Persistent Fetal Vasculature With Elongated Ciliary Processes in Children

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• PURPOSE: **Outcomes and reoperation rates in infants with unilateral persistent fetal vasculature (PFV) with elongated or stretched ciliary processes have not been extensively studied and were excluded from the Infant Aphakia Treatment Study (IATS). The purpose of this study is to analyze the preoperative measurements, reoperation rates, and complications after surgery in unilateral anterior PFV with stretched processes.**

• DESIGN: **Retrospective case series.**

• METHODS: **Inclusion criteria consisted of unilateral cataract surgery prior to 7 months of age and anterior PFV with elongated ciliary processes. Eyes with posterior retinal involvement and less than 6 months of follow-up were excluded. All patients underwent lensectomy, posterior capsulectomy, and vitrectomy.**

• RESULTS: **Eleven eyes of 11 patients were included. Patients had a mean age at surgery of 2.4 ± 1.4 months. Average follow-up was 4.5 ± 3.7years. Globe axial lengths were 18.6 ± 1.9 mm. Ten patients (91%) were initially left aphakic. Three patients (27%) later received a secondary intraocular lens (IOL), and 1 patient underwent an IOL exchange. Six out of 10 (60%) aphakic patients developed visual axis opacification. One aphakic patient required topical therapy for glaucoma. One additional patient developed neovascular glaucoma and retinal detachment. The eye was subsequently enucleated. Three patients underwent strabismus surgery. Two patients underwent pupilloplasty. Two patients (18.2%) had a final visual acuity better than 20/200.**

• CONCLUSION: **Eyes operated for PFV with elongated ciliary processes are unlikely to have a final visual acuity greater than 20/200 and many will need additional surgery. Postoperative visual axis opacification occurred in 60% and glaucoma developed in 18%. (Am J Ophthalmol 2019;198:25–29. © 2018 Elsevier Inc. All rights reserved.)**

PERSISTENT FETAL VASCULATURE (PFV) IS commonly associated with unilateral congenital cataracts. The pathogenesis of PFV is attributable to incomplete regression of fetal hyaloid vasculature, whose persistent remnants create a wide spectrum of pathology. The clinical subtypes can be divided into anterior, posterior, or combined depending on which structures are involved.[1] The constellation of clinical features was first named "persistent hyperplastic primary vitreous" by Reese in 1955.[2] Goldberg coined the term PFV in 1997 and suggested that it more accurately describes the anatomic and pathologic features of the disease. There are a limited number of studies that have been done regarding surgical outcomes of patients with anterior PFV.[3–12] Often, cataract surgery outcome studies exclude those with stretched or elongated ciliary processes owing to anticipated poor visual potential.

The Infant Aphakia Treatment Study (IATS) evaluated patients with mild anterior PFV, excluding eyes with elongated ciliary processes.[3] These mild PFV patients were found to have higher rates of adverse outcomes when compared to other types of unilateral cataracts.[3] Currently to date, to the best of our knowledge, there have been no published data detailing measurements, reoperation rates, and outcomes in patients operated for unilateral congenital cataract–associated and anterior PFV with elongated ciliary processes.[3,4,6,9,12,13]

METHODS

THIS WAS A RETROSPECTIVE CASE SERIES. CHARTS FROM A single institution, the Medical University of South Carolina, were queried with the diagnosis of unilateral congenital capsular cataract and congenital vitreous anomaly following cataract surgery with and without intraocular lens (IOL) implantation. This study was approved by the Institutional Review Board of the Medical University of South Carolina and conformed to the requirements of the United States Health Insurance Portability and Privacy Act. Inclusion criteria consisted of anterior PFV, defined as the presence of a visible persistent hyaloid artery or a retrolental membrane with or without visible vessels, and elongated ciliary processes but no retinal involvement. Our cases also all had unilateral cataract presenting prior to 7 months of age, at least 6 months of follow-up, and no systemic conditions associated with cataract. These were consecutive cases meeting inclusion criteria. Exclusion criteria consisted of active uveitis, prematurity (<36 gestational weeks), and posterior disease with tractional

