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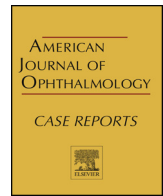
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Idiopathic elevated episcleral venous pressure in a teenager

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ABSTRACT

Purpose: To report a case of unilateral idiopathic elevated episcleral venous pressure (IEEVP) in a 15-year-old patient. We reviewed and summarized published case reports of IEEVP to determine how to manage this challenging and rare condition.

Observations: A 15-year-old Caucasian male presented with elevated intraocular pressures (IOP), blood in Schlemm canal in the left eye, and asymmetric cupping with corresponding glaucomatous findings on testing. We diagnosed the patient with IEEVP and describe successful surgical intervention with deep sclerectomy and viscocanalostomy.

Conclusions and Importance: IEEVP is a diagnosis of exclusion and based on clinical findings of dilated episcleral veins, blood in Schlemm canal and glaucomatous changes. If glaucomatous progression occurs with medication, filtration surgery is usually required, and most patients have good results in the literature. Care should be taken to prevent post-operative hypotony and serous choroidal detachment.

1. Introduction

Elevated episcleral venous pressure (EVP) is a rare condition that can present with elevated intraocular pressure (IOP) and blood in Schlemm canal. The causes of increased EVP include arteriovenous malformations, such as carotid-cavernous sinus fistula, orbital varix and Sturge-Weber syndrome, and conditions that lead to venous obstruction such as retrobulbar tumor, thyroid eye disease and superior vena cava syndrome.¹ It can also be idiopathic, referred to as idiopathic elevated EVP (IEEVP), idiopathic dilated episcleral veins or Radius-Maumenee syndrome,² and considered a diagnosis of exclusion. Here, we present a case of IEEVP in a teenager.

1.1. Case report

A 15 year-old healthy male was referred to our glaucoma service for elevated IOP in both eyes. He was born full-term and met all developmental milestones. His past medical history was only notable for childhood asthma. He had no prior ocular surgery, laser or trauma, and no family history of glaucoma. Prior to presentation, he was seen by an outside ophthalmologist with IOP of 30 mmHg in the right eye (OD) and 32 mmHg in the left eye (OS) on no medications, and subsequently started on brinzolamide and brimonidine two times a day in both eyes (OU). At his presenting visit, his vision was 20/20 OU with IOP of

15 mmHg OD and 18 mmHg OS on brinzolamide and brimonidine OU, with no afferent pupillary defect. His central corneal thicknesses were 588 μ m OD and 570 μ m OS. Slit lamp exam revealed normal anterior segments OU except for a few dilated episcleral vessels OS (Fig. 1A). On gonioscopy, his angles were open to ciliary body band 360°, and there was blood in Schlemm canal 270° in the left eye only (Fig. 1B). In the right eye, he had a cup-to-disc ratio of 0.3 with no disc hemorrhage. In the left eye, his cup-to-disc ratio was 0.6 with no disc hemorrhage or focal notching (Fig. 2). His dilated fundus exam was normal OU.

At presentation, his Humphrey visual field (HVF) was full in the right eye and showed early superior and inferior arcuate changes in the left eye (Fig. 3A). Optical coherence tomography (OCT) of the circumpapillary retinal nerve fiber layer (RNFL) showed normal thickness in the right optic nerve, and superior and inferior thinning in the left optic nerve (Fig. 3B). Magnetic resonance imaging (MRI) of the orbit and magnetic resonance angiogram (MRA) of the head were unremarkable with no evidence of intracranial aneurysm, vascular stenosis, venous sinus thrombosis, or cavernous carotid fistula.

The patient was diagnosed with Radius-Maumenee syndrome OS based on the elevated IOP, optic nerve cupping, blood in Schlemm canal, and unremarkable head and orbit imaging. He required escalation of medical therapy in his left eye. At his 1.5 year follow-up from presentation, his IOP was 28 mmHg OS on maximum topical medical therapy including brimonidine, brinzolamide, latanoprost, betaxolol

