• Clinical Research •

Subtypes and corneal astigmatism in pediatric persistent fetal vasculature

Shao-Hua Zhang[1,2,3], *Ning Lyu*[1,2,3], *Ying-Ying Hong*[1,2,3], *Yi Lu*[1,2,3], *Ying-Hong Ji*[1,2,3]

[1]Eye Institute and Department of Ophthalmology, Eye & ENT Hospital, Fudan University, Shanghai 200031, China
[2]NHC Key Laboratory of Myopia and Related Eye Diseases; Key Laboratory of Myopia and Related Eye Diseases, Chinese Academy of Medical Sciences, Shanghai 200031, China
[3]Shanghai Key Laboratory of Visual Impairment and Restoration, Shanghai 200031, China
Co-first Authors: Shao-Hua Zhang and Ning Lyu
Correspondence to: Yi Lu and Ying-Hong Ji. Eye Institute and Department of Ophthalmology, Eye & ENT Hospital, Fudan University, Shanghai 200031, China. luyieent@126.com; jiyh_eent@163.com
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Abstract

• **AIM:** To characterize the distribution of persistent fetal vasculature (PFV) subtypes and to evaluate corneal astigmatism (CA) in children with unilateral PFV.

• **METHODS:** The medical records of patients diagnosed with PFV between January 2014 and August 2021 were retrospectively reviewed. Corneal keratometry parameters were measured using IOLMaster or a handheld keratometer. Differences in CA between the affected and fellow eyes were analyzed in 52 unilateral PFV patients with available examination data.

• **RESULTS:** Totally 133 patients diagnosed with PFV were retrospectively reviewed. The male-to-female ratio was 73/60. Median age at surgery was 38.03mo (interquartile range 58.27mo). Among the PFV patients, 32 (24.06%) had anterior PFV, 2 (1.50%) had posterior PFV, and 99 (74.44%) had combined anterior-posterior PFV. Mild combined PFV was the most common subtype. In unilateral PFV cases, the mean CA in the affected eye was 2.29±1.11 D, and 59.62% (31 eyes) had CA≥2.0 D. The mean CA in the affected eyes was significantly higher than in the fellow eyes (1.37±0.77 D; P<0.001). Among PFV-affected eyes with CA≥2.0 D, the steepest corneal meridian was vertically oriented in 30 cases (96.77%), while only 1 case (3.23%) had the steepest meridian oriented horizontally.

• **CONCLUSION:** In children with unilateral PFV, CA is significantly higher in the affected eyes than in the fellow eyes, and the steepest corneal meridian was predominantly oriented vertically.

• **KEYWORDS:** persistent fetal vasculature; corneal astigmatism; corneal keratometry; children
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INTRODUCTION

Persistent fetal vasculature (PFV) is a developmental malformation caused by incomplete regression of the embryonic hyaloid vascular system[1]. It was first described as persistent hyperplastic primary vitreous (PHPV) by Reese[2] and later re-termed as PFV by Goldberg[3]. Most cases of PFV are unilateral, with only approximately 10% of cases being bilateral[2]. PFV has a spectrum of subtypes, classified according to the location of the vasculature remnants in the anterior or posterior segment, or both[4-5]. The anterior subtype is characterized by retrolental opacity, elongated ciliary processes, without a stalk or attachment to the posterior segment[6-7]. The posterior subtype presents with a fibrous stalk extending from the optic nerve head to the retrolental region, causing elevation of the vitreous membrane from the optic nerve, inducing retinal folds, dysplasia, or retinal detachment[8-10]. The combined subtype of PFV exhibits features of both anterior and posterior subtypes. In some severe cases, the PFV may be associated with corneal abnormalities, such as corneal opacification[11]. In addition to corneal opacity, abnormalities in corneal size and thickness may also coexist with PFV[12-13]. Furthermore, a subset of patients has been reported to present with a syndrome characterized by bilateral microcornea, posterior megalolenticonus, PFV, and chorioretinal coloboma[14-15].