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TABLE. Summary of Outcomes in Patients Operated for Unilateral Persistent Fetal Vasculature With Stretched Ciliary Processes

Case #	Age at Surgery (Months)	Axial Length (mm)	IOL Implantation	VAO (Months After Lensectomy)	Glaucoma	Strabismus	Pupilloplasty	Complications	Follow-up (Years)	Final VA
1	1.2	17.1	No	2.3	No		No	Vitreous hemorrhage	4.03	20/30
2	2.8		No		No	ET BMR rec 6 mm	No	Dense amblyopia	3.08	CF
3	3	21.11	Primary		No		No	Macular staphyloma, foveal hypoplasia	1.49	CSM
4	4.4	20.69	Secondary IOL (at age 1.3 years)	43.6	No	BMR rec 3.5 OU	No	Corectopia, synechiolysis, IOL exchange	3.55	20/800
5	2.5	17.38	No	3.7	Yes		Yes	NVG, RD, enucleated	13.92	N/A
6	1.8	16.47	No	1.4	No		No		3.41	FF
7	5.1	20.34	No		Yes	LET, LHT, LIO rec and LMR 5 mm rec	No		8.50	LP
8	1.2	18.45	No	4.6	No		No		3.38	LP
9	1.4	18.55	Secondary IOL (at age 4.37 years)	3.9	No		No		4.26	FF
10	1.8	15.79	No		No		Yes		0.73	FF
11	1.2	20.13	Secondary IOL (at age 2.9 years)		No		No		3.02	20/150

BMR rec = bilateral medial rectus recession; CF = count fingers; CSM = central steady maintained; ET = esotropia; FF = fix and follow; IOL = intraocular lens; LET = left esotropia; LHT = left hypertropia; LIO = left inferior oblique recession; LMR rec = left medial rectus recession; LP = light perception; N/A = not applicable; NVG = neovascular glaucoma; RD = retinal detachment; VA = visual acuity; VAO = visual axis opacification.

involvement of the retina, retina dysplasia, or optic nerve dysplasia. Diagnosis of PFV was made based on a combination of clinical, examination under anesthesia, operative note, and ultrasound findings (if performed). Preoperative ancillary testing included visual acuity using age-appropriate tests, intraocular pressure using Reichert Tono-Pen XL, Goldmann applanation, or iCare tonometer; corneal diameter measurement using calipers under microscope magnification; and A-scan ultrasonography. All study patients underwent lensectomy, posterior capsulectomy, and vitrectomy by a single surgeon. Patients also received amblyopia therapy, contact lens fittings, and spectacle correction as indicated.

RESULTS

A TOTAL OF 20 EYES OF 20 PATIENTS WERE OPERATED ON FOR unilateral anterior PFV at under 7 months of age, as determined from office examinations and examination under anesthesia. Of those, 9 patients did not have elongated ciliary processes. Eleven eyes of 11 patients having stretched ciliary processes met inclusion criteria and were analyzed. Six patients were male (55%) and 5 were female (45%). Racial demographics consist of 2 African Americans (18%) and 9 white patients (82%). Patients had a mean age at surgery 2.4 ± 1.4 months.

Mean corneal diameters were 10.7 ± 0.9 mm. Mean steep keratometry values were 54.9 ± 5.1 diopters (D). Globe axial lengths were 18.6 ± 1.9 mm. The majority of patients (10/11) were initially left aphakic. One patient received a primary IOL at 3 months of age. She did not develop visual axis opacification (VAO) or pseudophakic glaucoma. Three patients received a secondary IOL, and 1 patient underwent an IOL exchange.

Average follow-up duration was 4.5 ± 3.7 years (range 0.7-13.9 years). Six out of 10 aphakic eyes (60%) developed VAO. Two aphakic patients developed glaucoma: 1 developed a hyphema, neovascular glaucoma, and retinal detachment and was enucleated; the other patient received topical glaucoma therapy. Three patients also developed strabismus. Two of the patients developed esotropia and underwent bilateral medial rectus muscle recessions. A

third patient developed left esotropia and hypertropia and underwent multiple strabismus surgeries. Two patients underwent pupilloplasty. Eighteen percent (2/11) had a final visual acuity better than 20/200 (Table).

