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Case Report

Bilateral Persistent Fetal Vasculature With Multiple Fibrovascular Stalks.

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Running head: Bilateral PFV with multiple stalks.

Abstract

We report a case of a patient with bilateral partial retrolental leukocoria and clear lenses. Binocular indirect ophthalmoscopy identified peripheral retina atrophy in both eyes and laser photocoagulation was performed in order to prevent retinal detachment in both eyes. Ophthalmic B-scan ultrasound and doppler echography disclosed bilateral microphthalmia with axial length 17.85 mm in the right eye and 18.27 mm in the left eye, and bilateral multiple tubular stalks of fibrovascular tissue, attached to the posterior surface of the lens, connecting to the optic nerve, consistent with bilateral persistent fetal vasculature. Due to a progressive lens opacification and consequent visual impairment in both eyes a 25gauge sutureless lensectomy technique (TT–Technique) was performed in both eyes. At last follow up the best corrected visual acuity was 20/600 in the right eye and light perception in the left eye.

Keywords : *bilateral persistent fetal vasculature, multiple fibrovascular stalks, 4q-Syndrome, lensectomy, pediatric cataract, leukocoria.*

INTRODUCTION

Bilateral cases of persistent fetal vasculature (PFV) account for less than 10% of the cases, and may be associated with systemic syndromes and genetic testing is advisible.1 The patient reported here presented a rare ocular condition with bilateral combined form of PFV with multiple fibrovascular stalks challenging surgical treatment.

CASE REPORT

We report a case of a male patient with bilateral partial retrolental leukocoria **(Figures 1 and 2)** and clear lenses present at birth. Ophthalmic B-scan ultrasound disclosed bilateral microphthalmia, presenting axial length of 17.85 mm in the right eye and 18.27 mm in the left eye **(Figure 3)**. Ocular doppler echography disclosed bilateral multiple tubular stalks of fibrovascular tissue attached to the posterior surface of the lens connecting to the optic nerve consistent with bilateral PFV **(Figure 4)**. Blood flow inside the stalks vessels was detected in both eyes. Right eye showed four fibro vascular stalks and the left eye showed five. The fibrovascular stalks were connected to the optic nerve and nasal retina **(Figure 5)**. Binocular indirect ophthalmoscopy identified retinal dysplasia and peripheral retina atrophy and, at nine months of age, a 360 degrees peripheral laser photocoagulation was performed in order to prevent retinal detachment in both eyes. Anterior segment evaluation revealed partial peripheral leukocoria due to the fibrovascular sheath attachment to the posterior lens capsule **(Figure 2)**. Due to a progressive lens opacification developing total cataracts and consequent visual impairment, a 25 gauge sutureless lensectomy technique (TT-Tartarella Technique) was performed in both eyes at the age of 20 months for left eye and 26 months for the right eye **(Figure 6)**.

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Figure 1. Bilateral leukocoria.

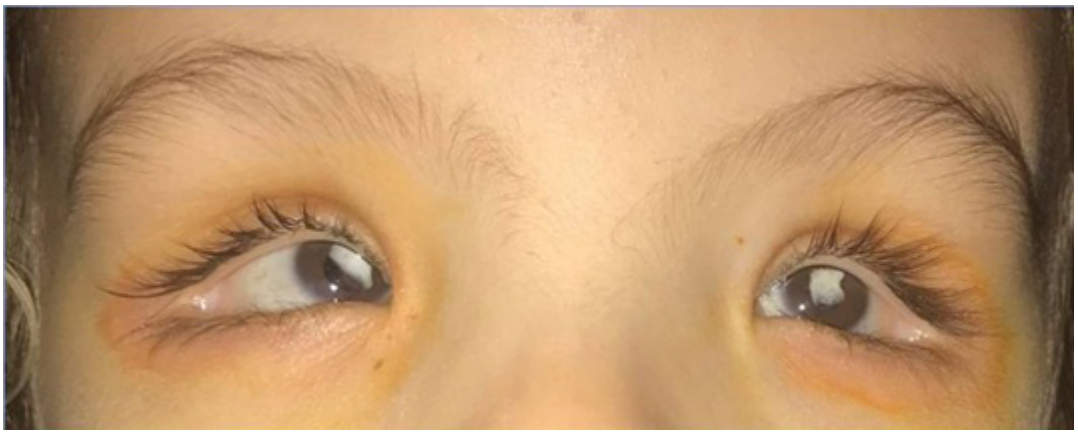


Figure 2. Partial peripheral and vascularized leukocoria in the right eye.

