Clinical characteristics and surgical approach to visually significant persistent pupillary membranes

Courtney L. Kraus, MD, and Gregg T. Lueder, MD

BACKGROUND
Infants with hyperplastic persistent pupillary membranes (PPM) may be at risk for deprivation amblyopia due to obstructions of the visual axis. We describe the long-term visual and anatomic outcomes of a surgical technique for their removal.

METHODS
The medical records of consecutive patients <3 years of age who underwent surgical removal of PPMs between December 1998 and May 2012 were retrospectively reviewed. Each PPM was judged to be visually significant based on poor visual acuity, poor retinoscopic reflex, or inability to visualize the fundus. The surgical technique included injection of a viscoelastic agent beneath the pupillary strands to bow them anteriorly, careful peeling of residual adherent strands from the anterior lens capsule, and lysis of the strands at the pupillary margin with intraocular scissors. Pre- and postoperative visual and anatomic results were recorded.

RESULTS
This case series included 10 eyes of 6 patients: PPMs were bilateral in 4 patients and unilateral in 2. The patient age at time of surgery ranged from 2.5 months to 2.5 years (mean, 14 months). Mean postoperative follow-up was 5.3 years (range, 2.5-8 years). All patients had successful clearance of the visual axis and good visual acuity. No intraoperative complications occurred.

CONCLUSIONS
All patients in this series had excellent visual and structural outcomes, with no significant complications. The technique described here may be considered for patients with visually significant PPMs to improve visual function and pupil appearance. (J AAPOS 2014;18:596-599)

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ersistent pupillary membranes (PPMs) range in appearance from fine, diaphanous strands to densely pigmented membranes that almost fully cover the pupil, obscuring the red reflex and impairing vision. In most cases they regress during the first year of life; however, in a small percentage of cases PPMs remain visible, and in an even smaller percentage some form of intervention is required. Pharmacologic therapy, laser treatments, and surgery have been employed to clear the visual axis and optimize visual development.

Surgical intervention is reserved for large, dense PPMs that obscure the pupillary axis (Figure 1). We present a series of patients treated for clinically significant PPMs using modifications of previously reported surgical techniques.

Subjects and Methods
This study was approved by the Washington University School of Medicine Institutional Review Board. In this interventional study, the medical records of consecutive patients <3 years of age who underwent surgical removal of a PPM at St. Louis Children’s Hospital between December 1998 and May 2012 were retrospectively reviewed. One surgeon (GTL) performed all operations. Age, sex, associated ocular abnormalities, and preoperative visual acuity were noted. Visual acuity was assessed preoperatively by ocular behavior or spatial-sweep visual evoked potentials (SSVEP). Preoperative interventions included patching and optical penalization. All patients underwent a detailed anterior segment evaluation, intraocular pressure (IOP) recording, and fundus evaluation by direct ophthalmoscopy. Indications for surgery included decreased visual acuity, poor retinoscopic reflex, or impaired visualization of the fundus. Cases of bilateral PPM underwent sequential surgery, with the more visually significant membrane removed first.

Surgical Technique
Informed consent was obtained from the parents of all patients. The operations were performed under general anesthesia. Cyclopentolate 1% drops were placed prior to surgery. The anterior chamber was entered via a clear corneal incision using an MVR blade. An ophthalmic viscosurgical device (OVD) was injected into the anterior chamber. Additional OVD was placed posterior to the PPM to achieve anterior displacement of the membrane,
with the exception of the central attachments to the anterior lens capsule. This central adherence was gently peeled off the capsule using a Sinskey hook or the blunt tip of the OVD cannula. Care was taken to avoid disruption of the lens capsule. Additional OVD was injected beneath the iris strands after the central adhesions were lysed. The attachments of the membrane near the ciliary body were then cut using intraocular scissors. The pupillary membrane was removed with Kelman-McPherson forceps. See Video 1, available at jaapos.org. OVD was irrigated from the eye using balanced salt solution. Intracameral 0.01% carbachol was injected to induce miosis, which allowed for identification of residual attachments by observing the movement of the iris. The corneal incision was closed with a 10-0 nylon suture for the first eye of a binocular case or 10-0 polyglactin 910 suture if a second surgery was not planned.

