Persistent Fetal Vasculature

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11.1 Introduction

Persistent Fetal Vasculature (PFV) is a congenital anomaly of the eye resulting from failure of the embryological, primary vitreous, and hyaloid vasculature to regress [1, 2]. Cloquet reported it for the first time as early as 1818 [3]. It was named persistent hyperplastic primary vitreous (PHPV) based on the embryology and origin and described in detail by Reese in 1955 [1]. This term was used for a long time till Goldberg introduced the term persistent fetal vasculature or PFV combining the anterior, posterior, and mixed presentation to the terminology [3]. This is a more inclusive term and is currently the most accepted terminology for the condition.

PFV may be present as congenital cataract or leukocoria. It may involve some or all the component of hyaloid and fetal vasculature. Based on the structures involved, it can be classified as anterior, posterior, or combined variety. The surgical management and prognosis depend on the structures involved by this anomaly. It is usually unilateral but may be bilateral in rare cases [4].

11.2 Pathogenesis

During intrauterine life, the fetal vasculature is important for development of iris, lens, vitreous and retina. The vascular system comprises of three parts: vasa hyaloidea propria (VHP) or hyaloid vessels in vitreous, tunica vasculosa lentis (TVL), or hyaloid vessels covering posterior surface of lens and anterior pupillary vessels covering anterior surface of the lens which anastomose with TVL. These vessels start to grow in first month of gestation and begin to involute by 5 months of gestation via the process of apoptosis [5, 6]. These vessels usually disappear completely by birth leaving behind an acellular hyaloid canal called Cloquet's canal [2, 3, 5]. Any abnormality in the involutional process can result in partial or complete failure of vascular regression. This causes different clinical presentations ranging from mild to severe; anterior, posterior or combined variety.

Cause for the abnormality in regression of fetal vasculature is not completely understood [7]. Most cases are sporadic in nature [1, 2, 7]. Some have been reported to be associated with genetic abnormalities, but no specific locus has been identified in humans [8]. In animal models, gene mutations have been documented in bilateral

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PFV [9, 10]. In the current literature, various animal models have been used to study the role of angiogenic and antiangiogenic factors [6, 11–13]. Vascular endothelial growth factor (VEGF) and placental growth factor (PGF) have been reported to have a regulatory role in involution of hyaloid vessels [13]. An association of PFV has been reported in p53-deficient and Bax/Bak proapoptotic Bcl-2-deficient mice [14]. This suggests a disturbance in proapoptotic, and antiapoptotic factors may have an influence in pathogenesis of PFV.

11.3 Clinical Presentation

PFV presents with a wide spectrum of presentation [15, 16] (Table 11.1). Anterior presentation may involve presence of pupillary membrane, 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. Combined variety, which contains the combination of the two, is the commonest presentation (Fig. 11.1).

Following are the important clinical presentation of PFV:

11.3.1 Anterior Presentation

11.3.1.1 Persistent Pupillary Membrane (PPM)

PPM is considered a mild presentation of anterior PFV where anterior TVL fails to regress. Most cases have a small PPM with no visual implications. In some cases, it may be vascularized and associated with other abnormalities such as anterior pyramidal cataract [17] or capsular plaque,

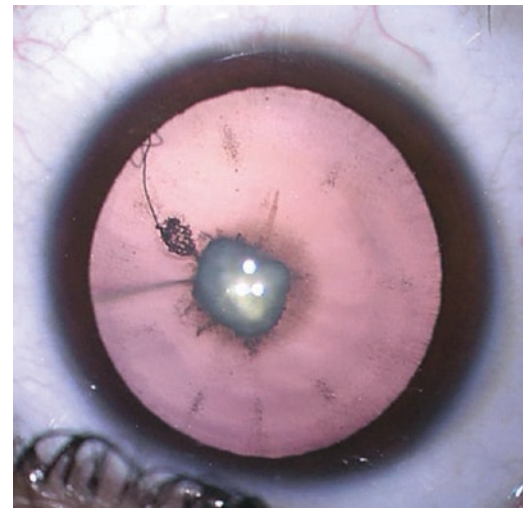

Fig. 11.1 Mixed variety of PFV with PPM, central cataract and stalk of PFV seen on the left

Table 11.1 Classification of PFV on the basis of clinical presentation [15, 16]

