Outcomes of small gauge pars plicata vitrectomy for patients with persistent fetal vasculature: a report of 105 cases

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Abstract

- AIM: To evaluate the surgical outcomes in eyes with persistent fetal vasculatures (PFV) managed by small gauge pars plicata vitrectomy.
- METHODS: Consecutive patients with PFV treated by small gauge pars plicata vitrectomy at Beijing Tongren Eye Center between January 2010 and January 2013 were retrospectively reviewed.
- RESULTS: A total of 118 eyes of 105 patients with PFV were included and undergone small gauge pars plicata vitrectomy, of which 84 (71.2%) eyes had lensectomy and 16 (13.6%) eyes had lens aspiration and immediate intraocular lens implantation. The percentage of sutured scleral incision of 23 gauge vitrectomy (71.7%, 33/46) was higher than that of the 25 gauge vitrectomy (18.1%, 13/72). At last follow-up, visual acuity remained stable in 34 eyes (28.8%) and improved in 84 eyes (71.2%). Age at surgery (less than 2y), anterior type of PFV, and immediate IOL implantation were associated with postoperative improved visual acuity. Sixty five (55.1%) eyes had retinal detachment preoperatively, among which 33 (50.8%, 33/65) eyes had retinal reattachment or partial retinal reattachment.
- CONCLUSION: The results suggest that cases with PFV have a potential for developing good visual acuity after small gauge pars plicata vitrectomy with favorable anatomic outcomes and acceptable rate of serious surgical complications.
- KEYWORDS: vitrectomy; persistent fetal vasculature

INTRODUCTION

Persistent fetal vasculature (PFV) has been described as an sporadic and congenital syndrome of cataract and progressive retinal detachment in a microphthalmic eye with eventual phthisis if left untreated[1]. Cases can further be segregated on the basis of location and classified as anterior, posterior and combined types[2]. The heterogeneity of clinical presentations of PFV makes it challenging to manage surgically, however, it is possible to eliminate amblyogenic media opacities and relieve tractional forces using a closed-system intraocular approach[3-6]. More recent studies reported improved visual and anatomical outcomes, probably the result of earlier diagnosis, more careful case selection for surgery, improved surgical techniques, and aggressive postoperative amblyopia therapy[7-8].

Small gauge vitrectomy, a transconjunctival sutureless vitrectomy, was introduced in recent years[9-10], with its advantages including faster healing, increased patient comfort, and faster visual recovery[11-13]. In this retrospective study, we reported the surgical procedures and clinical outcomes of 105 cases of PFV treated with small gauge pars plicata vitrectomy.

SUBJECTS AND METHODS

Records of 105 patients with PFV treated by small gauge pars plicata vitrectomy between January 2010 and January 2013 at the Beijing Tongren Eye Centre were retrospectively reviewed. The study protocol was reviewed and approved by the Ethnic Committee of Beijing Tongren Hospital.

Anterior PFV was defined as fibrovascular membrane around the lens or attached to the posterior capsule involving the pupillary zone (Figure 1A), presence of retrolental vitreous membrane and stalk reaching the optic nerve without retinal traction or detachment. Posterior PFV was characterized by the presence of retrolental vitreous membrane and stalk reaching the optic nerve with retinal traction or detachment (Figure 1B). Cases in which there was posterior lens capsule opacification or fibrovascular membrane without pupillary zone involvement were also defined as posterior type. Combined type of PFV included both anterior and posterior PFV features.

Patients were all operated on by a single surgeon (Lu H). Three surgical approaches were applied to these patients: 1) vitrectomy (23 or 25 gauge) through the pars plicata for posterior PFV; 2) lensectomy and vitrectomy (L+V) through
pars plicata for anterior PFV of younger than 2 years old and for combined PFV with severe pathological changes of the macular, peripheral anterior lens capsule were kept intact with central anterior capsulotomy using vitrectomy probe in most eyes (Figure 2A, 2B); 3) vitrectomy through pars plicata combined with aspiration of lens cortex, intraocular lens (IOL) implantation through clear cornea incision (LA+V+IOL) and central posterior capsulotomy for eyes with anterior PFV in cases of older than 2y or combined PFV without severe pathological changes of the macular. The scleral incisions were sutured in eyes of scleral leakage (bubbles visible at the site of scleral incision after removal of trocar). Adjunctive surgical procedures included electric coagulation of the primary hyaloidal vasculature (21 eyes) and partial air-fluid exchange (87 eyes).