Postoperative medications included topical prednisolone acetate 1% drops 4 times daily, cyclopentolate 1% drops 2 times daily, and neomycin/polymyxin B/dexamethasone ophthalmic ointment 2 times a day tapered over 3-4 weeks. Postoperative examinations were performed 1 day, 1 week, and 1 month following surgery, including assessment of visual acuity; pupil size, symmetry, and reactivity; anterior chamber depth; and IOP when possible. SSVEP was performed on children who were unable to perform optotype visual acuity testing. Photographs with pupil analysis were obtained during postoperative appointments when possible.

Results

A total of 10 eyes of 6 children (4 males) were included (Table 1). These 10 eyes represent all PPMs considered to be compromising visual development and necessitating surgical removal in our practice. Mean age at presentation was 7 months (range, 1-18 months). Mean patient age at time of surgery was 14 months (range, 2.5 months-2.5 years). Associated ocular abnormalities included nasolacrimal duct obstruction that resolved prior to surgery in patient 5 and Pallister-Killian syndrome in patient 6, who also had ptosis and bilateral small anterior polar cataracts that were not visually significant. Patient 6 had nystagmus at presentation and was found to have pigmentary retinopathy after his membranes were removed. Electretinogram at age 2 years revealed rod dystrophy.

Retinoscopy was difficult to assess accurately before surgery because of the presence of the membranes. Preoperatively, 5 children were treated briefly for presumed deprivation amblyopia with patching or optical penalization. All had resolution of amblyopia following surgery and amblyopia therapy was discontinued. None of the patients had significant abnormal refractive errors postoperatively except patient 2. He had surgery at age 7 and 7.5 months, with normal vision by SSVEP in both eyes at age 11 months. At age 2.5 years he developed 1.5 D anisometropic astigmatism with amblyopia in the right eye (SSVEP 20/78 in the right eye and 20/41 in the left eye). Vision normalized following spectacles and occlusion therapy.

Patients 3 and 5 initially had worse vision in their more affected eyes by SSVEP (patient 3: 20/46 in the right eye and 20/63 in the left eye; patient 5: 20/61 in the right eye and 20/73 in the left eye). Visual acuity improved following surgery in both children. Subsequently, relatively worse vision by SSVEP developed in the fellow eyes (patient 3: 20/40 in the right eye and 20/30 in the left eye; patient 5: 20/65 in the right eye and 20/54 in the left eye). The vision equalized in both patients following surgery on the second eyes (Table 1). Patient 6 had rod dystrophy and developmental delay. SSVEP was nonrecordable prior to surgery and improved to 20/51 in both eyes postoperatively. All of the patients had good final visual acuities (Table 1). Mean follow-up for all patients was 5.3 years (range, 2.5-8 years).

There were no intraoperative complications. All patients had successful clearance of the visual axis (Figures 2-3) and good visual acuity. Slight corectopia was noted in 2 eyes and 1 patient with unilateral PPM had 1 mm of anisocoria postoperatively.

Discussion

During the early weeks of gestation, the developing eye has a rich vascular supply, which largely involutes by birth. Persistent fetal vasculature (previously known as persistent hyperplastic primary vitreous) was initially described in 1946 as a failure of this early blood system to regress. It was later recognized that some or all of the intraocular fetal vessels may fail to resorb, creating a spectrum of disease affecting the anterior, posterior, or both compartments of the eye. In the growing fetus, the anterior tunica vasculosa lentis stretches forward from the hyaloid artery and envelopes the lens. This structure typically involutes during the third trimester. Incomplete regression leads to a
PPM. This is usually a sporadic occurrence, but familial cases have been reported. The severity and associated ocular findings vary.

The majority of PPMs are fine, wispy iris strands that regress within the first weeks of life. They may also be large and dense enough to affect vision, necessitating intervention. In older children, monitoring for the development of amblyopia can usually be achieved in the clinic. However, owing to the congenital nature of the anomaly, patients with PPMs are usually preverbal on presentation. Therefore, the decision to operate hinges on identification of those patients with PPMs substantial enough to potentially compromise visual development. The 10 eyes in the current study that required surgery were found to have one or more of the following characteristics: impaired retinoscopic reflex, obscured view of the fundus through the lesion, and decreased visual acuity as measured by SSVEP.

