

# A Case Report of Persistent Fetal Vasculature with Posterior Staphyloma and its Management

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**Abstract:** *Persistent fetal vasculature (PFV) is an ocular developmental disorder resulting from incomplete apoptosis of the embryonic hyaloid vasculature. Unilateral PFV is traditionally associated with a poor prognosis because of the challenges associated with its management. We report a child with unilateral PFV with posterior staphyloma who underwent lens aspiration, primary posterior capsulotomy, anterior vitrectomy, excision of the anterior portion of the stalk of PFV, with cauterization of the hyaloid stalk, and intraocular lens (IOL) implantation. The final best corrected visual acuity was 6/9(P) in the affected eye and without evidence of glaucomatous optic neuropathy. This case illustrates the excellent visual outcome possible in a child with complex, unilateral PFV using an intensive management approach comprising early surgical intervention, glaucoma monitoring, meticulous monitoring of refraction, visual acuity and intraocular pressure, and motivated parents who engaged in the management.*

**Keywords:** Persistent fetal vasculature, posterior staphyloma, lens aspiration, primary posterior capsulotomy, anterior vitrectomy, cauterization of hyaloid stalk, intraocular pressure, myopia, refraction

## 1. Background

Persistent fetal vasculature (PFV) results from persistence of the embryonic hyaloid vasculature manifesting variably with a combination of anterior and posterior segment abnormalities. Unilateral PFV was often managed conservatively or associated with poor outcomes due to difficulties in the long-term management of progressive amblyopia. We report a child with unilateral PFV with posterior staphyloma, who had an excellent visual outcome beyond visual maturity, with lens aspiration, primary posterior capsulotomy, anterior vitrectomy, excision of the anterior portion of the stalk of PFV, with cauterization of the hyaloid stalk, and IOL implantation. This case demonstrates the excellent long-term visual outcome in children with unilateral PFV with an intensive management approach. We suggest a similar approach be considered in this patient group.

## 2. Case Presentation

A 9 years old female child and having non-consanguineous parents presented to the Ophthalmology out-patient department (OPD) with the complaints of diminution of vision and presence of white opacity in the right eye since 2-3 years, informant being the patient's mother. There was neither any history of local or systemic disease nor any history of operative procedure or trauma to the right eye. The antenatal, intranatal and postnatal birth history was normal. Immunization was up to date according to Indian Academy of Pediatrics guidelines. There was no family history of any congenital disease. General and systemic examinations were normal.

**Ocular Examination:** Eye examination revealed right eye leukocoria with vision of hand movements close to face (perception of light present and projection of rays accurate) in right eye and 6/6 in left eye. Pupillary light reflexes were normal and there was no relative afferent pupillary defect. Orthophoria was present. The intraocular pressure was 12 mm Hg in both eyes, measured with Perkin's applanation tonometer. Dilated fundus examination revealed a stalk like structure extending from adjacent to the optic disc to the white opacity seen in papillary area,

suggestive of persistent fetal vasculature, with retinal excavation seen superonasal to the optic disc suggestive of posterior staphyloma. Patient had an improvement in vision in the right eye when vision was checked again with both eyes dilated, i.e. she gave a vision of 6/60 in the right eye improving to 6/24 on refraction with -4.0 Diopters (D) spherical concave lens.

**Investigations:** B scan ultrasonography (USG) was done and PFV and posterior staphyloma were confirmed with axial length of 25.4 mm in right eye and 21.8 mm in left eye. Colour doppler did not show the presence of blood flow within the stalk seen in the vitreous cavity (i.e. PFV). Chest X-ray was normal. Routine blood examination including ESR was normal. Ultrasound abdomen showed normal study. Serological tests and TORCH titres were found to be negative.

Keratometry readings were obtained using autokeratometry and axial length was determined using A-scan USG (25.9 mm right eye). Biometry calculations were done using the A-constant of 118.5 for the Aurolab's3 piece IOL (made from ultraviolet light absorbing high quality clinically proven polymethyl methacrylate material).



