Surgical Results of Persistent Fetal Vasculature

Li-Sheng Cheng, MD; Hsi-Kung Kuo, MD; Sue-Ann Lin, MD; Ming-Lun Kuo, MD

Background: To evaluate the surgical results of patients with persistent fetal vasculature (PFV), also known as persistent hyperplastic primary vitreous (PHPV).

Methods: From 1991 to 2001, a retrospective, noncomparative study of 7 eyes of 7 patients diagnosed with PFV was conducted. In each case, type of anterior and/or posterior PHPV findings, preoperative testing, surgical procedures, and visual outcomes were obtained from the records. Follow-up ranged from 12 to 43 months, with a mean of 15 months.

Results: Of the 7 eyes, one (14%) had strictly anterior PFV, 1 (14%) had strictly posterior PFV, and 5 (71%) had components of both anterior and posterior disease. Initial lens aspiration only was performed in 3 (43%) eyes. Initial vitrectomy only was performed in 1 eye (14%). Initial lensectomy and vitrectomy was performed in 3 (42.8%) eyes. The reoperation rate was 43% for membrane reproliferation, glaucoma, vitreous hemorrhage, and retinal detachment. Final best-corrected visual acuity ranged from light perception only to 20/70 on the Snellen chart .

Conclusions: Functional vision is possible in selected patients. However, poor final visual outcome despite adequate anatomic success were noted in this study. The poor outcomes might have been due to patients delaying vitrectomy until retinal detachment developed or that the patients had poor compliance with postoperative ocular rehabilitation.

(Chang Gung Med J 2004;27:602-8)

Key words: persistent fetal vasculature (PFV), persistent hyperplastic primary vitreous (PHPV).

Persistent fetal vasculature (PFV), also known as persistent hyperplastic primary vitreous (PHPV) was first defined histopathologically as a failure of embryologic regression by Reese in 1949.[1] In 1955, these phenomena were further specified in Reese's famous Jackson Memorial Lecture to describe an idiopathic sporadic congenital syndrome and usually isolated monocular findings.[2] Since then, it has been subclassified into anterior, posterior and combined forms, although the etiology seems to be identical.[3]

PFV usually presents in an otherwise healthy newborn, and the clinical ocular manifestations include microphthalmia, progressive cataract, retrolental fibrovascular tissue, persistent hyaloid vessel remnants, and tunica vasculosa lentis remnants. The degree of severity from this failure of the persistent fetal vasculature to regress may be as mild as a Mittendorf dot and Bergmeister's papilla, for which surgical intervention is not necessary, or may be so severe as to require enucleation for secondary com-

From the Department of Ophthalmology, Chang Gung Memorial Hospital, Kaohsiung.
Received: Apr. 9, 2004; Accepted: Jun. 7, 2004
Address for reprints: Dr. Ming-Lun Kuo, Department of Ophthalmology, Chang Gung Memorial Hospital. 123, Dabi Road, Niaosung Shiang, Kaohsiung, Taiwan 833, R.O.C. Tel.: 886-7-7317123 ext. 2801; Fax: 886-7-7317123 ext. 2831; E-mail: kufer@pchome.com.tw

plications of hemorrhage, glaucoma, and phthisis.[4,5] Surgical intervention is recommended for selected cases to achieve functional visual acuity or preservation of the globe. Visual outcomes vary considerably depending on the severity of PFV. We retrospectively reviewed 7 eyes of 7 patients with PFV. Preoperative testing, surgical intervention, and visual and anatomic outcomes were investigated.

METHODS

We performed a retrospective medical record review of all patients with the diagnosis of PFV who underwent vitreoretinal surgical rehabilitation after appropriate informed consent in the Department of Ophthalmology, Chang Gung Memorial Hospital, Kaohsiung, from June 1991 to June 2001. We collected the following information from each record when available: patient name, date of birth, medical history including any history of prematurity, sex, age at diagnosis, type of PFV, initial visual acuity, surgical procedure performed, age at time of surgical procedure, intraoperative and postoperative complications, location and number of sclerotomy sites, postoperative appearance, type of aphakic rehabilitation, amount of amblyopic patching therapy, compliance with amblyopia therapy, final visual acuity, and length of follow-up.