	Anterior PFV	Posterior PFV	Combined PFV
Clinical presentation	• Microphthalmia • Leucocoria • Shallow anterior chamber • Persistent pupillary membrane • Cataract • Elongated ciliary processes • Retrolental fibrovascular membrane • Intralenticular hemorrhage • Chronic inflammation • Glaucoma • Strabismus • Ectropion uvea • Coloboma Iridis	• Microphthalmia • Leucocoria • Mittendorf Dot • Vitreous membranes and stalk • Bergmeister papilla • Retinal fold • Tractional retinal detachment of posterior pole • Hypoplastic optic nerve • Dysplastic optic nerve • Pigment maculopathy • Hypoplastic macula • Clear lens • Strabismus	Clinical features from both anterior and posterior PFV

aniridia [17] or posterior stalk of PFV. If cataract is visually significant, surgery may be required.

In severe cases of PFV (Fig. 11.2), visual acuity may be affected. It can cause changes in the capsular curvature and lead to lenticular myopia. If visual acuity is significantly affected, surgery may be required. Lim et al. [18] reported surgical intervention in five eyes with PPM. They recommended the use of vitrectomy scissors instead of Vannas scissors for removal of PPM. After 22.6 months of follow up, no complication was reported, and visual acuity improved in 60% cases [18].

11.3.1.2 Cataract

PFV is considered one of the most important cause for unilateral cataract across the world, although it may cause bilateral cataract as well. The incidence of PFV in unilateral cataract varies. We detected PFV in 27.6% cases of unilateral cataract and it was the most common identifiable cause [19]. *Unilateral cataract* associated with ***microphthalmos*** should raise suspicion of PFV [20].

Morphology of cataract may vary from posterior capsular plaque, partially absorbed cataract or total cataract. Posterior capsular plaques have been hypothesized to develop from regressing hyaloid vasculature which may or may not be associated with stalk of PFV [21]. Posterior capsular plaque may be the remnant of the strong adhesions formed by capsule and hyaloid vessels which regress later [21]. Posterior lenticonus has also been hypothesized to have causal association with PFV [22].

Associated features of PFV should be looked for in all cases of unilateral cataract. Significant cataract associated with combined or anterior PFV requires surgical intervention [23]. Since surgical intervention is more difficult and requires additional instrumentation, timely clinical diagnosis is important.

11.3.1.3 Enlarged Ciliary Processes

Prominent and elongated ciliary processes may be associated with cataractous lens or retrolental membrane. They may result from traction caused by contraction of the fibrovascular membrane. Prominent ciliary processes can be visualized following pupillary dilatation. They are considered a telltale sign of PFV (Fig. 11.3).

11.3.2 Posterior Presentation

11.3.2.1 Mittendorf Dot

This is also a mild presentation of PFV where there is near completer regression hyaloid artery, only a small white dot or cone is left on posterior capsule or just behind it. It is generally slightly nasal to the visual axis and is visually insignificant. It may be seen in 0.7–2% of the normal population [5, 23].