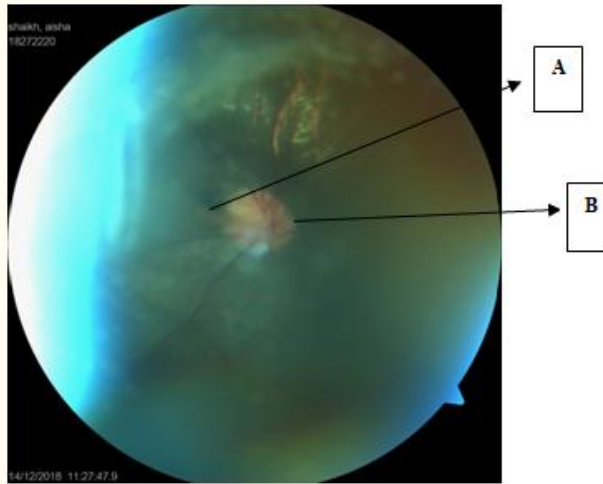
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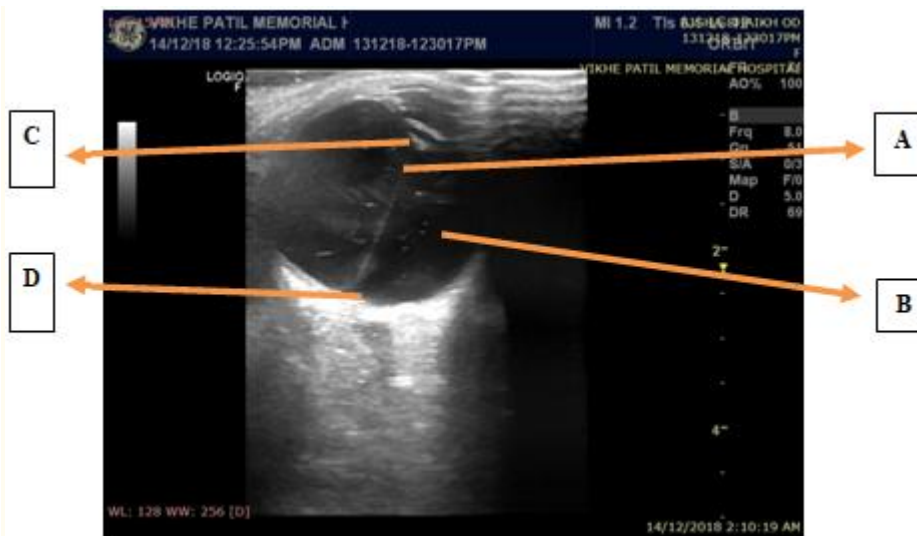
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Anterior Segment Examination with Dilated Pupil Showing  
Retrolental Mass Attached on the Posterior Surface of the