DISCUSSION

THIS STUDY IS UNIQUE IN THAT IT IS THE FIRST TO ANALYZE patients operated for congenital cataract and anterior PFV with stretched ciliary processes. Of previous studies analyzing PFV, the overall conclusion was that patients with PFV have poor visual outcomes and high rates of complications.[3,4,7,8]

Pollard[5] found that 17% of the 48 patients reviewed had a visual acuity 20/100 or better after surgery, contact lens fitting, and amblyopia therapy. Those with anterior PFV had the most successful visual outcomes. Solebo and associates conducted an observational population-based cohort study[4] and concluded that 24% of children with unilateral PFV achieved normal vision of the operative eye at 1 year following surgery. Alexandrakis and associates evaluated 42 eyes with unilateral PFV, 30 of which underwent vitreoretinal surgery for media opacity, vitreoretinal traction, or retinal detachment.[7] Fourteen of the surgical eyes (47%) achieved a final visual acuity of 20/400 or better at their last follow-up. Five aphakic children had visual acuity of 20/80 or better. Risk factors for a poor visual outcome (visual acuity of <20/400) were microphthalmia, preoperative retinal detachment, retinal or optic nerve abnormalities, or both, as also echoed in the study by Sisk and associates.[8] The IATS evaluated children with anterior PFV without stretched ciliary processes. Sixty-one percent (11 of 18) of these children achieved a final visual acuity of logMAR 1.0 (20/200 Snellen equivalent).[3,14,15]

In the present study, final visual acuity was better than 20/200 in 18.2% of patients with stretched processes. PFV with stretched ciliary processes had a mean age of 2.4 months, whereas the IATS patients with PFV had surgery at 2.0 months.[3] The IATS study operated on PFV patients at a slightly earlier age but excluded elongated ciliary processes, which may explain the better visual acuities. Patients with elongated ciliary processes in our study were unlikely to achieve final visual acuity better than 20/200. Sixty-one PFV cases without elongated ciliary processes enrolled in the IATS had a visual acuity of 20/200.[3]

In the IATS, 2 children with stretched ciliary processes that were not recognized prior to surgery were included in the outcome results but were not separately analyzed. Intraoperative complications in IATS were similar for the PFV and non-PFV groups. However, adverse events were reported in the first year after cataract surgery in 67% of eyes in the PFV group, compared to 46% in the group without PFV. Patients with unilateral PFV who received an IOL had similar adverse event rates compared to eyes left aphakic. However, unilateral cataract without PFV had fewer adverse events (20%) than eyes with PFV (55%) and this difference was statistically significant.[3]

In the IATS trial comparing contact lenses to IOL, 37% of patients in the contact lens group and 43% in the IOL group underwent strabismus surgery within 5 years.[15] David and associates[16] analyzed 90 eyes that underwent unilateral and bilateral cataract surgery, including 13.3% with PFV, and found that 46.7% of aphakic children and 58.7% of pseudophakic patients developed strabismus ($P = .283$). These patients had at least 1 year of follow-up, with average follow-up being 51 months. Eyes with PFV were not analyzed separately. In this study 27% (3 out of 11) developed strabismus that underwent surgery. Patients with PFV tend to have poor visual acuities, so it is interesting that the rates of strabismus in patients with elongated ciliary processes are low.[3] This is likely secondary to our inclusion of only patients who underwent strabismus surgery, whereas David and associates reported rates of strabismus whether or not they received strabismus surgery.[16] Seventeen percent of patients with PFV in the IATS developed glaucoma. Of the 18 PFV patients, 2 developed aphakic glaucoma and 1 developed pseudophakic glaucoma 1 year after cataract surgery.[3] In this study, 2 patients (18%) developed aphakic glaucoma.