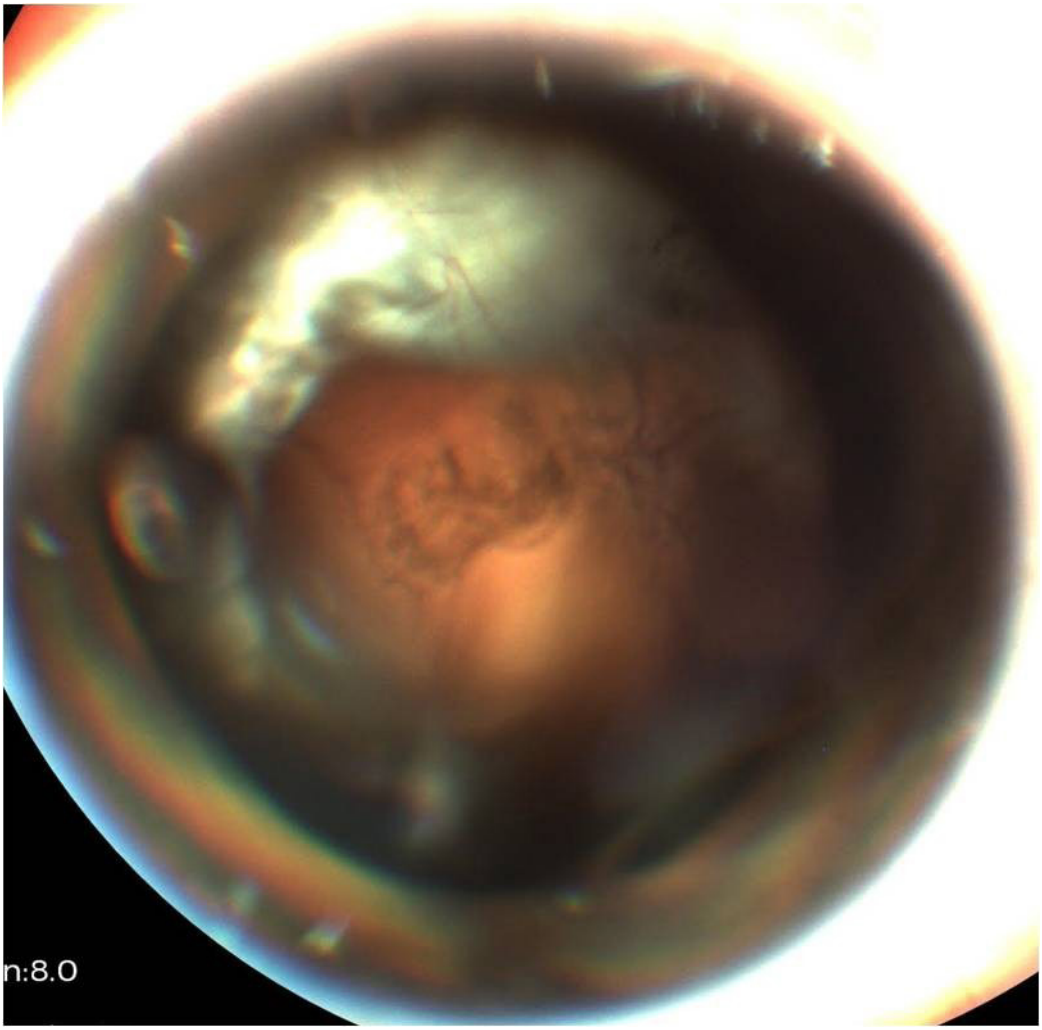


Figure 3. Ultrasonographic imaging of the right and left eyes.

