



# Pediatric Surgical Outcomes of Persistent Fetal Vasculature: A Five-Year Retrospective Study at a Tertiary Eye Center in Kazakhstan

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## Abstract:

**Objective:** To present the surgical outcomes of Persistent Fetal Vasculature (PFV) in a pediatric cohort over five years at the main referral tertiary eye care center in Kazakhstan.

**Methods:** A retrospective analysis was performed on the medical records of 23 pediatric patients (24 eyes) under 18 years old who were treated for PFV at the Kazakh Eye Research Institute between 2017 and 2021. Clinical presentations of PFV, types of surgery performed, and postoperative functional outcomes were analyzed.

**Results:** The average age at the time of surgery was  $4.49 \pm 4.24$  years, ranging from 5 months to 16 years, with 39.1% boys and 60.9% girls. The anterior, posterior, and mixed forms of PFV were observed in 12.5%, 4.2%, and 83.3% of cases, respectively. PFV was most commonly accompanied by cataracts (95.8%), followed by strabismus (33.3%), microphthalmia (25%), and both nystagmus and microcornea (16.7% each). There was a significant overall improvement in visual acuity, from  $2.24 \pm 0.26$  to  $1.11 \pm 0.66$  logMAR ( $p < 0.001$ ).

**Conclusion:** The findings of this study confirm that timely surgery leads to improved anatomical and functional outcomes in PFV, influenced by a range of factors, both evident and less apparent.

**Keywords:** Persistent fetal vasculature, Persistent hyperplastic primary vitreous, Vascular malformations, Congenital abnormalities, Cataract, Pars plana vitrectomy.

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## 1. INTRODUCTION

Persistent Fetal Vasculature (PFV), formerly known as Persistent Hyperplastic Primary Vitreous, is a severe congenital anomaly that affects an infant's eye due to the failure of the hyaloid vessels to regress [1]. Normally, during the fourth month of embryonic development, the hyaloid vasculature system should naturally regress as the eye and its vascular system mature. However, in cases of PFV, the hyaloid artery and its proximal retinal branches typically persist in the newborn's eye as the central retinal artery. The primary objective of our study is to analyze and report the results of PFV surgery.

## 2. MATERIALS AND METHODS

The study was approved by Local Ethics Committee and conducted in accordance with the Declaration of Helsinki. A retrospective analysis was performed on the medical records of 24 eyes of 23 children with PFV who were admitted to the main referral tertiary eye care center, Kazakh Eye Research Institute, Kazakhstan, between 2017 and 2021. Data collection included information on age, gender, ophthalmic findings, PFV type, visual acuity at admission and at the last visit represented in logarithmic minimum angle of resolution

(logMAR), as well as any ocular comorbidities, the types of surgeries performed, and postoperative complications. All data are presented as means  $\pm$  standard deviation and range.

We classified PFV based on its localization: Anterior PFV is associated with cataracts and a retrolental plaque. Posterior PFV is characterized by its connection to the retinal surface, which may or may not result in optic nerve or macular distortion. Mixed PFV is characterized by the presence of a connection between the posterior pole and the lens, often leading to the development of cataracts and/or distortion of the optic nerve or macula (Table 1).

**Table 1. Comorbidities and incidence rates in patients with PFV.**

Ocular Comorbidities	%
Cataract	95.8%
Strabismus	33.3%
Corneal astigmatism ( $\geq 1.75$ diopters)	29.2%
Microphthalmia	25%
Nystagmus	16.7%
Microcornea	16.7%
Myopia	12.5%
Uveitis	12.5%
Vitreous hemorrhage	8.3%
Retinal detachment	8.3%
Lens subluxation	4.2%
Retinal degeneration	4.2%
Optic disc anomaly	4.2%

### 3. RESULTS

The average age at the time of surgery for the 23 children (24 eyes) was  $4.49 \pm 4.24$  years (ranging from 5 months to 16 years): 9 boys (39.1%) and 14 girls (60.9%). We observed the anterior form of PFV in 3 eyes (12.5%), the posterior form in 1 eye (4.2%), and the mixed form in 20 eyes (83.3%). One case of bilateral mixed-form PFV was observed in a three-year-old girl. Ocular comorbidities are detailed in Table 1. In most cases, PFV was accompanied by cataracts in 23 eyes (95.8%), followed by strabismus in 8 eyes (33.3%), corneal astigmatism more than 1.75 diopters in 7 eyes (29.2%), microphthalmia in 6 eyes (25%), nystagmus in 4 eyes (16.7%), and microcornea in 4 eyes (16.7%).

