



Pseudo-hyaloidal Stalk in Anterior Persistent Fetal Vasculature: A Report of Two Cases

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Abstract

Persistent fetal vasculature (PFV) syndrome is characterized by abnormal regression of the fetal hyaloid system and may occur in various forms. In this report, two atypical cases associated with posterior capsular defect and ectopic lens material located along Cloquet's canal are discussed. Ultrasonography of these patients presenting with bilateral total cataracts revealed a hyaloidal stalk extending from the optic nerve head to the retrolental area. During lensectomy, it was observed that lens particles were moving anteriorly from the central mid-vitreous to the aspiration port and that the posterior capsule was developmentally defective. There was no pathological vascular remnant, rather the lens material partially filled Cloquet's canal through the opening in the posterior capsule and created a pseudo-stalk appearance on the preoperative ultrasonography. We aim to discuss possible mechanisms underlying these cases, which may help to improve our understanding of the PFV spectrum.

Keywords: Congenital cataract, Cloquet's canal, persistent fetal vasculature, persistent hyperplastic primary vitreous, anatomical variation

Introduction

Persistent fetal vasculature (PFV), previously known as persistent hyperplastic primary vitreous, is a congenital developmental abnormality caused by failed regression of the primary vitreous and hyaloid vasculature.¹ Although typically characterized by the presence of a vascularized retrolental plaque or a hyaloidal stalk extending from the optic disc to the posterior lens capsule, PFV refers to a much broader spectrum of ocular abnormalities with varied clinical presentations and numerous anatomical variations.^{1,2,3} This variation in the spectrum may even include regression of the hyaloid vasculature after causing various pathologies in the eye, as shown in the literature.⁴

Here, we report two cases with bilateral congenital cataracts associated with developmental posterior capsule defects and

ectopic lens material located in Cloquet's canal, which we interpret as a possible expression of an abnormal fetal hyaloid system in the gestational period. We aimed to discuss the possible underlying mechanisms, which may help to improve our understanding of the PFV spectrum.

Case Report

Patient 1 was a 2-month-old boy who was referred to our clinic due to bilateral congenital cataract. He was born full term via normal spontaneous delivery. There were no fetomaternal complications in the pre- or perinatal period nor was he confined to the hospital. Family history was remarkable for an older sibling with bilateral congenital cataracts, the clinical and surgical details of which were not known. Laboratory work-up for

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TORCH (*Toxoplasma*, other agents, rubella, cytomegalovirus, and herpes simplex virus) titers, serum glucose, phosphate, calcium, and urine studies were negative and pediatric evaluation yielded no associated systemic anomalies. Ocular examination revealed near-total white cataract with only a thin peripheral rim of clarity in both eyes (Figure 1a). There was no fibrovascular structure visible within or behind the lens. The corneas were of equal and normal size, and no associated microphthalmia was present. B-scan ultrasonography showed a hyperechoic band extending from the optic nerve head to the posterior lens surface, representing a persistent hyaloidal stalk and leading to the diagnosis of anterior PFV (Figure 1b).

Patient 2 was a 3-month-old boy who was similarly referred due to bilateral congenital cataract. The patient was born full term via cesarean section. The parents were consanguineous and the mother had an infection of unknown cause during pregnancy. The family history was otherwise insignificant. Blood and urine work-up were unremarkable and no systemic abnormalities were identified. Ocular examination showed near-total cataract with a thin peripheral clear zone in both eyes and there was no visible fibrovascular structure within or behind the lens, microcornea, or microphthalmia, similar to patient 1 (Figure 2a, b). B-scan ultrasonography revealed a small stalk emerging from the optic nerve head towards the anterior vitreous in both eyes. Based on these findings, the diagnosis of anterior PFV was made (Figure 2c, d).

