Persistent Fetal Vasculature

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Persistent fetal vasculature (PFV) is a congenital anomaly of the eye occurring from failure of the embryological, primary vitreous, and hyaloid vasculature to regress. The previous term persistent hyperplastic primary vitreous (PHPV) [1] has been replaced by PFV, coined by Goldberg [2]. Presentation in most cases is unilateral and varies from primarily anterior, posterior, or mixed variety.

4.1 Clinical Presentation

Anterior presentation typically involves the presence of persistent pupillary membrane (PPM), Mittendorf dot, cataract, vessels over lens, enlarged ciliary processes, glaucoma, and/or retrolental membranes. Posterior presentation may contain Bergmeister papilla, stalk of PFV, falciform fold, and/or retinal detachment. Mixed variety, which contains the combination of the two, is the commonest variant (Fig. 4.1).

Persistent pupillary membrane (PPM): It is a mild variant of PFV in which anterior tunica vasculosa lentis fails to regress leaving stands of iris or regressed vessels attached to lens. In most cases, it is visually insignificant but in rare cases may be associated with posterior stalk especially when associated with cataract or retrolental membrane. Sometimes, PPM may be severe and may cause lenticular myopia (Fig. 4.2). Such cases may require surgery.

Mittendorf dot: It is seen in 0.7–2.0% of the population [3]. Typically, it is small white dot on the posterior capsule on the nasal side representing incomplete regression of the hyaloid artery from its point of attachment (Fig. 4.3).

Enlarged ciliary processes: They may be seen in association with cataract or retrolental membrane. The contraction of membrane may be responsible for elongation of the ciliary processes, which may become visible as the pupil is dilated (Fig. 4.4).

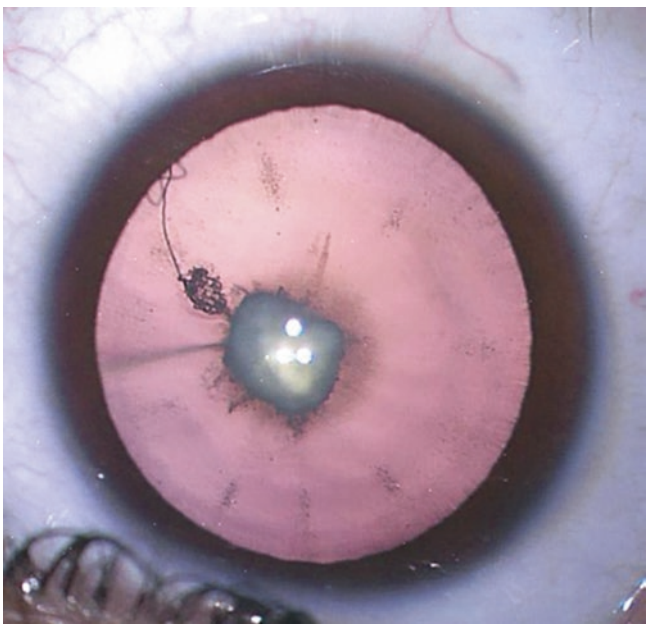

Fig. 4.1 Mixed variety of PFV with PPM, central cataract and stalk of PFV seen on the left

Fibrovascular membrane: The lens may be cataractous or partially absorbed or rarely completely absorbed. Sometimes only a part of lens is involved (Fig. 4.5). Usually, it is associated with small vessels over the fibrosed lens which may or may not bleed during surgery. They constitute a vascular connection between the posterior and the anterior tunica vasculosa lentis (Fig. 4.6).

Salmon patch sign: The lens may be associated with posterior capsular plaque where the vessels seem to have regressed. In some cases, there may be pink hue seen from the plaque which may predict the presence of PFV [4] (Fig. 4.7a–d).