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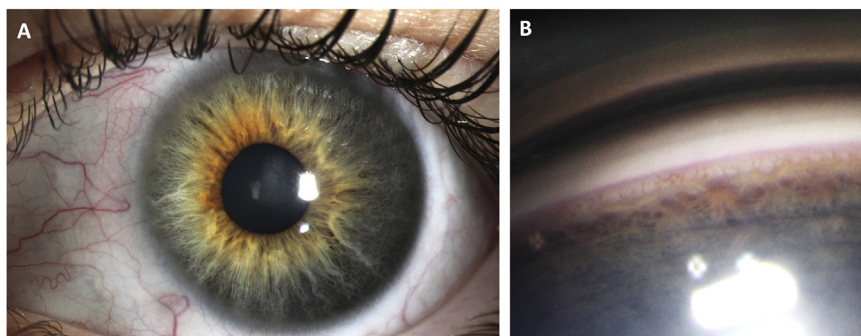


Fig. 1. A) Slit lamp photo of left eye (OS) showing several dilated, tortuous episcleral vessels. B) Gonioscopy photo OS showing blood in Schlemm canal.



Fig. 2. Fundus photos of right eye (OD) and left eye (OS) showing asymmetric cup-to-disc ratio.

and netarsudil. His OCT demonstrated progressive RNFL thinning from an average RNFL thickness of 76 μm to 63 μm over 1.5 years, which corresponded to a progressive inferior arcuate defect on HVF OS (Fig. 3C and D). The patient underwent a deep sclerectomy and viscocanalostomy OS. His IOP was 9 mmHg on post-operative day 1 on no drops and remained at 16 mmHg on no drops at post-operative month 3.

2. Discussion

IEEVP was first described by Minas and Podos in 1968 when they described two cases of unilateral dilated episcleral veins, elevated EVP and blood in Schlemm canal in a mother and daughter.³ In 1978, Radius and Maumenee reported 4 additional cases of idiopathic dilated episcleral vessels and open-angle glaucoma.² Approximately 55 cases of IEEVP have been reported in the literature in all languages.^{4–23} We conducted a PubMed search using the following search terms: idiopathic elevated EVP, idiopathic dilated episcleral veins or Radius-Maumenee syndrome (date 1/29/20). In Table 1, we present the cases reported in the literature in English that required surgical intervention for glaucoma control, grouped by surgery type.

Our patient presented at age 15, matching the youngest age of presentation reported in the literature.⁵ The reported age of onset ranges from mid-teens to 70s, and most cases appear to be unilateral or bilateral with asymmetric involvement.^{4–20} The pathogenesis is currently unknown, though congenital vascular abnormalities and genetic predisposition have been proposed.^{2,6} Mechanistically, increased EVP can prevent normal aqueous outflow and lead to elevated IOP. The normal range for EVP is 8–10 mmHg, and based on the Goldmann equation, there should theoretically be a linear relationship between EVP and IOP.¹

Since the diagnosis of IEEVP is one of exclusion, it is necessary to rule out other causes of increased EVP, namely pathologic conditions such as carotid-cavernous sinus fistulas that lead to arteriovenous communication and venous obstructive disorders such as thyroid eye disease. Computed tomography (CT) or MRI can help identify any

