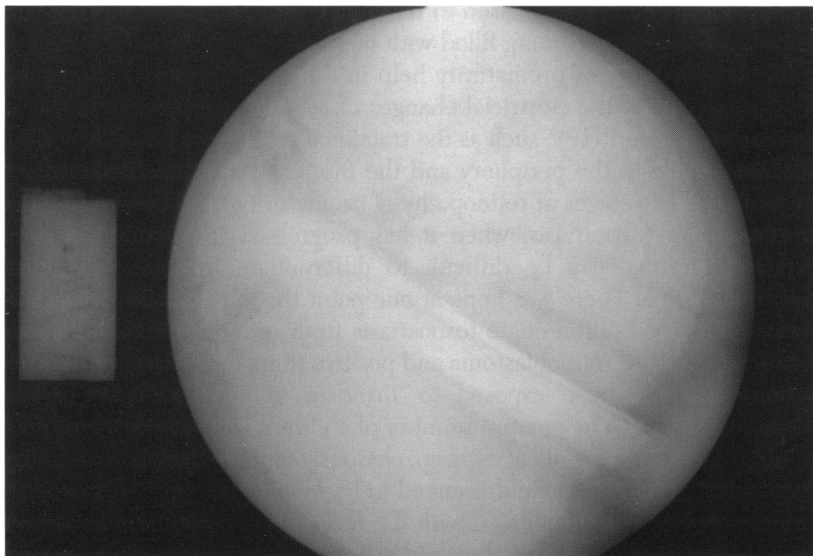


FIGURE 17

Patient with retinal fold secondary to ocular toxocariasis.



FIGURE 18

Patient with large glial mass extending off optic nerve into vitreous. Patient had ocular toxocariasis but was initially referred with a diagnosis of posterior PHPV.

TABLE IV. ENTITIES CONFUSED WITH PHPV

Diseases readily confused with PHPV:

- Retinoblastoma
- Retinopathy of prematurity
- Coats' disease
- Ocular toxocariasis

Diseases rarely confused with PHPV:

- Coloboma of optic nerve
 - Coloboma of posterior pole
 - Uveitis
 - Cataract
 - Myelinated nerve fibers
 - Retinal dysplasia
 - Juvenile xanthogranuloma
 - Norrie's disease
-

have been a few reported patients who have PHPV in adulthood. Zalta and associates⁷⁰ reported on a 61-year-old man with a unilateral PHPV who developed rubeosis iridis and central retinal vein occlusion. The rubeosis predated the central vein occlusion by 6 weeks. Mason and Huamonte⁷¹ performed a pars plana vitrectomy in a 29-year-old man with PHPV. They mentioned increasing the hydrostatic pressure or using diathermy to stop fresh bleeding from the iris, ciliary processes, or the fibrovascular membrane. Spaulding and Naumann⁷² reported on the enucleation of an eye with PHPV in a 22-year-old woman in order to rule out a neoplasm in a blind, painful eye. The eye contained PHPV. Consul and associates⁷³ reported on PHPV in an 18-year-old man with a unilateral PHPV who underwent surgery for strabismus. No glaucoma was present. Vision consisted of only counting fingers in the involved eye.

Maurer and Hiles⁷⁴ reported on 2 cases not treated surgically. The first patient was a 5-year-old with a posterior subcapsular cataract overlying a fibrovascular stalk, which went from the lens to the disk. The posterior pole had only mild distortion. The amblyopic eye with the PHPV improved in vision from 20/80 to 20/30 with patching. Since the cataract was mild and nonprogressive, no surgery was performed. A 3-year follow-up was documented. The second patient was first seen at 2 weeks of age with PHPV involving a unilaterally microphthalmic eye. A total retinal detachment was present. The anterior chamber was shallow, but the lens remained clear during a 4-year follow-up. At that time, no glaucoma had developed. I have followed a patient from age 2 until age 11 who had severe amblyopia and a posterior PHPV in one eye only. The anterior chamber has remained at normal depth. For the first time at age 11, this patient has developed open-angle glaucoma, which has been easily controlled with drops for 2 years. After being followed for 11 years, the patient had no cataract, and the anterior chamber remains fully formed.

Reese recommended a needling and aspiration of the lens followed later by a discission of the membrane. As early as 1908, Collins⁷⁵ recommended a needling of the lens and a couching of the persistent primary hyperplastic vitreous. Acers and Coston⁷⁶ in 1967 operated on a patient with PHPV, performing an aspiration of the lens and then a punch capsulotomy of the fibrovascular membrane at the same surgical setting.

In 1970, Gass⁷⁷ approached the persistent hyperplastic primary vitreous in 2 separate surgical settings. First, he performed an incision and aspiration of the lens. He waited for 1 week for the dilated vessels in the iris and PHPV membrane to decrease in prominence. He then used a limbal entrance to excise the membrane with DeWecker scissors. He believed that if the blood vessels in the membrane were engorged, it was prudent to approach the membrane at a second surgical setting when the vessels had become much less prominent.

In 1976 Peyman and associates⁷⁸ discussed the management of PHPV using a pars plana approach to the vitrectomy. They discussed 2 patients under 4 months of age who underwent this procedure successfully. They advocated early intervention before complications such as bleeding into the lens or vitreous or traction developed on the ciliary processes. They stressed that while visual results may be poor, the release of the antero-posterior traction caused by the PHPV could allow the eye to grow and achieve reasonable cosmesis.

Nankin and Scott⁷⁹ reported on the use of the Douvass rotoextractor to treat PHPV. They used a limbal approach on some eyes and the pars plana approach on others. They stressed certain absolute indications for surgical intervention. They included shallowing of the anterior chamber, intralenticular hemorrhage, progressive traction on the ciliary body, and elevation of intraocular pressure. They stressed that severance of the hyaloid artery by the rotoextractor can aid in hemostasis, because the resulting contraction of the vessel leads to a removal of the blood source of the fibrovascular membrane.

Stark and colleagues⁸⁰ in 1983 recommended a limbal approach to the treatment of persistent hyperplastic primary vitreous. They recommended this approach because they believed that the pars plana was often narrow and that there may be an anterior retinal insertion or incarceration of retinal tissue within the retrolental membrane. In the only eye to develop a retinal detachment after surgery in their series, a pars plana approach had been used. They used vitreous suction-cutting instruments to remove the intracular membranes.

Karr and Scott² reported on a series of 48 PHPV patients from the University of Iowa. Twenty-five were managed nonsurgically, with 23 obtaining a poor visual result ($\leq 5/200$). Twenty-three underwent surgery. Five had no postoperative rehabilitation of vision. Eight of the remaining 18 surgical patients achieved 20/200 or better. A few of their patients obtained 20/30 vision with aggressive contact lens and amblyopia therapy. The investigators stressed that the most important indicator for future visual potential is the patient's age at presentation, since the younger patients obtained the best results. The group with an average age of 2.4 months all obtained 20/200 or better, while the group with the average age of 4.3 months obtained 20/300 or worse.

Scott,⁸¹ in 1986, stressed that PHPV is second only to juvenile xanthogranuloma in causes for spontaneous hyphema in children. These two were followed by retinoblastoma, retrolental fibroplasia, blood dyscrasias, contusion, vascular anomalies of the iris, and iridocyclitis. He pointed out that some of the good visual results are attributed to the fact that the lens was clear for a period of time after birth, allowing normal or fairly normal vision to develop. As the membrane invades the lens and causes a cataract,

then vision which may have been fairly good, drops off. In 1989, Scott and associates⁸² updated his follow-up of 48 patients with PHPV. He had several in the 20/30 and 20/40 range.

Volcker and colleagues⁸³ also stressed an anterior approach via the limbus in the surgical treatment of PHPV, since they believed that the retina can extend up to the pars plicata in some of these patients. In one of their patients treated surgically for bilateral PHPV, 1 eye obtained 20/30 and the other 20/200.

Frezzotti and coworkers⁸⁴ chose the pars plana approach in 2 infants with PHPV. They believed that even with microphthalmic eyes, the retina would not be injured if the surgeon made the sclerotomy only 3 mm posterior to the limbus. Federman, and associates⁸⁵ reported on 15 patients with PHPV. At the beginning of each surgery, an attempt was made to localize the pars plana with transillumination. This was usually unsuccessful. The sclerotomy was made between 2.0 mm and 3.5 mm posterior to the limbus. In 1 case, a portion of peripheral retina prolapsed through the sclerotomy. In a second, 2 holes were made in the peripheral retina where the vitrectomy machine had passed. In a third, a 200° dialysis was produced. They believed that these complications were due to the fact that the retina inserted directly into the pars plicata in these cases. In one bilateral case, results were good and the patient was wearing contact lenses bilaterally. They concluded that surgery was not reasonable in eyes with a corneal diameter less than 8 mm. In the surgically treated group, all the eyes showed some growth postoperatively. They believed that this was because surgery released the anterior-posterior tractional component, stopping posterior-tractional detachments. The surgery also released the transverse and circumferential traction on the ciliary body, preventing phthisis. All but 1 of the 9 eyes in the nonsurgical group showed progressive deterioration. They used the SITE vitrectomy instrument in this series.