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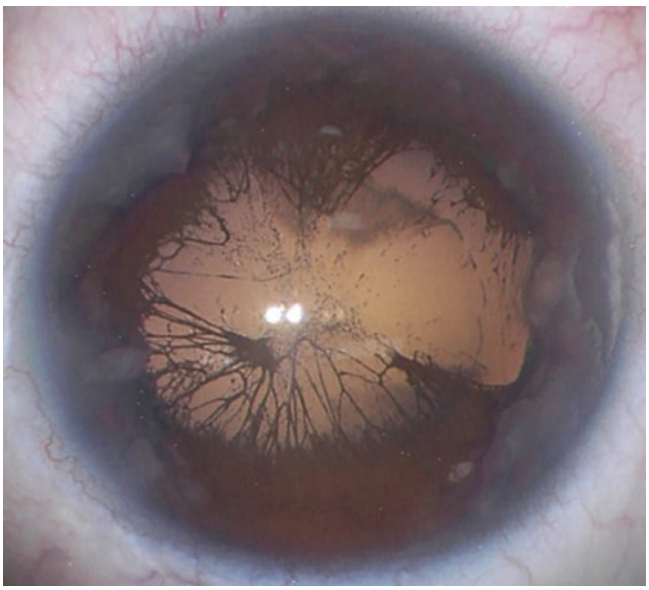

Fig. 4.2 Extensive PPM seen in association with minimal lamellar cataract

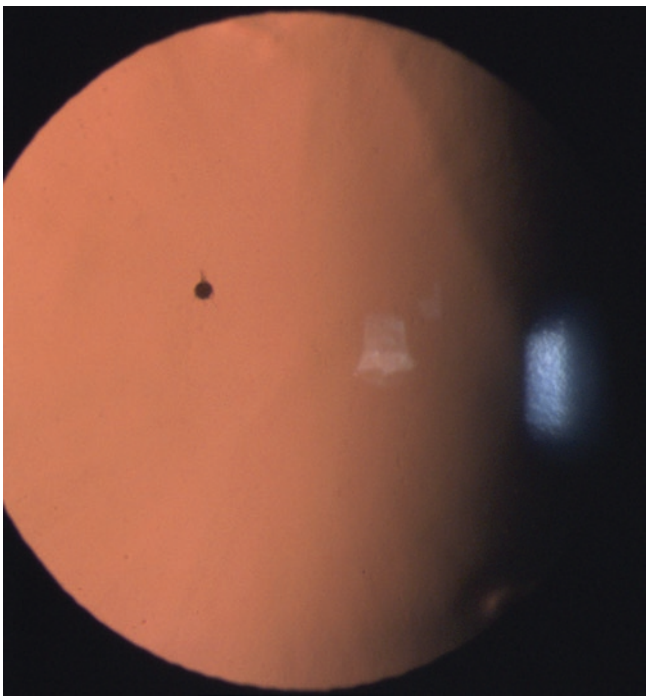

Fig. 4.3 Mittendorf dot seen in the left eye (nasal to the center)