Cases were reviewed for demographic and ophthalmic data, including age at surgery, gender, laterality, chief complaints, PFV type, associated eye abnormalities, visual acuity (VA), intraocular pressure, surgical procedures performed, findings during the examinations, complications, and length of follow-up. Ancillary studies included A and/or B-scan or color doppler ultrasonography and Retcam fundus photography (Clarity Medical Systems, Pleasanton, California, USA). VA records included perception of light, fixation, and follow ability of light, presence of nystagmus, and determination of VA using the Teller card, E-chart, or Snellen acuity cards depending on the age and cooperation of the child. Postoperative VA were compared with preoperative values and classified as stable, improve, or deteriorate. When documentable VA data were not available, visual improvement was arbitrarily defined as a change in vision from an inability to an ability to fixate on light and unsteady/centrific fixation to central/steady fixation. To achieve standardization, documented VA was converted to logarithm of the minimum angle of resolution (logMAR) values. Pediatric ophthalmologists and optometrists would be involved if indicated for deprivational, anisometropic, and refractive amblyopia with cycloplegic correction, spectacles and aphakic contact lens fitting, or occlusion therapy. Amblyopia therapy consisted of patching the better eye for several waking hours every day depending on the age with placement of contact lens or spectacles on the involved eye. The lens power was checked every 3 to 6mo and replaced if necessary.

**Statistical Analysis** Statistical analysis was performed using SPSS version 17.0 (SPSS Inc., Chicago, IL, USA). Testing of normality was performed by the Kolmogorov-Smirnov method. Dichotomous variables were compared using Chi-square test.

**RESULTS**
A total of 118 eyes of 105 patients were identified with PFV and underwent vitrectomy during the study period, with a median follow-up of 17mo. Thirteen patients (12.4%) were bilateral. The median age at surgery was 22mo (range, 2mo-20y), with 59.0% (62/105) of the patients aged less than 2y, one female patients aged 20y complained with blurred vision because of vitreous hemorrhage caused by PFV. No systemic abnormalities were present in any of the patients. Associated eye abnormalities included microphthalmia (10 eyes), morning-glory syndrome (1 eye), macular hypoplasia and schisis (4 eyes), pupillary membrane (2 eyes), vitreous hemorrhage (2 eyes), and retinal detachment (65 eyes). Baseline characteristics of patients are summarized in Table 1. Surgical information of eyes with PFV is listed in Table 2. We performed L+V in 71.2% (84/118) of the eyes. Two eyes with anterior PFV and one eye with combined PFV received lens-sparing vitrectomy because of the eccentric pupillary zone opacification of the posterior lens capsule. Four eyes with combined PFV underwent LA+V+IOL for minor tractional retinal detachment around the optic nerve with comparatively normal macula.

We performed 23 gauge vitrectomy in the earlier period since we had no 25 gauge vitrectomy system then, and we found that the percentage of sutured scleral incision of 23 gauge vitrectomy (71.7%, 33/46) was higher than that of the 25 gauge vitrectomy. Peripheral anterior lens capsule were kept intact with central curvilinear capsulotomy using the vitrectomy probe in 59 of
84 eyes (70.2%) underwent L+V. Sixteen eyes received central curvilinear posterior capsulotomy after implantation of IOL. No eye with capsular tearing or IOL dislocation was found during the study period.

Only 10 eyes (8.5%) of 10 patients (aged≥4y) have preoperative objective records of corrected VA ranged from hand move to 0.7 logMAR, VA of 54 eyes (45.8%) were recorded as having no fix-and-follow, 25 eyes (21.2%) could follow light, 17 (14.4%) had central unsteady fixation, and 12 (10.2%) had central steady fixation on a light source. At last follow-up, VA remained stable in 34 eyes (28.8%) and improved in 84 eyes (71.2%), age at surgery (less than 2y), anterior type of PFV, and immediate IOL implantation were associated with postoperative improved VA (P=0.000, 0.000, 0.002, respectively; Table 3).

