Case Report

# Bilateral persistent fetal vasculature: A rare entity

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**Abstract**

Persistent Fetal Vasculature (PFV), previously known as Persistent hyperplastic primary vitreous (PHPV), is a rare congenital developmental malformation of the eye, caused by the failure of regression of the primary vitreous. It can occur in isolation, or in association with other ocular disorders and rarely as a part of systemic disorder. Although genes responsible for PFV are unknown, the lack of inheritance suggests that idiopathic, isolated, sporadic mutations may be responsible for the disease; a developmental defect caused by environmental factors during embryogenesis rather than genetic factors. Characteristic features include microphthalmic eye, white vascularised retrolental tissue with or without a persistent hyaloid artery, centrally dragged ciliary processes, an anteriorly shifted and (or) swollen lens, and varying degrees of lenticular opacification. Radiological investigations (ultrasound, computerised tomography, magnetic resonance imaging) aid in the diagnosis and differentiation from other causes of leukocoria like retinoblastoma. This case report discusses an 8 month old male child with bilateral Persistent Fetal Vasculature.
**Keywords:** Persistent Fetal Vasculature, leukocoria, microphthalmos.

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## INTRODUCTION

Persistent hyperplastic primary vitreous (PHPV) is a congenital anomaly caused by failure of regression of the primitive hyaloid vascular system first described by Reese.[1] Goldberg later renamed the entity persistent fetal vasculature (PFV) since this term more accurately describes the anatomic and pathologic features of the disease.[2] The condition is almost always unilateral and usually presents as leukocoria, microphthalmia, and cataract. Bilateral cases are very rare[5,6]. Although most cases of PFV are sporadic, it can be inherited as an autosomal dominant or recessive trait. Primary vitreous forms around 7th week of intrauterine life and starts involuting around 20th week and nearly always disappears at the time of birth. Failure of regression of primary vitreous results in many of the abnormalities seen in PFV.[7] In these cases, the child usually presents due to poor vision and white reflex. [8] The entity often presents a diagnostic challenge as it is often confused with retinoblastoma.[9] PFV can occur with other disorders such as Axenfeld Rieger syndrome, Peter's anomaly, myopia, osteoporosis pseudoglioma syndrome, morning glory syndrome, megalocornea, bilateral retinal vascular hypoplasia, neurological abnormalities, neurofibromatosis, Aicardi syndrome. Pupillary strands and a Mittendorf's dot represent the mildest manifestations and leukocoria with a dense retrolenticular membrane and or retinal detachment the most severe. Depending on which intraocular structures are involved it is divided into anterior, posterior or a combination of anterior and posterior.[8] The anterior type of PFV includes a shallow or collapsed anterior chamber, a retrolental vascular membrane, cataract and anterior chamber anomalies.[10] Abnormalities of lens and anterior chamber are signs of combined anterior and posterior variant of PFV.[11] Posterior PFV consists of a prominent vitreous fibrovascular stalk that emanates from the optic nerve and courses anteriorly. Preretinal membranes at the base of the stalk are common. Tractional retinal folds and traction retinal detachment may be present. Retinal dysplasia and optic nerve hypoplasia have also been described. Various degrees of microphthalmia and leukokoria may occur in

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both anterior and posterior PFV. Complications of PFV can be rupture of lens capsule, cataract, intraocular hemorrhage, secondary glaucoma, traction retinal fold, and subsequent phthisis bulbi.[8] Differential diagnosis of PFV includes retinoblastoma, retinopathy of prematurity, Coat's disease, toxocariasis, Norrie's disease, familial exudative vitreoretinopathy, congenital cataract, uveitis, and incontinentia pigmenti.[10]

## CASE REPORT

An 8-month-old male child was referred to the department of ophthalmology. The informant was the mother. She complained of the baby having poor vision and a white reflex in both the pupillary area since birth. The baby was born at 36 weeks of gestation with birth weight being 1.700 kgs. There was history of NICU admission for 1 month after birth due to low birth weight. The mother's age was 24years and this was her first child, born out of a non consanguineous marriage. There was no history of trauma. Family history was also negative.