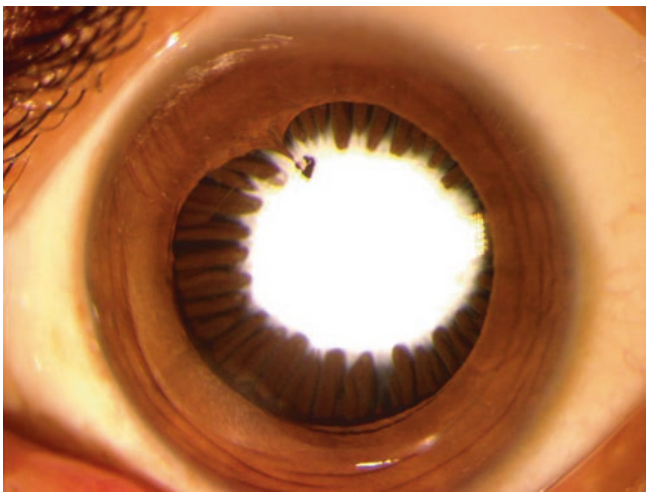

Fig. 4.4 Anterior variety of PFV with enlarged ciliary processes

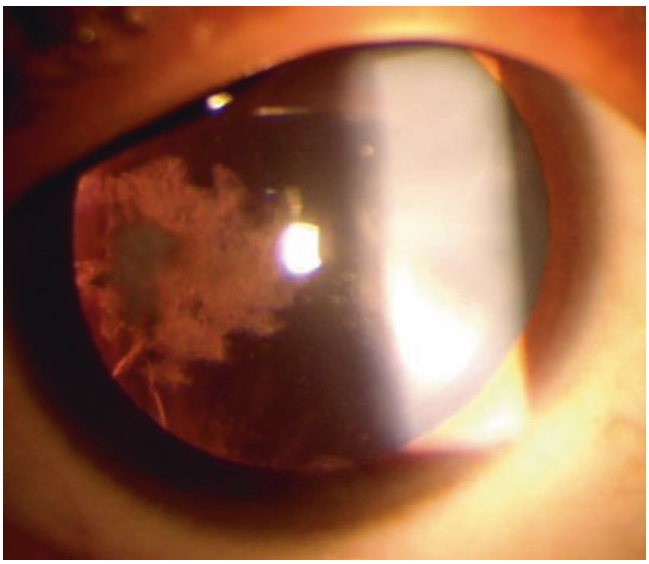

Fig. 4.5 Posterior subcapsular cataract with plaque with faint vessels seen

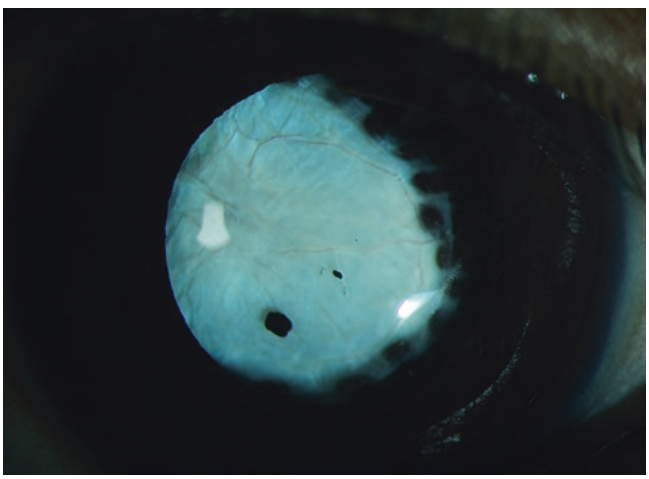

Fig. 4.6 Anterior PFV with fibrovascular membrane with visible multiple vessels crossing pupil and enlarged ciliary processes