Corneal astigmatism (CA) is a common refractive error resulting from meridional asymmetry in corneal curvature, leading to asymmetric refraction of light rays[16]. Regular astigmatism can be classified as with-the-rule, against-the-rule, and oblique based on the orientation of the steepest corneal meridian[17]. Moreover, corneal curvature is a key parameter

in intraocular lens (IOL) power calculation[18-19]. If CA is not properly identified and corrected following cataract surgery in PFV patients at the appropriate age, it may lead to meridional amblyopia in pediatric patients and ultimately compromise visual outcomes[20]. Notably, a similar degree of astigmatism may have a greater visual impact in eyes implanted with multifocal IOLs compared with monofocal IOLs[21]. Therefore, corneal shape abnormalities should be carefully considered. Existing studies on the prevalence and distribution of CA in PFV are limited, with most research primarily focusing on the diagnostic and therapeutic aspects of PFV.

In this study, we investigated the prevalence of CA and the distribution of keratometric values in children with unilateral PFV.

PARTICIPANTS AND METHODS

Ethical Approval All procedures conformed to the tenets of the Declaration of Helsinki, and written informed consent was obtained from at least one parent or legal guardian of each participant. The study protocol was approved by the Ethics Committee of Eye and Ear, Nose, and Throat (EENT) Hospital of Fudan University (Shanghai, China) (approved No.2022119).

Participants Patients aged 18y or younger who presented with PFV at the EENT Hospital of Fudan University from January 2014 to August 2021 were enrolled in this study. The exclusion criteria included any history of ocular surgery, ocular trauma, or corneal abnormalities, such as corneal opacity, infection, or edema.

Persistent Fetal Vasculature Classification PFV was classified according to both location—isolated anterior, isolated posterior, or combined—and severity (mild *vs* severe), as previously described[8]. Briefly, mild or severe isolated anterior PFV was characterized by a retrolental plaque with or without ciliary process elongation and without a stalk or attachment to the posterior segment. In isolated posterior PFV, the distinction between mild and severe cases was based on the presence of moderate to severe distortion of the optic nerve or macula. In the combined type, severe cases were defined by the degree of optic nerve or macular distortion and ciliary process elongation.

Ocular Examination and Parameters Corneal parameters—including CA, flat keratometry (Kf), steep keratometry (Ks), and mean keratometry (Km)—were measured under undilated pupil conditions before surgery using either the IOLMaster (Carl Zeiss Meditec AG) or the Nidek HandyRef-K (Tokyo, Japan). Axial length (AL) was measured using the IOLMaster or an A-scan ultrasound biometer (AL-3000, Tomey Corp., Japan). B-mode ultrasound and color Doppler imaging (CDI) were performed using the Aviso platform (Quantel Medical, France) and the HDI 5000 CDI (Philips Ultrasound, USA) with a 7.5 MHz linear probe, respectively. Patients who were unable to cooperate during the preoperative examination were sedated with 10% chloral hydrate (0.4 mL/kg, orally). AL and keratometry were assessed intraoperatively using the A-scan ultrasound biometer and the Nidek HandyRef-K. For each parameter the mean of three measurements meeting quality standards was calculated.

CA was defined as the difference between Kf and Ks, and Km was defined as the arithmetic mean of Kf and Ks. CA was categorized using a threshold of 2.0 D[22]. Age was stratified into two subgroups using 6y—the typical age of school entry—as the cutoff.

Statistical Analysis Statistical analyses were performed using IBM SPSS Statistics version 26.0. Qualitative variables are presented as absolute counts (*n*) and percentages (%). Age is reported as the median and interquartile range (IQR). The normality of all other continuous variables was assessed using the Shapiro-Wilk test, and normally distributed variables were expressed as mean±standard deviation (SD). An independent-samples *t* test was used to analyze variables of PFV-affected eyes, and a paired *t*-test was used to evaluate the differences between the affected and fellow eyes of patients with unilateral PFV. A P value <0.05 was considered statistically significant.