A diagnosis of anterior PFV was made if two or more of the following features were described: microphthalmia, shallow anterior chamber, elongated ciliary processes, cataract, or retrolental opacity or persistent hyaloid artery. A diagnosis of posterior PFV was made if a retinal fold was detected emanating from the disc. Patients presenting with both anterior and posterior features were diagnosed with the combined form of PFV. Indications for surgery included media opacity (cataract), vitreoretinal traction, and retinal detachment.

RESULTS

Seven eyes of 7 patients were managed surgically (Table 1). Of the 7 patients, 5 were male and 2 were female. The right eye was involved in 4 patients, and the left eye, in 3 patients. All patients had unilateral involvement. The presenting age ranged from 1 month to 68 months, with an average of 7 months. Presenting complaints included 6patients with leukocoria, 4 with strabismus, 3 with microphthalmos, and 1 with nystagmus. Neither systemic abnormalities nor prematurity was documented.

One of the patients (patient 4, 1 eye) had strictly posterior PFV and was treated with primary vitrectomy and dissection of the PFV. Of the remaining 6 eyes, 1 (patient 5) had strictly anterior PFV and 5 had components of both anterior and posterior disease. Four patients had optic nerve hypoplasia or dysplasia.

Three patients had primary vitrectomy procedures performed (2 via pars plana and 1 via limbus). In 2 of these 3 patients trans pars plana lensectomy was performed at the same time as vitrectomy. In two of seven patients the diagnosis of PFV was made at the same time as surgery for unilateral congenital cataracts, and the operation was terminated after lens removal only. In 5 of 7 patients, tractional retinal detachment was not detected on initial examination but was diagnosed preoperatively at a follow-up visit. The 5 patients did not undergo vitrectomy initially, until retinal detachment was detected.

The location of vitrectomy port varied according to the age of the patient and surgeons' preferences, from limbus to 4 mm posterior to limbus. One patient had a posterior chamber intraocular lens placed at the time of surgery and finally achieved a best corrected visual acuity of 20/70 on the Snellen chart.

Surgical complications of posterior vitrectomy included 2 large retinal tears in 1 patient and intraoperative retinal breaks in2 patients. The patient suffered from large retinal tears intraoperatively had a massive tractional retinal detachment preoperatively, and after the lens was removed, the retina inserted directly into pars plicata with no visible pars plana. The instrument placed through the inferotemporal sclerotomy (3 mm posterior to limbus) caused enough traction on the nearby retina to cause a retinal tear. Posterior capsular opacity developed after lens aspiration or lensectomy in 4 patients. One patient (patient 1) underwent a scleral buckling procedure for postoperative retinal detachment in which the visual acuity was undetermined (the patient was

Table 1. Patient Information

Number	1	2	3	4	5	6	7
Gender	M	M	M	M	F	M	F
Age at diagnosis	7 mo	58 mo	5 mo	33 mo	47 mo	1MO	68MO
Prematurity	Nil	Nil	Nil	Nil	Nil	Nil	Nil
Type of PFV	Anterior OD	Posterior OD	Combined OS	Combined OD	Combined OS	Combined OS	Combined OD
Initial presentation	Leukocoria	Leukocoria	Leukocoria	Leukocoria	Leukocoria	Leukocoria	Leukocoria
Other ocular associations	Cataract, microphthalmia	Nystagmus, strabismus, optic disc hypoplasia	Cataract, microphthalmia	Cataract, microphthalmia, strabismus	Cataract, strabismus	Cataract	Cataract, strabismus
Initial VA	Undetermined	0.04	Undetermined	Undetermined	0.01	Undetermined	HM 1M
Final BCVA	CF 50 cm	CF 60 cm	Undetermined	CF 40 cm	20/70	Undetermined	LP
Interval between diagnosis and surgery	5 days	12 days	2 mo	1 mo	9 mo	1 mo	48 mo
Surgical procedures	LA at 7 mo TLPC at 16 mo	TPPV at 58 mo	LA at 3 mo TPPV at 5 mo ESB+TPPV at 10 mo	LA at 34 mo TPPV at 47 mo	TPPV+TPPL +PCIOL at 56 mo	LA at 2 mo TPPV at 4 mo	TPPV+TPPL at 9 years
Intra-operative break	No	No	Yes	No	Yes	No	2 large retinal tears
Complications	After first surgery PC opacity and pupillary membrane and TRD detected	TRD detected preoperatively	After first surgery, PC opacity, pupillary membrane and TRD detected	After first surgery, PC opacity and TRD detected	TRD detected preoperatively	After first surgery, pupillary membrane. 2nd glaucoma, VH and TRD detected	TRD detected preoperatively. PC opacity, rubiosis iridis and cyclic membrane noted postoperatively
Type of amblyopic therapy	Occlusion	Occlusion	Occlusion	None	IOL	None	None
Length of follow-up (mo)	36	15	12	43	12	29	11
Macula involvement	No	Yes	No	Yes	No	Yes	Yes
Optic stalk	No	Yes	No	Yes	No	Yes	Yes
Final anatomic status	Aphakia Attached retina	Attached retina Pseudophakia	Aphakia Attached retina	Attached retina Aphakia	Aphakia Total RD with PVR	Attached retina Aphakia	Aphakia Retina tear with PVR