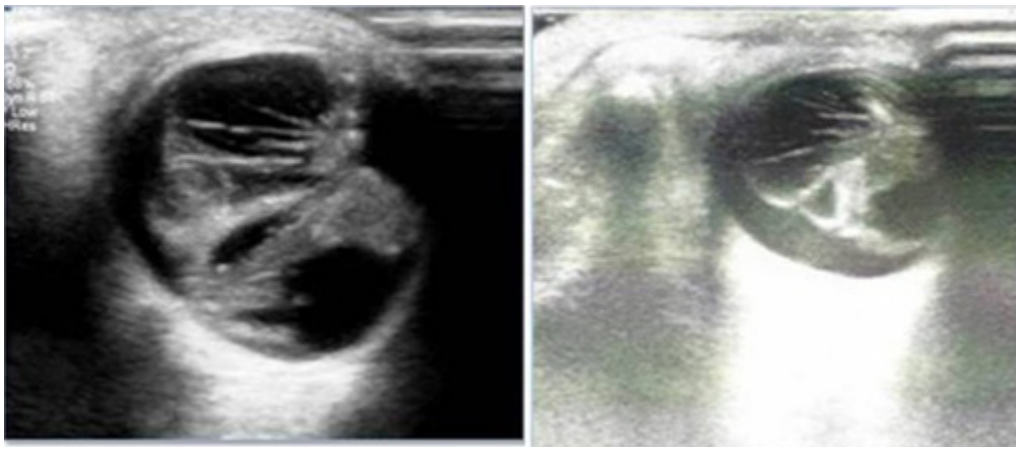


Figure 4. Ocular doppler echography of the left eye disclosed multiple tubular stalks of fibrovascular tissue attached to the posterior surface of the lens connecting to the optic nerve with blood flow inside the stalk vessels.

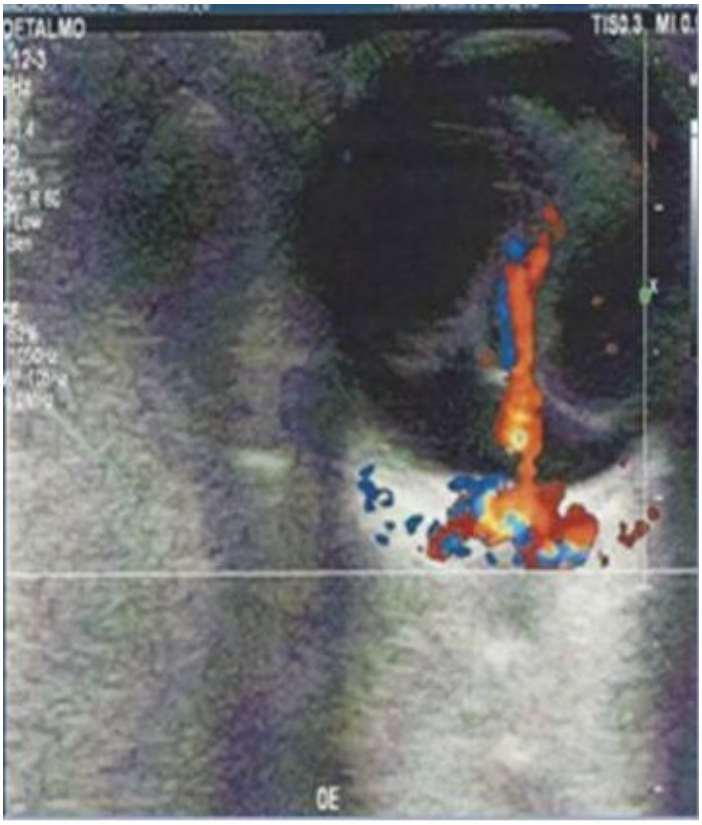


Figure 5. Fibrovascular stalks were connected to the optic nerve and nasal retina.