RESULTS

The medical records of 133 patients diagnosed with PFV were retrospectively reviewed. Representative preoperative B-scan ultrasonography, CDI images, and intraoperative photographs of the included patients are shown in Figure 1. Patient demographics and baseline characteristics are summarized in Table 1. The median age at the time of surgery was 38.03mo. The IQR for the time from initial detection to surgical intervention was 58.27mo. The male-to-female ratio was 73/60. Among these patients, 124 (93.23%) had unilateral PFV (54 cases involving the right eye and 70 involving the left eye), while 9 patients (6.77%) had bilateral involvement. In 52 of the 124 patients with unilateral PFV, appropriate measurements were obtained for both eyes, allowing comparison between the affected and fellow eyes.

Of the 133 patients with PFV, 32 (24.06%) were diagnosed with anterior PFV, 2 (1.50%) with posterior PFV, and the remaining 99 (74.44%) with combined anterior-posterior PFV (Figure 2A). Mild combined PFV was the most frequently observed subtype, accounting for 49.62% (66 cases), followed by severe combined PFV (24.81%, 33 cases) and mild anterior PFV (20.30%, 27 cases). Among the 52 unilateral PFV patients with available examination data included in the comparative analysis, 14 (26.92%) had isolated anterior PFV and 38 (73.08%) had combined anterior-posterior PFV (Figure 2B).

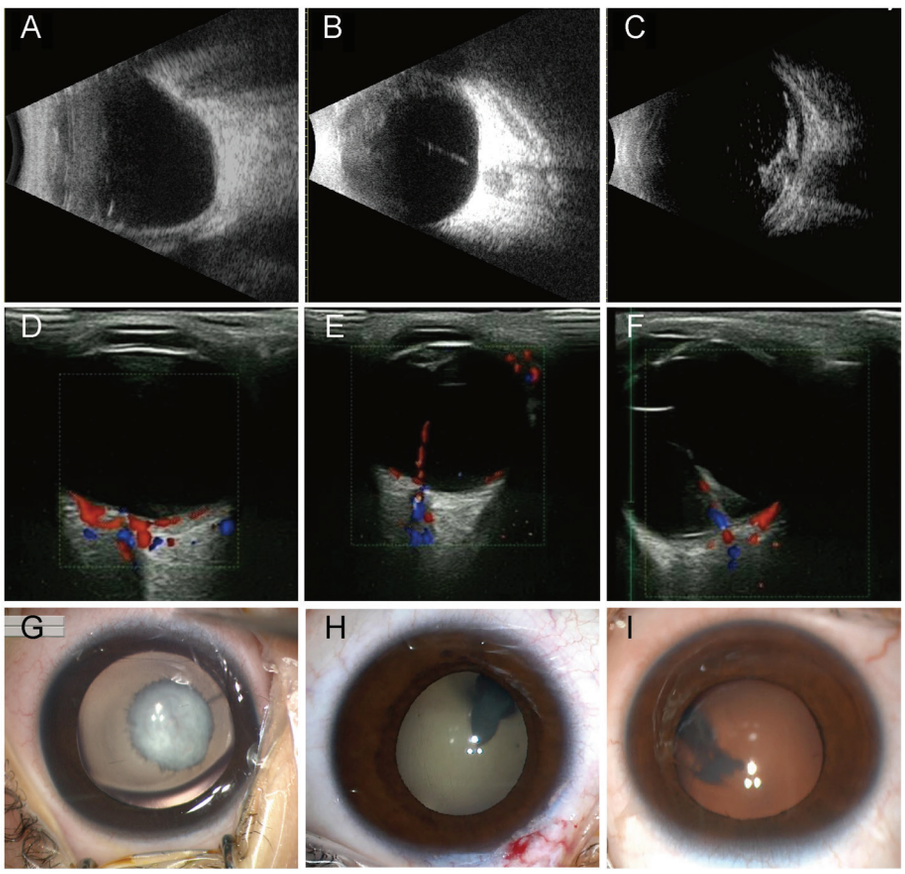

Figure 1 Representative images of preoperative B-scan ultrasonography (A–C), color Doppler imaging (D–F), and intraoperative photographs (G–I) in patients with PFV PFV: Persistent fetal vasculature.