Shami and coworkers⁸⁶ presented a limbal approach for surgery on a patient with PHPV. In the 6-month-old boy, the lens was removed followed by the removal of a retrolental membrane. An open funnel-shaped retinal detachment with a fibrovascular stalk extending to the posterior pole was seen. One month postoperatively, a previously detached retina was completely flat, as the anteroposterior traction had been removed.

Kingham⁸⁷ discussed operating on an 11-year-old girl who had a tractional retinal detachment associated with a persistent hyaloid artery. The artery, contained in a stalk, went from the optic disk to the lens. Upon severing the attachment of the stalk at the level of the posterior lens capsule and applying a No. 276 Silastic exoplant with a No. 40 band, the retina reattached. Visual acuity improved from 3/200 to 20/200. No retinal breaks were seen.

The controversy of the anterior approach versus the pars plana approach does not confine itself to the United States. Raspiller, and associates⁸⁸ discussed the anterior approach for PHPV treatment in the French literature in 1981. In 1992, Mannot and Assi⁸⁹ reported in the French literature on the pars plana approach for lensectomy and vitrectomy for PHPV. Both papers reported good results.

Not only does the physical obstruction of the visual media present a problem in persistent hyperplastic primary vitreous, but once the media has been cleared, the fight with amblyopia begins. In 1977, Ryan and Maumenee⁹⁰ reported disappointing results in 8 cases of unilateral cataract operated on in the first 2 years of life. These patients were treated with contact lenses within 3 weeks of the surgery and then treated with patching. None achieved better than 3/200 vision. However, only 2 were operated on in the first 4 months of life. Frey and colleagues⁹¹ reported on 50 cases of monocular cataract in children operated on by the aspiration technique followed by contact lens fitting and occlusion of the phakic eye. While some of the eyes did not have good visual results (especially rubella cataracts), they did report 1 child who obtained 20/30 and 2 who obtained 20/40 vision.

Even if good vision is not obtained after the surgery for pediatric monocular cataract, the optical correction of the aphakia can produce a significant improvement in the binocular alignment. Kodsí, and associates⁹² reported that the optical correction of pediatric monocular aphakia with a contact lens improved the binocular alignment in 28% of cases. They concluded that the binocular alignment should be taken into consideration when deciding about the value of prescribing a contact lens for aphakia, even if the vision is not significantly improved. Two of their patients operated on in the first month of life fused at near the Worth 4-dot test when wearing their aphakic correction and suppressed without their contact lens. None of their 25 patients demonstrated any stereopsis. Figure 19 shows a 5-year-old girl with orthophoria who is wearing a soft contact lens on her operated left eye. She is aphakic from surgery for PHPV. Her vision is only 20/400 in this PHPV eye, but the eyes are straight with her contact lens. She has esotropia without her contact lens. Figures 20 and 21 show a 6-year-old boy aphakic in his left eye with exotropia without his contact lens (Fig 20) and orthophoric with his contact lens (Fig 21). He has 20/25 vision in his aphakic eye. Gregg and Parks⁹³ reported on an 8-year-old patient who had undergone surgery for a congenital cataract at day 1 of life. The patient had been fitted at the time of surgery with a hydrophilic lens, and patching was started. Visual acuity at age 8 was 20/25 in the operated eye with contact lens correction, and the patient was tested to have 50 seconds of arc of stereo acuity via the Titmus test.

I am currently using two different contact lenses for the rehabilitation



FIGURE 19

A 5-year-old patient with orthophoria. She is wearing an extended-wear soft contact lens on left eye. She has had a vitrectomy with a stalk dissection for PHPV. Her vision is only 20/400 owing to abnormalities of posterior pole.



FIGURE 20

Six-year-old patient with left exotropia. He has aphakia OS and is not wearing contact lens.



FIGURE 21

Same patient as in Fig 20 is now orthophoric wearing his aphakic correction in a gas-permeable hard contact lens OS.

of vision during the newborn period. The first is the Silsoft lens, which is a 100% silicone polymer, and is used as an extended-wear lens. It is much more rigid than most soft lenses, and therefore handling is easier for most parents. It is available in base curves of 7.5, 7.7, 7.9, 8.1, and 8.3 mm. It comes in 2 diameters, 11.3 mm and 12.5 mm. It is available in 1-diopter steps from +12 to +20, and then it is available in 3-diopter steps from +23 to +32 diopters. Levin and coworkers⁹⁴ have reported good results in using this lens for pediatric aphakia. I have also been satisfied with this lens and have used it in children who have a normal-sized cornea with moderate corneal readings in the 4200 to 4700 range. If the cornea is small, as is often the case in PHPV, then I use a gas-permeable hard contact lens. If the cornea is steep, then a gas-permeable contact lens can be designed at any steepness. I have seen many newborns with corneal readings in the 5400 to 5900 range. I try to obtain a lens with a high Dk value. Oxygen permeability is an intrinsic quality of a lens that describes how well oxygen can get into that particular material. It is called Dk. Oxygen transmission Dk/L is an extrinsic quality that varies with lens thickness.⁹⁵ As the aphakic lens is quite thick, I choose a lens with a high Dk, such as the fluroperm 92, which has a Dk of 92. This lens is a fluorinated silicone acrylate con-

sisting of a combination of silicone, polymethylmethacrylate, and fluorine. I have also used the Boston IV lens with a Dk of 26.3.

The measurements for the contact lens fitting are taken at the time of the original surgery with the infant placed either in the lateral decubitus position or in an infant carrying seat (Fig 22). The patient is intubated and secured sitting up so that keratometry readings can be taken. An infant speculum is used to keep the lids open. Balanced salt solution is added frequently to keep the cornea moist. Lately we have been using the Alcon automated keratometer, which can be used with the patient asleep in the supine position, as in Fig 23. Frequent changes in the lens parameters need to be made in the first year of life. Inagaki⁹⁶ found the mean keratometry value for several prematures to be 49.50 and for mature babies to be 47.00. He followed 8 newborns for 12 weeks and found the average keratometry readings to change from 49.01 \pm 2.18 at 2 weeks of age to 45.98 \pm 2.15 at 4 weeks of age to 44.60 \pm 1.97 at 8 weeks of age to 44.05 \pm 1.70 at 12 weeks of age. Isenberg and coworkers⁹⁷ showed a mean axial length of 16.2 \pm 0.7 mm for 16 infants measured at 39 to 41 weeks after conception. The axial length ranged from 11.9 mm at 28.5 weeks to 18.8 mm at 48 weeks postconception age. Most of the growth in the eye was in the length of the vitreous cavity. Since the axial length and the corneal curvatures are changing so much in the first few months of life, it is vital that frequent refractions and corneal measurements be made in order to keep the child in the proper lens. I have seen as much as 10 diopters in change of refraction occur in the first year of life as less hyperopic correction was required. This has been observed without the eye becoming glaucomatous. Of course, if one sees a large loss in the refractive power required in the lens during the first year of life, one must watch for glaucoma associated with a rapid increase in the length of the eye.

The fitting of a contact lens and the beginning of amblyopia therapy early in life are critical if a reasonable visual result is to be obtained. The work of von Noorden^{98,99} with monkeys showed that between birth and 9 weeks of life, an irreversible amblyopia could be produced by suturing the lids closed. Lid closure at 12 weeks of age did not produce amblyopia. He showed that brief closure of the lids for only 2 to 4 weeks during the susceptible period could produce severe amblyopia. He showed histologic evidence of significant reduction in cell section areas of the lateral geniculate nuclei that received input from the deprived eye. These studies have been extrapolated to the human to make us aware of the profound amblyopia that can be produced from occlusion of the visual axis early in life. Parks¹⁰⁰ felt that the amblyogenic problems from a scarred posterior capsule were so prevalent that he recommended a primary capsulectomy at the time of surgery for cataracts in infants and children. He pointed out that the incidence of cystoid macular edema might be greater in doing a

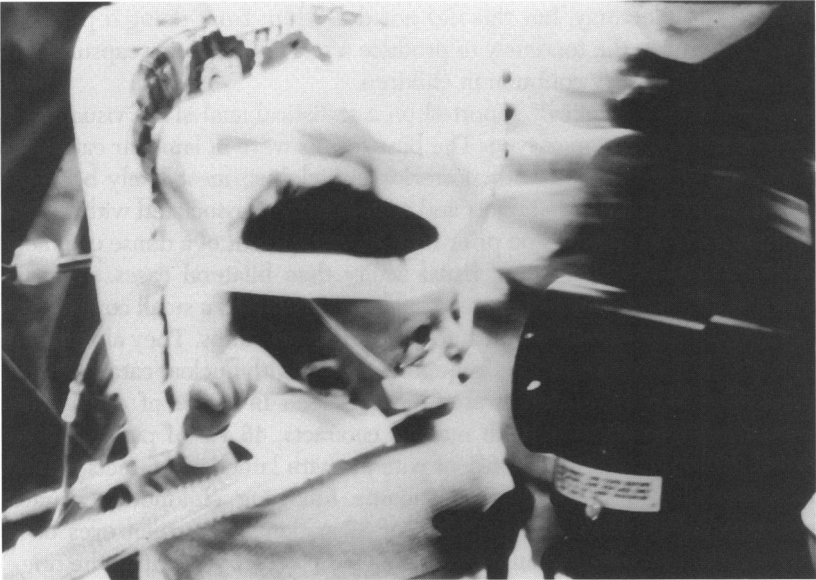


FIGURE 22

Keratometry readings being taken in 3-week-old infant while under general anesthesia. Patient is secured in infant carrying seat while intubated. After measurements were taken, surgery for PHPV was performed.

