Persistent fetal vasculature (PFV) is a condition caused by failure of apoptosis of the primary hyaloidal vasculature system, incomplete ocular vascular development, or a combination of both. PFV may present with retinal detachment and secondary glaucoma, with a wide range of microphthalmia from an almost normal sized eye to a very small eye. Because PFV has a wide spectrum at the time of presentation, surgical management continues to be challenging.

**RT:** Dr. Berrocal, how often do you see PFV?

**Dr. Berrocal:** I see PFV quite often. For example, last month I had three cases presenting within 2 weeks. Practicing at a major referral center, these cases are more common.

**RT:** How are these patients picked up?

**Dr. Berrocal:** Usually one of the parents or a relative notices an abnormality in one of the baby’s eyes. After visiting a pediatrician, who refers to a pediatric ophthalmologist, the family is ultimately referred to a retina specialist.

**RT:** What do you tell the parents?

**Dr. Berrocal:** I actually sit and chat with them. I prefer parents who do preliminary research on the disease because it makes it much easier for me. I examine the baby in the clinic and usually do a baseline ultrasound to have an idea of axial length and retinal involvement. If the eye is very small and the asymmetry between the two eyes is large, I know that the prognosis for visual development and surgical success will be more limited. When eyes are smaller than 15 mm or there is a disparity between the eyes larger than 3.5 mm, these eyes do not tend to achieve better than light perception vision despite our best efforts.
What this says is that the more normal the eye in size, the better the prognosis. Also, we have to take into account the retinal findings. These can range from a persistent hyaloideal stalk with no vascular flow to complete tractional retinal detachment. Again, the more normal the retina, the better the prognosis. The last factor is the anterior chamber involvement. We usually find cataracts with a thick posterior plate, displacement of the ciliary body, and a shallow anterior chamber. Depending on the severity of these factors, the better or worse the prognosis. Many of these children will eventually develop aphakic glaucoma requiring surgical intervention. Parents must be aware of all these issues prior to surgical intervention.

**RT:** *What is your surgical approach?*

**Dr. Berrocal:** After having been in practice for 10 years, I have to say these are among my favorite surgical cases because they are always different and very challenging. I decide my surgical approach in the OR after we perform an examination under anesthesia, repeat the ultrasound, and take photographs of the eye. If it seems that the eye is almost normal, there is no anterior traction noted, and I can be sure (either by exam or by ultrasound) that the anterior retina is normal and there is a pars plana/plicata, then I will use a posterior approach with 23-gauge instrumentation. If I cannot confirm these things, I will use a limbal approach with 23-gauge instrumentation. In these cases, it is important to minimize vitreous traction. The goal is to remove the cataract and release the traction on the retina.
23-gauge Surgery for Persistent Fetal Vasculature
By Audina M. Berrocal, MD

direct link to video:
http://eyetube.net/video/23-gauge-surgery-for-persistent-fetal-vasculature/

RT: Are there any new surgical techniques in this condition?

Dr. Berrocal: Small-gauge surgery for pediatric cases is a real advantage in many ways. I sometimes use 23-gauge, with or without trocars, depending on the case. If I use trocars in small eyes with thin scleras, I like to go straight in with the trocar, and then I always suture the sclerotomies. If I do not use trocars, I use the trocar inserter blade to make the sclerotomies. I like 23 gauge vs 25 or 25+ (Alcon Laboratories, Inc., Fort Worth, TX) because the instruments are a bit stiffer, and if the lens plate in PFV is calcified, 23 gauge works better to remove this tissue. Recently, I have begun performing intraoperative optical coherence tomography (OCT) in these cases, enabling me to look at the posterior portion of the lens. With OCT, we may be able to determine how involved the lens is and whether sacrifice is necessary. There are PFV cases in which the lens is almost normal with slight opacity. OCT can determine whether the opacity involves the lens or if it just goes to the anterior face of the hyaloid, a very important difference for the development of vision in those eyes.

RT: What other surgical pearls can you offer with regard to PFV?

Dr. Berrocal: In some more infrequent cases of PFV, the tunica vasculosa is still present and seems very vascular. Once you infuse the eye and the intraocular pressure goes up, these vessels tend to disappear—and when they are cut with the vitrector they rarely bleed, so one need not worry if they are present. I like to take anterior angle photographs with a digital camera (RetCam, Clarity Medical, Pleasanton, CA) in the OR. Often, the degree to which the angle is affected can be assessed preoperatively, which is good for documentation.

Last but not least, if there is an intraoperative retinal detachment, it is important to perform a thorough vitrectomy, to apply laser, and to use intraocular gas. I do not recommend scleral buckling rather, most patients do well with intraocular gas, with parental compliance regarding head positioning.

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