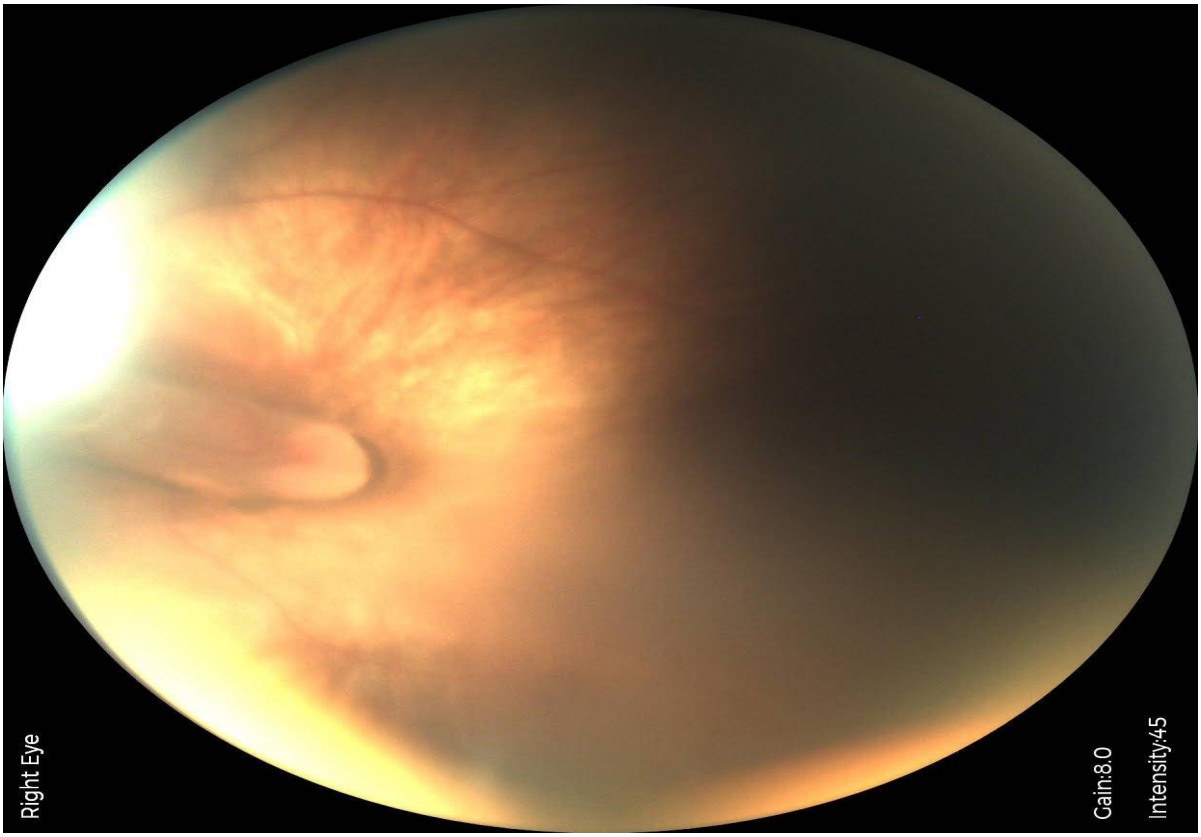
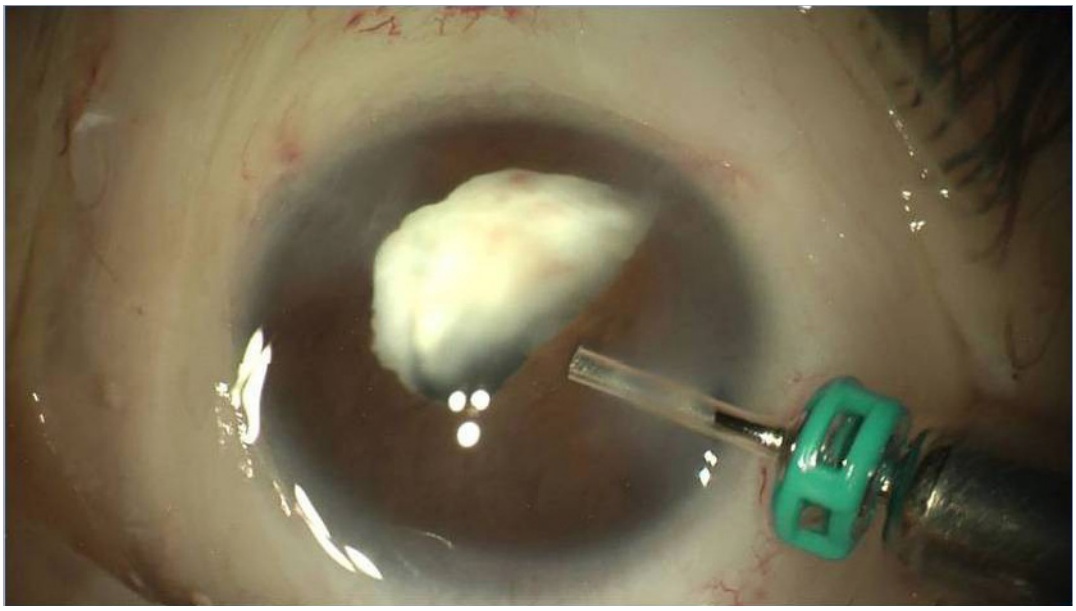


Figure 6. 25 gauge sutureless lensectomy technique (TT-Tartarella Technique) was performed in both eyes.



Surgical procedure was challenging. Blood vessels were cauterized with endo-diathermy, and no intraocular bleeding or hemorrhage occurred. There were total cataracts in both eyes with a dense plaque adhered to the anterior end of the fibrovascular stalk. Intra-vitreous anti-VEGF Ranibizumab was injected at the end of the surgical procedure. At the age of 50 months, both visual axes were partially clear with the presence of red reflex. Both eyes were left aphakic and glasses were prescribed. At last follow up by 50 months of age the best corrected visual acuity was 20/600 in the right eye and light perception in the left eye. Nystagmus, esotropia and low vision were detected.