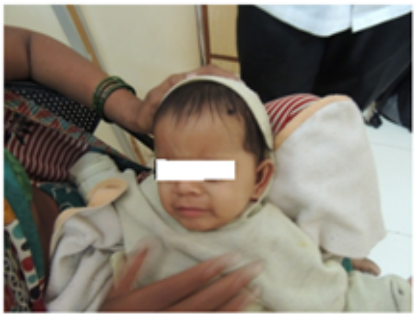
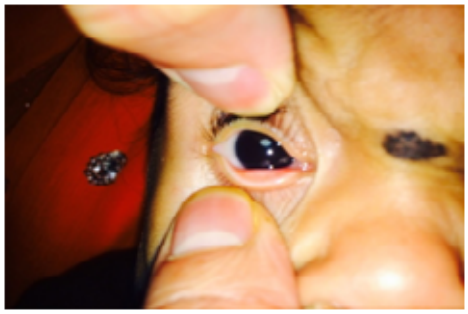


Figure 1:



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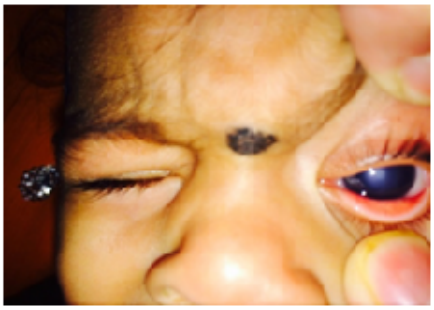
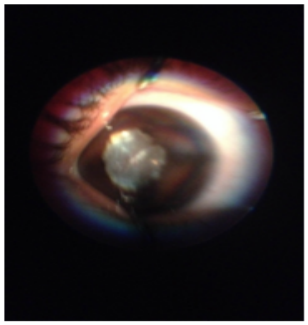


Figure 3:

The baby's general condition was stable. Baby was afebrile and irritable. There was no evidence of any cyanosis, pallor, icterus, oedema or any evident lymphadenopathy on palpation of pre auricular and submandibular lymph nodes. The general appearance, head, back, spine and other extremities were normal. On ocular examination, during vision assessment, the patient did not follow light. On torch light examination, both pupils were normal in size, not reacting to light. There was a whitish reflex in the pupillary area, occupying the entire pupillary area (Leukocoria). No fundus glow was seen on distant direct ophthalmoscopy. Intra ocular pressure was measured using Schiotz's tonometer. Tension in the right eye was 31.8 mmHg and in the left eye was 27.2. For dialted fundus examination, tropicamide and phenylephrine eye drops were instilled in both the eyes. Examination under sedation was performed. On dilatation, both pupils were irregular in shape approximately 4mm dilating. A whitish mass occupying the entire pupillary area was noted, which was dirty white in colour. No fundal glow was seen using the direct or indirect ophthalmoscope. On slit lamp examinatiom, the anterior chamber was shallow in both eyes. Both corneas had an even surface and was normal in shape and transparency. Corneal diameter of the of the right eye was 8.9 mm and left eye was 9.1 mm. Posterior synechiae was noted in both the eyes. The iris-lens diaphragm appeared to be pushed forward.



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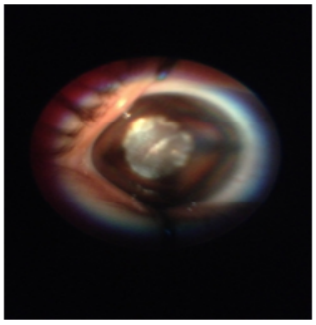

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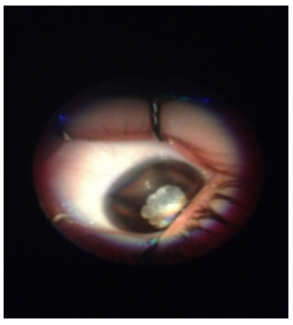

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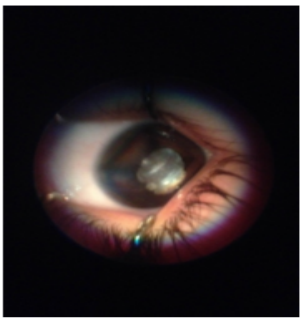

Figure 7:

Magnetic resonance imaging (MRI) of brain and orbit was done which showed the following:

- Bilateral microphthalmos
- Axial lengths of right and left eye are 16.2 and 16.1 mm, respectively
- Bilateral orbit shows crescentric smotthly marginated isointensity on T1W1; hyperintensity

on T2W1 in posterior chamber extending along retinal attachment circumferentially reaching up to lens without evidence of diffuse restriction or blooming on SW1.