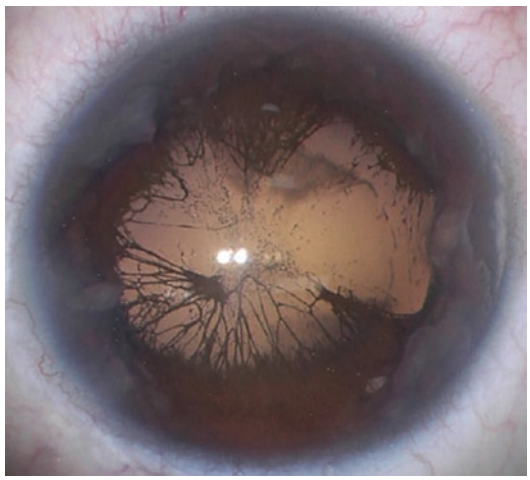

Fig. 11.2 PPM seen in association with minimal lamellar cataract

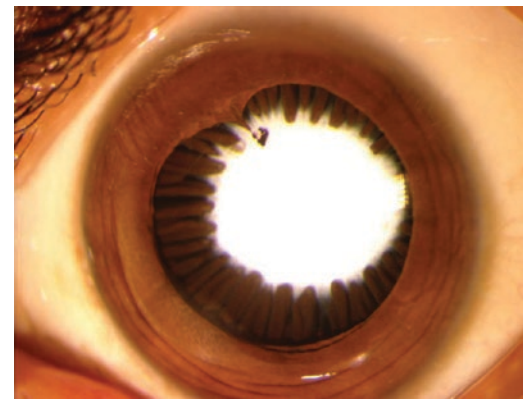

Fig. 11.3 Anterior variety of PFV with enlarged ciliary processes

11.3.2.2 Fibrovascular Membrane

Retrolental fibrovascular membrane has been described in literature as an important sign of PFV by Reese in 1955 [1]. It is considered the remnant of posterior TVL and may vary from small thin membrane to complete membrane covering posterior surface of the lens. The lens may be clear and this membrane may be the cause of leukocoria in children. This is not pathognomic of PFV and may be seen in retinopathy of prematurity or retinal detachment [24].

11.3.2.3 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. 11.4).

11.3.2.4 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.

11.3.2.5 Tractional Detachment

PFV may be associated with congenital tent shaped detachment of retina where it may adhere to lens or ciliary body. If the detachment is severe, visual prognosis is poor.

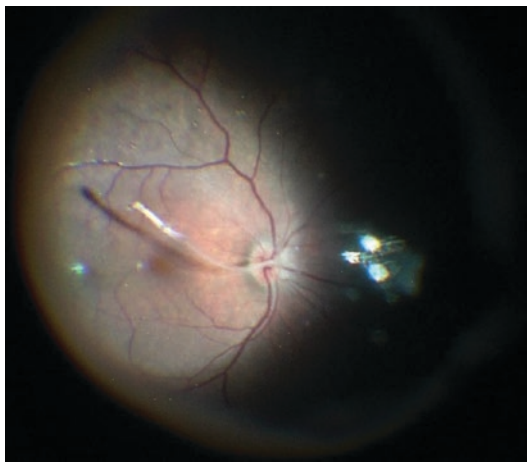

Fig. 11.4 Stalk of hyaloid artery remnant seen intraoperatively using wide angle viewing system

11.3.3 Associations

PFV may be associated with retinal folds, optic disc hypoplasia, macular hypoplasia, anterior segment dysgenesis, lens subluxation, ocular coloboma, etc. [25–30] These are relatively rare as compared to the other features of PFV. Systemic association include cleft palate and lip, polydactyly, and microcephaly [31]; and trisomy 13 [2].

11.3.4 Complications

PFV is associated with arrest in the normal of growth of eye ball and is usually associated with microphthalmia [15, 16]. Secondary glaucoma is a common sequalae to untreated PFV. This may be associated vitreous hemorrhage, hyphema, corneal edema and/or buphthalmos [32, 33]. These complications can occur suddenly and may result in eventual phthisis bulbi or painful blind eye. In the past, even in eyes with very poor prognosis, surgery has been recommended to avoid these complications [34].

11.4 Investigations

Diagnosis of PFV solely bases on clinical findings, may be challenging. A high suspicion of PFV should be kept in all cases with unilateral cataract. A differential diagnosis of leukocoria should be kept in mind when differentiating PFV from other entities (Table 11.2).

The following investigations are useful in confirming the diagnosis of PFV:

11.4.1 Ultrasonography (USG)

Ultrasonography needs to be performed in all cases of leukocoria where fundus is not visible. It is a noninvasive and inexpensive tool which can be useful in diagnosis of PFV. USG has been useful in diagnosis PFV and ruling out mass lesions such as retinoblastoma [35, 36]. Depending on the presentation, USG may show a thin stalk of remnant hyaloid vessel in the vitreous cavity between the disc and posterior lens capsule

Table 11.2 Differential diagnosis of leukocoria [15, 20, 23]

- Retinoblastoma
- Coat's disease
- Familial exudative vitreoretinopathy
- Retinopathy of prematurity
- Norrie's disease
- Ocular toxocariasis
- Retinal dysplasia
- Incontinentia pigmenti
- Uveitis
- Congenital cataract
- Fundal coloboma
- Juvenile xanthogranuloma
- Myelinated nerve fibers
- Endopthalmitis

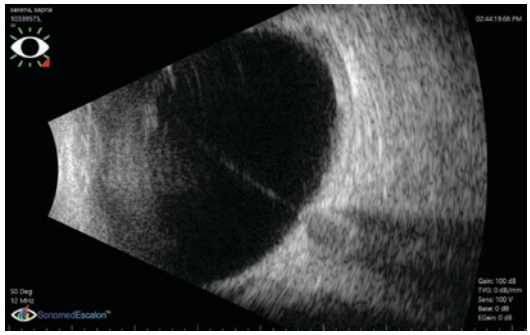

Fig. 11.5 USG picture showing hyperechoic stalk seen attached from disc to posterior surface of lens

(Fig. 11.5). USG may reveal presence or absence of retinal detachment. Although effective, sensitivity ranges from 70 to 80% [20].

