Visual Outcomes Following Lensectomy and Vitrectomy for Combined Anterior and Posterior Persistent Hyperplastic Primary Vitreous

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**Objective:** To determine the visual outcome after surgery for persistent hyperplastic primary vitreous using modern vitreoretinal techniques.

**Design:** Retrospective medical record review during a 5-year period (June 1992 to June 1997). Information recorded for each patient included age, medical history, sex, results of preoperative ocular examination, age at diagnosis, procedure performed, intraoperative and postoperative complications, location and number of sclerotomy sites, type of aphakic rehabilitation, amblyopic therapy given, final visual acuity, and length of follow-up.

**Results:** Fourteen patients who underwent surgical management of combined anterior and posterior persistent hyperplastic primary vitreous were identified. Eleven patients underwent aphakic rehabilitation and aggressive amblyopic therapy consisting of occlusive therapy for several waking hours each day. One additional older patient received aphakic rehabilitation only. Ten eyes (71%) achieved a visual acuity of 20/300 or better, and 8 (57%) obtained a final visual acuity of 20/100 or better. Average length of follow-up was 22 months (range, 4-57 months). Nine patients were fitted with an aphakic soft contact lens, 2 older patients had a posterior chamber intraocular lens placed at the time of vitrectomy, and 1 patient wore aphakic spectacles.

**Conclusions:** With modern vitreoretinal techniques, aphakic rehabilitation, and aggressive amblyopic therapy, useful vision can be obtained in the majority of patients with combined anterior and posterior persistent hyperplastic primary vitreous.


Persist**

Persistent hyperplastic primary vitreous (PHPV), also known as persistent fetal vasculature, is a congenital anomaly in which the normal regression of the primary vitreous and hyaloid vasculature does not occur. Classically, PHPV appears unilaterally in an otherwise healthy, full-term newborn. A retrolental fibrovascular mass is seen on ocular examination. Contraction of this mass can draw the ciliary processes into the visual axis or cause recurrent intraocular hemorrhage. Most eyes are microphthalmic, and while the lens is clear initially, a progressive cataract develops with time in most cases. With this lens opacity present, diagnosis is often difficult without the use of ultrasonography, computed tomography, or magnetic resonance imaging. If left untreated, many eyes with severe PHPV will develop severe glaucoma, retinal detachment, or phthisis early in life.

Some authors have separated PHPV into anterior and posterior forms. Ocular findings in purely anterior PHPV can include microphthalmia, cataract, elongated ciliary processes, retrolental fibrovascular membrane, and glaucoma. Posterior PHPV can consist of microphthalmia, usually without cataract, vitreous membrane and stalk, retinal fold, traction retinal detachment, and hypoplastic optic nerve and macula. The separation of PHPV into these distinct anterior and posterior varieties is arbitrary, as many cases will exhibit some features of both.

In an effort to counteract the dismal natural history of PHPV, numerous operative techniques for removal of the cataract and retrolental membrane have been used during the last several decades. Initially, an open-sky cataract wound was fashioned and the cataract and membrane were evacuated in a 1- or 2-stage procedure. With the advent of mechanical suction-cutting devices, the visual axis could be cleared using a closed-system intraocular approach that provided superior control of any bleeding encountered while dissecting the retrolental tissue.
SUBJECTS AND METHODS

We undertook a retrospective record review of all patients with the diagnosis of PHPV who underwent vitreoretinal surgical rehabilitation after appropriate informed consent at the Eye Institute, Medical College of Wisconsin, Milwaukee, from June 1992 to June 1997. We recorded the following information from each record when available: patient name, date of birth, medical history including any history of prematurity, sex, age at diagnosis, type of PHPV, initial visual acuity, surgical procedure performed, age at time of surgical procedure, intraoperative and postoperative complications, location and number of sclerotomy sites, postoperative appearance, type of aphakic rehabilitation, amount of amblyopic patching therapy, final visual acuity, and length of follow-up. Visual acuity was assessed with age-appropriate tests such as the Teller and Snellen-equivalent methods. Those 20 years of age or older had Snellen acuity measured. If the patient was 6 years of age or older, visual acuity was assessed using the Teller acuity card.