During the last follow-up visit, best corrected VA was able to be assessed in 57 (48.3%) eyes (32 with anterior PFV, 8 with posterior PFV and 17 with combined PFV). VA was better than 0.5 logMAR in 15.8% (9/57) eyes, 1.3-0.5 logMAR in 35.1% (20/57) eyes, and worse than 1.3 logMAR in 49.1% (28/57) eyes. One eye with combined PFV and 2 eyes with posterior PFV had VA records better than 0.5 logMAR at last visit. Sixty-five eyes (55.1%) had retinal detachment preoperatively (Figure 3A, 3C), among which 33 eyes (50.8%) had retinal reattachment (Figure 3B) or partial retinal reattachment after surgery (Figure 3D), and 17 eyes (26.2%) had macular reattachment at last visit, 94.1% eyes (96/102) regained clear visual axis after surgical repair. Postoperative anatomy and complications are listed in Table 4. Temporary higher intraocular pressure were controlled after medical treatment; pupillary membrane formation and secondary cataract required additional surgery and silicon oil tamponade had retinal attachment at last visit. No eyes developed endophthalmitis or required enucleation.

**DISCUSSION**

Over the years, the goal of early surgery recommended for PFV has been shifted from preventing future glaucoma and
hemorrhagic complications to achieving a comparatively better visual prognosis as knowledge of the disease and surgical instrumentation have advanced. Several reports have discussed the advantages and safety of small gauge vitrectomy in patients with proliferative diabetic retinopathy, retinal detachment and retinopathy of prematurity, but very few patients with PFV were included in these reports. Our focus in the current study was to determine the surgical management and short-term postoperative outcomes in eyes with PFV using small gauge pars plicata vitrectomy.

A total of 118 eyes of 105 patients were identified with PFV and undergone small gauge pars plicata vitrectomy during the study period, which 12.4% of the patients were bilateral, but the percentage found in this study cannot be generalized given the selection bias resulting from the referral setting. Small gauge vitrectomy is advantageous due to its less surgical trauma, faster healing, increased patient comfort and perhaps faster visual recovery [14-16], and it obviates the need for suturing of the scleral incisions, but postoperative hypotony is a common safety problem, especially for children. Lam et al. [17] reported that 40% of the eyes which undergone small gauge vitrectomy but without suturing of the scleral incisions had an intraocular pressure less than 5 mm Hg on postoperative day 1. Postoperative hypotony may be of a concern in children, as they are more likely to rub their eyes after surgery, additionally, in children with vasoproliferative disorders, hypotony in the immediate postoperative period may result in an increased risk of bleeding. In our series, all the patients had PFV that might be associated with active neovascular tissue and the sclerotomies were sutured for 38.9% eyes to prevent postoperative intraocular hemorrhage induced by postoperative hypotony, of which 71.7% eyes undergone 23 gauge vitrectomy had suturing of the scleral incision compared to 18.1% eyes undergone 25 gauge vitrectomy, but further study is needed to compare the exact percentage of leakage of scleral incisions for 23 or 25 gauge vitrectomy referring patient age, PFV type or surgical time. Although it takes more time to dissect the conjunctiva and/or suture at the close of the case, we believe that these modifications improve the safety of small gauge vitrectomy in these patients. Indeed, in our series, there was no hypotony observed after surgery.

There has been consensus about the surgical modality for patients with PFV, lens-sparing vitrectomy for predominately posterior PFV without axial lens opacification, and vitrectomy combined with lens surgery for anterior or combined PFV [18]. However, there has been some disagreement among authors over the preferred location of incisions for lens surgery in PFV. Authors who favor a limbal approach argue that the peripheral retina and the ciliary body can be dragged anteriorly and/or centrally, and pars plicata approach could make iatrogenic injury to these structures more likely [19-20]. Other authors advocate a pars plicata approach for less disturbance of the cornea and anterior chamber angle, improved ability to remove all of the lens cortex, and reduced anterior traction on posterior retina intraoperatively by earlier transection of the persistent hyaloid [21-24]. A study showed that the choice of limbal versus pars plicata incisions produced similar safety and visual outcomes when careful case selection and perioperative planning are employed [25]. We did lensectomy through pars plicata pars plana incisions using the vitrectomy probe for eyes without immediate IOL implantation, for less disturbance of the anterior chamber and postoperative inflammatory response.