Patient's phenotype included mild facial dysmorphism with hypertelorism, short upturned nose and depressed nasal bridge.

He was referred for genetic analysis (microarray testing) that revealed deletion of the long arm of chromosome 4 (4q31.3-q32.3).

DISCUSSION

Persistent fetal vasculature is a congenital developmental anomaly of the eye resulting from failure of the embryological primary vitreous and hyaloid vasculature to regress. Most cases are unilateral. Bilateral cases of PFV account for less than 10% of the cases, and may be associated with rare systemic syndromes.1,2 A genetic study would be advisable, although no gene has been reported to be directly involved with PFV.1 Other bilateral neonatal ocular pathologies have to be excluded as differential diagnosis as Familial Exsudative Vitreous Retinopathy and Norrie disease.3

The case reported here presented an atypical ocular condition with bilateral combined form of PFV with multiple fibrovascular stalks in both eyes. Partial peripheral retro-lental leukocoria was due to the fibrovascular sheath attachment to the posterior lens capsule.4 Genetic study was undertaken and detected an anomaly in the long arm of chromosome 4. A few cases of 4q-Syndrome have been reported in the literature.5. Further analysis is been carried out to better understand the 4q-Syndrome in this patient.

The heterogeneity of clinical presentation makes PFV a challenge to surgical management. A novel classification of PFV based on high-resolution B-mode ultrasound and color doppler imaging was proposed by Hu et al. in order to help in the planning of surgical treatment.6 Recent advances in surgical instrumentation, vitrectomy equipments and techniques have changed the indications for PFV surgery.7,8

Cataract surgery in patients with PFV needs retinal surgery skills and is related to higher rates of intra or postoperative complications as: retinal detachment, hyphema, intraocular hemorrhage, glaucoma, opacification of the visual axis and extensive inflammatory response with synechiae and pupillary block.9-11 Initial surgical planning may alter depending on the pre and intraoperative ocular conditions. Ocular eco-doppler findings detecting blood flow from the optic nerve to the posterior face of the lens is a predictive factor of intraoperative hemorrhage. In some cases, endo-diathermy of the blood vessels is necessary. Intraocular hemorrhage is a common cause of bad prognosis and poor anatomical outcomes.1,11.

Eyes with PFV and cataract may be associated with variable degrees of microphthalmia that could interfere in the visual improvement. Early detection of ophthalmic pathology in children is important in order to minimize visual impairment. Congenital cataract is an amblyogenic condition and when associated with PFV can cause secondary glaucoma, and intraocular hemorrhages that interfere in the final functional vision.1 The patient underwent a 25G sutureless vitrectomy and lensectomy (TT-Technique) at 20 and 26 months of life. The surgical TT-Technique was previously described.,7,8 Early surgical intervention with minimal invasive 25G lensectomy technique combined with anti-amblyopic therapy may result in favorable visual outcomes. The use of intraocular anti-VEGF drugs in order to avoid excessive post operative inflammatory reaction and minimize complications in cases with intra-ocular vascularization, as in PFV, is being prescribed.12 Further studies concerning cumulative factors that could predict surgical prognosis for PFV cases are necessary.

Few studies have discussed the possibility of a patient having more than one vascular stalk. There is a previous article from Shah and Moshfeghi relating a PFV case having two well-delineated stalks in only one eye in a 5-year-old female.13 Rossin et al. published a case in an 8-week-old boy with two distinct vascular stalks, evaluated by angiofluoresceinography. They discuss the ocular development and pathogenesis of PFV. Their hypothesis is that the aberrant additional stalk may represent failure of the vasa hyaloidea propria (tributaries of the hyaloid artery) to regress.14

Retinal anomalies and deprivation amblyopia contributed to visual impairment in this patient, on the other hand, the child is able to find the way to walk and perform daily tasks for his age. But he prefers to play the keyboard, which means his hearing sensitivity is overtaking his visual impairment. He exhibits speech delay and he demonstrates autism-like behavior due to his low vision. He presents mannerism by rubbing his eyes frequently. He is attending an intensive program of visual and global early intervention to achieve the best results of his sensorial potentials.

Further systematic follow-up is required to evaluate anatomical and functional results after surgery, as retinal detachment can occur due to the retinal traction caused by the fibrovascular stalks in an atrophic peripheral retina despite previous photocoagulation.