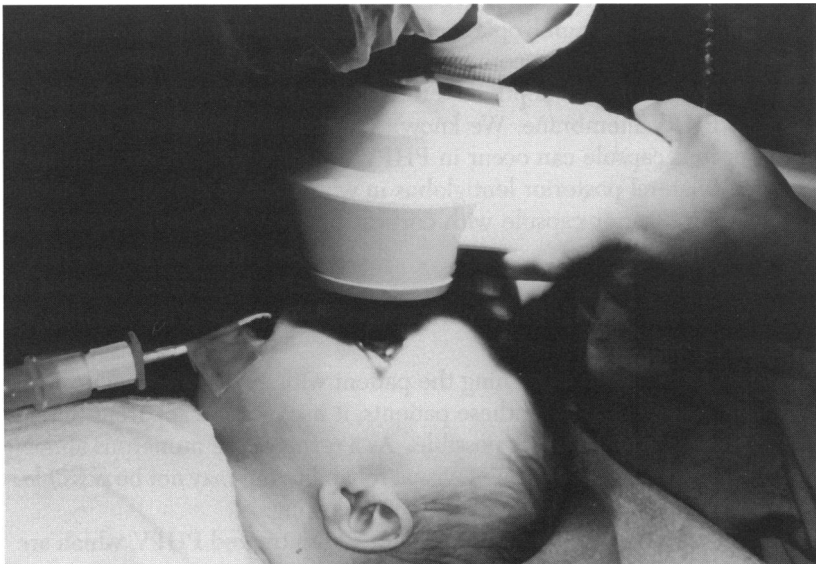


FIGURE 23

Alcon automated keratometer is used to take corneal measurements with infant intubated and in supine position.

primary capsulectomy, but this did not deter him from doing a primary capsulectomy, as the tendency to produce a scarred posterior capsule and amblyopia were very common in children.

Parks and associates¹⁰¹ reported on a statistical analysis of visual outcomes related to cataract type. The best results were in lamellar cataracts and cataracts associated with posterior lentiglobus, most likely because these were progressive cataracts and probably were associated with some period of good central vision prior to the development of a dense cataract. Unilateral cases had a lower visual acuity than bilateral cases. A small cornea was associated with poorer visual outcomes, and a small cornea was also associated with a nuclear cataract and PHPV groups. They also found that glaucoma occurred more frequently in eyes with nuclear cataracts and PHPV. The prevalence of strabismus was seen in 100% of cases with PHPV, 65.5% of patients with nuclear cataracts, 48.4% of patients with posterior lentiglobus, and 21.8% of patients with lamellar cataracts. Mills and Robb¹⁰² also reported on glaucoma following childhood cataract surgery. They reported on 125 eyes of 82 patients. Fourteen eyes of 9 patients developed open-angle glaucoma 5.3 to 13.1 years after the original surgery. The greatest risk for developing glaucoma was in children under age 1 at operation. Additional risks included eyes with microcornea, congenital rubella syndrome, and eyes with poor dilation to cycloplegics. The investigators stressed that these children need to be followed for a long time after surgery. I am currently following a child who had a purely posterior PHPV with amblyopia secondary to a dysplastic disc. At age 8, he developed open-angle glaucoma for the first time. The anterior chamber has remained of normal depth, and there has been no invasion of the lens with the PHPV membrane. We know that a spontaneous rupture of the posterior lens capsule can occur in PHPV. This has also been reported in a case of bilateral posterior lentiglobus in which both lenses showed rupture of the posterior capsule with cortical material in the anterior vitreous.¹⁰³

GOALS IN THE TREATMENT OF PHPV

While all physicians approaching the patient with PHPV desire a positive visual result after managing these patients, it has become apparent that in many patients this goal is not possible. As a result of the numerous abnormalities of the posterior segment, visual rehabilitation may not be possible. Therefore the goals in treatment are the following:

1. Save the eye from the complications of untreated PHPV, which are mainly glaucoma and phthisis.
2. Try to give the patient a black pupil so that the eye will look as normal as possible. Here, a lensectomy to remove a cataractous lens and a

membranectomy are suggested to produce the normal pupil. In patients in whom this is not possible, the use of a cosmetic contact lens to give a black pupil is necessary. Strabismus surgery is performed mainly for cosmesis, since fusional potential is very poor in these patients.

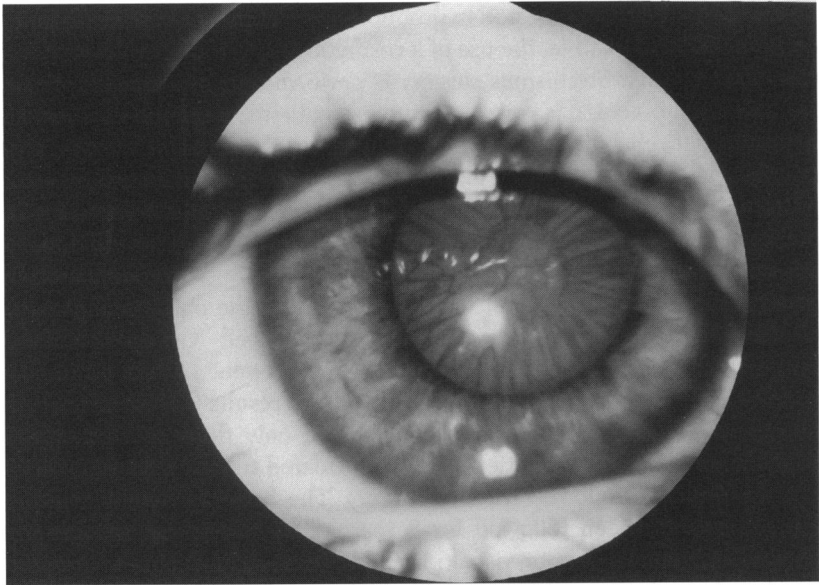
3. Visual rehabilitation is possible with eyes that are fairly normal in structure after the lensectomy and membranectomy. In these patients the posterior pole has a normal anatomy or fairly normal anatomy. Use of an aphakic contact lens and amblyopia therapy are the guidelines for achieving this objective.

The single most important factor in predicting the outcome in these patients is whether or not the posterior pole is involved. Counting fingers vision was the best result for any patient who underwent a lensectomy and vitrectomy if the posterior pole was involved. All results that were 20/100 or better were observed in patients who had only the anterior PHPV involvement. Of these 83 patients, 80 were treated surgically. There were 3 who have been followed without surgery. The first was a 1-month-old patient who has been followed for 8 years. She had a purely posterior PHPV but had a very poor visual prognosis, since she had a retinal fold running through the macula. She also had a dysplastic optic nerve. Her anterior chamber has remained full, and there is no evidence of glaucoma. Her vision in this eye is only hand motions at 1 foot.

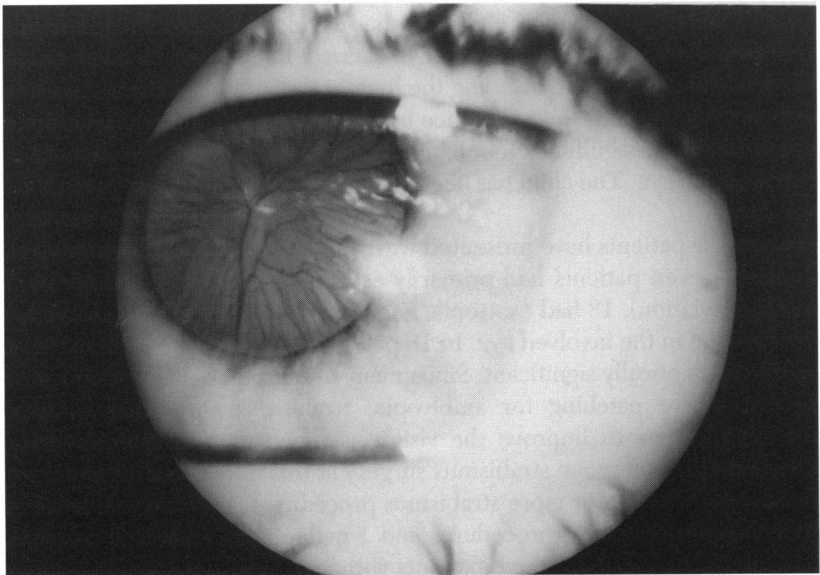
The second patient is now 13 years old and also had only a purely posterior PHPV with a dysplastic optic nerve. The anterior chamber has remained well formed. This child now has developed open-angle glaucoma, which is controlled with drops. No cataract has developed, and there is no anterior membrane invading the lens.