The affected eyes of our patients with stretched ciliary processes had steep corneas. The mean steep keratometry was 54.9 ± 5.1 D, compared to a mean of 48.37 D reported for a cohort of children with unilateral cataracts aged 0-6 months.[17] There are sparse data on PFV, none of which comment on average steep keratometries in PFV.

Anteby and associates reported the largest series of IOL implantation in children with PFV, including stretched processes, anterior, posterior, and combined. Formation of VAO was found in 25.8% (8 of 31) of aphakic eyes and 31.0% (9 of 30) of pseudophakic eyes.[9] Vasavada and associates reported postoperative outcomes in eyes with anterior PFV, 4 of which had prominent ciliary processes but were not separately analyzed. Thirty-three patients were analyzed; 16 eyes received an IOL. Six patients developed VAO, 2 were aphakic, and 4 were pseudophakic. VAO developed at a mean follow-up of 7.03 ± 4.5 months. VAO developed in the majority between 1 and 6 months after cataract surgery.[11,12,18]

Six out of 10 (60%) aphakic patients developed VAO. This is clinically important. It also raises the question of how soon patients with stretched ciliary processes develop VAO. In the IATS, 1 aphakic patient (9%) with PFV developed VAO, compared to 3 PFV patients (43%) who received an IOL. VAO occurred in 60% of the eyes left aphakic, even after attempts were made to remove all of the vascular retrolental plaque and completely relieve the circumferential traction and ciliary process traction (Figure). In each of our cases, ciliary processes returned to their normal anatomic location during surgery once

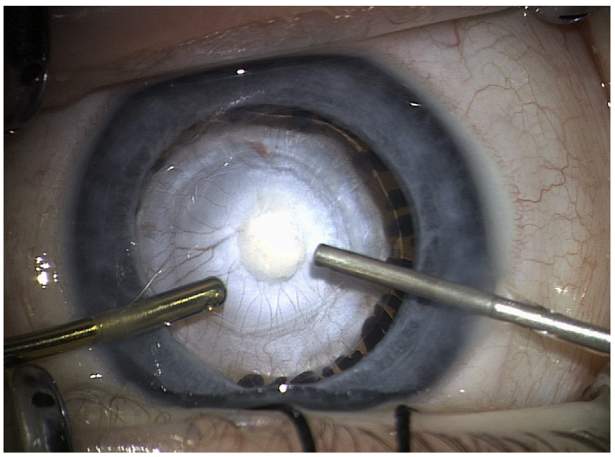

FIGURE. Intraoperative photograph of lensectomy in the right eye of an infant using bimanual vitrector and irrigation. Following removal of the lens nucleus and cortex, the vitrector was used to meticulously remove all the circumferential traction, revealing approximately 245 degrees of stretched ciliary processes and retrolental fibrovascular plaque.

the traction was reduced. This often required intraocular scissors to cut the plaque tissue between each stretched ciliary process prior to complete removal using the vitrector. It is possible that without this meticulous removal of traction forces, the VAO rate would have been even higher. It is also noteworthy that we had no retinal tears or early retinal detachments after surgery. The 1 eye that developed hyphema and neovascular glaucoma developed a retinal detachment but not in the perioperative period. We approach these surgeries from an anterior approach since the retina may be pulled forward with the stretched ciliary processes. In eyes with PFV and stretched processes, the pars plana approach may carry a higher risk of inadvertent retinal injury.

This study had several limitations. The study has a small sample size. Also, some of our operated patients were comanaged by a referring pediatric ophthalmologist and did not return to us for enough follow-up to be included in the study. Other limitations include variable compliance in amblyopia therapy and the retrospective nature of the study.

In conclusion, patients with PFV with stretched ciliary processes are unlikely to have visual acuity greater than 20/200, and complications, such as the need for reoperation, are common. Since infants presenting with PFV cataracts and stretched ciliary processes were excluded from the IATS, reporting them here helps surgeons to predict outcomes when initiating treatment for this group of children.

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