CONCLUSIONS

The related case presents an atypical condition of bilateral PFV associated with multiple fibrovascular stalks in both eyes and with an anomaly of chromosome 4. Surgical intervention with vitrectomy and endo-diatermy was necessary due to cataract formation. Early visual intervention program was mandatory due to visual impairment.

Declaration of Patient Consent

The authors certify that they have obtained appropriate patient consent form. In the form the patient s parents have given consent for the images and other clinical information to be reported in the journal. The patient s family understand that their names and initials will not be published and due

efforts will be made to conceal their identity

Acknowledgment

The authors certify that the content has not been published or submitted for publication elsewhere. Authors also certify that the protocol for the research project has been approved by a suitably constituted Ethics Committee of the institution within which the work was undertaken, and that it conforms to the provisions of the Declaration of Helsinki in 1995 (as revised in Edinburgh 2000).

Disclosures

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REFERENCES

1. Ramasubramanian A, Duff-Lynes SM. Persistent Hyperplastic Primary Vitreous in American Academy of Ophthalmology EyeWiki. 2024 Nov 19.

2. Avila Morales MS, Tartarella MB, Gouveia EB, Mandello MH, Allemann N. Ophthalmic Doppler in persistent hyperplastic primary vitreous atypical presentation: case report. Arq Bras Oftalmol 2015; 78(5):320-322.

3. KumarA, Jethani J, Shetty S, Vijayalakhmi P. Bilateral persistent fetal vasculature: a study of 11 cases. J AAPOS 2010;14(4):345-348.

4. Tartarella MB, Britez-Colombi GF, Fortes Filho JB. Proposal of a novel classification of leukocorias. Clin Ophthalmol. 2012;6:991-995. doi: 10.2147/OPTH.S31469. Epub 2012 Jun 29. PMID: 22815641; PMCID: PMC3399387.

5. Sarda P, Lefort G, Fryns JP, Humeau C, Rieu D. Interstitial deletion of the distal long arm of chromosome 4. J Med Gent. 1992 Apr;29(4):259-261. doi: 10.1136/jmg.29.4.259. PMID: 1583648; PMCID: PMC1015928.

6. Hu A, Pei X, Ding X, Li J, Li Y, Liu F et al. Combined persistent fetal vasculature. A classification based on high-resolution B-mode ultrasound and color doppler imaging. Ophthalmology 2016;123:19-24.

7. Tartarella MB, Fortes Filho JB. Twenty-five-gauge sutureless lensectomy in infants with congenital cataract. J AAPOS. 2017 Oct;21(5):393-396. doi: 10.1016/j.jaapos.2017.07.203. Epub 2017 Sep 6.

8. Tartarella MB, Takahagi RU, Braga AP, Fortes Filho JB. Persistent fetal vasculature: ocular features, management of cataract and outcomes. Arq Bras Oftalmol 2013;76(3):185-188.

9. Müllner-Eidenböck A, Amon M, Moser E, Klebermass N. Persistent fetal vasculature and minimal fetal vascular remnants: a frequent cause of unilateral congenital cataracts. Ophthalmology 2004;111(5):906-913.

10. Kuhli-Hattenbach C, Hofmann C, Wenner Y, Koch F, Kohnen T. Congenital cataract surgery without intraocular lens implantation in persistent fetal vasculature syndrome: Long-term clinical and functional results. J Cataract Refract Surg 2016;42:759-767.

11. Vasavada AR, Vasavada SA, Bobrova N, Praveen MR, Shah SK, Vasavada VA, et al. Outcomes of pediatric cataract surgery in anterior persistent fetal vasculature. J Cataract Refract Surg. 2012;38(5):849-857.

12. Elhusseiny AM, Hennein L, VanderVeen DK. Bevacizumab as adjunctive therapy in anterior persistent fetal vasculature. Eur J Ophthalmol. 2024 Jan;34(1):NP18-NP21. doi: 10;1177/11206721231187428. Epub 2023 Jul 9. PMID 37424268.

13. Shah SM, Moshfeghi DM. Multiple vascular stalks in a patient with persistent fetal vasculature. Ophthalmic Surg Lasers Imaging Retina 2019;50(5)330-331. doi.org/10.3928/23258160-20190503-12.

14. Rossin EJ, VanderVeen DK, Yonekawa Y. Anterior–Posterior Persistent Fetal Vasculature with multiple stalks: Persistent vasa hyaloidea propria. J of VitreoRetinal Dis. 2(4): doi.org/10.1177/2474126418776072.