Abbreviations: mo: months; PFV: persistent fetal vasculature; BCVA: best correct visual acuity; VA: visual acuity; LP: light perception; NLP: no light perception; CF: counting fingers; LA: lens aspiration; TPPV: trans pars plana vitrectomy; TPPL: trans pars plana lensectomy; PCIOL: posterior chamber intraocular lens; ESB: encircling scleral buckle; TLPC: trans limbal posterior capsulectomy; PC: posterior capsule; IOL: intraocular lens; RD: retinal detachment; TRD: traction retinal detachment; VH: vitreous hemorrhage; PVR: proliferative vitreoretinopathy.

too young to determine visual acuity). Four patients had secondary pupillary membranes after surgical intervention. In 1 of these 4 patients, increased intraocular pressure was noted postoperatively. On further follow-up, a retinal detachment with proliferative vitreoretinopathy, judged inoperable was noted in 2 patients (patients 6 and 7).

The differential diagnosis of PFV includes congenital cataract, retinopathy of prematurity (ROP), Norrie's disease and retinoblastoma. PFV can be distinguished from an uncomplicated congenital cataract by the presence of a fibrovascular stalk and other features such as elongated ciliary processes and a shallow anterior chamber. PFV can be differentiated from ROP by its unilateral occurrence in full-term healthy infants. Norrie's disease can be differentiated from PFV by its X-linked inheritance, associated systemic manifestations, and bilaterality.[16] Retinoblastoma is commonly distinguished from PFV by the lack of microphthalmia and cataract.

Indications for surgical intervention in PFV have changed over the years as knowledge of the disease and surgical instrumentation advanced. A decision to operate depends on a variety of the factors such as the severity of the disease, the patient's age and the visual prognosis. Surgical treatment may not be effective in patients with severe microphthalmia or advanced posterior PFV, such as marked foveal hypoplasia, dysplasia or retinal detachment. These patients (as well as patients with only mild clinical PFV findings, such as a partial vitreous stalk) may need only to be observed.

When surgical intervetion described by Reese in 1955, a two-staged approach was proposed, consisting of needling the lens, followed by later discission of the retrolental membranes. Others also recommended variations of the two-staged technique, including open-sky dissection to minimize hemorrhagic complications.

Vitreoretinal surgery techniques have advanced over the years. Multiport vitrectomy, closed system and single-staged procedures became available and have many advantages to previous two-staged techniques.

In 1991, Pollard[17] reported that 17% of patients achieved a final visual acuity of 20/100 or better after surgery, contact lens fitting, and amblyopia therapy. In 1998, Mittra et al.[18] reported the visual outcomes of 14 patients with combined anterior and posterior PFV after lensectomy and vitrectomy; 10 eyes (71%) achieved a visual acuity of 20/300 or better and eight eyes (57%) achieved a final visual acuity of 20/100 or better. In 1999, Dass and Tress[19] reported the outcomes of PFV in 35 eyes of 27 patients; 19 of the eyes had components of both anterior and posterior disease. Initial lensectomy and vitrectomy were performed in 24 eyes (68%). Six eyes (17%) achieved a final visual acuity of 20/800 or better. In 2000, George et al reported the outcomes of PFV in 42 eyes of 42 patients; 35 of these eyes had components of anterior and posterior disease.[20] Initial lensectomy and vitrectomy were performed in 26 eyes, 12 eyes (47%) achieved a final visual outcome of 20/400 or better.