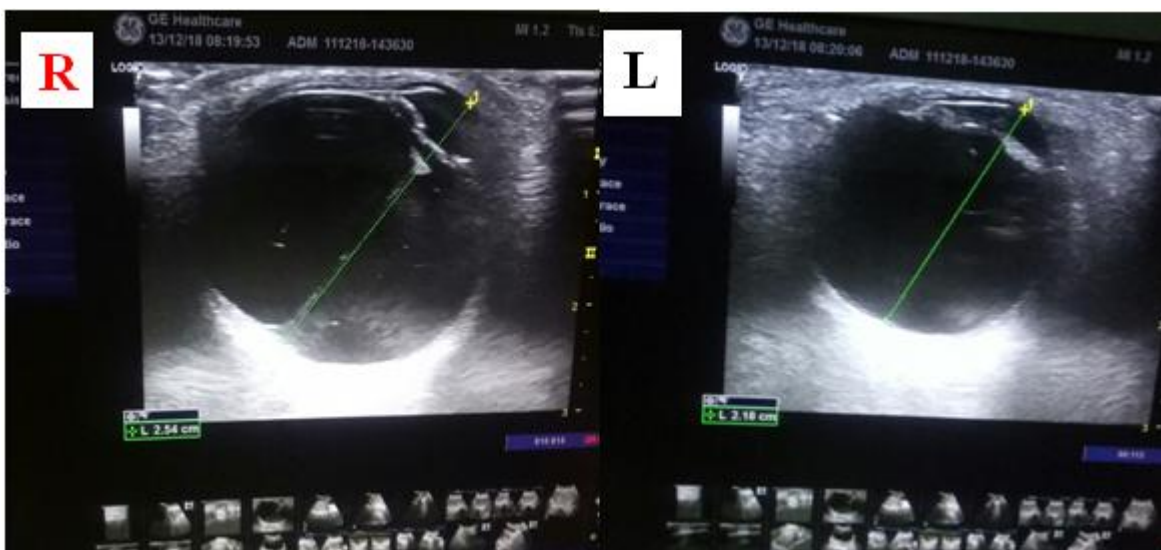
Lens Capsule in the Right Eye



Pre-Operative Fundus Photograph showing a Falciform Fold (A) arising from the optic disc (B) in the right eye.



Pre-Operative USG Scan Showing A Hyperchoic Structure (A) in vitreous cavity (B) extending from posterior lens CAPSULE (C) to the temporal edge of optic DISC (D) suggestive of PFV.



Ocular USG showing PHPV in right eye with axial length of 25.4 MM and normal left eye with Axial Length of 21.8 MM



Pre-Operative Evaluation Showing Orthophoria



C. Implantation of Aurolab's 3 Piece Iol in the Ciliary Sulcus

**Management**

Right lens aspiration with primary posterior capsulorhexis, anterior vitrectomy and excision of the anterior portion of the stalk of PHPV was done, with cauterization of the persistent anterior hyaloid stalk, and implantation of a +18.0 D intraocular lens (3 piece IOL, Aurolab).



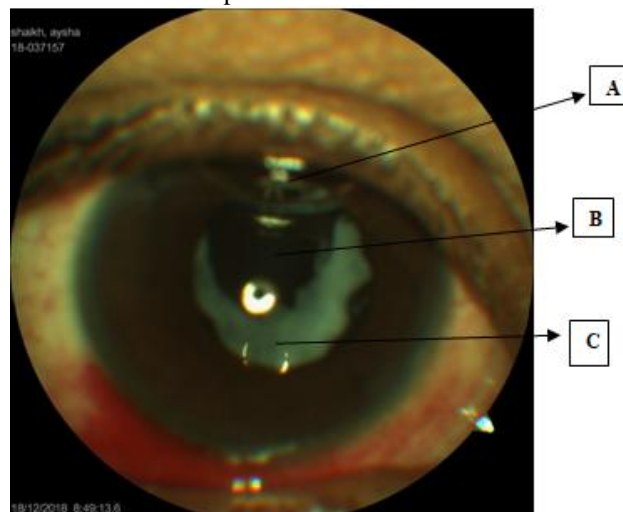
A. Excision of the Anterior Portion of the Stalk of PHPV, after Lens Aspiration, Primary Posterior Capsulotomy and Anterior Vitrectomy were done



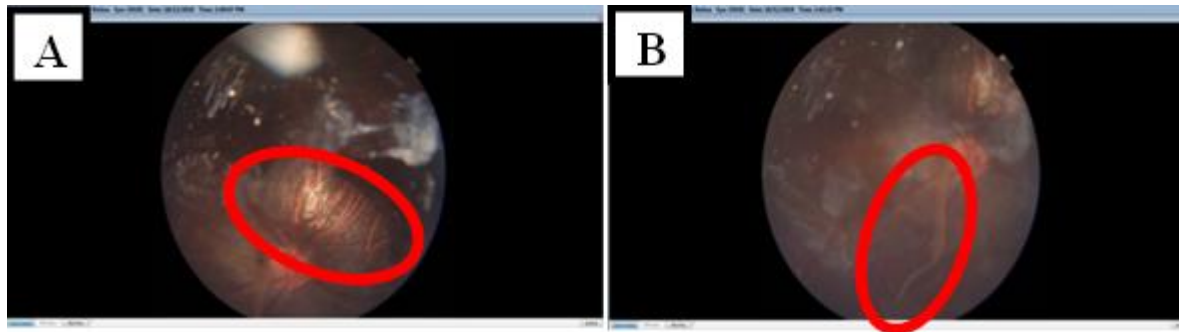
B. Cauterization of the Hyaloid Stalk after Excision of its Anterior Portion