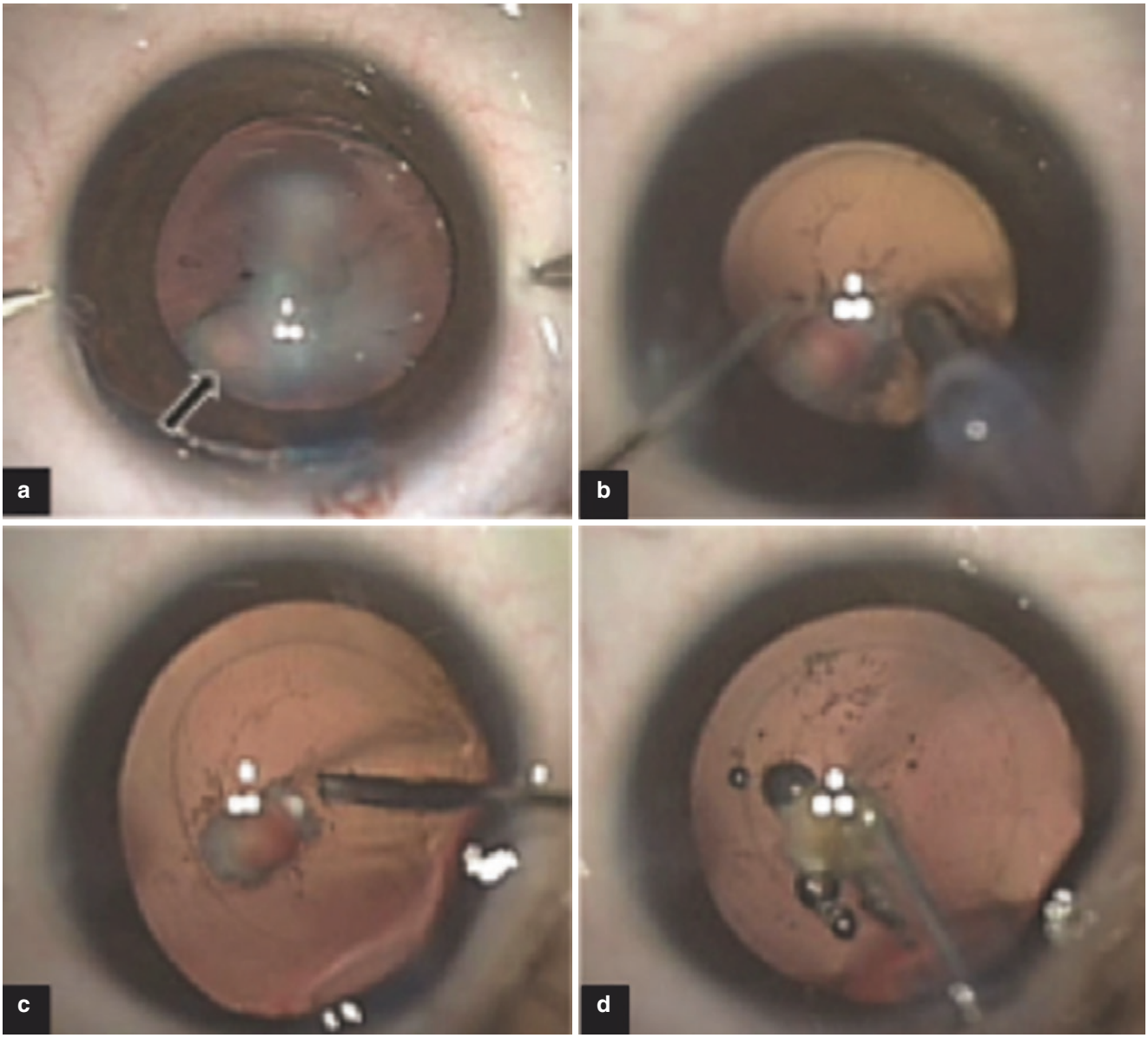

Fig. 4.7 (**a**) A congenital cataract showing the presence of an eccentric salmon pink sign intraoperatively (arrow). (**b**) Irrigation aspiration of the lens matter further enhances the pinkish hue from the retrocapsular plaque. (**c**) Posterior capsulorhexis is being performed with a bent 26 gauge needle. (**d**) The vascularized mass is coagulated with Fugo plasma blade PC. Source S Khokhar et al. [4]

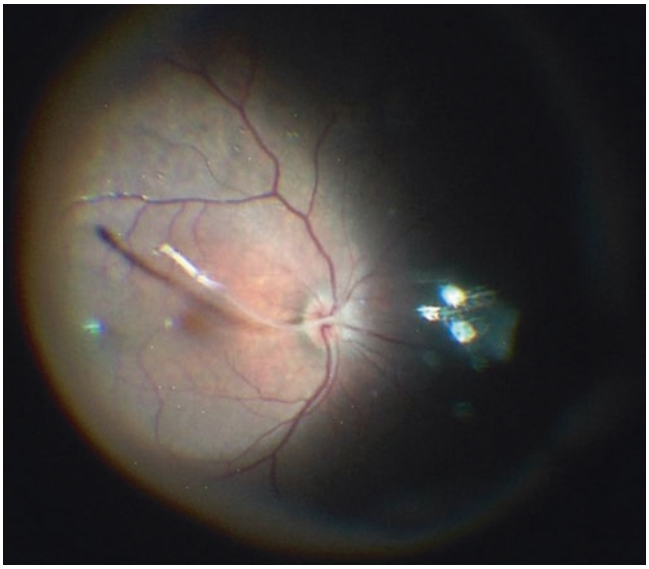

Fig. 4.8 Stalk of hyaloid artery remnant seen intraoperatively using wide angle viewing system in a case of posterior PFV

Persistent hyaloid artery: The fetal hyaloid artery lies within the Cloquet canal and regresses around the seventh month of gestation. If it persists, a stalk may be seen arising from the disc to behind the lens (Fig. 4.8).

Bergmeister papilla: Refers to remnant of hyaloid artery to disc and can be seen as fibrovascular tuft at disc. It may be associated with other disc or macular abnormalities.

4.2 Tools for Diagnosis

Despite the clinical manifestations, diagnosing PFV may sometimes be challenging. Any child with a cataract, especially unilateral, should be suspected of having PFV. The differential diagnosis includes diseases causing leukocoria, including retinoblastoma, Norrie's disease, ocular toxocariasis, retinal dysplasia, incontinentia pigmenti, uveitis, congenital cataract, coat's disease, retinopathy of prematurity, familial exudative vitreoretinopathy or endophthalmitis.