The third patient, followed since infancy, has a remnant of the tunica vasculosa lentis engulfing the lens bilaterally (Figs 24 and 25). Vision is 20/60 in each eye. The child has been followed for 9 years with no cataract formation.

All these patients have presented with or developed strabismus (Table VI). Fifty-seven patients had primarily esotropia (some also had a small vertical deviation), 18 had exotropia, and 8 presented mainly with a vertical deviation in the involved eye. In 18 patients, the deviation is very small and not cosmetically significant. Since many of these patients have undergone extensive patching for amblyopia, strabismus surgery has been delayed in order to improve the vision as much as possible. Only 30 patients have undergone strabismus surgery at this time, and 10 of the 30 have undergone two or more strabismus procedures. Three patients have undergone 3 strabismus procedures and 1 patient has undergone 4 strabismus operations. All of the 8 patients with vertical deviations had a dissociative hypertropia. Dissociative hypertropia is a condition that is almost always present bilaterally. In many of the bilateral cases, it is not unusual

**FIGURE 24**

Right eye showing remnant of the tunica vasculosa lentis and 20/60 vision.

**FIGURE 25**

Left eye of same patient as in Fig 24 showing remnant of tunica vasculosa lentis and 20/60 vision. No surgery has been performed.

to have the dissociation more prominent in 1 eye. In the series of 8 patients with a dissociative hypertropia, 7 showed the dissociative hypertropia only in the PHPV eye. One patient also showed the dissociative hypertropia in the uninvolved eye. This was only present when covering the normal eye while having the patient fixate with the PHPV eye. While none of these patients have developed any depth perception as measured on the Titmus test, the parents of these patients continue to be satisfied with the surgical procedures to straighten the eyes.

Of the 80 operated cases, only 15 have 20/200 or better. There was 1 case with 20/200, 2 cases with 20/100, 2 with 20/70, 1 with 20/50, 3 with 20/40, 5 with 20/30, and 1 with 20/20. All these patients had the anterior form of PHPV only. All 15 were operated on in the first month of life and were fitted with an aphakic contact lens within 2 weeks of surgery. Nine of these 15 were fitted with the aphakic contact lens within 1 week of surgery. The other 6 had to wait 2 weeks because their postoperative inflammation was greater, and 2 weeks were required to obtain a clear enough media to fit the contact lens and begin amblyopia therapy. Amblyopia therapy was started with patching all but 1 or 2 waking hours during the first month of life, with half-time patching continued until age 9. All 15 have been followed for at least 9 years, and 3 have been followed for 18, 20, and 22 years, respectively. Only 2 of the 15 had a normal-sized eye; 13 were microphthalmic. In all, a limbal approach with the bimanual technique was used, with an infusion needle in one hand and a vitreous suction-cutting instrument in the other hand. A total of 21 patients presented with a purely anterior PHPV. Only 1 was followed without surgery. The results of 15 have been discussed above. The remaining 5 who underwent surgery have visual acuities in the counting-fingers-at-one-foot range because of amblyopia.

A total of 10 patients presented with a purely posterior PHPV. Two were followed without surgery, as mentioned previously. One is now 13 years old and has developed open-angle glaucoma controlled with drops. He has dense amblyopia in his involved eye. Another patient refused surgery but has not developed glaucoma. The other 8 patients were treated with a pars plana lensectomy, membranectomy, and vitrectomy by a vitreoretinal surgeon. All of these patients have retained their eyes without glaucoma, but none has vision better than hand motions. This was because of the pathology of the posterior pole, which prevented the development of usable vision.

This series included 52 patients with a combination of anterior and posterior PHPV. This was the most common presentation for PHPV. All in this group were treated with surgery. Ten have undergone a pars plana lensectomy, membranectomy, and vitrectomy. Forty-two have undergone a limbal approach involving a lensectomy, membranectomy, and anterior

vitrectomy. All 52 have visual acuities in the range of no light perception to hand motion vision only. Two have developed phthisis and wear a prosthetic shell over their eye. They are pain-free. Two have uncontrolled glaucoma but are tolerating pressures in the 50s with only light perception vision.

Only 2 patients in this series have the PHPV involvement of both eyes (Table VII). These 2 children are otherwise normal in their mental and physical development. They each have only light perception vision in both eyes.

Twelve patients had a normal-sized eye. One patient had a buphthalmic eye, because secondary glaucoma had set in before any treatment was started. The remaining 70 patients had a small eye (Table VIII). The criterion for micropthalmia was based on corneal diameter only. The upper limits of normal at birth was placed at 10.5 mm. Hence, micropthalmia was considered with a corneal diameter of 9.5 mm or less with the smaller eye measuring at least 1 mm less than the normal-sized eye. From 1973 to 1992, this was the method used. Since 1992, axial lengths are routinely taken in the operating room at the time of the surgery using biometry based on ultrasonography. Since performing A-scan biometry, all eyes (except one) believed to be smaller via corneal measurements were also found to measure at least .65 mm less axially than the fellow eye. The one exception was in a PHPV eye whose corneal diameter measured 1 mm less than the uninvolved eye, but axially actually measured .12 mm longer.

CASE PRESENTATIONS

Case 1

A 1-week-old baby girl had a unilateral PHPV with a purely anterior presentation. Surgery was performed at 2 weeks of age, and on the first postoperative day only an injected sclera was seen (Fig 26). A limbal approach had been used for her lensectomy, membranectomy, and anterior vitrectomy. Four days postoperatively she was fitted with a gas-permeable contact lens (Fig 27), and patching was begun on her normal right eye (Fig 28). She wore the patch full-time for 1 week and then one-half time. At a 4-year follow-up visit, orthophoria was present and fixation was found to be good, central, and maintained with the operated left eye. At age 6, her vision was measured at 20/30 in each eye.

Case 2

A 3-month-old baby girl had surgery via a limbal approach for an anterior PHPV. On the second postoperative day she presented with straight eyes (Fig 29). Her involved eye was of normal size. One week later she was wearing a continuous-wear soft lens (Fig 30), and patching was started for amblyopia therapy (Fig 31). At a follow-up visit at age 19, she had clinically straight eyes (Fig 32). Her best corrected visual acuity was only 20/200. She has only a 2-diopter esotropia and no glaucoma.



FIGURE 26

A 2-week-old infant on first postoperative day. She has undergone lensectomy, membranectomy, and anterior vitrectomy for an anterior PHPV in left eye.



FIGURE 27

Same patient as in Fig 26 on fourth postoperative day wearing a gas-permeable hard contact lens on left eye.



FIGURE 28

Same patient as in Fig 26 and Fig 27 with patching of normal right eye started on fourth post-operative day. She has contact lens on left eye.

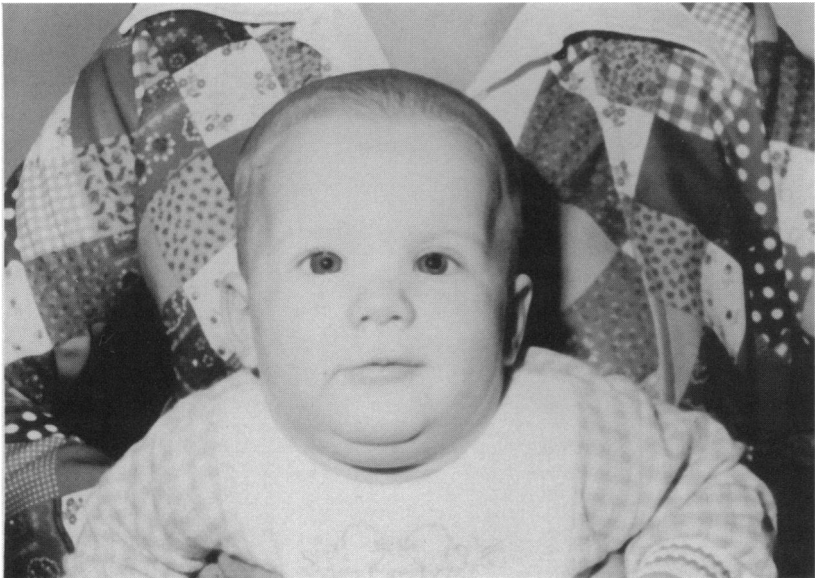


FIGURE 29

This 3-month-old child 2 days following surgery for anterior PHPV in left eye.