**Table 2. Clinical characteristics of pfv patients with preoperative and postoperative visual acuity, and surgery types.**

No.	Age (Years)	Gender (Male/Female - 1/0)	Type of PFV	Preoperative Visual Acuity	Postoperative Visual Acuity	Ocular Comorbidities	Surgery Type
1	0.42	1	Anterior	fix and follow light	fix and follow objects	Cataract, uveitis, strabismus, microcornea, microphthalmia, corneal astigmatism	Phaco + AV
2	0.5	0	Mixed	no light sensitivity	fix and follow light	Cataract, strabismus, microcornea, microphthalmia, corneal astigmatism	Phaco + IOL + AV
3	0.83	0	Mixed	light sensitivity	fix and follow objects	Cataract, nystagmus, strabismus	Phaco + IOL + AV
4	0.83	0	Posterior	no light sensitivity	fix and follow light	Microphthalmia, corneal astigmatism	LS-PPV
5	1	1	Anterior	fix and follow light	fix and follow objects	Cataract, nystagmus, strabismus	Phaco + IOL
6	1.12	0	Anterior	fix and follow light	fix and follow objects	Cataract, strabismus	Phaco + IOL

### 3.1. Surgery Types

Surgery for PFV was performed on all patients, which included phacoaspiration with an intraocular lens (IOL) implantation (2 eyes - 8.3%), phacoaspiration without IOL implantation (1 eye - 4.2%), phacoaspiration with IOL implantation and anterior vitrectomy (14 eyes - 58.3%), phacoaspiration with IOL implantation and pars plana vitrectomy (6 eyes - 25%), and lens-sparing pars plana vitrectomy (1 eye - 4.2%). IOL implantation was performed in 22 eyes (91.7%), while it was not performed in 2 eyes (8.3%): one eye due to microphthalmia, and the other due to a lens-sparing vitrectomy.

Postoperatively, patients were prescribed topical antimicrobial and anti-inflammatory treatment for one month, which included topical steroids and antibiotics (0.1% dexamethasone and 0.3% tobramycin eye drops, respectively). The mandatory follow-up period was three months.

In one case, a 5 years old girl with mixed form PFV and microphthalmia, who underwent phacoaspiration with IOL implantation and anterior vitrectomy, had closed funnel rhyematogenous retinal detachment four years after surgery. A pars plana vitrectomy with silicone oil tamponade was performed.

### 3.2. Visual Acuity

Table 2 contains the preoperative and postoperative visual acuity data for each individual child. Nine patients (9 eyes, 37.5%) were younger than 3 years and not cooperative for optotype visual acuity testing, with the youngest being 5 months old and the oldest being 2 years old. Among these younger patients, before surgery, 3 children showed no light sensitivity, 2 children showed light sensitivity, and the other 4 children were fixed and followed light. After the surgery, improvements in visual acuity were observed in all eyes: 3 children who initially showed no sensitivity to light as well as 1 child with limited light sensitivity developed the ability to fix and follow light, while the other 4 children initially had the ability to fix and follow light, and 1 child with limited light sensitivity achieved the ability to fix and follow objects at near distances.

(Table 4) contd....