11.4.2 Ultrasound Biomicroscopy (UBM)

UBM is particularly useful in cases of anterior presentation or combined presentation. Lens may be swollen or partially absorbed with prominent and enlarged ciliary processes (Fig. 11.6). In some cases, a stalk attached to posterior capsule may also be visible.

11.4.3 Color Doppler Imaging

It can detect a stalk of PFV based on the flow [37]. It can also differentiate between arterial or

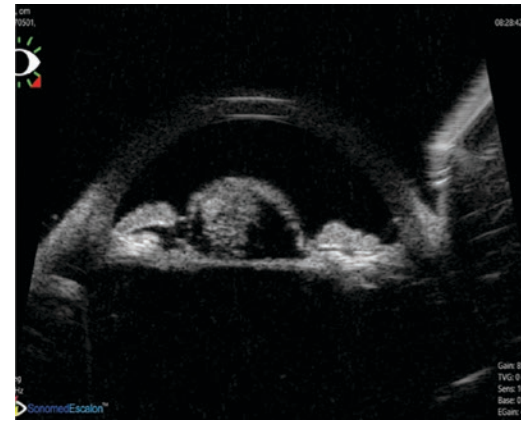

Fig. 11.6 UBM showing swollen anterior lens with posterior capsular plaque and enlarged and centrally displaced ciliary processes

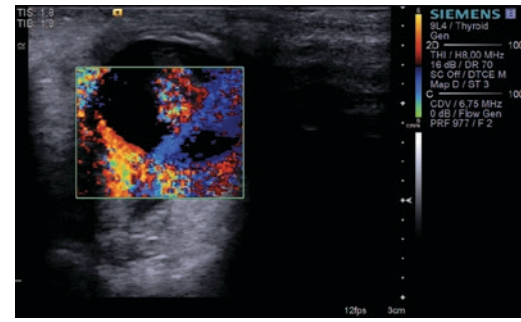

Fig. 11.7 Color Doppler of a 1 year old child suggestive of arterial blood flow in the stalk

venous flow (Fig. 11.7). It is not useful in avascular cases or cases with no flow.

11.4.4 Magnetic Resonance Imaging (MRI)

MRI is considered highly sensitive and specific for diagnosis for PFV [38–40]. It is more expensive and may require anesthesia or sedation for the child to be performed accurately. Sensitivity is almost 100% (Fig. 11.8).

Computerized Tomography (CT scan) have also been reported as diagnostic tool [39]. It helps in ruling out more severe differential diagnosis such as retinoblastoma.

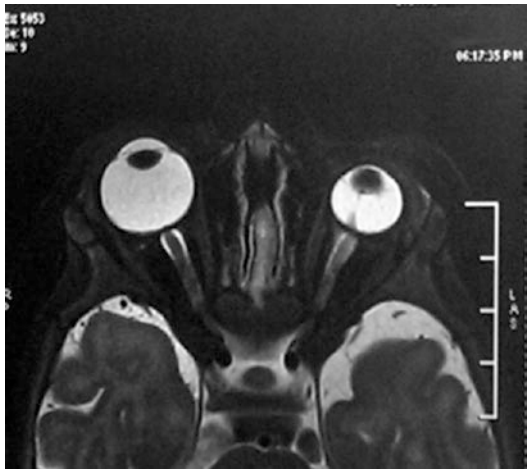

Fig. 11.8 T-2W image of MRI of a 3 month old baby showing left eye smaller in size with hypointense stalk attached from disc to lens confirm PFV

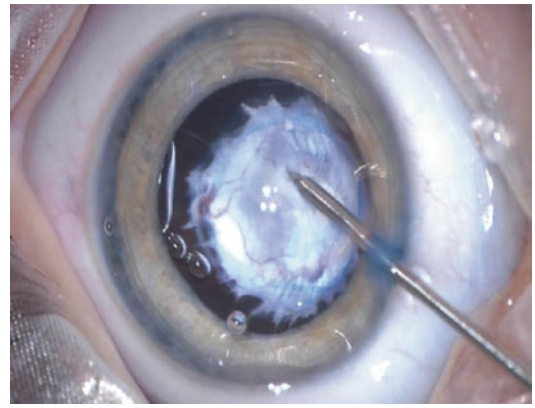

Fig. 11.9 Intraoperative image of anterior PFV where vessels are being cauterized using diathermy

