

Surgical Technique

Ex-PRESS Shunt for Choroidal Fluid Drainage in Uveal Effusion Syndrome Type 2 A Potentially Novel Technique

Juan B. Yopez, MD; J. Fernando Arevalo, MD

Uveal effusion syndrome (UES) was reported by Schepens and Brockhurst in 1963,¹ and later described as a nanophthalmic disorder with a scleral abnormality.² Gass³ described idiopathic UES and hypothesized that the cause was a congenital anomaly of the sclera



Video at
jamaophthalmology.com



Supplemental content at
jamaophthalmology.com

and vortex veins that was related to aging, hormonal changes, or impairment of the permeability of the sclera. Using histological methods, Trelstad et al⁴ reported the scleral abnormalities in UES. In addition, Forrester et al⁵ observed migra-

tion of retinal pigment epithelial cells into the subretinal space.

For the treatment of UES, Brockhurst⁶ described good surgical results with decompression of the vortex veins by scleral resection with a sclerotomy. Gass³ reported an effective surgical procedure with a sclerectomy and sclerostomy without decompression of the vortex veins owing to the difficulty of isolating the vortex veins. Uyama et al⁷ treated 19 eyes of 16 patients with UES by making a two-third-thickness scleral flap and performing a scleral excision to expose the underlying choroid.

The Ex-PRESS shunt (Alcon Laboratories) glaucoma implant is a small (2- to 3-mm-long and 0.4-mm-diameter tube) stainless steel nonvalved device that was designed to lower intraocular pressure by shunting aqueous humor from the anterior chamber into the subconjunctival space under a partial-thickness scleral flap space adjacent to the limbus.⁸

In the current study, we report the surgical outcomes of the use of an Ex-PRESS shunt device for choroidal drainage without vortex vein decompression for the management of 3 eyes with UES.

Procedures

A retrospective medical record review was performed of 2 consecutive patients (3 eyes) with UES without nanophthalmos who underwent Ex-PRESS shunt implantation at the Clinica de Ojos de Mara-

caibo, Maracaibo, Venezuela, from March 2012 through March 2014 (Table). Institutional review board/ethics committee approval was obtained from Clinica de Ojos de Maracaibo and the patients signed a standard informed consent. This study adhered to the tenets of the Declaration of Helsinki for research involving human participants.

Surgical Technique

After peribulbar anesthesia and standard asepsis and antisepsis in the operating room, a conjunctival incision was performed, bare sclera was exposed in the quadrant with more accumulation of fluid according to ultrasonography, and an oblique sclerotomy was performed with a 25-gauge needle 13 mm posterior to the limbus (to decrease the risk for infection), at which time, a small amount of suprachoroidal fluid was spontaneously drained. The Ex-PRESS shunt (model P-50) was then inserted obliquely in the sclerotomy to facilitate the continuous drainage of the suprachoroidal fluid (eFigure 1 in the Supplement). Balanced salt solution was injected into the anterior chamber with a 30-gauge needle to reconstitute intraocular pressure. The conjunctiva was closed with 7-0 polyglactin 910 suture and subconjunctival antibiotics were injected.

Report of Patients

Patient 1

A woman in her 60s presented to the emergency department with a 15-day history of a "shadow" that affected the upper part of the visual field in her left eye. She had undergone cataract surgery in both eyes 3 years prior to presentation. Her best-corrected visual acuity (BCVA) without any refractive error was 20/20 OD and 20/200 OS. Fundus examination revealed choroidal detachment and shifting subretinal fluid underneath the exudative retinal detachment of the left eye. Ultrasonography was performed demonstrating thick sclera (1.5 mm), choroidal detachment, and exudative retinal detachment (eFigure 2 in the Supplement). The axial length was 22 mm in the right

Table. Demographic and Baseline Characteristics, Functional and Anatomic Results, and Complications in Eyes With Ex-PRESS Shunt for Choroidal Drainage in UES