No.	Age (Years)	Gender (Male/Female - 1/0)	Type of PFV	Preoperative Visual Acuity	Postoperative Visual Acuity	Ocular Comorbidities	Surgery Type
7	1.25	0	Mixed	light sensitivity	fix and follow light	Cataract, corneal astigmatism	Phaco + IOL + AV
8	1.25	1	Mixed	no light sensitivity	fix and follow light	Cataract, uveitis, microphthalmia	Phaco + IOL + PPV
9	2	1	Mixed	fix and follow light	fix and follow objects	Cataract, microcornea, myopia, corneal astigmatism	Phaco + IOL + AV
10	3	0	Mixed	2.0	1.3	Cataract, nystagmus, myopia	Phaco + IOL + AV
11	3	0	Mixed	2.0	1.0	Cataract, nystagmus	Phaco + IOL + AV
12	3	0	Mixed	light perception	1.0	Cataract, lens subluxation, strabismus	Phaco + IOL + AV
13	3	0	Mixed	no light perception	2.1	Cataract, microcornea	Phaco + IOL + AV
14	3	0	Mixed	light perception	1.5	Cataract, corneal astigmatism	Phaco + IOL + AV
15	3	1	Mixed	no light perception	2.1	Cataract, microphthalmia	Phaco + IOL + PPV
16	3	0	Mixed	2.0	0.398	Cataract, strabismus, corneal astigmatism	Phaco + IOL + AV
17	5	1	Mixed	light perception	1.5	Cataract, microphthalmia	Phaco + IOL + AV
18	7	0	Mixed	2.3	1.7	Cataract, retinal detachment	Phaco + IOL + PPV
19	8	1	Mixed	2.0	1.7	Cataract	Phaco + IOL + AV
20	9	1	Mixed	2.0	1.0	Cataract, optic nerve hypoplasia	Phaco + IOL + AV
21	10	0	Mixed	1.5	0.5229	Cataract, uveitis, strabismus	Phaco + IOL + PPV
22	10	1	Mixed	2.0	0.0969	Cataract, myopia, retinal degeneration	Phaco + IOL + AV
23	12	0	Mixed	2.0	0.0969	Cataract, vitreous hemorrhage, retinal detachment	Phaco + IOL + PPV
24	16	0	Mixed	1.7	0.6989	Cataract, vitreous hemorrhage	Phaco + IOL + PPV

**Notes:** Phaco: phacoaspiration; AV: anterior vitrectomy; IOL: intraocular lens implantation; PPV: pars plana vitrectomy; LS-PPV: lens sparing-pars plana vitrectomy.

Fourteen patients (15 eyes, 62.5%) (6.57 ± 4.13 years, ranging from 3 to 16 years) were cooperative for optotype visual acuity testing at admission. Table 3 shows preoperative and postoperative improvements in visual acuity for children older than 3 years, categorized into two age groups: younger (3 - 6 years) and older (7 - 16 years). Although the younger children showed a slightly greater improvement in visual acuity compared to the older ones after surgery (by 1.15 ± 0.35 and 1.09 ± 0.61, respectively), this difference was not statistically significant (p = 0.887; Mann-Whitney U test).

**Table 3. Preoperative and postoperative visual acuity in children over 3 years, categorized by age group (3-6 Years and 7-16 Years).**

Age	Visual Acuity Preoperative*	Visual Acuity Postoperative*	p-value <sup>†</sup>
3-6 years	2.5 ± 0.44 (2.7; 2.0 - 3.0)	1.36 ± 0.58 (1.4; 0.39 - 2.1)	0.005 <sup>‡</sup>
7-16 years	1.93 ± 0.26 (2.0; 1.5 - 2.3)	0.83 ± 0.67 (0.69; 0.09 - 1.7)	0.004 <sup>‡</sup>
Overall	2.24 ± 0.26 (2.0; 1.5 - 3.0)	1.11 ± 0.66 (1.0; 0.09 - 2.1)	< 0.001 <sup>‡</sup>

**Note:** \* visual acuity represented in mean ± standard deviation (median; range)

<sup>†</sup>Wilcoxon signed-rank test

<sup>‡</sup>statistically significant

**4. DISCUSSION**

PFV manifests in different forms based on the localization of fetal fibrovascular tissue. Although there is no standardized classification for PFV, some authors

classify it as anterior or posterior, while others offer a more comprehensive classification that includes anterior, posterior, mixed, and sometimes borderline forms [2, 3]. The anterior form is characterized by persistence of vasculature in the lens, retrolental membrane, forward displacement of the iridocrystalline diaphragm, abnormal ciliary processes, iridocorneocrystalline adhesions, and the development of cataracts. On the other hand, the posterior form is associated with retinal folds, vitreous membranes, optic disc hypoplasia, and the presence of a dense fibrous band running around the optic disc. The mixed form of PFV combines characteristics of both anterior and posterior PFV, typically presenting as the presence of a dense fibrous band extending from the optic disc to the lens.