Surgical Procedure and Anatomical Findings

The patients underwent combined lensectomy-vitreotomy in both eyes. A limbal approach was utilized in the first patient, and pars plana entries were used in the second patient. Procedural steps and surgical findings were similar in all four eyes, as follows: Following entries, the anterior lens capsule was opened centrally and the lens material was aspirated using a vitrector (Figure 1c). During lensectomy, we noticed lens particles were moving anteriorly from the central mid-vitreous towards the aspiration port, and the central part of posterior capsule was observed to be developmentally defective, with margins demarcated and slightly fibrosed, and have coalescent white opacities (Figure 1d, 2e, f). Additionally, the distance between the anterior and posterior capsule was reduced. The posterior capsule defect (PCD) was gently trimmed with the vitrector to clear the visual axis, and epithelial debris on the capsular leaflets was cleared. In patient 1, it was impossible to clear all of the dot-like opacities that were firmly adhered to the capsules and some of them had to be left in place. Complete aspiration of the lens material revealed that there was no fetal vascular hyaloidal structure extending from the disc, but instead lens material had partially filled Cloquet's canal through the opening in the posterior capsule and created a pseudo-stalk appearance on the preoperative ultrasonography (Figure 1e). The retina was normally attached; hence only core vitrectomy was performed (Figure 1f, 2g, h). The peripheral retina, pars plana, and plicata were normal, free of any pathology in both patients. The eyes were left aphakic and fitted with bilateral

contact lenses in the postoperative period and followed up for 16 months (Patient 1) and 8 months (Patient 2). Patient 1 had a secondary proliferation of the capsular epithelium blocking the pupillary axis in the right eye within 4 months and had a second operation to clear the opacities, which resulted in a clear axis for a 12-month follow-up period. There was no problem or complication noted in either eye of patient 2 during follow-up. Visual acuity was noted as central, steady, and maintained bilaterally in both patients, and nystagmus was observed in patient 2. Both patients had esotropia in the right eye, and 2 hours/day occlusion therapy to the left eye was initiated but discontinued after observing the development of alternating deviation.

Discussion

Developmental defects of the posterior capsule are rare and have been reported in association with congenital cataracts in a few studies.^{5,6,7} Vajpayee and Sandramouli⁵ were the first in the literature to report a pre-existing PCD in a patient with congenital cataract. More recently, Vasavada et al.⁶ reported on 400 eyes that had congenital cataract surgery, 6.75% of which had a preexisting PCD. The authors demonstrated several features commonly found in these eyes to ease the identification of a preexisting PCD, such as well-demarcated, thick defect margins and white dots on the capsule and anterior vitreous. However, all these reports described the location of the lens in its natural position, anterior to the posterior capsule. In contrast, we observed a significant amount of lens material displaced posterior to the PCD and along Cloquet's canal, mimicking the presence of a hyaloid stalk. These findings were similar to those observed by Tandon et al.⁷ in a recent case report. The authors presented an 8-week-old patient with bilateral congenital cataracts that were displaced into the anterior/middle vitreous in association with a pre-existing PCD. Similarly, they reported a small posterior stalk in one eye. However, unlike our observations, it was poorly defined and did not extend to the anteriorly located lenticular opacity. The cataract type was less dense as well, mainly in the form of subcapsular opacities. The presence of a denser, more diffuse cataract and a more prominent stalk on ultrasound may suggest an earlier onset in our patients.

The exact underlying mechanism of developmental PCDs is unknown. Several studies pointed out that PCDs may initially begin as posterior lenticonus.^{4,6,7} Therefore, proposed mechanisms to explain the development of posterior lenticonus (i.e., embryologic hyaloid artery traction on the posterior capsule, inherent weakness of the capsular wall) are likely to be triggering factors in the formation of PCD as well.^{4,6,8} However, unlike the classical posterior lenticonus formation, PCD appears to occur at an accelerated pace and results in a full-thickness defect.

We believe that in the presented cases, an abnormal hyaloid artery and tunica vasculosa lentis system exerted some traction on the posterior capsule, causing the capsule

to stretch outwards and weaken, and as the axial length of the globe increases, proportionally increased traction on the capsule finally led to the development of a PCD. The possible migration of the lens material through the PCD into Cloquet's canal in the presented cases suggests a somewhat different course from the previously reported cases of PCD where the lens material is in its natural position or slightly displaced in the retrolental space or anterior vitreous. The underlying mechanism might be PCD formation much earlier in the gestational period, before lenticular development is complete, either due to inherent weakness of the posterior capsule or stronger traction or both. Indeed, the dense total cataract with slightly fibrosed edges of the PCD in the presented cases may reflect a more chronic time course. Although the lack of a persistent hyaloid artery remnant in

our patients appears to contradict the mechanism proposed here, the literature suggests that as the eye continues to grow, the hyaloid vasculature may resorb even after causing lenticular and capsular changes, leaving no evidence of its involvement in the pathology.⁴ Additionally, our cases did not have microphthalmos or microcornea. As the normal growth of the eye depends on expansion of the secondary vitreous along with involution of the hyaloid vasculature, we can speculate that the hyaloid system formed a PCD early in development, then regressed without interrupting the growth process of the eye, and as the hyaloid vasculature regressed, lens material filled Cloquet's canal through the PCD.