Eye No./Sex/Age, y	Diagnosis	UES Duration, d	Previous Surgery	BCVA		CD and SRF		Total FU, mo	Complications
				Baseline	Last FU	Baseline	Last FU		
1/F/65	UES	14	Cataract surgery	20/200	20/30	Yes	None	24	None
2/F/55	UES	21	None	20/400	20/40	Yes	None	24	None
3/F/55	UES	14	None	20/200	20/40	Yes	None	12	None

Abbreviations: BCVA, best-corrected visual acuity; CD, choroidal detachment; F, female; FU, follow-up; SRF, subretinal fluid; UES, uveal effusion syndrome.

eye and 22.5 mm in the left eye. She was diagnosed as having UES in the left eye. Despite treatment with steroids (oral prednisolone, 50 mg/d), the retinal findings and BCVA did not improve. Ex-PRESS shunt drainage surgery was performed in the inferotemporal quadrant (eFigure 3 in the Supplement and Video) without vortex vein decompression without complications. Two days postoperatively, a flat retina was seen on fundus examination. The retina remained attached during the 24-month follow-up, and an improved BCVA of 20/30 was achieved. We did not observe subretinal fluid or choroidal effusion by ultrasonography after surgery at 24 months (eFigure 4 in the Supplement).

Patient 2

A woman in her 50s was referred owing to decreased visual acuity in her left eye for 3 weeks' duration. She had been wearing glasses for hyperopia since childhood. Her BCVA was 20/25 OD and 20/400 OS, and her spherical equivalents were +1 diopters (D) and +1.50 D in the left and right eyes, respectively. Fundus examination revealed uveal effusion involving the macula in the left eye. Axial length was 21 mm in the left eye and 22 mm in the right eye on A-scan ultrasonography. B-scan ultrasonography demonstrated thick sclera (1.6 mm), an exudative choroidal, and retinal detachment.

An Ex-PRESS shunt was implanted in the inferotemporal sclera quadrant in the left eye. One week postoperatively, a flat retina was seen on fundus examination and BCVA improved to 20/40. The retina remained attached during the 24 months of follow-up. Six months later, she presented with impaired visual acuity in her right eye. Her BCVA was 20/200 and the intraocular pressure was 16 mm Hg in the right eye and 18 mm Hg in the left eye. Fundus examination demonstrated supraciliary effusion and exudative retinal detachment. Ex-PRESS shunt implantation was performed in the inferotemporal quadrant. The subretinal fluid and the choroidal effusion disappeared 48 hours after surgery and BCVA improved to 20/30. The retina and choroid have remained attached during 12 months of follow-up.

Ex-PRESS Shunt for UES

Uveal effusion syndrome is difficult to manage and often follows a relapsing course. Our 3 cases (eyes) can be classified as type 2 according to the classification by Uyama and colleagues⁷ because they were not associated with nanophthalmos or hyperopia but had a thick sclera. Although Brockhurst⁶ described good surgical results with decompression of vortex veins, other investigators³ have questioned the usefulness of this technique because vortex veins are difficult to isolate. Uyama et al⁷ reported that sclerectomy with a small sclerostomy under the scleral flap could be effective in both type 1 and type 2 UES because the abnormal sclera and increased trans-scleral outflow of intraocular fluid are thought to be the main causes of these disorders. However, this technique was not effective in type 3 UES, which develops in non-nanophthalmic eyes with normal eyeball size and normal scleral thickness.

We believe that isolation of the vortex veins is complicated and that vortex vein decompression is technically difficult to perform without complications such as vein rupture. Therefore, we elected to perform the Ex-PRESS shunt technique instead of full-thickness sclerotomy or scleral windows because it seemed likely to have a sufficient effect for resolving UES. We suspect that our technique may work by facilitating a constant drainage of uveal exudation from the suprachoroidal space to the subconjunctival space.