FIGURE 30

Same patient as in Fig 29 1 week after surgery. She is wearing a continuous-wear soft contact lens on left eye.



FIGURE 31

Same patient as in Fig 29 and Fig 30 with amblyopia therapy starting 1 week after surgery with patching of normal right eye.



FIGURE 32

Same patient as in Fig 29 through 31 at age 19. She has only a 2-diopter esotropia and straight eyes clinically.

Case 3

This patient was first seen at 2 weeks of age and was found to have an anterior PHPV in a microphthalmic eye. The child was held tightly and examined with a gonio lens to look at the angle (Fig 33). This method is feasible with cooperative parents who will restrain the child during the examination. Typically, the child cries, but if held firmly, a good look at the angle is possible. This patient underwent a limbal lensectomy, membranectomy, and anterior vitrectomy. Keratometry readings were taken with the patient in the left lateral decubitus position under general anesthesia (Fig 34). The forehead rest was removed to make the procedure easier. This patient was treated with a gas-permeable contact lens for his microphthalmic left eye. Patching was continued until age 9, when his vision was 20/30. He obtained straight eyes after esotropia surgery by wearing a gas-permeable hard contact lens (Fig 35).

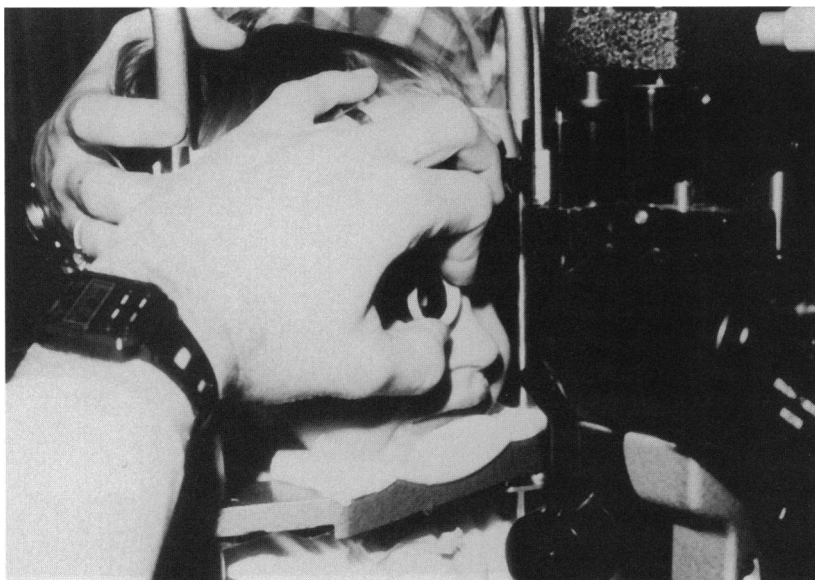


FIGURE 33

Gonioscopy performed in infant who is firmly held in parent's lap.

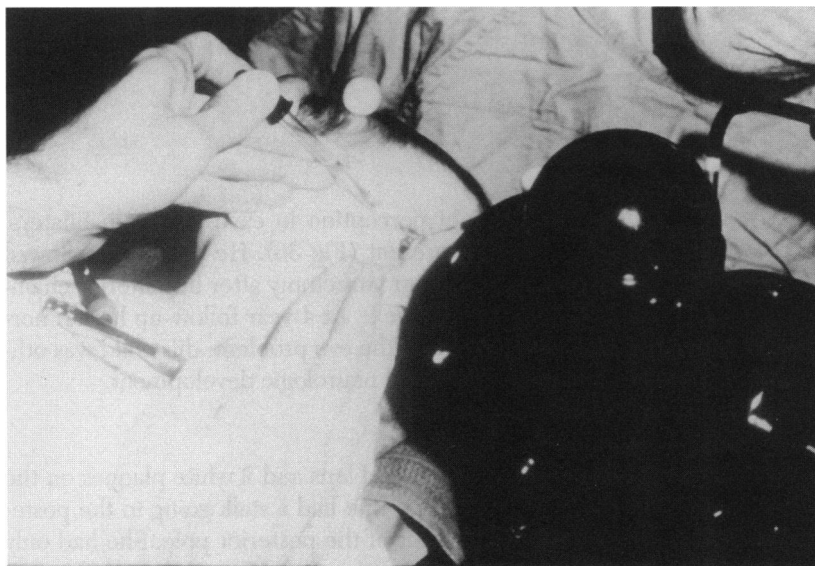


FIGURE 34

Keratometry readings are taken under general anesthesia with patient placed in left lateral decubitus position while intubated. Forehead rest has been removed to make readings more accessible.



FIGURE 35

A 9-year-old patient who had surgery for PHPV at 3 weeks of age. His eyes are straight after esotropia surgery, and visual acuity is 20/30 in his microphthalmic left eye.

Case 4

A 1-year-old child had only light perception in each eye from bilateral PHPV. Bilateral leukokoria was present (Fig 36). He has been followed with bilateral lensectomy and anterior vitrectomy after his anterior chambers became almost nonexistent at age 2. At 4-year follow-up he has normal pressures. With the exception of the eye problem, this child was otherwise totally normal and has a normal neurologic development.

Case 5

A 9-year-old presented with an absorbed lens and 2 white plaques on the posterior capsule secondary to PHPV. She had a stalk going to the posterior pole and a tractional detachment of the posterior pole. She had only hand motion vision at 1 foot. Her pressure was 17 in this eye. She presented with 2 plaques on the posterior capsule and a membranous absorbed lens (Fig 37). No treatment was offered other than polycarbonate plastic lenses for protective purposes for the good eye.



FIGURE 36

A 1-year-old patient with bilateral leukokoria secondary to bilateral PHPV.

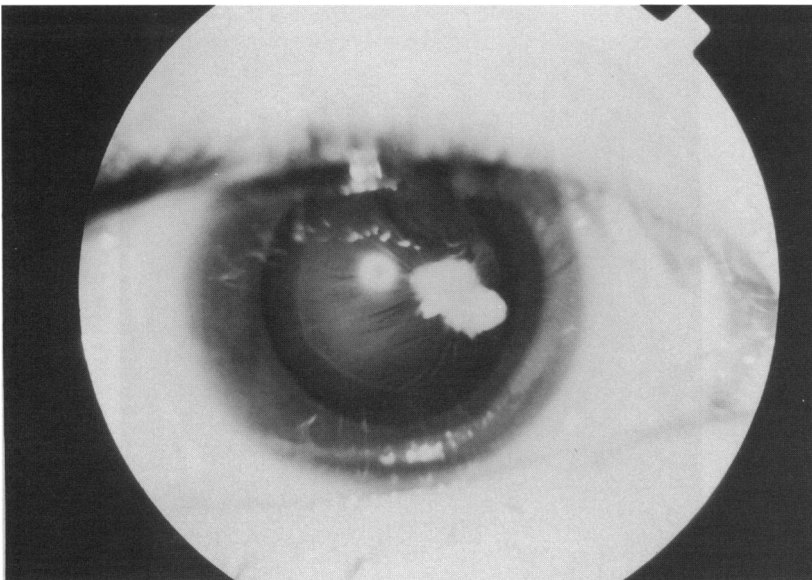
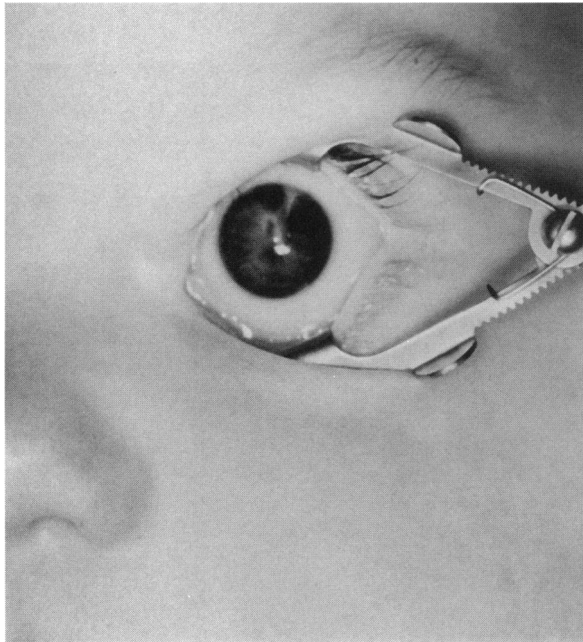


FIGURE 37

A 9-year-old patient with membranous lens after absorption due to PHPV. Two plaques are seen on posterior capsule.