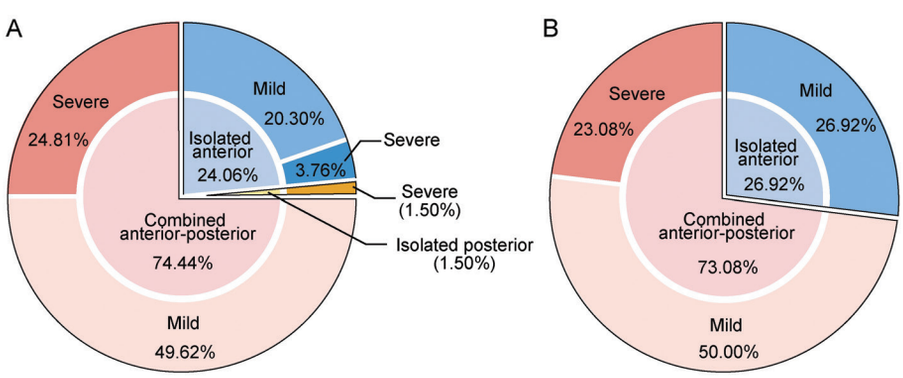

Figure 2 Sunburst charts illustrating the classification of PFV The inner ring shows the proportions of the three PFV subtypes—isolated anterior, isolated posterior, and combined—arranged clockwise from the top. The outer ring represents the corresponding severity subcategories (mild and severe). A: PFV classification among all patients; B: PFV classification among patients included in the subsequent comparative analysis. PFV: Persistent fetal vasculature.

Table 1 Patient demographics and baseline characteristics

Parameters	Total	Eligible
Age at surgery (mo), median (IQR)	38.03 (58.27)	63.10 (32.49)
Time from detection to treatment (mo), median (IQR)	7.65 (19.47)	15.25 (22.94)
Gender (M/F), *n* (%)	73/60 (54.89/45.11)	27/25 (51.92/48.08)
Eyes (unilateral/bilateral), *n* (%)	124/9 (93.23/6.77)	52/0 (100/0)
Eyes (OD/OS), *n* (%)	54/70 (43.55/56.45)	28/24 (53.85/46.13)

IQR: Interquartile range; OD:Right eye; OS: Left eye.

The mean values of CA, Kf, Ks, Km, and AL for the PFV-affected and fellow eyes are presented in Table 2. The mean CA in the affected eyes was 2.29±1.11 D (range, 0.11–3.32 D), which was significantly higher than that in the fellow eyes

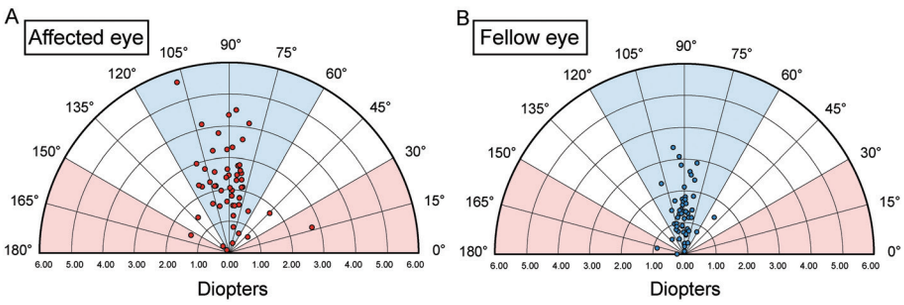

Figure 3 Single-angle polar plots of corneal astigmatism in PFV-affected eyes (A) and fellow eyes (B) PFV: Persistent fetal vasculature.