Patient fixation patterns were then extrapolated to visual acuity measurements using a previously described technique modified from Zipf,31 as follows:

<table>
<thead>
<tr>
<th>Fixation Pattern</th>
<th>Visual Acuity</th>
</tr>
</thead>
<tbody>
<tr>
<td>CSM</td>
<td>≤20/30</td>
</tr>
<tr>
<td>CSUM</td>
<td>20/30-20/100</td>
</tr>
<tr>
<td>CUSUM</td>
<td>≤20/300</td>
</tr>
<tr>
<td>UCUSUM</td>
<td>≤5/20</td>
</tr>
</tbody>
</table>

where C indicates central; S, steady; M, maintained; UC, uncentral; US, unsteady; and UM, unmaintained.

Amblyopia therapy consisted of patching the better eye for several (2 hours to full-time) waking hours each day. An attempt was made to limit patching to no more than 80% to 90% of waking hours for patients younger than 8 years. Contact lens power was checked every 4 months and replaced if more than 6 diopters (D) over-plus for infants and more than 3 D over-plus for older children.

Studies surgery was used solely to save the eye from enucleation, and visual results were poor. Outcomes have improved as surgical techniques and instrumentation have advanced, aphakic soft contact lenses in pediatric sizes developed, and greater importance placed on early amblyopic occlusive therapy.30

In an effort to determine the current visual results following surgery for PHPV, we undertook a retrospective review to evaluate visual outcomes after modern vitrectomy surgery for combined anterior and posterior PHPV with the use of prompt aphakic rehabilitation and aggressive amblyopic therapy.

RESULTS

Fourteen patients were identified from the surgical records of the retina service at the Eye Institute (Table). Six patients were male and 8 were female. The age at diagnosis varied from 1 week to 7 years, although 13 patients were diagnosed prior to 9 months of age. Five patients were born prematurely, but only 2 patients were more than 1 month premature. No patient had a significant systemic illness. All patients had features of combined anterior and posterior PHPV with cataract formation and a fibrovascular stalk extending from the optic nerve to posterior lens in one eye only. In addition, 2 patients had a tractional retinal detachment on initial examination, and 1 patient exhibited neovascularization of the iris, vitreous hemorrhage, and advanced glaucoma.

The initial visual acuity in the affected eye was light perception in 12 younger patients and 20/200 and 20/200 in the 2 older patients, aged 7 and 17 years, respectively. Average follow-up after operation was 22 months (range, 4-57 months).

Nine patients had a primary lens extraction and vitrectomy procedure performed (7 performed through the pars plana and 2 through the limbus), and the 2 older patients had a combined phacoemulsiﬁcation/posterior chamber intraocular lens placement surgery with pars plana vitrectomy. The 3 remaining patients had an initial cataract extraction performed with subsequent posterior vitrectomy. In 2 of these patients the diagnosis of PHPV was made at the time of surgery for unilateral congenital cataract, and the operation was terminated after lens removal only. In the remaining patient, who preoperatively was thought to have a mild form of PHPV, a posterior capsulotomy and pupillary membrane removal with an anterior vitrectomy unit was performed at the time of cataract surgery. Postoperatively, this patient developed a dense vitreous hemorrhage, an organized secondary membrane surrounding the residual posterior stalk, and hemolytic glaucoma necessitating a complete posterior vitrectomy. The age at surgery varied from 2 weeks to 17 years.

Surgical complications of posterior vitrectomy included a retinal tear in 1 patient. This patient had a massive tractional retinal detachment preoperatively, and after the lens was removed it was noted that the retina inserted directly into the pars plicata with no visible pars plana. The instrument placed through the superotemporal sclerotomy (2.5 mm posterior to limbus) caused enough traction on the nearby retina to cause a retinal tear. This necessitated placement of a scleral buckle with endolaser treatment and an air-fluid exchange. The retina has remained attached postoperatively. Another patient developed a secondary pupillary membrane with increased intraocular pressure and a vitreous hemorrhage. On further follow-up, an organized retinal detachment, deemed to be inoperable, was noted. Both of these patients were thought to have insufficient visual potential to warrant aphakic rehabilitation and amblyopic therapy; however, neither patient has gone into phthisis or has had to have an enucleation as of the last follow-up visit. Vitrectomy port location varied according to the age of the patient and surgeon preference from the limbus to 2.75 mm posterior to the limbus. Posterior placement of the vitrectomy ports seemed to cause a complication in only 1 case.