In our study, microphthalmia (patients 1, 3 and 4) and preoperative retinal detachment (patients 1-5 and 7) or retinal or optic nerve abnormalities (patients 2, 4, 6, and 7), such as hypoplasia, folds or an indistinct macula with hypopigmentation, were associated with poor visual outcomes. Although these associations did not reach statistical significance due to the small sample size, we believe they are clinically relevant. These associated poor risk factors were also published by George et al in 2000.[20]

Comparison with different series mentioned above, relatively poor visual outcome seems noted in our study. Aggressive contact lens fitting with amblyopic therapy for postoperatively aphakic status is the important method we should consider.

The best visual result happened in the patient 5 is owing to the less extent of macula involvement preoperatively and appropriate intraocular lens implantation after lens aspiration procedure. Beside, good compliance with amblyopic therapy is also noticed to the patient at the regular follow-up visit.

Prior to surgery, the posterior segment of the eye must be adequately evaluated. When there is a limited or absent view of the fundus, echography can provide information concerning the axial length, status of the lens, presence of a vitreous stalk or opacities, and presence of retinal detachment or tumor. Nonetheless, ultrasonography may not demonstrate subtle dysplastic nerve and macular changes.

Definite abnormal posterior segment findings often preclude visual rehabilitation and permit surgical decision making to be based on considerations related to the preservation of the globe and the comfort of the patient.

The one major surgical complication noted during our study was a retinal tear (patient 7) that resulted partially from the posterior placement of a sclerotomy incision in a patient with a preoperative tractional retinal detachment. Anterior placement of incisions to avoid this type of complication has previously been suggested for small eyes that frequently have a poorly developed pars plana region. Our incision site placement varied according to surgeon's preference and the age of the patient.

The other surgical complication noted after lens aspiration was posterior capsular opacity (4/4 eyes, 100%).

Parents play a crucial role in the success or failure of visual development therapy. We find it necessary to explain in detail, more than once, that the surgery is only the first step in a program of vision development that will extend for years and require frequent, extended clinic visits, multiple lens changes, and long-term occlusion therapy. Parents must, therefore, understand the process, be in agreement, be dependable and importantly, be compliant with the therapy.

In summary, we found that functional vision is possible in selected patients. However, poor visual outcomes after surgical intervention were encountered despite good anatomic appearance of the eye. Our experience shows that early diagnosis and surgical intervention, followed by aggressive visual rehabilitation with adequate parental compliance cannot be overemphasized. Otherwise, our experience shows that patients with PFV may be at risk for development of glaucoma, retinal detachment and vitreous hemorrhage after surgical repair. Therefore, continued long-term follow-up of these patients is important to ensure proper treatment of complications if they occur.