Table 2 Corneal curvature parameters and AL in unilateral PFV patients

Parameters	Total (*n*=52)	≤6y (*n*=33)	>6y (*n*=19)	P_1
CA				
Affected eye	2.29±1.11	2.27±1.15	2.31±1.07	0.924
Fellow eye	1.37±0.77	1.28±0.70	1.52±0.87	0.280
P_2	<0.001	<0.001	<0.001	
Kf				
Affected eye	43.14±2.19	43.16±2.36	43.04±1.99	0.809
Fellow eye	42.87±1.30	42.8±1.45	42.98±1.02	0.637
P_2	0.225	0.195	0.855	
Ks				
Affected eye	45.38±2.35	45.47±2.53	45.24±2.03	0.735
Fellow eye	44.21±1.46	44.05±1.59	44.50±1.18	0.287
P_2	<0.001	<0.001	0.042	
Km				
Affected eye	44.26±2.20	44.33±2.37	44.14±1.95	0.764
Fellow eye	43.54±1.32	43.42±1.47	43.74±1.02	0.414
P_2	0.001	0.003	0.229	
AL				
Affected eye	22.28±1.71	21.86±1.59	22.98±1.71	0.026
Fellow eye	22.38±1.12	21.95±0.89	23.08±1.11	<0.001
P_2	0.649	0.716	0.792	

PFV: Persistent fetal vasculature; CA: Corneal astigmatism; Kf: Flattest keratometry; Ks: Steepest keratometry; Km: Mean keratometry; AL: Axial length. P_1: Patients aged <6y *vs* those aged >6y; P_2: Affected eye *vs* fellow eye.

(1.37±0.77 D, P<0.001). The Ks value in the affected eyes (45.38±2.35 D) was also significantly steeper than that in the fellow eyes (44.21±1.46 D, P<0.001). No statistically significant differences were observed in Kf or AL between the affected and fellow eyes. Additionally, corneal keratometric values did not differ significantly between patients aged ≤6y and those aged >6y.

The distributions of CA in PFV-affected eyes and fellow eyes are illustrated in Figure 3. High CA (≥2.0 D) was observed in 59.62% (31 eyes) of the affected eyes and in 19.23% (10 eyes) of the fellow eyes. Among PFV-affected eyes with high CA (≥2.0 D), the steepest corneal meridian was vertically oriented in 30 eyes (96.77%), whereas a horizontally oriented steepest corneal meridian was observed in 1 eye (3.23%).

To evaluate the effects of PFV subtypes and severity on

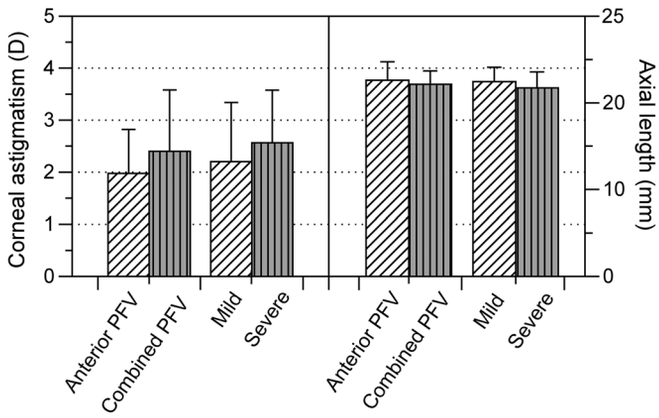

Figure 4 Bar diagrams comparing corneal astigmatism and axial length between patients with anterior or combined PFV, and between those with mild or severe PFV. Bars present mean±standard deviation. PFV: Persistent fetal vasculature.

CA and AL, we compared the corresponding data among the groups (Figure 4). The mean CA was slightly higher in patients with combined PFV than in those with anterior PFV (2.40±1.18 *vs* 1.98±0.85 D, respectively). In terms of severity, the mean CA was 2.56±1.02 D in patients with severe PFV and 2.20±1.14 D in those with mild PFV. However, none of these differences reached statistical significance. The mean AL was 22.62±2.12 mm in patients with anterior PFV and 22.15±1.54 mm in those with combined PFV. In patients with mild and severe PFV, the mean AL was 22.45±1.54 mm and 21.72±1.84 mm, respectively. These differences were also not statistically significant.