11.5 Management

Historically, surgery was indicated to primarily avoid severe complications such as secondary glaucoma, buphthalmias, painful blind eye or phthisis bulbi with guarded to nil visual prognosis [1, 2, 31, 41, 42]. With time there have been advances in the surgical technique which has improved the anatomical and functional outcomes to a certain extent [43–46]. Surgery can be performed via anterior or posterior approach. Each has its merits and demerits.

11.5.1 Anterior Approach

Anterior approach has been commonly utilized by congenital cataract surgeon for management of PFV [31, 43]. It has certain advantages over posterior approach. IOL implantation can be performed with this method in the same surgery allowing better visual rehabilitation and cosmetic results [43]. This technique provides better visualization of small bleeders on the posterior capsule which can be tackled better. ***Salmon patch sign*** has been described to identify PFV in cases of posterior capsular plaque [47]. Intraoperatively, appearance pink hue on the plaque suggests leaking vessels, which can help in diagnosis of a previously missed PFV.

The principle of PFV surgery, in addition to providing clear media, is achieving *hemostasis*. In addition to the usual steps performed for congenital cataract surgery including lensectomy with anterior vitrectomy, additional steps are required to avoid intraoperative bleeding.

After making 2 MVR entry, 180° apart, ACCC and lens aspiration are performed as usual in cases with posterior capsular plaque. In absorbed cataract, there may be a vascularized membrane. Hemostasis is achieved by a variety of maneuvers before cutting the plaque or vascularized membrane. Heavy viscosity OVD is used and IOP is maintained/increased throughout surgery.

Additional instruments may be required to manage vascular component. **Diathermy** is an easily available instrument which can be used to coagulate the blood vessels (Fig. 11.9). Following this, a microincision scissors and forceps can be used for removal of plaque. Caution is required while cutting plaque as stalk of PFV may be present underneath. If stalk is present, it needs to be cauterized before cutting the anterior end. Following hemostasis, stalk is allowed to fall back and anterior vitrectomy can be performed.

Plasma knife or Fugo blade is an ingenious alternate to diathermy [48]. 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" [48, 49]. The advantage of this technique is no heat production and minimal damage to adjacent tissue. Figure 11.10a–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 finally we are left with intact rim of sulcus for placement of IOL.

11.5.2 Posterior Approach

It is commonly employed by retinal surgeons. The advantage of this technique is if associated vitreous hemorrhage, vitreous membranes, retinal folds or retinal detachment is present, it can