Case 6

At 3 weeks of age, this patient was operated on for a unilateral anterior PHPV via a limbal approach. At 4 weeks of age, she was wearing a gas-permeable soft contact lens, and patching for amblyopia was started. A large opening was made with the ocutome in the posterior capsule at the time of the original lensectomy, membranectomy, and anterior vitrectomy. At 3 months of age, a dense membrane had covered the entire pupil (occlusio pupil) and posterior synechia had totally shut off the anterior chamber from the posterior chamber with a seclusio pupil (Fig 38). She was taken back to the operating room, where a membranectomy was performed with the ocutome. The pupil was enlarged with the ocutome, as seen on postoperative day 1 (Fig 39). Three days later, therapy with the contact lens was restarted, along with patching for amblyopia. At 2-year follow-up she was doing well with 6 hours of patching per day. She can count fingers at 1 foot, but I am optimistic that her vision will be measured at a higher level when she becomes more cooperative. Half-time patching will be continued until age 9. I have seen 4 children develop a similar membrane within 3 months of the original surgical procedure for PHPV. Children often have a lot of postoperative inflammation and should be treated aggressively with topical steroids for several weeks.

**FIGURE 38**

A 3-month-old patient with dense secondary membrane 9 weeks postoperative from original surgery for PHPV.

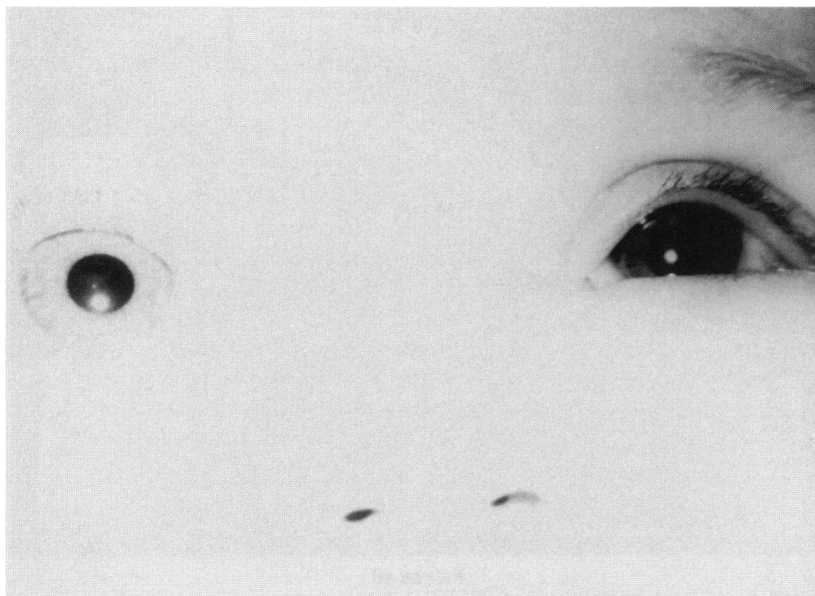


FIGURE 39

Same patient as in Fig 38 1 day following membranectomy. A large opening has been made with ocutome.

Case 7

An 11-year-old patient who had undergone surgery during infancy for a unilateral congenital cataract associated with microphthalmia and anterior PHPV now had 20/20 vision in her operated eye. She had received amblyopia therapy and a continuous-wear soft contact lens for the first month after surgery. For several years she came to the office once a month to have her lens removed and sterilized because her mother could not remove her lens. Finally the mother was able to insert and remove the lens, which she did monthly. This case is presented to show what can be done by the family with persistence in their efforts to treat amblyopia. At age 11 she was still wearing the soft contact lens on her microphthalmic right eye (Fig 40). This is the only patient in this series with 20/20 in the operated eye.

Case 8

A 2 month-old baby boy underwent a pars plana lensectomy, membranectomy, and vitrectomy for a combined anterior and posterior PHPV. He had a dissection of a stalk from the posterior aspect of the lens down to the surface of the optic nerve. Six years postoperatively, his visual acuity was only

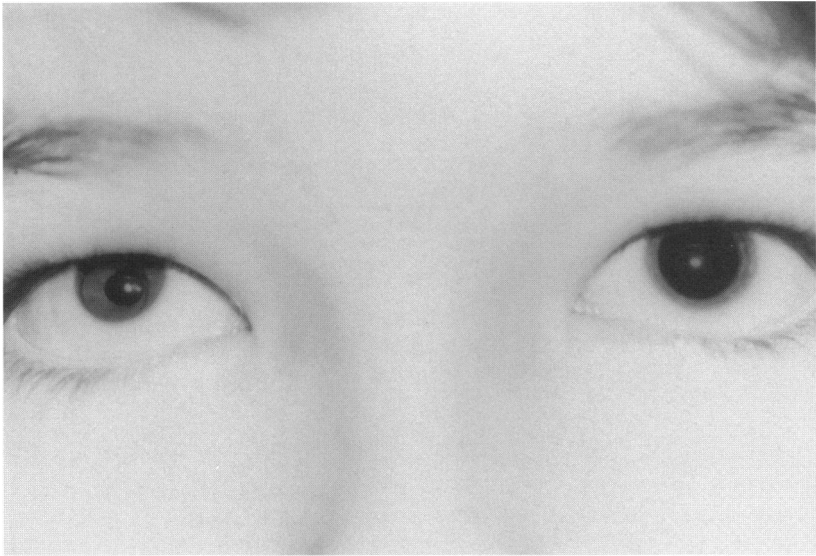


FIGURE 40

An 11-year-old with 20/20 vision in her microphthalmic, aphakic right eye. She had cataract and anterior PHPV surgery in the first month of life and amblyopia therapy until age 9.

hand motion at 1 foot. He has a perfectly clear pupil and vitreous (Fig 41). The view of the posterior pole (Fig 42) shows scarring of the retina between the macula and the optic nerve as well as superior to the optic nerve. This prevented the development of central vision. This case shows that while anatomically a clean vitrectomy can be performed in these difficult cases, the results are usually dismal because of the involvement of the posterior pole with either a dysplastic disc, tractional retinal detachment, or maculopathy.

Case 9

A 4-year-old had had surgery for PHPV in a microphthalmic right eye at age 3 months and had been treated with a gas-permeable hard contact lens and patching from infancy. Her mother had the use of only one hand (her right arm and hand had been paralyzed since infancy) but was determined to help her daughter learn how to handle and wear the contact lens. The mother learned to handle the lens placement, and she taught the child how to handle the lens very early in life. At age 4 the child could insert and remove the lens by herself (Figs 43 and 44). While this case is truly the exception, it is mentioned only to show what can be accomplished with excellent parental support.

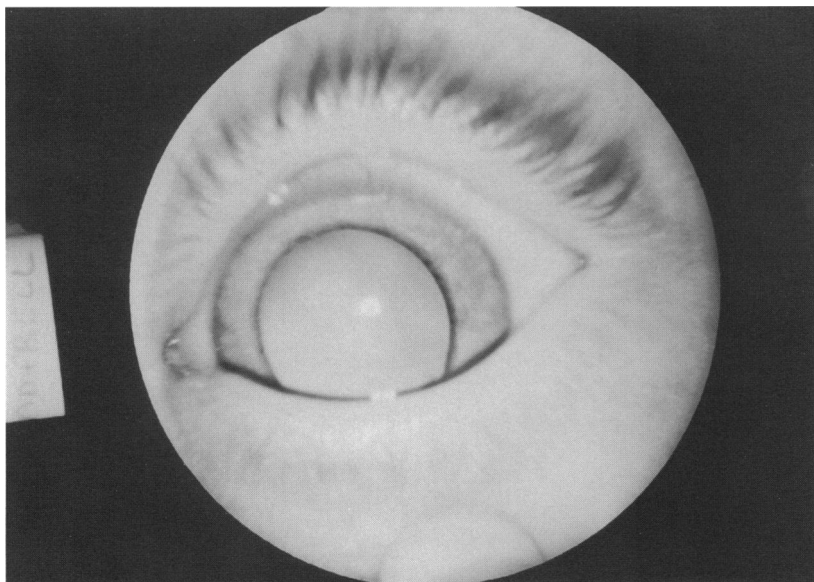


FIGURE 41

Clear pupillary space after lensectomy, membranectomy, and deep vitrectomy via a pars plana approach.