In conclusion, persistent fetal vasculature may present with minimal or even no visible fetal vascular remnants.^{4,9} We

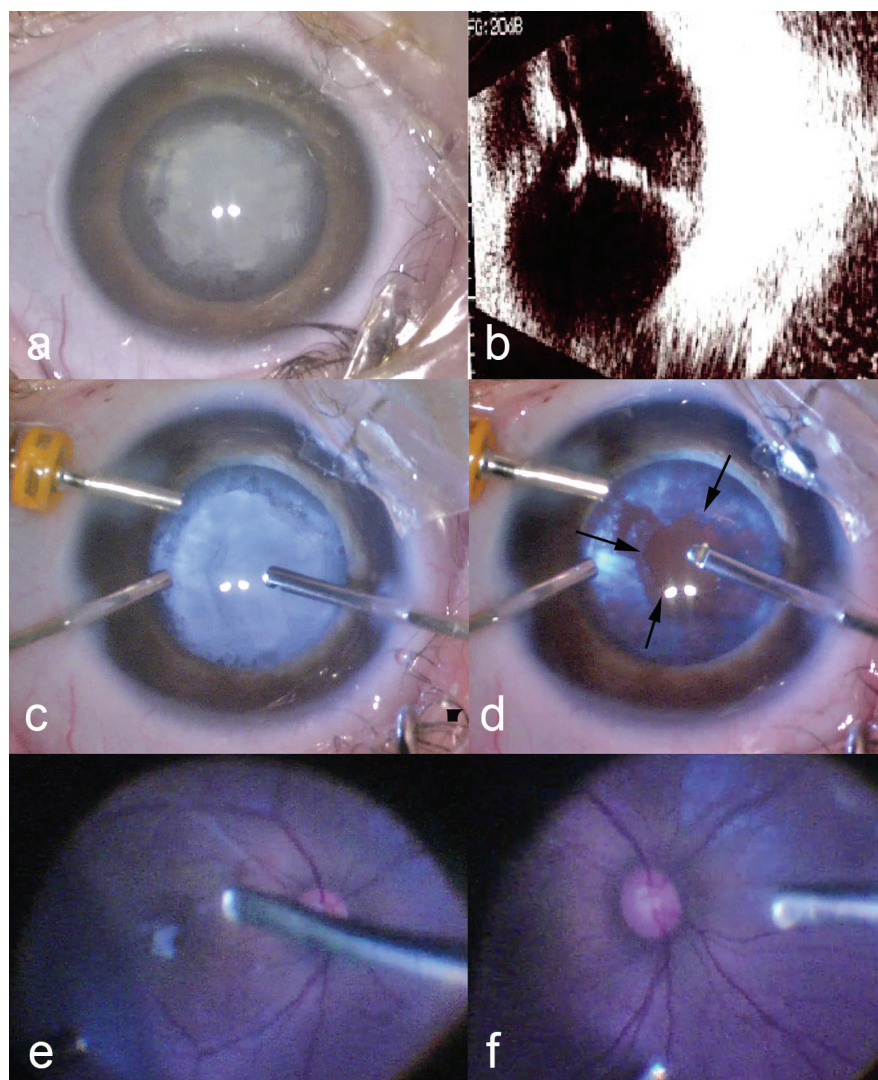


Figure 1. (a) Images of the right eye of patient 1 under the operating microscope showing white cataract in the center surrounded by a relatively clear zone in the periphery. (b) B-scan ultrasonography indicates a highly prominent hyperechoic stalk extending from the optic disc to the posteriorly bulging posterior lens surface. (c) The cataractous lens is located relatively posteriorly in the anterior vitreous. (d) A well-demarcated posterior capsular defect (arrows) and accompanying white dots are seen. (e) Lens particles along Cloquet's canal are removed during central core vitrectomy. (f) The retina is attached and the optic disc is normal without any stalk