<table>
<thead>
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<th>Variables</th>
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<tr>
<td>Preoperative</td>
<td>65/118 (55.1%)</td>
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<tr>
<td>Postoperative</td>
<td>32/118 (27.1%)</td>
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<td>Visual axis opacification</td>
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<tr>
<td>Preoperative</td>
<td>102/118 (86.4%)</td>
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<tr>
<td>Postoperative</td>
<td>6/118 (5.1%)</td>
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<td>Complications</td>
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<tr>
<td>High intraocular pressure</td>
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<tr>
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<td>Rhegmatogenous retinal detachment</td>
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Figure 3 Fundus photographs of patients preoperatively and 6mo postoperatively A: Two-year-old girl with anterior PFV presented with membranous vitreous stalk (white arrow) starting from the optic nerve with local tractional retinal detachment (red arrow) without macular involvement; B: Retinal reattachment and normal foveal reflex 6mo postoperatively; C: Seven-year-old girl with posterior PFV presented with membranous vitreous stalk starting from the optic nerve (white arrow), tractional retinal detachment (red arrow) involving the macular; D: Relieved retinal and macular detachment 6mo postoperatively.

Table 4 Postoperative anatomy and complications of eyes with PFV

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and less corneal distortion that reduces visibility during vitreous surgery; and for eyes that needed immediate IOL implantation, cortical lens were aspirated through clear corneal incision in order to safely implant the IOL into the capsule bag. VA outcome in patients with PFV seems to be affected by the type and extent of PFV, age at surgery, and postoperative refractive status. In our study, VA remained stable in 34 eyes (28.8%) and improved in 84 eyes (71.2%) at last visit, which is similar to that of Vasavada et al’s study of 33 patients with anterior PFV. Kuhl-Hattenbach et al. reported the result of visual outcome in 23 patients with PFV who underwent surgery, in which 34.7% had a VA of 6/60 or better and most of the patients were classified as anterior PFV. Others, however, reported a more modest success rate in surgically managed cases with PFV of the combined type. In this current report, we found that postoperative improved VA was mostly seen in patients with anterior PFV. However, eyes with posterior or combined PFV could have a potential to gain better VA if the macula is relatively normal. Surgery at an early age is also reported to be a predictor for better visual outcomes, and we also reported in this study that age at surgery (less than 2y) was associated with improved postoperative VA.

Authors who preferred immediate IOL implantation argued that these eyes have unilateral cataract and unilateral aphakia can induce anisometropia, and IOL implantation in such eyes would provide a stable and consistent optical focus. Others, however, reported higher postoperative complications such as secondary glaucoma or pupil membrane formation after immediate IOL implantation for PFV eyes. We performed immediate IOL implantation in 16 eyes for patients older than 2y and 4 eyes had VA better than 0.5 logMAR whereas 1 eye complicated with pupillary membrane formation and no eye ended with glaucoma at last visit. But further study is still needed to document the optimum timing of IOL implantation for cases with PFV.

Anatomically, 94.1% (96/102) eyes in this study had regained clear visual axis after surgical repair, which is similar to Sisk et al’s study; and we attribute low rate of postoperative visual axis obscuration (5.1%, 6/118) to using of vitrectomy probe to do central curvilinear posterior or anterior capsulotomy. Fifty-eight percent (33/56) of the eyes had relieved retinal detachment at last visit, of which 26.2% (17/65) had macular reattachment, and further study using optical coherence tomography is still needed to document the anatomic postoperative macular changes.

Potential limitations of the study include relatively short time of follow-up, limited data of objective VA, and its retrospective design. However, favorable anatomic and preliminary postoperative visual outcomes had been achieved with low rate of serious postoperative complications, demonstrating the feasibility of using this approach in patients with PFV. Long-term follow-up of VA results and final anatomical outcomes would still be needed in the future.

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Pars plicata vitrectomy for persistent fetal vasculature