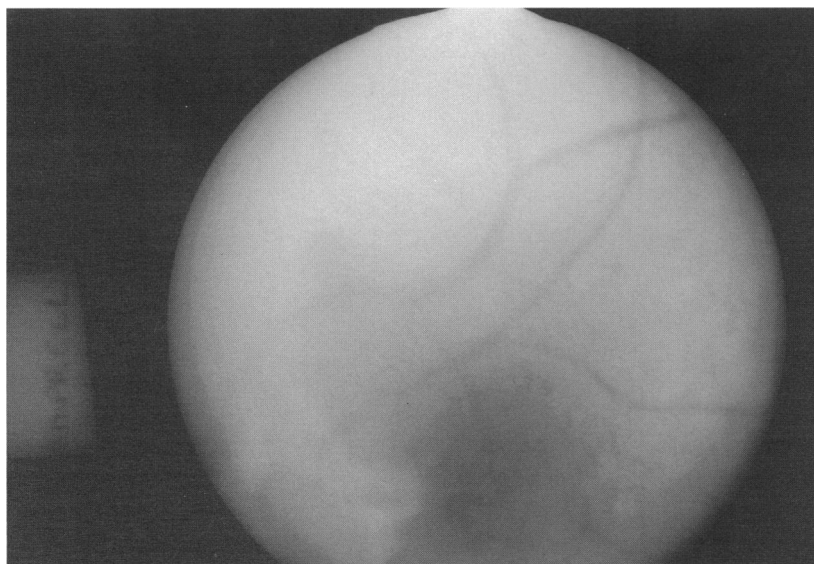


FIGURE 42

Same patient as in Fig 41 showing clear vitreous but poor visual result owing to scarring of posterior pole.

**FIGURE 43**

A 4-year-old patient holding a contact lens on her right index finger preparing to insert lens.

**FIGURE 44**

Same patient as in Fig 43 inserting lens herself onto her cornea.

Case 10

A 10-year-old boy underwent a limbal lensectomy, membranectomy, and anterior vitrectomy for a unilateral anterior PHPV at 3 weeks of age. He was the product of a full gestational pregnancy and had been delivered vaginally with a birth weight of 6 lb, 7 oz. Horizontal corneal diameter at the time of surgery was 11.0 mm in his normal right eye and 9 mm in his PHPV left eye. Surgery was uneventful, and he was fit with a gas-permeable hard contact lens 1 week after surgery. His keratometry readings at the time of surgery were 5000 horizontally and 4850 vertically. These are normal parameters for a newborn. Amblyopia treatment was started at once with full-time patching for the first week followed by half-time patching until age 9.

During the first 3 years of life, he had several examinations under anesthesia to obtain keratometry readings, because the base curve of his lens had to be flattened as he grew older. After age 3 he would allow accurate readings to be taken in the office while awake. His eyes were straight horizontally, but he showed a small dissociative left hypertropia, which did not require surgical repair. At age 3 he developed accommodative esotropia, which responded quite nicely to glasses; the hyperopic correction was +4.00 in his normal right eye (Figs 45 and 46). His aphakic left eye refracted to +9.00 +1.00 axis 90. He continued to wear the contact lens on his left eye. The glasses were +4.00 on the right and plano on the left. At last follow-up at age 10, he had orthophoria at distance and a small esotropia of 4 prism diopters at near with his glasses. His right corneal diameter measured 12 mm, and the left measured 10 mm. Axial length measurements showed the right eye to be 22.32 mm and the left eye to be 21.54 mm. Without glasses, he measured 16 prism diopters at distance of esotropia and 20 prism diopters at near. Visual acuity is 20/20 in the right eye and 20/200 in the left eye. Patching was stopped at age 9.

This case shows that children with PHPV can develop accommodative esotropia just like other children. If a significant amount of hyperopia is present (at least 2 diopters), then a consideration of accommodative esotropia should be given as the etiology of the strabismus. This should be entertained even with the knowledge that the child had PHPV as the original insult to the oculomotor balance system.

Case 11

A 3-year-old girl had first presented at 3 days of age with a shallow chamber in the right eye. A diagnosis of anterior PHPV was made as a membrane with vessels was seen behind the lens. Ultrasound showed a normal posterior segment. While she was taking her bottle, the tonopen was easily used to measure her pressure under topical anesthesia. Her pressure was 14 in each eye (Fig 47). She underwent an uneventful lensectomy,

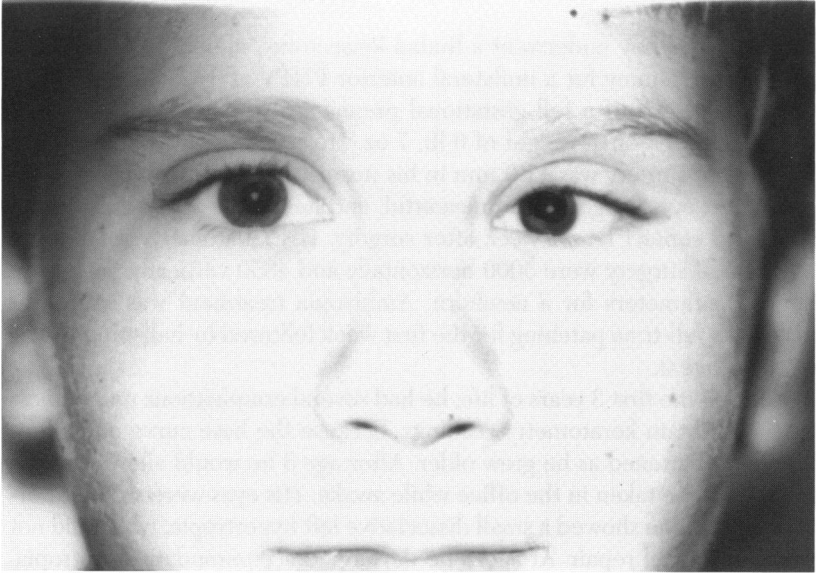


FIGURE 45

Case 10. Patient has accommodative esotropia showing a left esotropia without glasses. As an infant, patient had surgery for PHPV of left eye.

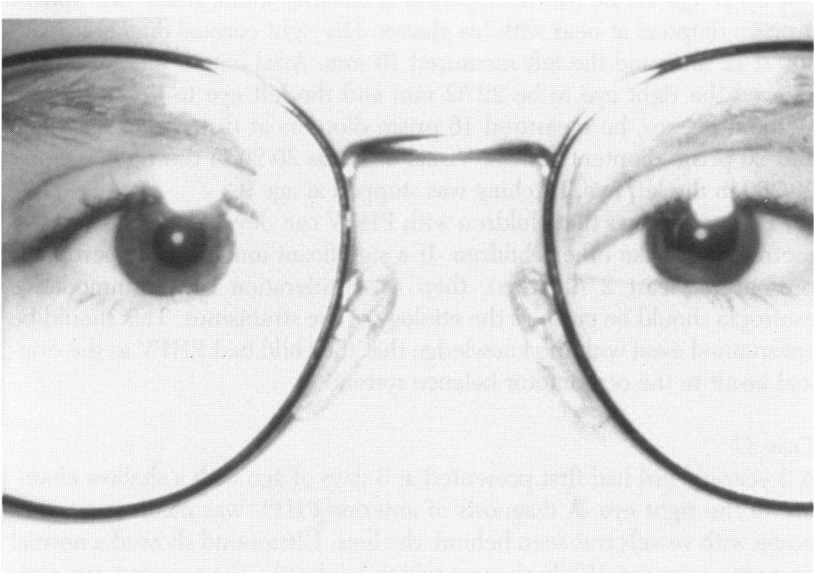


FIGURE 46

Same patient as in Fig 45 with accommodative esotropia controlled wearing his glasses. He has a gas-permeable hard contact lens in left eye.

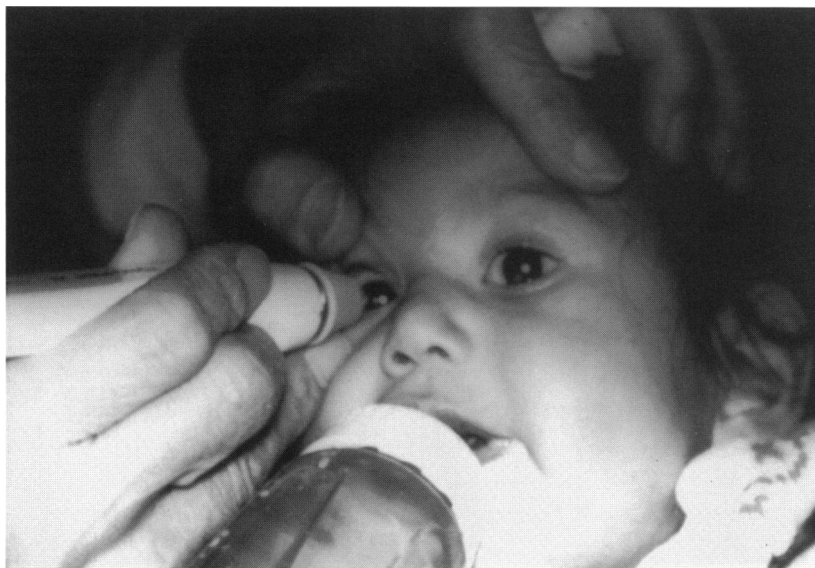


FIGURE 47

Case 11. Newborn is having her pressure measured under topical anesthesia with tonopen while she was feeding.

membranectomy, and anterior vitrectomy via the limbal approach at 2 weeks of age and was fitted with a gas-permeable hard contact lens at age 3 weeks. Amblyopia therapy was carried out until age 2, when she suffered child abuse and presented with bilateral retinal hemorrhages. Unfortunately, the hemorrhage in the right eye involved the macula. During the 3-month period needed to clear the macular hemorrhage, the patient could not be patched. After the blood finally cleared, the patient became more amblyopic, because she would not allow patching. When seen at age 4, she had counting fingers vision at 1 foot in the operated eye. A marked dissociative right hypertropia was present. She underwent a retroequatorial myopexy (faden) to the right superior rectus at 15 mm from the insertion combined with a 5-mm recession of the right superior rectus. At 3-year follow-up, the eyes were essentially straight with a very small dissociative right hypertropia present only on covering the right eye. She does continue to wear the contact lens, but she will not allow any patching. This case shows that even though the visual result was less than desirable, use of the contact lens together with strabismus surgery has given her a very nice cosmetic result (Figs 48 and 49).