**Post-Operative Status**

Postoperative cycloplegic refraction revealed an error of -0.50/-1.50X 180 in the right eye. The right eye was prescribed -0.50/-1.5X180 with +2.00 D spherical near additonto aid in near fixation, the child's vision improving to 6/9(P) and N6. Intraocular pressure monitoring and optic nerve head evaluation was done in every visitpostoperatively. The child's motivated parents gave regular follow-ups which was required to manage and monitor the visual response of the child.



Anterior Segment Photograph of First Post-Operative Day Showing Air Bubble(A), PCIOL in Situ(B) and Peripheral Retrolental Plaque of PFV Stalk(C) Left Undisturbed.

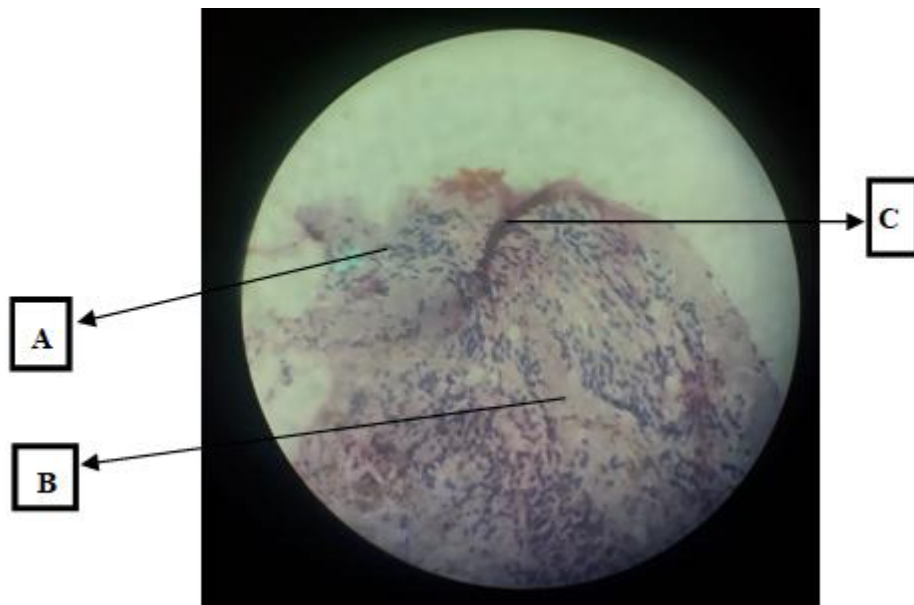


Fundus Photograph of First Post-Operative Day showing Posterior Staphyloma Superonasally (A), and Stalk of PFV is Seen Lying in the Vitreous Cavity (B)

### Histopathology

Light microscopic evaluation of histological sections stained with hematoxylin-eosin, confirmed the presence of PFV. It

was composed of fibroblasts, a meshwork of collagen fibres and a dense fibrous tissue suggestive of blood vessel (hyaloid vasculature).



Histological Section Stained with Hematoxylin-Eosin showing the Presence of Fibroblasts(A), A Meshwork of Collagen Fibres (B) and A Dense Fibromuscular Tissue Suggestive of Blood Vessel (Hyaloid Vasculature: C)

### 3. Discussion

Persistent hyperplastic primary vitreous (PHPV), also referred to in recent years as persistent fetal vasculature (PFV), is an uncommon congenital developmental anomaly of the eye first described in detail by Reese AB in 1955.<sup>(1)</sup> It was renamed by Goldberg M as Persistent Fetal Vasculature Syndrome (PFVS) in 1997 which refers to the persistence of embryonic primary vitreous and fetal hyaloid vasculature due to arrest in the process of their normal involution by the stipulated time.<sup>(2)</sup> It is a developmental disorder of the eye characterized by failure of regression of the embryological intravitreal and perilental vasculature, proliferation of fibroblasts associated with that vasculature, and ultimately impairment of normal ocular growth.<sup>(3)</sup> During embryonic development of the eye, the compartment between the retina and crystalline lens contains a vascular system (hyaloid artery) that provides nutrients for the developing eye. The hyaloid vessels and the primary vitreous are supposed to regress in the third trimester of pregnancy.