REFERENCES

1. Reese AB. Persistence and hyperplasia of primary vitreous; retrolental fibroplasia- 2entities. Arch Ophthalmol 1949;41:527-52.
2. Reese AB. Persistent hyperplastic primary vitreous. The Jackson Memorial Lecture. Am J Ophthalmol 1955;40: 317-31.
3. Pruett RC. The pleomorphism and complications of posterior hyperplastic primary vitreous. Am J Ophthalmol 1975;80:625-9.
4. Haddad R, Font RL. Reeser F. Persistent hyperplastic primary vitreous. A clinicopathologic study of 62 cases and review of the literature. Surv Ophthalmol 1978;23:123-34.
5. Alward WL, Krasnow MA, Keech RV.l. Persistent hyperplastic primary vitreous with glaucoma presenting in infancy. Arch Ophthalmol 1991;109:1063-4.
6. Goldberg MF. Persistent fetal vasculecture (PFV): an integrated interpretation of signs and symptoms associated with persistent hyperplastic primary vitreous (PHPV). LIV Edward Jackson Memorial Lecture. review. Am J Ophthalmol 1997;124:587-626.
7. Brown GC, Gonder J, Levin A. Persistence of the primary vitreous in association with the morning glory disc anomaly. J Pediatr Ophthalmol Strabismus 1984;21:5-7.
8. Joseph N, Iver M. Persistent hyperplastic primary vitreous at the optic nerve head. Am J Ophthalmol 1972;73:580-3.
9. Levine RA, Gray DL, Could N, Pergament E, Stillerman ML. Warburg syndrome. Ophthalmology 1983;90:1600-3.
10. Traboulsi EI, Faris BM, Der Kaloustian VM. Persistent hyperplastic primary vitreous and recessive oculo-dento-osseous dysplasia. Am J Med Genet 1986; 24:95-100.
11. Frydman M, Kauschansky A, Leshem I, Savir H. Oculoplato-cerebral dwarfism: a new syndrome. Clin Genet 1985;27:414-9.
12. Marsh man WE, Jan JE, Lyons CJ. Neurologic abnormalities associated with persistent hyperplastic primary vitreous. Can J Ophthalmol 1999;34:17-22.
13. Mafee MF, Goldberg MF. Persistent hyperplastic primary vitreous: role of computed tomography and magnetic resonance. Radiol Clin North Am 1987;25:683-92.
14. Mafee MF, Goldberg MF, Valsassori GE, Capek V. Computed tomography in the evaluation of patients with persistent hyperplastic primary vitreous (PHPV). Radiology 1982;145:713-7.
15. Kase SC, Jenkins JJ. Meyer D, Fontanesi J, Pratt CB. Persistent hyperplastic primary vitreous of the eye: imaging findings with pathologic correlation. AJR Am J Radiology 1994;16:437-40.
16. Ravia Y, Braier-Goldstein O, Katznelson M, Erlich S Barkai G, Goodman B. X-linked recessive primary retinal dysplasia is linked to the Norrie disease locus. Hum Molec Genet 1993;2:1295-7.
17. Pollard ZP. Results of treatment of persistent hyperplastic

primary vitreous. Ophthalmic Surg 1991;22:48-52.
18. Mittra RA. Huynh LT. Ruttum MS. Visual outcomes following lensectomy and vitrectomy for combined anterior and posterior persistent hyperplastic primary vitreous. Arch Ophthalmol 1998;116:1190-4.
19. Dass AB, Trese MT. Surgical results of persistent hyperplastic primary vitreous. Ophthalmolgy 1999;106:280-4.
20. George AL, Ingrid U.Scott, Harr WF. Visual acuity outcomes with and without surgery in patients with persistent fetal vasculature. Ophthalmolgy 2000;107:1068-72.

殘留性胎兒血管組織的手術結果

鄭力升 郭錫恭 林淑妍 郭明倫

背 景： 評估患有殘留性胎兒血管組織的病患接受手術治療後的結果。

方 法： 迴顧及分析高雄長庚醫院於1991年到2001年10年間內，對於患有殘留性胎兒血管組織的病患接受手術治療後的結果，特別針對這些病患治療前後的視力，臨床表現及眼球的併發症加以探討。

結 果： 共有7位病人，7隻眼睛接受手術治療。其中，1位爲完全前位型態、1位爲完全後位型態、5位爲混合型態。所有的病患皆爲單一患眼。初期治療包括水晶體吸除者有3隻眼睛、僅接受玻璃體切除術者僅有1隻眼睛、合併水晶體及玻璃體切除術者有3位。因增生膜、或青光眼、或玻璃體出血、或視網膜剝離而再次手術率爲42.8%，治療術後最佳矯正視力從20/70到僅餘光覺。

結 論： 在這次回顧中發現，針對特定的病患手術後可以達到具有功能性的視力。然而，大多數的病患儘管術後達到解剖學上結構成功，然而視力仍然不好。推論原因可能爲手術治療的介入時間點延遲、及術後病人接受視力復健的耐受性不佳，所以早期的診斷、治療以及手術後積極的視力復建是此類患者可以達到良好視覺功能的關鍵。
(長庚醫誌 2004;27:602-8)

關鍵字： 殘留性胎兒血管組織，殘留性增殖型原始玻璃體。

長庚紀念醫院　高雄院區　眼科
受文日期：民國93年4月9日；接受刊載：民國93年6月7日。
索取抽印本處：郭明倫醫師，長庚紀念醫院　眼科。高雄縣833鳥松鄉大埤路123號。Tel.: (07)7317123轉2801; Fax: (07) 7317123轉2831; E-mail: kufer@pchome.com.tw