In the majority of cases in our series, PFV was unilateral (95.8%), and most cases presented as a mixed form (83.3%). Moreover, 61% of PFV cases were girls, while other authors have reported that PFV is less common in girls (38% - 48.5%) [2, 3]. According to other authors, anterior PFV was observed in the range of 8.7% to 37.1%, the mixed form in the range of 60% to 89.1%, and the posterior cases in the range of 2.2% to 13% [2-6]. It is noteworthy that 71% of posterior PFV cases often had neurological problems [2]. In our study, the prevalence of anterior, mixed, and posterior forms of PFV was 12.5%, 83.3%, and 4.2% of cases, respectively, which is consistent with other studies. Bilateral cases were observed in the range of 4.3% to 24.1%, which is consistent with our study (4.3%) [2, 6, 7].

In PFV, microphthalmia is one of the observable

concurrent conditions, and it was found to be present in 20% to 44.2% of cases, with a higher incidence (34% to 71.4%) in cases of mixed form [2, 3, 5]. The prevalence of microphthalmia (25%) in our study is consistent with findings from other studies. Specifically, in our study, one out of three cases of anterior PFV, four out of twenty cases of mixed PFV, and the only case of posterior PFV had microphthalmia. Other authors reported that microphthalmic eyes were often associated with more severe posterior PFV and resulted in significantly worse visual and anatomical outcomes postoperatively [2, 3, 6, 8]. Sisk *et al.* noted that no eye with an axial length of less than 15 mm had light perception after surgery [2]. In our study, we had two children with microphthalmia who were cooperative for optotype visual acuity testing and their visual acuity increased from light perception and no light perception to 20/600 and counting fingers after surgery, respectively. According to other authors, PFV associated with retinal detachment is observed in 18.2% to 37.1% of cases, primarily in mixed forms [2, 5]. However, in case of retinal detachment surgery, complete reattachment after surgery is achievable in nearly two-third of cases [2].

Postoperative complications are a significant concern in cases of PFV, with reported rates up to 50% [2-5, 7]. Among these complications, glaucoma is a significant issue, with its occurrence ranging from 10.5% to 25% [2-4, 7]. Retinal detachment, which was observed in our study at a rate of 4.2%, is another worrisome complication, with reported occurrences ranging from 5.3% to 21.4% of cases [2-5, 7]. Macular holes are less common but still present, accounting for approximately 1.4% of cases [2]. Pupillary membrane formation is also a widely seen complication, affecting about 12.9% to 25% of cases [2, 4]. Posterior capsular opacification is another concern, with a prevalence ranging from 12.5% to 21% [4, 7]. Considering that the surgery aims to remove the unregressed vessel, there is insufficient data regarding postoperative vitreous hemorrhage. However, according to limited data, it occurs in approximately 5.3% of cases [4]. A noteworthy complication is the occurrence of postoperative phthisis bulbi, which ranges from 5.7% to 12%, with varying degrees of severity [2-5].

Our observations, consistent with findings from our colleagues, suggest that mixed and posterior forms of PFV are associated with less favorable functional outcomes after surgery [2, 6, 9]. This correlation is closely related to the extent of the posterior pole involvement, which depends on the site of stalk attachment and the width of its base. Smaller stalk base diameters and exclusive attachment to the optic nerve head are associated with better postoperative visual outcomes. Conversely, wider stalk base attachments located closer to the macula result in significantly poorer visual outcomes as a consequence of macular distortion. Anatomical outcomes are influenced by variables such as stalk width, the type of PFV, and the presence of microphthalmia. These variables are associated with a higher incidence of postoperative complications.