Aphakic rehabilitation was carried out in 12 patients. Nine received an aphakic soft contact lens after...
surgery (1-12 weeks postoperatively), while 2 patients had a posterior chamber intraocular lens placed at the time of surgery. One patient was bilaterally aphakic and wore aphakic spectacles. Amblyopic therapy was instituted in 11 of the 12 patients and consisted of occlusion of the better eye from 2 hours to full-time patching for the waking hours each day. Compliance with the contact lens and patching regimen was good for patients 1, 3, 7 through 10, 12, and 13 and poor for patients 2, 5, and 14 (Table). Only 1 of the patients with poor compliance had a final visual acuity worse than 20/300.

Final visual acuity in the 12 patients who had visual potential and underwent amblyopic therapy varied from less than 20/300 to 20/30. No patients had a visual acuity less than 5/200. Ten (83%) of these 12 patients achieved a visual acuity of 20/300 or better and 8 (66%) obtained a final vision of 20/100 or better (Table). Elevated intraocular pressure (>20 mm Hg) was present at the initial visit in 1 patient and developed later in 2 patients (Table).

### COMMENT

Hiles and Reynolds in 1983 and Pollard in 1991 and 1997 reported on their respective series of PHPV patients managed with a variety of surgical techniques. Their conclusions were that patients who had a purely anterior form of PHPV (ie, no stalk or retinal abnormalities) had visual potential, while those with combined or posterior forms of the disease did not. Our study clearly illustrates that patients with combined anterior and posterior disease can achieve functional visual acuities when managed with modern vitreoretinal techniques and aggressive amblyopic therapy. In another large series of patients in which PHPV was not classified into anterior, combined, or posterior forms, 8 (44%) of 18 patients with visual potential achieved a final visual acuity of 20/200 or better. This study included 4 patients managed with an older technique of lens needling and aspiration with a scissors membranectomy. We attribute the improved outcomes seen in our study to advances in instrumentation for vitrectomy and cataract surgery that were applied uniformly to all the patients and to prompt aphakic rehabili-
tation and amblyopic therapy instituted in all patients with visual potential.

The diagnosis of PHPV can be difficult when a dense cataract is present. This scenario was demonstrated by 2 of our patients in whom the correct diagnosis was not made until the time of cataract extraction. Ultrasonography in these small infants is challenging, and the stalk can be thin and thus overlooked, as it was in these 2 patients. In another patient, a very thin, partial stalk was noted on ultrasonography and it was felt that cataract extraction alone would be sufficient to clear the visual axis; however, at surgery a substantial, thick fibrovascular stalk was uncovered. Even though an anterior vitrectomy with partial membranectomy was performed on this patient at the time of cataract surgery, eventually a posterior vitrectomy was required for a dense postoperative vitreous hemorrhage with vitreous organization around the residual stalk and a secondary pupillary membrane. After this case, no further cases of combined anterior and posterior PHPV were primarily handled with anterior segment surgical techniques alone for fear of postoperative hemorrhagic complications. If any posterior fibrovascular stalk is visible on ultrasonography, regardless of its perceived thickness, posterior vitrectomy and lensectomy should probably be performed. Computed tomography and/or magnetic resonance imaging may have helped in the diagnosis of PHPV in these 3 patients and might have enabled us to eliminate the additional surgical procedure that each required. Unfortunately, computed tomography or magnetic resonance imaging in these patients usually requires heavy sedation or general anesthesia.

Although historical comparison among different studies is problematic, the visual results obtained in our study seem to approximate those acquired after surgery for unilateral congenital cataracts without PHPV. This similarity implies that the presence of PHPV alone does not necessarily confer a poor prognosis. Two eyes in our study were operated on at 5 and 10 months of age, respectively. Despite the enhanced susceptibility to irreversible amblyopia, visual acuities of 20/100 and 20/70 were obtained. Similar results have been achieved by others for late surgery for unilateral congenital cataract, both with and without PHPV. These findings suggest that