4.2.1 Ultrasonography (USG)

It is noninvasive, inexpensive tool of great use in cases of identifying PFV as well as ruling out other causes of leukocoria. Both USG and ultrasound biomicroscopy (UBM) of the anterior segment are valuable [5, 6]. USG typically shows a small globe and a stalk in vitreous extending from the posterior lens capsule to the disc area [6]. It can also reveal whether a retinal detachment is present or not. UBM may show swollen lens with a resultant shallow anterior chamber or partially absorbed lens with enlarged ciliary processes and thickened anterior vitreous face [5]. Although effective, sensitivity ranges from 70% to 80% (Fig. 4.9).

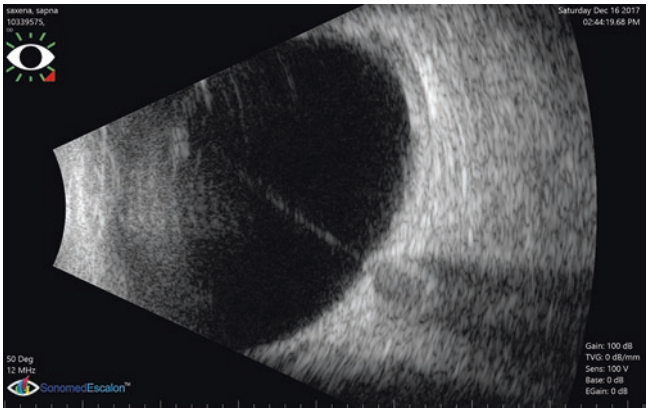

Fig. 4.9 USG picture showing hyperechoic stalk seen attached from disc to posterior surface of lens

4.2.2 Color Doppler Imaging

It detects blood flow within the stalk. It can also differentiate between arterial or venous flow (Fig. 4.10).

4.2.3 Magnetic Resonance Imaging (MRI)

MRI as well as computerized tomography (CT scan) have also been reported as excellent tool in the diagnosis of PFV [7, 8]. It helps in ruling out more severe differential diagnosis such as retinoblastoma. Sensitivity is almost 100% (Fig. 4.11).

4.2.4 Histopathology

Histopathology and electron microscopy can be used in confirming the diagnosis. It can be performed on enucleated eyes or tissue retrieved from PFV structures intraoperatively. Blood vessels associated with loose connective tissue can be identified in remnants of PFV (Fig. 4.12).

4.3 Management

Surgical is indicated in patients with visually significant cataract or fibrovascular membrane. Previously, surgery was indicated only to avoid complications and visual

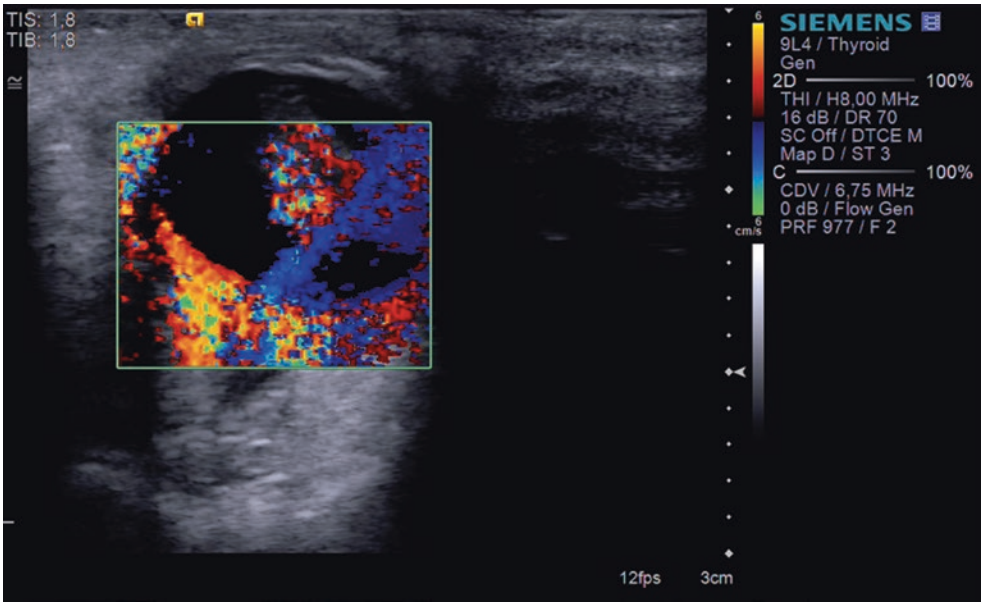

Fig. 4.10 Color Doppler of a 1-year-old child suggestive of arterial blood flow in the stalk

