



Persistent Fetal Vasculature (PFV)

WHAT IS PERSISTENT FETAL VASCULATURE?

Persistent fetal vasculature (PFV), also called persistent hyperplastic primary vitreous (PHPV), is a problem with eye development. When the eye is forming, it has blood vessels (arteries and veins) in the early stages. Normally, these blood vessels go away when the eye has fully formed. However, in PFV, these blood vessels do not fully go away. This can lead to the lens in the eye becoming cloudy, which is called a cataract. A cataract from PFV has a specific appearance that your eye doctor can often detect.

HOW COMMON IS PFV?

Many people have mild signs of these early blood vessels. For the most part these are not harmful but may be seen by an eye doctor (ophthalmologist) on a routine eye checkup. More severe forms of PFV can cause vision problems but this is quite rare.

DOES PFV RUN IN FAMILIES?

Although a few cases PFV have been found running in families, most cases of PFV are not inherited, it does not usually get passed down from parent to child.

DOES PFV AFFECT ONE OR BOTH EYES?

PFV usually affects only one eye (unilateral). In about 5-10 percent of cases, it can affect both eyes (bilateral). When both eyes have PFV, there may be an underlying larger medical problem like Norrie disease or brain problems.

HOW WOULD I KNOW IF MY CHILD HAS PFV?

The signs of PFV can be different from person to person and are often noticeable a few weeks after birth. Parents or pediatrician may notice that one eye is smaller. During a checkup, the pediatrician might also see a white spot in the pupil (the dark spot in the center of the colored part of the eye). Sometimes the eye may not be straight (strabismus) in PFV. An ophthalmologist can do a full eye checkup and see if PFV is there.



Figure 1. PFV cataract showing a white spot in the pupil. (Source: <https://www.aao.org/education/bcscsnippetdetail.aspx?id=ba8b8e2c-857b-444a-b7d7-30a12c6e9124>, last accessed 11/2/2023)

HOW MIGHT PFV AFFECT MY CHILD'S VISION?

In severe cases of PFV, some big eye problems can happen like include a cloudy lens (cataract), poor vision/lazy eye (amblyopia), eye crossing or drifting (strabismus), high eye pressure (glaucoma), and/or retinal detachment (where the lining of the back wall of the eye falls off). These problems can cause bad vision and may need treatment and lifelong eye checkups.

HOW IS PFV TREATED?

Treatment of PFV is different from person to person, depends on serious the problem is and what problems are present. Mild cases might just need checkups with a pediatric ophthalmologist. Treatment may include wearing glasses or contact lenses and an eye patch to help vision develop. Sometimes, your ophthalmologist might recommend a special eye exam under anesthesia (EUA). More severe cases may need surgery to fix the problems from PFV, like: cataract surgery, eye muscle surgery, or retina surgery.

HOW DO CHILDREN WITH PFV DO AS THE GET OLDER?

How well children with PFV see, is different from person to person. In severe PFV cases, vision can be really poor. Even with surgery, can still be really poor in some people with PFV. Some problems with PFV, like glaucoma or a retina detachment, can show up later in life. Regular eye checkups are very important to catch and treat problems as they develop. Your pediatric ophthalmologist will check for these problems at each visit.



MORE SCIENTIFIC INFORMATION ON PFV CAN BE FOUND:

https://eyewiki.org/Persistent_Hyperplastic_Primary_Vitreous

Updated 11/2023