



Persistent Fetal Vasculature (PFV)

WHAT IS PERSISTENT FETAL VASCULATURE?

Persistent fetal vasculature (PFV), previously known as persistent hyperplastic primary vitreous (PHPV), is a developmental disorder of the eye. As the eye matures it is nourished by a set of embryonic blood vessels. Normally, these blood vessels completely disappear when the eye has finished its development. Failure of these vessels to regress causes this disorder.

HOW COMMON IS PFV?

Many people have ocular signs of these embryonic vessels. For the most part these are benign and can be seen by an eye doctor under magnification. Your child's doctor may comment on them during an eye examination. More severe variants of PFV are quite rare. Unfortunately, these severe variations may profoundly affect the child's visual development.

IS PFV HERITABLE?

Most cases of PFV are sporadic. However, with the advancement of molecular genetics, some familial cases of PFV have been identified.

DOES PFV AFFECT ONE OR BOTH EYES?

PFV usually affects only one eye (e.g. unilateral). Both eyes (e.g. bilateral) may be affected in about 5-10 percent of cases. When bilateral cases occur, there may be an underlying systemic and/or genetic condition. These conditions include Norrie disease and other central nervous system disorders.

HOW MIGHT I KNOW IF MY CHILD HAS A SEVERE VARIANT OF PFV?

Clinical signs vary from case to case and usually appear within a few weeks of birth. The parents or pediatrician may notice that one eye is smaller. The pediatrician might also notice a white pupil when checking for the baby's red reflex in the office (e.g. leukocoria). An eye misalignment (strabismus) may also be present.

HOW MIGHT PFV AFFECT MY CHILD'S VISUAL DEVELOPMENT?

Severe manifestations of PFV include a cataract, which is a clouding of the crystalline lens inside the eye. Other manifestations include poor visual development (e.g. amblyopia), strabismus (crossing or drifting of the eyes), high pressure inside the eye (e.g. glaucoma), and/or retinal detachments.

HOW IS PFV TREATED?

Treatment of PFV varies by patient and depends on the severity and presence of certain features. Less severe cases may be simply monitored by a pediatric ophthalmologist. Treatment may include the use of glasses and patching to ensure appropriate visual development. Examination in the office may be



difficult, in which case your doctor might recommend an examination under anesthesia (EUA). More severe cases may require cataract surgery to replace a cloudy lens, eye muscle surgery to correct strabismus, and retina surgery if the retina becomes detached (separated) from the back of the eye.

WHAT IS THE PROGNOSIS FOR PFV?

Visual outcomes are variable and can be quite poor in severe PFV. Even after successful surgery, visual development may still be severely affected. Continued follow-up is essential and may include the use of glasses, contact lenses, eye patching to treat amblyopia, monitoring of intraocular pressure to detect the development of glaucoma, and further surgery if other problems arise. Your pediatric ophthalmologist will check for these problems at each visit.