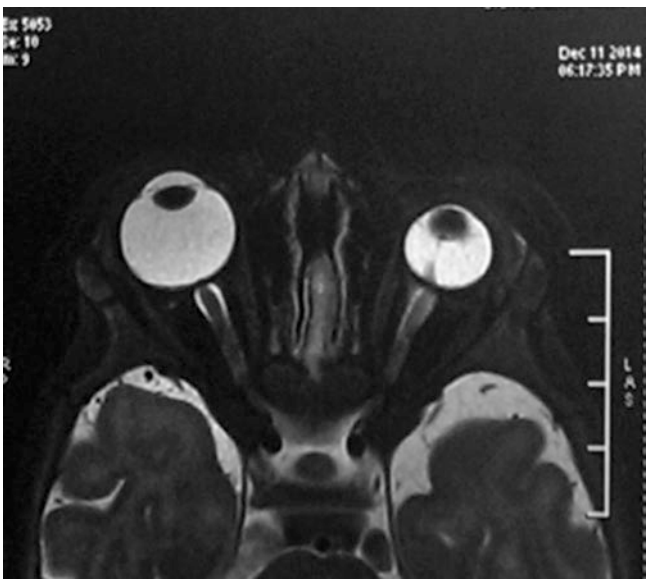

Fig. 4.11 T-2 W image of MRI of a 3-month-old baby showing left eye smaller in size with hypointense stalk attached from disc to lens confirm PFV

prognosis was very guarded [1, 9–11]. Various advances have come since then and successful management of cataract with posterior stalk has been possible [12–15]. Surgical approach can be anterior or posterior. The common steps like any congenital cataract include lensectomy along with anterior vitrectomy with management of the vascular structures. Hemostasis can be achieved by raised IOP, diathermy of vessels (Fig. 4.13), or using plasma knife [16]. The plasma knife (Fugo blade) is a radiofrequency electrosurgical incising instrument that uses electromagnetic energy to perform cutting and provides non-cauterizing hemostasis called "autostasis" [17]. The advantage of anterior approach is that IOL can be placed at the time of surgery.

Figure 4.14a–d shows the steps of surgery where after lensectomy, vascular stalk is visible. The posterior capsular plaque with attached stalk is cut using plasma knife (Fugo blade) avoiding inadvertent bleed. Afterward, the stalk is cut with microincision forceps. We can see the intact rim of sulcus for placement of IOL.