**FIGURE 48**

Case 11. Dissociative right hypertropia in eye that had undergone PHPV surgery.

**FIGURE 49**

Same patient as in Fig 48 who has retroequatorial myopexy (faden) to right superior rectus muscle.

Case 12

This case discusses a legal issue that arose concerning PHPV. I recently was asked by the defendant's lawyer to evaluate a case about to come to court. This case involved a child who was operated on at 7 months of age for a combined anterior and posterior PHPV. The child has a poor visual result, and the child's parents had filed suit against the pediatrician who had taken care of the child since birth. The parents claimed that as the PHPV had been present since birth, the pediatrician missed the diagnosis until the child was 6 months of age. The family attributed the poor visual result to the pediatrician's delay in diagnosis. After the surgery for PHPV at 7 months of age, the parents closely examined the child's newborn photograph, which had been taken at the hospital. They noticed a small white opacity in the pupil of the PHPV left eye, which they interpreted as either a cataract or a PHPV membrane.

I reviewed the preoperative and operative notes, which clearly stated that the patient had a retinal fold traversing the macula as well as a pigmented maculopathy. At the time of surgery, a lensectomy and membranectomy were performed along with cutting of the anterior part of the PHPV stalk. The postoperative course has been quiet with no evidence of glaucoma, but the visual result has been very poor because of the posterior segment abnormalities. Had the case only involved amblyopia, the delay in diagnosis would have been detrimental to the child's visual prognosis. However, the presence of the posterior segment involvement as evidenced by the macular fold and pigmentation, dictated a poor visual result regardless of the delay in diagnosis. Therefore, the pediatrician was not held liable for causing the poor visual result. Significantly, the plaintiff's attorney reviewed my deposition, which stated my experience with PHPV, and subsequently recommended that the child's parents withdraw the lawsuit.

SUMMARY AND CONCLUSIONS

While the great majority of patients with persistent hyperplastic primary vitreous never obtain useful vision, it is encouraging that 18.07% of all the patients in this series did achieve 20/200 vision or better (Table V). In the whole group, 12 of 83, or 14.45%, obtained 20/70 vision or better with treatment. All of these successful results were in eyes with the purely anterior PHPV only. In fact, when looking at the results of the anterior PHPV cases alone, 15 of 21 achieved 20/200 or better, which was 71.4% of the patients with anterior presentation only. In this anterior group, 12 of 21 (57.1%) attained 20/70 vision or better with treatment.

The goals of treatment with PHPV should always be kept in mind when confronted with an infant who has this entity. First, an effort should be

TABLE V. SURGERY AND VISUAL RESULTS IN PHPV PATIENTS

Anterior PHPV: 21 patients, 20 treated with limbal lensectomy, membranectomy, and anterior vitrectomy. 20/200-1 patient, 20/100-2 patients, 20/70-2 patients, 20/50-1 patients, 20/40-3 patients, 20/30-5 patients, 20/20-1 patient, counting fingers at one foot-5 patients, One patient unoperated mth 20/60 in each eye with the tunica vasculosa lentis engulfing each lens.

Posterior PHPV: 10 patients, 8 treated with pars plana lensectomy, membranectomy, and vitrectomy. All with hand motion vision or less. Two unoperated with hand-motion vision only.

Combined anterior-posterior PHPV: 52 patients, 10 with pars plana lensectomy, membranectomy, and vitrectomy. All in range of no light perception to hand- motion vision only. 42 with limbal lensectomy, membranectomy, and anterior vitrectomy. All in range of no light perception to hand-motion vision only.

made to save the eye from glaucoma or phthisis, which are the most devastating results of untreated PHPV. A lensectomy will usually prevent the secondary glaucoma, which results from the lens-iris diaphragm being pushed forward to cause a secondary angle-closure glaucoma. A secondary glaucoma can also be produced from recurrent hyphema, which scars the outflow passageways in the angle. By removing the PHPV membrane and reducing the tractional forces applied to the ciliary body by the membrane, one can lessen the possibility of phthisis. Surgery reduces the centripetal, as well as axial, forces generated by the membrane. The second goal is to produce a black pupil for cosmetic reasons, and the third goal of obtaining useful vision is reasonable to expect if the entity presents with only anterior involvement.

Bilaterality, while rare in this series (2:83 cases, or 2.4%), was a very poor prognosis for vision, since both patients had only light perception to hand motion vision in both eyes. With the exception of the eye problems, these two children were otherwise normal in their physical and mental development.

No measurable stereopsis or binocularity has been achieved in any of these patients. This was primarily because 100% of the patients had stra-

bismus and secondarily to the fact that in the patients with good surgical results, aggressive and prolonged patching may have precluded the development of binocular vision.

Two patients have uncontrolled glaucoma with only light perception vision. Both of these patients are comfortable and pain-free. Two patients have, in time, developed phthisis and wear a cosmetic shell. No cases have required enucleation in this series. (The pathological specimen from Figs 1 and 2 represents an enucleated eye during my residency.)

Two patients developed an extensive cyclitic membrane after surgery, which completely occluded the pupillary space. Both patients had originally undergone extensive membrane removal of their PHPV with the production of a nice clear pupillary space. The first patient developed the cyclitic membrane only 3 weeks after the original surgery. A second surgery has been successful in clearing the pupillary opening, but the patient has only hand motion vision at 1 foot due to amblyopia. The second patient had been fitted with a gas-permeable hard contact lens after original surgery for an anterior PHPV. An extensive cyclitic membrane developed 4 months later and required a secondary procedure to clear the pupillary space. The patient's pupillary space has remained open after this second procedure, and the patient is currently wearing a contact lens and undergoing amblyopia therapy.

The most important factor in the prognosis of patients with persistent hyperplastic primary vitreous is the extent of the membrane. Families should be told that if their child has only an anterior presentation, then surgical and amblyopia therapies offer a good chance for useful vision. Patients with a combination of anterior and posterior PHPV, or patients with only a posterior PHPV, should be advised that there is a reasonable opportunity to save the eye from glaucoma and phthisis, but that useful vision is usually not obtainable in these eyes. Three patients with a purely posterior PHPV and 21 with a combination of anterior and posterior PHPV had amblyopia treatment postoperatively, even though they had an abnormal posterior pole. The abnormalities consisted of a dysplastic optic nerve, retina fold through the macula, and/or a pigmented maculopathy. All 24 of these patients attained vision of hand motion or less. All of the other patients with the purely posterior PHPV or the combination of anterior and posterior PHPV had posterior segments too distorted to even try amblyopia therapy.

The second most important factor in having a favorable prognosis is to be able to diagnose and treat PHPV at an early age. All 15 patients who obtained 20/200 vision or better not only had a purely anterior presentation, but also all had their surgery in the first month of life. All had their contact lens fitting within 2 weeks of the surgery, and 6 were fit in the first week after surgery. This early rehabilitation of the eye with early ambly-

opia treatment helped to produce a successful visual outcome.

Families should be told of the long-term commitment to amblyopia therapy that is necessary if their child is to rehabilitate the involved eye. Extensive patching is required in the first few months after surgery. This is followed by part-time patching of 4 to 6 hours per day until age 9, when visual maturity is reached. Contact lens fitting is easier in the newborn period compared to fitting the child for the first time between ages 2 and 6. Infants are usually not strong enough to prevent their parents from inserting and removing the contact lens.

**TABLE VI. TYPES AND DISTRIBUTION OF STRABISMUS IN 83
PHPV PATIENTS**

Esotropia: 57 Patients
Exotropia: 18 Patients
Vertical strabismus: 8 patients

TABLE VII. PRESENTATION OF PHPV: UNILATERAL VS BILATERAL

Unilateral: 81 patients
Bilateral: 2 patients

TABLE VIII. SIZE OF EYES IN 83 PHPV PATIENTS

Normal-sized eye: 12 patients
Small-sized eye: 70 patients*
Buphthalmic eye: 1 patient

*The two patients with bilateral PHPV had microphthalmia bilaterally.

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