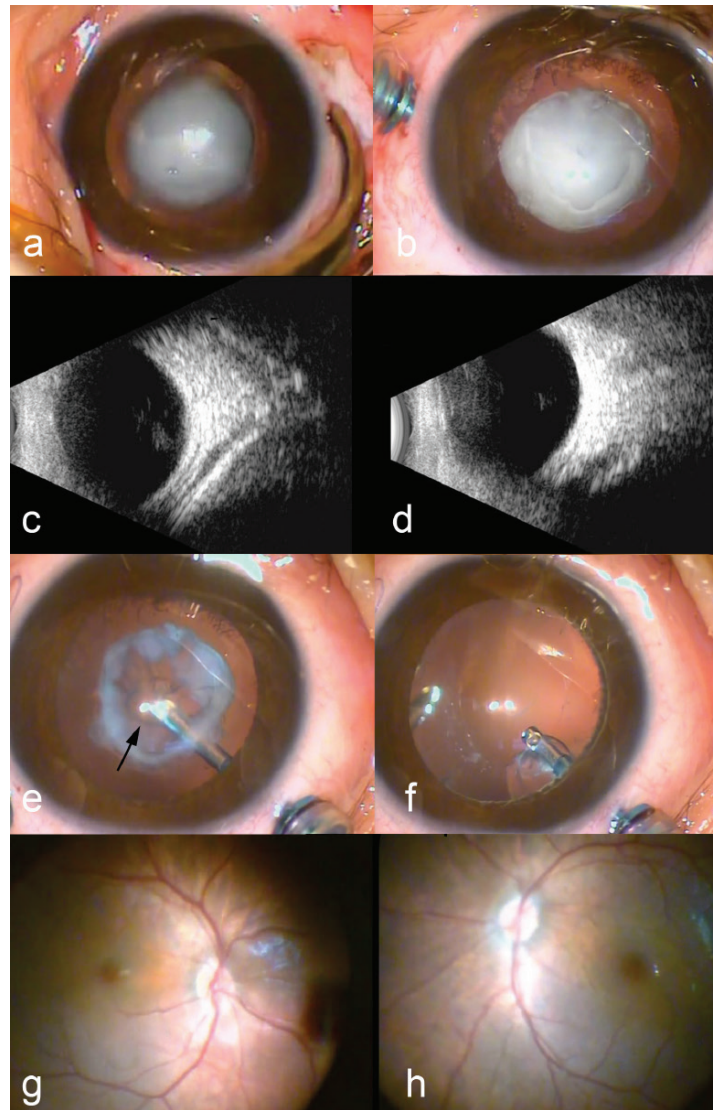


Figure 2. Images of the right (a, c, e, g) and left eye (b, d, f, h) of patient 2. (a, b) A white near-total cataract is seen in the center of the lens in both eyes with a thin clear zone at the periphery. A small hyperechoic stalk could be identified extending from the optic disc and the back of the lens (c, d). A posterior capsular defect (arrows) is evident in the right (e) and left eye (f). Following cataract removal, the retina is observed to be attached and optic discs are normal in both eyes (g, h)

hypothesize that ectopic congenital cataracts with PCD are on the milder end of the PFV spectrum, caused by abnormal regression of the fetal hyaloid system. More evidence is necessary to confirm this pathogenesis. As congenital cataracts may present with complex morphological variations, a meticulous assessment should be made preoperatively, and surgeons should be prepared for a possible vitreoretinal surgery.

Ethics

Informed Consent: Obtained.

Peer-review: Externally peer reviewed.

Authorship Contributions

Concept: E.Ö.Z., A.B.T., H.T.A., Ş.Ö., Design: E.Ö.Z., A.B.T., H.T.A., Ş.Ö., Data Collection or Processing: E.Ö.Z.,

A.B.T., H.T.A., Ş.Ö., Analysis or Interpretation: E.Ö.Z., A.B.T., H.T.A., Ş.Ö., Literature Search: E.Ö.Z., A.B.T., H.T.A., Ş.Ö., Writing: E.Ö.Z., A.B.T., H.T.A., Ş.Ö.

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