<table>
<thead>
<tr>
<th>Surgical Complications</th>
<th>No. and Location of Sclerotomy Sites</th>
<th>Type of Aphakic Rehabilitation</th>
<th>Duration of Amblyopic Therapy, h/d</th>
<th>Final Visual Acuity†</th>
<th>Length of Follow-up, mo</th>
</tr>
</thead>
<tbody>
<tr>
<td>None</td>
<td>2 ports, 1.0 mm posterior to limbus</td>
<td>CL at 2.5 wk</td>
<td>Patching 5 h</td>
<td>CUSUM = 20/300</td>
<td>18</td>
</tr>
<tr>
<td>Postoperative VH</td>
<td>3 ports, 2 mm posterior to limbus</td>
<td>CL at 2 mo</td>
<td>Patching 6 h</td>
<td>CUSUM = 20/300</td>
<td>25</td>
</tr>
<tr>
<td>VH, increased IOP; stalk/membrane after first surgery</td>
<td>3 ports, at limbus</td>
<td>CL at 5 mo</td>
<td>Patching 1-2 h</td>
<td>CSM ≥ 20/30</td>
<td>30</td>
</tr>
<tr>
<td>Secondary pupillary membrane remnant, increased IOP; VH, late RD with PVR</td>
<td>3 ports, at limbus</td>
<td>None</td>
<td>None</td>
<td>No LP</td>
<td>18</td>
</tr>
<tr>
<td>None</td>
<td>2 ports, at limbus</td>
<td>CL at 2 mo</td>
<td>Patching 6 h</td>
<td>CSUM = 20/30-20/100</td>
<td>8</td>
</tr>
<tr>
<td>None</td>
<td>3 ports, 2.5 mm posterior to limbus</td>
<td>Posterior chamber IOL</td>
<td>None</td>
<td>20/80</td>
<td>4</td>
</tr>
<tr>
<td>None</td>
<td>3 ports, 2.0 mm posterior to limbus</td>
<td>CL at 2.5 mo</td>
<td>Patching 6 h</td>
<td>20/300</td>
<td>5</td>
</tr>
<tr>
<td>None</td>
<td>3 ports, 1.5 mm posterior to limbus</td>
<td>CL at 3 mo</td>
<td>Patching 6 h</td>
<td>CSM ≥ 20/30</td>
<td>13</td>
</tr>
<tr>
<td>Mild VH</td>
<td>NA</td>
<td>Aphakic glasses at 3 mo</td>
<td>Patching 3 h</td>
<td>CSM ≥ 20/30</td>
<td>21</td>
</tr>
<tr>
<td>Retinal tear</td>
<td>3 ports, 2.5 mm posterior to limbus</td>
<td>None</td>
<td>None</td>
<td>LP</td>
<td>9</td>
</tr>
<tr>
<td>None</td>
<td>3 ports, 2 mm posterior to limbus</td>
<td>CL at 7 mo</td>
<td>Patching 6 h</td>
<td>20/100</td>
<td>57</td>
</tr>
<tr>
<td>None</td>
<td>3 ports, 2.75 mm posterior to limbus</td>
<td>CL at 11 mo</td>
<td>Patching 6 h</td>
<td>20/70</td>
<td>52</td>
</tr>
<tr>
<td>None</td>
<td>3 ports, 2.5 mm posterior to limbus</td>
<td>Posterior chamber IOL</td>
<td>Patching full-time</td>
<td>20/300</td>
<td>6</td>
</tr>
</tbody>
</table>
whenever the diagnosis of PHPV is made, evaluation for possible surgical rehabilitation should be undertaken if light perception acuity exists.

The one major surgical complication noted during our study was a retinal tear that resulted partially from the posterior placement of a sclerotomy incision in a patient with a preoperative tractional retinal detachment. Anterior placement of incisions to avoid this type of complication has previously been suggested for these small eyes that frequently have a poorly developed pars plana region.11 Our incision site placement varied according to surgeon preference and the age of the patient. Transillumination is occasionally helpful in locating the ora serrata, but is often unsuccessful.11 In our opinion, placement of incisions as anteriorly as possible is beneficial, especially if a retinal detachment is noted on preoperative ultrasonography.

Two of our patients were considered to have insufficient visual potential postoperatively to warrant amblyopic therapy. One had a secondary membrane developing before a late inoperable retinal detachment, and the other had extensive macular changes after repair of a complex tractional/rhegmatogenous retinal detachment. We believe that almost every child deserves a trial of occlusion therapy after removal of media opacity. In cases with advanced retinal or optic nerve pathology, however, amblyopic therapy should be promptly terminated after a short trial (~2 months) if no improvement is noted, to avoid undue psychosocial impairment.30 Eyes with PHPV have a lifetime risk of glaucoma and retinal detachment. While our study exhibited the attainment of useful vision in the majority of patients, the follow-up was brief. One study has shown that glaucoma develops in 32% of patients with PHPV at a mean of 64.6 months postoperatively.39 With our current intermediate follow-up, we have demonstrated that a visual acuity of 20/300 is obtainable in the majority of patients with a combined form of anterior and posterior PHPV using modern vitreoretinal surgical techniques and aggressive amblyopic therapy. Continued surveillance of these young patients will be necessary to ensure that their present vision is maintained for the future.

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REFERENCES