For select PPMs, there are nonsurgical management options, including pharmacologic mydriasis and close observation. There have been reports of good visual outcomes in asymptomatic patients who were observed. This has led some authors to suggest a trial of refractive correction and occlusion therapy for amblyopia-related vision loss before deciding on surgical or laser intervention. However, there have been no large studies detailing the natural history of PPMs, and it is therefore not possible to determine whether the patients who improved in these case reports are representative or outliers. Similar to congenital cataracts, the amblyogenic risk of a PPM can be difficult to quantify in infants, and there is a risk that delayed treatment could lead to irreversible vision loss. We felt that the membranes in the patients described herein were substantial enough to warrant surgical removal, based on the size and density of the membranes and the other criteria listed above.

The primary limitation of this study is that we do not know what the visual outcomes in our patients would have been without surgical intervention. The decision to operate in the 3 youngest patients (1, 2, and 4) was based on the density and size of the membranes alone, due to the concern of deprivation amblyopia with delayed treatment.

### Table 1. Patient data

<table>
<thead>
<tr>
<th>Patient</th>
<th>Eye</th>
<th>Age at surgery, months</th>
<th>Indications for surgery</th>
<th>Final VA</th>
<th>Follow-up, years</th>
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<td>20/20 OU</td>
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<tr>
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<td>5</td>
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<tr>
<td>2</td>
<td>OS</td>
<td>7</td>
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<td>30</td>
<td>Decreased SSVEP</td>
<td>20/25</td>
<td>3</td>
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<td>24</td>
<td>Decreased SSVEP</td>
<td>20/25</td>
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<tr>
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<td>OS</td>
<td>2.5</td>
<td>Obscured visual axis</td>
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<tr>
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<td>Nonrecordable SSVEP</td>
<td>20/50 (SSVEP)</td>
<td>7</td>
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</tr>
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</table>

**OD**, right eye; **OS**, left eye; **OU**, both eyes; **SSVEP**, spatial sweep visual evoked potential; **VA**, visual acuity.

![FIG 2. Preoperative membrane in patient 3.](image1)

![FIG 3. Postoperative appearance of patient 3.](image2)
In the other 3 patients (3, 5, and 6) there was SSVEP evidence of decreased vision. Although SSVEP does not precisely correlate with optotype acuity, the relative difference in visual acuity between eyes can help guide management. In patients 3 and 4 the relative visual acuity improved following surgery in both patients following surgery on the initially more affected eye. In patient 6, who was developmentally delayed, the initial SSVEP was nonrecordable and improved to 20/51 following surgery. The major strength of the study is the long-term follow-up, with all patients except patient 6 able to perform optotype acuity at their final visits, with good visual outcomes in all.

Use of Nd:YAG laser for membranectomy has been described, and successfully employed in older patients. This technique is conceptually appealing because it avoids incisional surgery and the associated risks. However, laser lysis is technically difficult in young children and may not be effective in cases of thick, fibrotic membranes. Moreover, because the membranes may have active vasculature, photodisruption could lead to hyphema. Although none of our patients developed hyphemas, it would be easier to manage this complication using an intraocular approach. In addition, laser lysis poses a risk of pigment dispersion.

Various intraocular surgical techniques for removing PPMs have been described. Reports differ with regard to incisions, instruments used to manipulate and remove the PPM, and incision closure. Our approach is modified from those previously described. Rather than a 4.0 mm incision or paired smaller incisions, we use a solitary stab-incision, which allows for a more stable chamber and closure with a single suture. Vitrectomy scissors have been recommended for separating the membrane from the iris. After gently peeling the membrane from the lens, we use intraocular scissors to cut the strands at their origins, thus eliminating the need for a separate infusion port. We believe that the use of a nonautomated instrument also reduces the risk of complications. It has been suggested that the use of intraocular scissors in the anterior chamber increases the risk of inducing a traumatic cataract; however, our patients had excellent outcomes and no induced lens opacities. We also use intraoperative 0.01% carbachol following removal of the membrane to assess pupillary mobility and look for residual adhesions.

There were no significant complications in any patient using our technique, which may be considered for patients with visually significant PPMs to improve visual function and pupil appearance.

References