DISCUSSION

CA is a common accompanying feature of PFV. In this study, we found that the mean CA in PFV-affected eyes was significantly higher than that in fellow eyes. This difference may be attributed to the steeper Ks observed in PFV-affected eyes. No significant association was found between CA and patient age. PFV is typically characterized by tractional forces, with fibrovascular strands extending between the ciliary body, retina, and posterior lens[23]. Such traction may contribute to alterations in corneal toricity. A previous study reported a mean CA of 3.17±0.23 D in patients with persistent fetal membranes[24]. Another study involving 21

patients with unilateral PFV found that the Km in affected eyes (46.48±3.16 D) was significantly higher than that in fellow eyes (44.38±2.26 D)[25].

In this study, the mean CA in the affected eyes of patients with unilateral PFV was 2.29±1.11 D, and 59.62% of patients had CA≥2.0 D. In contrast, a recent report indicated that PFV was associated with CA greater than 1.75 D in 29.2% of cases[26]. Several studies have focused on the distribution of CA in patients with unilateral congenital cataract before surgery and have found that affected eyes generally exhibit steeper keratometric values compared with fellow eyes[27-28]. For instance, Lin *et al*[27] reported a mean CA of 2.05±1.55 D in cataract-affected eyes among patients under 18 years of age. These findings suggest that cataract may be an important contributing factor to increased CA and should be taken into consideration. The presence of fibrovascular membranes can exert mechanical traction on the peripheral cornea, retina, and lens, potentially leading to structural deformation[23]. Our data showed a trend toward higher CA in patients with combined PFV compared to those with anterior PFV, and in severe cases compared with mild ones, although these differences were not statistically significant. These observations suggest that both fibrovascular traction and lens abnormalities may contribute to elevated CA in patients with unilateral PFV. Furthermore, CA may correlate with the severity of PFV.

Most PFV cases are sporadic and unilateral, although bilateral involvement has been reported in 4%–11% of patients[8,29]. Consistent with this, our study included nine patients (6.77%) with bilateral PFV. Combined PFV represents the most common and complex subtype of the disease. In this study, combined PFV was present in 74.44% of the 133 patients. We further classified PFV based on both anatomical location and disease severity, following the classification system proposed by Khandwala *et al*[8]. In their cohort of 45 patients, 50% had mild combined PFV, 39% had severe combined PFV, 2% had mild anterior PFV, and 7% had severe anterior PFV. Compared with that study, our cohort had a larger proportion of patients with anterior PFV (24.06% *vs* 9%), although this proportion still lower than the 48.3% reported in another single-center study[30]. These discrepancies may reflect differences in ethnicity, population characteristics, and surgical indications across centers, which may have introduced sampling bias.

There are several limitations to this study. First, some biometric data were unavailable for certain cases. Although the study included 133 patients, only 52 underwent comprehensive biometric examination. Second, the number of eyes in each subgroup was relatively small, which limited the statistical power for intergroup comparisons and precluded the identification of potential risk factors. Third, this study did not include a direct comparison of corneal CA between patients with congenital cataract and those with PFV, making it difficult to fully assess the impact of traction on corneal toricity in PFV. Further studies with larger sample sizes, including congenital cataract cases for comparison, are needed to clarify the relationship between CA and PFV subtypes.

In conclusion, this study provides a preliminary investigation into the distribution of PFV subtypes and highlights the high prevalence of CA in PFV-affected eyes. The observed biometric differences between PFV-affected and fellow eyes underscore the limitations of relying on corneal parameters derived from normative data or the fellow eye. Individualized assessment and refractive correction are essential in the clinical management of PFV patients.

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Conflicts of Interest: Zhang SH, None; **Lyu N,** None; **Hong YY,** None; **Lu Y,** None; **Ji YH,** None.

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