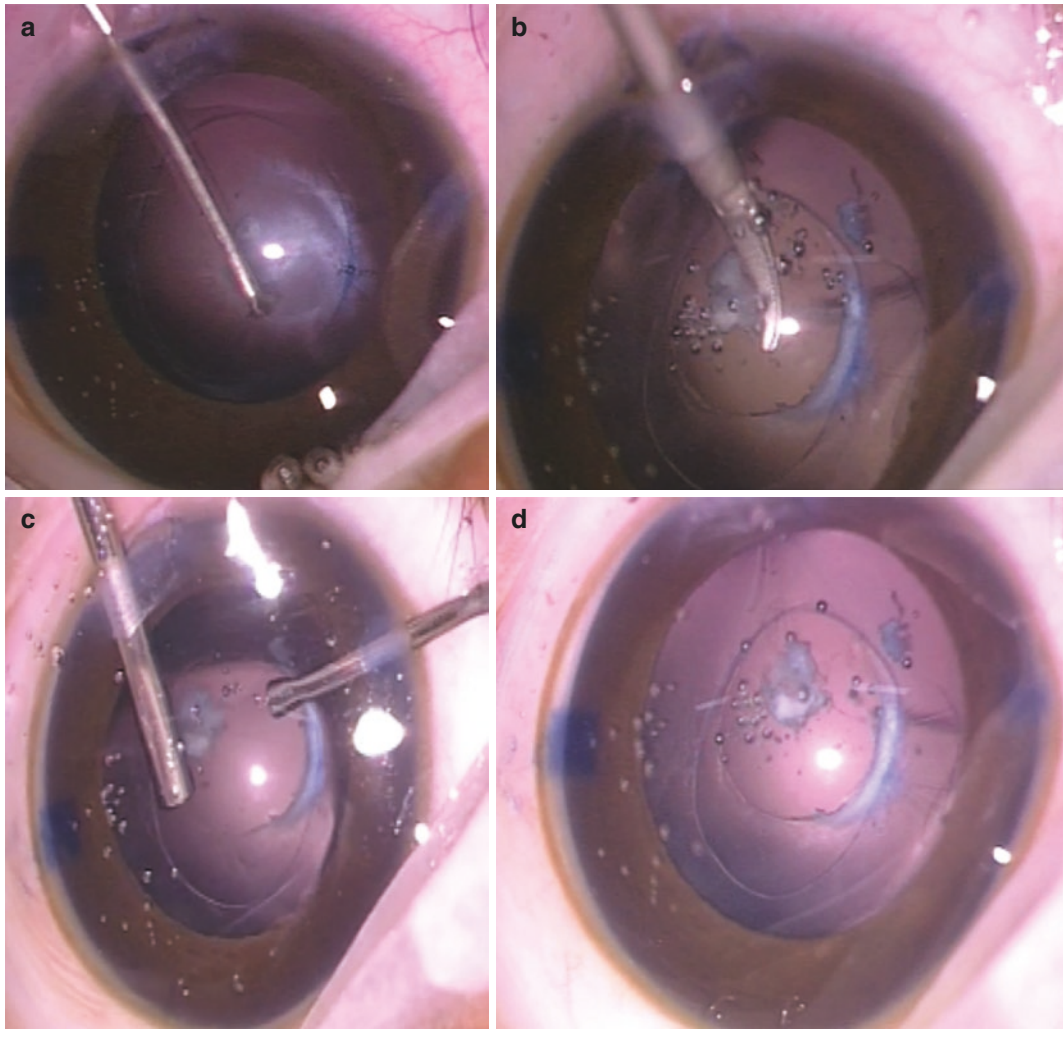

Fig. 11.10 (**a**) Fugo blade is used to cut the posterior capsule along with PFV stalk. (**b**) After hemostasis, stalk is cut with microincision forceps. (**c**) Stalk seen falling back. (**d**) After anterior vitrectomy, rim of sulcus is intact, IOL can be placed

be managed simultaneously. The disadvantage includes reduced feasibility for IOL implantation and requirement of vitreoretinal setup.

Two pars plana ports are made and lensectomy is performed starting from a stab in the posterior capsule. Following lensectomy, opening can be made of adequate size in anterior and posterior capsule. If bleeders are encountered, diathermy can be performed [50]. If thick plaque or membranes are encountered, intravitreal scissors can be used to cut the membranes [50]. A third port for illumination can be placed for associated retinal pathology. Prognosis is usually poor in cases of extensive retinal abnormalities.

11.6 Prognosis

Favorable outcomes may be achieved in children with PFV by early intervention followed by aggressive amblyopic treatment [43–46, 51]. Hunt et al. [34] found visual acuity of 6/60 or better in 18% of eyes with PFV in a long-term follow-up. Early surgery before 77 day was identified as good prognostic marker. Anteby et al. [43] found visual acuity of 20/200 (6/60) or better in 25% of the eyes. They implanted IOL in 30 eyes for better visual rehabilitation and found visual acuity of 20/50 or better in 20% and 20/200 or better in 33.3% eyes. Sisk et al. [52] found posterior manifestations of PFV, bilaterality, and microphthalmos were associated with poorer visual outcomes. They also compared the outcomes of limbal and pars plana approach and found similar visual and anatomical outcomes.

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 [43, 44, 53, 54].

11.7 Conclusion

PFV is a congenital anomaly caused by partial or complete failure of fetal vasculature to regress. It commonly presents as unilateral cataract. It has anterior, posterior or combined presentation. Diagnosis using clinical findings and USG (MRI if needed and available) is important as surgical approach is different than normal congenital cataract. Vascular hemostasis is required throughout surgery, diathermy, or plasma blade can be used for the same. Despite best efforts, visual outcomes remain lower than most pediatric cataracts. Nevertheless, a good surgical technique followed by refractive correction and amblyopia therapy provides hope for useful visual and cosmetic outcome.

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