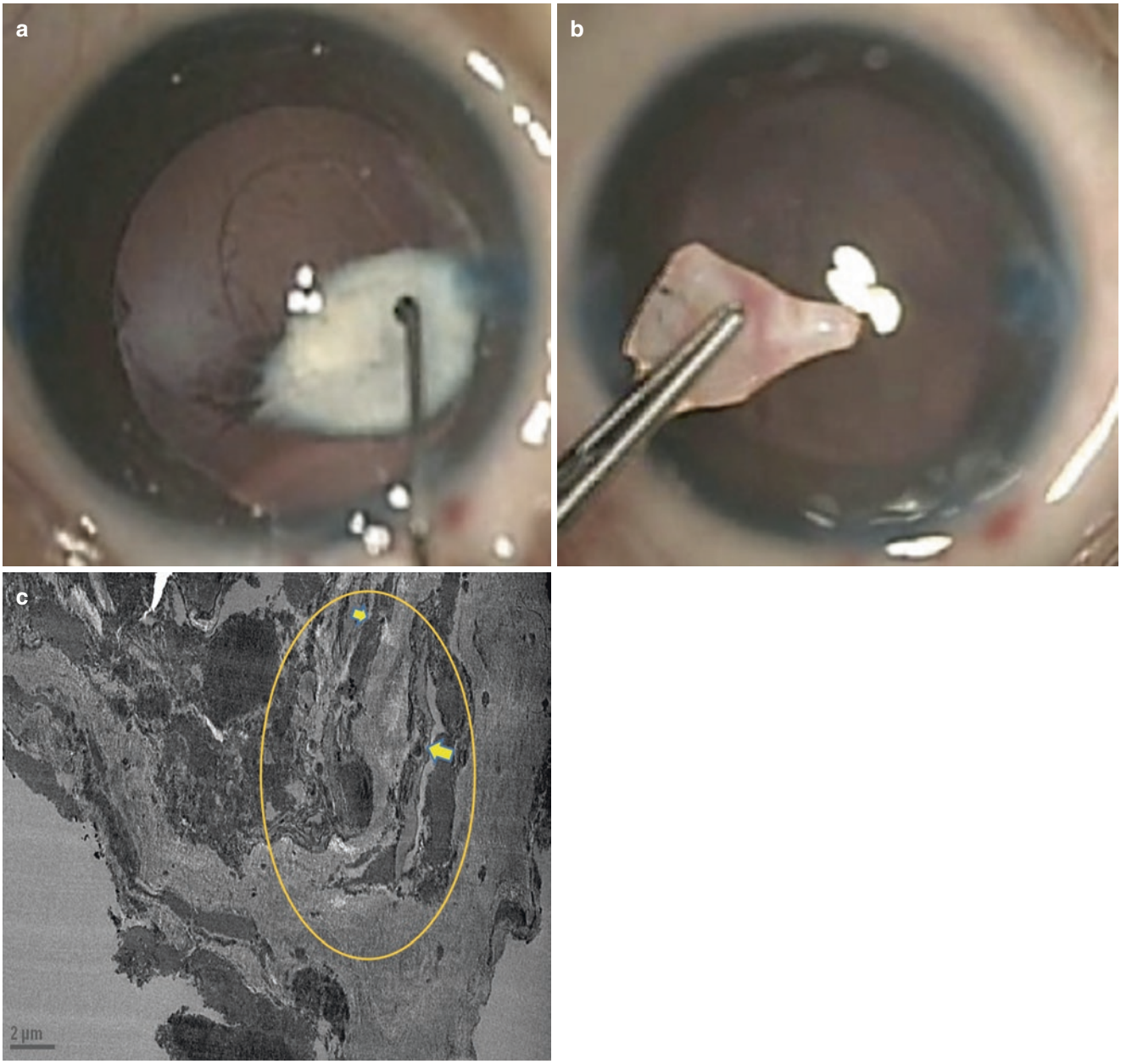

Fig. 4.12 (**a**) Intraoperative image of mixed variant of PFV after lens aspiration showing posterior capsular plaque with attached stalk. (**b**) Posterior plaque along with remnant of PFV removed. (**c**) Electron microscopy of the same showing loose elastic tissue (circle) with vessels (arrows)

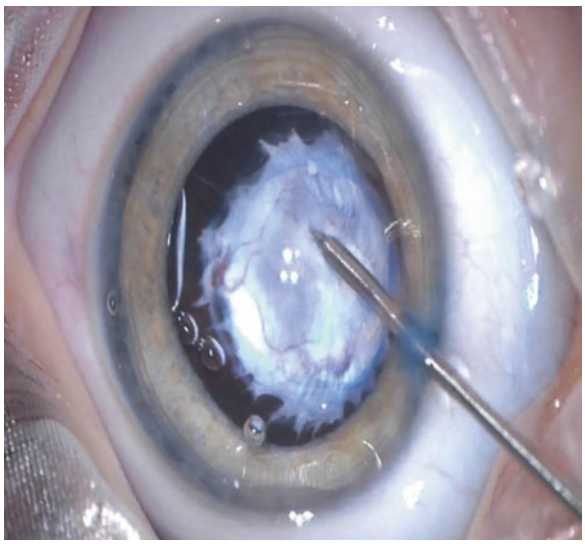

Fig. 4.13 Intraoperative image of anterior PFV where vessels are being cauterized using diathermy