PFV may be due to a defect in the regression of the primary

vitreous or in the formation of the secondary vitreous (that fills the developing second eye and is derived from the inner retinal cells starting in the 9th week of gestation) or to a combination of both. The persistent hyaloid vasculature and mesenchymal tissue from the embryonic primary vitreous in a microphthalmic eye leads to the clinical spectrum of PFV.<sup>(4)</sup> The exact cause and pathogenetical mechanisms, however, remain poorly understood.

Posterior staphyloma is diagnosed by ophthalmoscopy, which shows an area of retinal excavation in the region of the staphyloma. Staphyloma is the term for a thinning of the outer, white coat of the eye (the sclera) in which the underlying pigmented tissue then adds its color to the thinned sclera, giving an appearance of bluish to almost black color. Spaide clearly defines posterior staphyloma as “an outpouching of the wall of the eye that has a radius of curvature that is less than the surrounding curvature of the wall of the eye”.<sup>(5)</sup> The author added nasally distorted eye shape to the definition based on results of three-dimensional magnetic resonance imaging (3D MRI) study.<sup>(6)</sup>

PFV can lead to a wide spectrum of ocular complications as a sequelae to its subsequent contraction and opacification like shallow anterior chamber, cataracts, angle closure glaucoma, elongated ciliary processes, retrolental mass, microphthalmos, and retinal detachment eventually leading to phthisis bulbi and intractable glaucoma.<sup>(7)</sup> Depending upon the part and extent of involvement they are classified as anterior, posterior or mixed variety.<sup>(8)</sup>

Patient of PFV most commonly presents unilaterally with white pupillary reflex and microphthalmia without associated systemic finding.<sup>(9)</sup> Bilateral presentations are rare and are known to have systemic and syndromic associations such as trisomy 13, 15 or 18, Norrie's disease, Warburg's syndrome.<sup>(10)</sup> The case discussed here has a typical presentation in being unilateral but with myopic right eye and posterior staphyloma present superonasally and therefore an increased axial length as compared to the fellow normal eye. Moreover, it presented with white pupillary reflex although there was nicrophthalmos. Presence of white reflex makes the condition noticeable to the parents and timely diagnosis with early intervention enables preserving some productive vision.<sup>(11)</sup> Since PFV is a congenital anomaly, proper examination of suspected children with pupillary dilatation is must. Though in our patient media was sufficiently clear to visualise the vitreous band on simple pupillary dilatation, many a times opaque media due to cataract or retrolental mass warrants additional radiological investigations like ultrasonography or computed tomography (CT) scan.<sup>(12)</sup> It not only diagnoses PFV but also classifies it into anterior, posterior or mixed variety essential to plan further line of management. Nevertheless it rules out retinoblastoma, one of the important differentials with life threatening potentials.<sup>(13)</sup> Although visual prognosis of PFV is generally poor, but timing of intervention and amblyopia management are the most important determinants for final visual prognosis.<sup>(14)</sup>

Despite our patient presenting very late, the eyes were not amblyopic, probably due to the central involvement of the opacity leaving a clear margin at the periphery, which was not noticed by the parents until 9 years of age. It was noticed only when the central lenticular opacity began to occupy the entire pupillary area and the patient started experiencing diminution of vision in the affected eye. Though, she had mixed variety of PFV, there was only opacification of the lens at the anterior stalk but since it was involving the visual axis it caused marked visual loss. Presence of the posterior staphyloma superonasally not involving the macula or the optic nerve head, as in our case, helped the patient to achieve an excellent visual outcome of 6/9(P) post-operatively.

Improved surgical techniques, as done in our case, enable more favourable results in the management of PFV.

#### 4. Conclusion

PFV eyes have a potential for developing useful vision with favourable cosmetic outcome after surgery. Intraocular lens implantation may be a favourable and beneficial option for the management of children with unilateral PFV.<sup>(15)</sup>

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