The latest analysis performed by Sisk (2010) regarding

visual acuity outcomes after surgery for PFV (7 studies, 280 eyes) showed that 39.9% of eyes achieved a final visual acuity better than 20/400 [2]. These outcomes varied significantly among different studies, with percentages ranging from 11.4% to 71.4%. Furthermore, overall visual acuity improvement was observed in 72.02% of cases, ranging from 25.7% to 85.7% across the various studies. The most recent studies reported an improvement in visual acuity in up to 83% of cases after surgery, with up to 33% of eyes achieving a final visual acuity better than 20/200, indicating that not all cases experienced a substantial visual improvement [4, 5, 7, 8]. Another study reported that after surgery, visual acuity improved in 66% of eyes, remained stable in 25%, and worsened in 9% [7]. In our study, we observed visual acuity improvement in all cases, with a final visual acuity better than 20/200 in 53.3% of cases.

According to our colleagues' reports, anterior PFV has a significantly higher likelihood of achieving functional vision compared to eyes with posterior PFV [2-4, 7-9]. Furthermore, eyes that experienced preoperative traction on the retina and optic nerve generally showed poorer visual outcomes. As mentioned earlier, microphthalmic eyes were often associated with severe posterior malformations, leading to significantly lower visual and anatomical results. Pollard emphasized the importance of age at the time of presentation in achieving better visual outcomes. Apparently, the impact of PFV affecting the posterior segment of the eye, including the macular area, becomes more severe as the eye grows and matures [9]. Therefore, performing surgery at an earlier age may help prevent or lessen these changes, suggesting that children operated on at a young age are more likely to achieve better visual outcomes compared to those operated on at older ages. Khandwala *et al.* reported that children who underwent lens-sparing surgery had significantly better visual acuity compared to those who were aphakic or pseudophakic after surgery [9]. The differences in visual outcomes observed across these studies may be attributed to variations in surgical techniques, patient populations, the type of PFV, the age at the time of surgery, the presence of concurrent eye pathologies, amblyopia, the status of the lens (phakic or aphakic), and other variables. It is important to acknowledge that PFV is a complex condition with individual variations, and its management requires personalized treatment strategies.

The primary limitations of our study include a short follow-up period and a limited number of children. The results of our research may be specific to our population and the surgical approach of our center. As the main referral center, we observed that the majority of children with PFV were referred for surgery due to various degrees of lens opacity. Since we did not study the general population, we cannot accurately determine the prevalence of different types of PFV. Consequently, many cases of posterior PFV may remain undiagnosed, as it is challenging to diagnose in children.

## CONCLUSION

In evaluating treatment outcomes, it is important to note that modern micro-invasive techniques allow for simultaneous surgery on PFV, congenital cataracts, vitreous opacities, and retinal detachments, contributing to the restoration of ocular media transparency and the enhancement of visual function. The complex and multifactorial nature of visual outcomes in PFV cases is highlighted by the variations in outcomes across different studies, influenced by a range of patient-specific factors and the details of the surgical procedures. The results of our colleagues' studies, along with our own findings, confirm that surgery, in some cases, leads to better anatomical and functional outcomes.

## DECLARATION

All named authors meet the International Committee of Medical Journal Editors () criteria for authorship for this manuscript, take responsibility for the integrity of the work as a whole, and have given final approval to the version to be published.

## AUTHORS' CONTRIBUTIONS

It is hereby acknowledged that all authors have accepted responsibility for the manuscript's content and consented to its submission. They have meticulously reviewed all results and unanimously approved the final version of the manuscript.

## LIST OF ABBREVIATIONS

PFV = Persistent Fetal Vasculature  
 logMAR = logarithmic minimum angle of resolution  
 ICMJE = International Committee of Medical Journal Editors

## ETHICS APPROVAL AND CONSENT TO PARTICIPATE

The study has gained approval from the Local Research Ethics Committee, Kazakh Eye Research Institute (Approval no. 17-2022).

## HUMAN AND ANIMAL RIGHTS

This research is in accordance with the Helsinki Declaration (2013).

## CONSENT FOR PUBLICATION

Informed consent was obtained from the guardians of the patients.

## STANDARDS OF REPORTING

STROBE guidelines were followed.

## AVAILABILITY OF DATA AND MATERIALS

The data supporting the findings of the article is available in Zenodo at <https://zenodo.org/records/11242905>, reference number 11242905.

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None.

## CONFLICT OF INTEREST

The authors declare no conflict of interest.

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Declared none.

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