

Persistent Fetal Vasculature (PFV)

WHAT IS PERSISTENT FETAL VASCULATURE?

Persistent fetal vasculature (PFV) (also called persistent hyperplastic primary vitreous (PHPV)) is a developmental disorder of the eye. As the eye develops, it is fed by embryonic blood vessels. Normally, these blood vessels go away when the eye has fully formed. PFV is caused when these blood vessels do not fully go away.

HOW COMMON IS PFV?

Many people have mild signs of these embryonic vessels. For the most part these are not harmful and can be seen by an eye doctor, but do not cause symptoms or problems. More severe variants of PFV are quite rare. Unfortunately, these severe variations may severely affect a child's visual development.

IS PFV HERITABLE?

Most cases of PFV are not inherited. However, with recent advances, some cases of familial 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 findings of PFV include a cataract, which is a clouding of the crystalline lens inside the eye. Other findings 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. Sometimes 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?

Vision varies from person to person, but sometimes can be quite poor in severe PFV. Even after successful surgery, visual development may still be severely affected. Continued follow-up is very important 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.

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