The actual surgical technique and implantation of the device are very simple. The tip of the Ex-PRESS shunt is not sharp and it is inserted obliquely so the chance to cause retinal damage is unlikely. In addition, considering the reported persistent and recurrence rates (23%-50%) for type 2 UES,^{7,9,10} it seems that the cost may be worthwhile. Compared with other surgical procedures, our Ex-PRESS shunt technique could simplify the operative procedure, is less invasive, and reduces surgical time.

Limitations of our report include that we cannot know the long-term benefits and risks from 3 eyes and limited follow-up. At this point, we know that the procedure is feasible and safe for a limited time.

ARTICLE INFORMATION

Author Affiliations: Department of Vitreoretinal Surgery, Clinica de Ojos, Maracaibo, Venezuela (Yepez); Division of Retina, Wilmer Eye Institute, Johns Hopkins University School of Medicine, Baltimore, Maryland (Arevalo); Division of Vitreoretinal, King Khaled Eye Specialist Hospital, Riyadh, Kingdom of Saudi Arabia (Arevalo).

Corresponding Author: J. Fernando Arevalo, MD, Vitreoretinal Division, The King Khaled Eye Specialist Hospital, Al-Oruba St, PO Box 7191, Riyadh 11462, Kingdom of Saudi Arabia (arevalojf@jhmi.edu).

Submitted for Publication: August 17, 2014; final revision received November 2, 2014; accepted November 12, 2014.

Published Online: February 12, 2015.
doi:10.1001/jamaophthalmol.2014.5913.

Conflict of Interest Disclosures: All authors have completed and submitted the ICMJE Form for Disclosure of Potential Conflicts of Interest. Dr Arevalo has received personal fees from Iridex, Optos Inc, Novartis Pharmaceuticals Corp, and

Alimera Sciences Inc for lectures; from Second Sight LLC and Alcon Laboratories for serving as a consultant and lecturer; and from Springer SBM LLC related to a patent. No other disclosures were reported.

REFERENCES

- Schepens CL, Brockhurst RJ. Uveal effusion, 1: clinical picture. *Arch Ophthalmol*. 1963;70:189-201.
- Brockhurst RJ. Nanophthalmos with uveal effusion: a new clinical entity. *Arch Ophthalmol*. 1975;93(12):1989-1999.
- Gass JD. Uveal effusion syndrome: a new hypothesis concerning pathogenesis and technique of surgical treatment. *Retina*. 1983;3(3):159-163.
- Trelstad RL, Silbermann NN, Brockhurst RJ. Nanophthalmic sclera: ultrastructural, histochemical, and biochemical observations. *Arch Ophthalmol*. 1982;100(12):1935-1938.
- Forrester JV, Lee WR, Kerr PR, Dua HS. The uveal effusion syndrome and trans-scleral flow. *Eye (Lond)*. 1990;4(pt 2):354-365.
- Brockhurst RJ. Vortex vein decompression for nanophthalmic uveal effusion. *Arch Ophthalmol*. 1980;98(11):1987-1990.
- Uyama M, Takahashi K, Kozaki J, et al. Uveal effusion syndrome: clinical features, surgical treatment, histologic examination of the sclera, and pathophysiology. *Ophthalmology*. 2000;107(3):441-449.
- Kanner EM, Netland PA, Sarkisian SR Jr, Du H. Ex-PRESS miniature glaucoma device implanted under a scleral flap alone or combined with phacoemulsification cataract surgery. *J Glaucoma*. 2009;18(6):488-491.
- Johnson MW, Gass JDM. Surgical management of the idiopathic uveal effusion syndrome. *Ophthalmology*. 1990;97(6):778-785.
- Jackson TL, Hussain A, Morley AM, et al. Scleral hydraulic conductivity and macromolecular diffusion in patients with uveal effusion syndrome. *Invest Ophthalmol Vis Sci*. 2008;49(11):5033-5040.