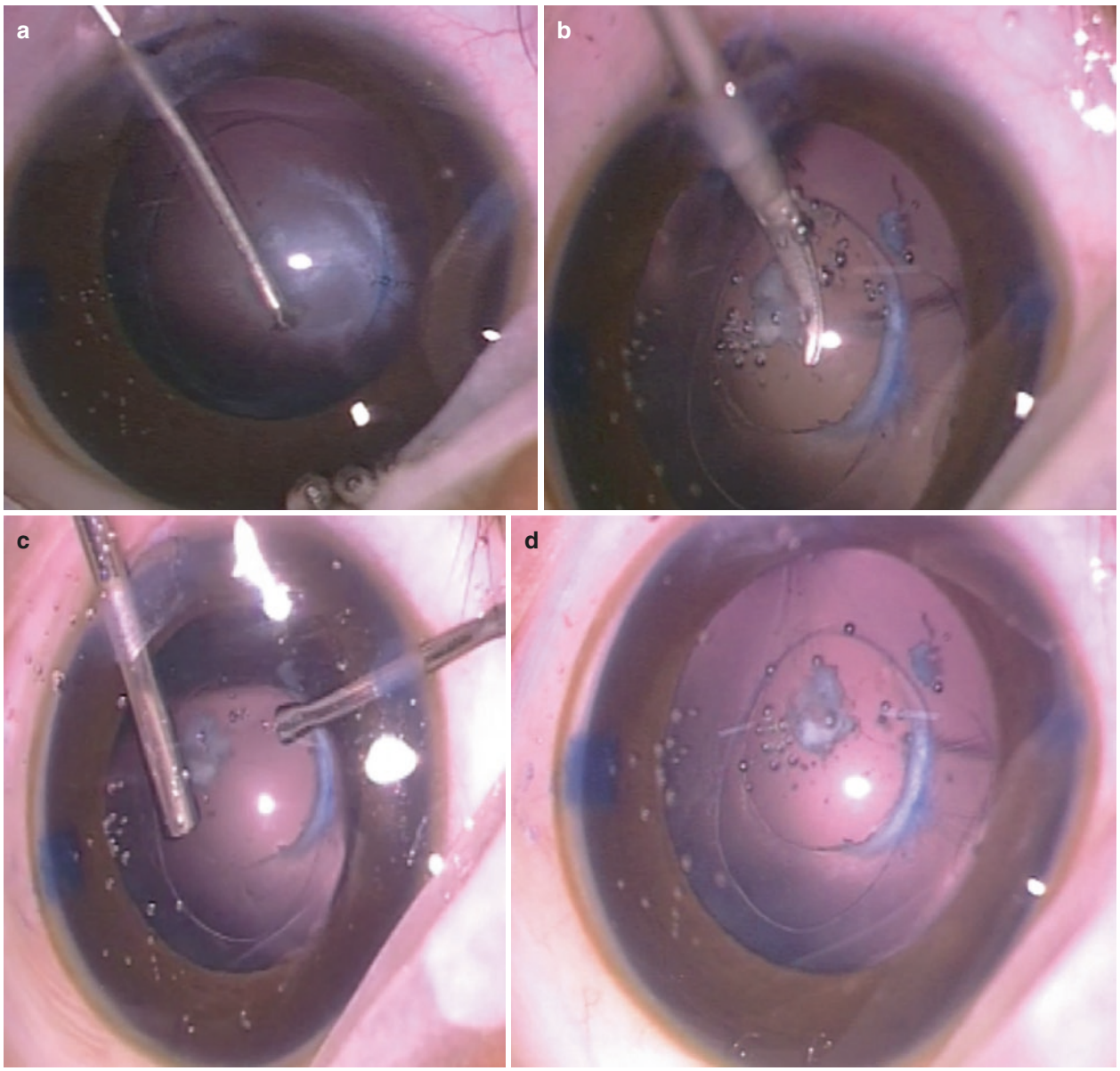

Fig. 4.14 (**a**) Fugo blade is used to cut the posterior capsule along with PFV stalk. (**b**) After hemostasis, stalk is cut with microincision forceps. (**c**) Stalk seen falling back. (**d**) After anterior vitrectomy, rim of sulcus is intact, IOL can be placed

4.4 Prognosis

Favorable outcomes may be achieved in children with PFV, irrespective of surgical approach by early intervention followed by aggressive amblyopic treatment [12–15, 18]. Still outcomes remain inferior compared to other children with unilateral cataract without PFV. It may be due to higher percentage of complications including glaucoma, visual axis opacification, vitreous hemorrhage, and retinal detachment [12, 13, 19, 20].

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