- No evidence of intra ocular calcification or haemorrhage seen.
- Irregular curvilinear hypointensity on T1W1 and hyperintense on T2W1 displaced anterior to crescentric hyperintensity (descrbed above), likely to be proliferation of fibrovascular tissue and retinal folds which are displaced and mobile.
- Linear tubular hypointensity extending from optic nerve up to the lens seen likely due to the persistence of hyaloid canal ( proliferation of fibrovascular tissue more of embryonic type of connective tissue and creating a retro lental fibrovascular mass)
- Optic nerve appears normal bilaterally.

MRI brain was within normal limits.All the above features were suggestive of Bilateral Persistent Fetal Vasculature.

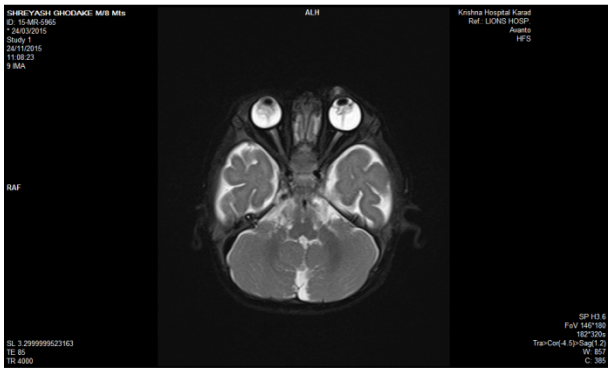


**Figure 8:**

## DISCUSSION

Persistent Fetal Vasculature is a congenital anomaly of the eye that results following failure of embryological primary vitreous and hyaloid vasculature to regress. It is characterized by persistence of various portions of the primary vitreous (embryonic hyaloid vascular system) with hyperplasia of the associated embryonic connective tissue. The primary vitreous is formed during the first month of development and contains branches of the hyaloid artery. This hyaloid artery begins to regress during the formation of the avascular secondary vitreous at 9 weeks. By the third month, the secondary vitreous, which ultimately forms the adult vitreous, fills most of the developing vitreous cavity. The primary vitreous becomes condensed into a narrow band (Cloquet's canal), running from the optic disc to the posterior aspect of the lens. The most common presenting signs are leukoria (white papillary reflex due to a dense retrolenticular membrane or cataractous lens), microphthalmia and cataract. PFV is classified into three types: anterior, posterior, or a combination of the two. Mild PFV can run a relatively benign natural course without surgery. Surgery may be avoided if the visual axis is clear, anatomical anomalies are not progressive, and the anterior chamber angle is not compromised**.** Recent advances in surgical techniques have improved the prognosis of PFV. The standard surgical method for lensectomy in infants, described in textbooks and literatures, is pars plana vitrectomy, or more accurately, pars plicata vitrectomy, since the anterior-to-posterior width of the pars plana is narrow in infants. In case of implanting the intraocular lens, cataract surgery can be done as usual as in adults, and then posterior capsulotomy with anterior vitrectomy can be additionally performed through the pars plana or plicata.[14] Surgery for severe posterior PFV is rarely undertaken. Although the fibrovascular tissue can be dissected and the accompanying retinal detachment may approximate to the retinal pigment epithelium (RPE), visual outcomes are quite poor.[12] This is a result of the associated retinal dysplasia. Bilateral disease is also typically associated with a poor outcome due to the high prevalence of posterior component. It is important to exclude retinoblastoma in all cases of leukokoria. PFV can be differentiated from retinoblastoma by the absence of a calcified mass, artery running through Cloquet's canal, and typical signal characteristics of retinoblastoma on MRI. Differentiation from advanced retinopathy of prematurity (ROP) can be difficult on imaging alone. History of a premature, low birth weight infant undergoing prolonged supplemental oxygen therapy helps to distinguish it from bilateral PHPV. It is differentiated from vitreoretinal dysplasia by a patent hyaloid artery at it is not a feature of vitreoretinal dysplasia. Although rare,

bilateral PFV should be considered in the differential diagnosis while evaluating a case of bilateral leukocoria. Typical imaging features of ultrasound, CT, and MRI can be helpful in the diagnosis and are important in differentiating this entity from the retinoblastoma.

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