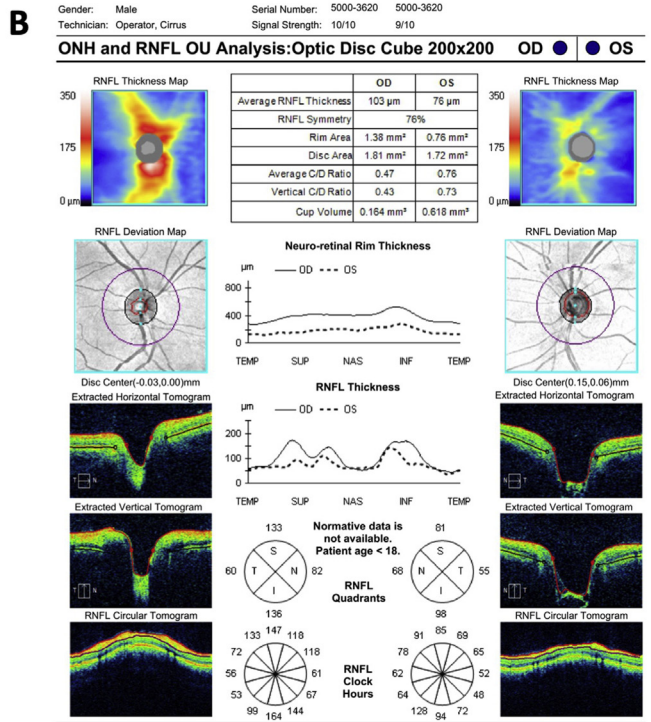
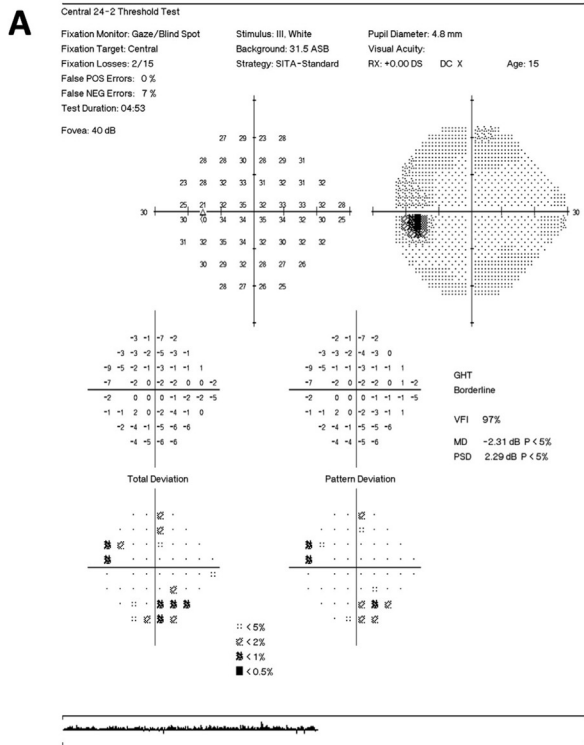
intraorbital or intracranial pathology, and CT or MR angiography is necessary to exclude intracranial vascular abnormalities. If identified, management of the primary neurovascular disorders usually results in the normalization of IOP.¹ Our patient had a negative MRI/MRA ruling out a neurovascular abnormality.

The diagnosis of IEEVP is clinical based on findings of dilated episcleral veins, open angle and elevated IOP with corresponding optic nerve and visual field findings consistent with glaucoma. Gonioscopy typically shows an open angle with blood in Schlemm canal. Our patient presented with unilateral dilated episcleral veins and blood in Schlemm canal suggesting elevated episcleral venous pressure. The patient's episcleral veins were noted to be more prominently dilated during surgery due to masking by thick Tenon's capsule. The measurement of EVP can be performed through direct and indirect methods, but no commercially available device is available, making it difficult to perform in the routine clinical setting.^{17,24,25} Glaucoma status should be assessed with automated perimetry, and OCT of the RNFL and ganglion cell complex. Optic nerve photos can be taken at the time of presentation to allow for future comparison.

Treatment can be challenging in these cases. Typically, topical medications are started, but most patients in the literature required maximum medical therapy, and few were stable only on medications.^{4,14,17} The efficacy of laser trabeculoplasty in this patient population has not been well studied. There are reports of argon laser trabeculoplasty and micropulse diode laser trabeculoplasty having minimal effect on IOP in IEEVP.^{9,18} If there is continued progression on maximum-tolerated medical therapy, glaucoma surgery should be performed. Most cases of IEEVP were managed with filtration procedures, including trabeculectomy, penetrating cyclodiathermy, sinusotomy, deep sclerectomy and viscocanalostomy, with favorable outcomes reported in the English literature (Table 1).^{2,3,5,7,9,12,13,15–19,23} There is a risk of choroidal effusion or suprachoroidal hemorrhage with rapid reduction of IOP after filtration surgery.^{15–17,19,22} Most of these cases resolved spontaneously with conservative management.^{15–17,19,22} One case required choroidal drainage on post-operative day 4 after trabeculectomy with mitomycin C.¹⁵ Prophylactic sclerectomies could be considered at the time of filtration surgery to prevent intraoperative and post-operative serous choroidal detachments.

Alternative therapies include glaucoma drainage device or bleb-forming minimally invasive glaucoma surgery (MIGS), such as the XEN45 gel stent, but use of these devices for IEEVP has not been reported in the literature. Cyclodestructive procedures may require high amounts of energy to achieve near-complete ciliary body shut-down to reach target IOP and cause post-operative complications such as inflammation and macular edema in a well-seeing eye. In patients with limited visual potential, it may be a reasonable first-line option given the serious complications that can occur with filtration surgery. Trabecular meshwork-bypassing MIGS (e.g. iStent, Kahook dual blade, gonioscopy-assisted transluminal trabeculotomy) likely will not lower the IOP enough or resolve the issue of elevated EVP if the obstruction is

At Presentation



After 1.5 years

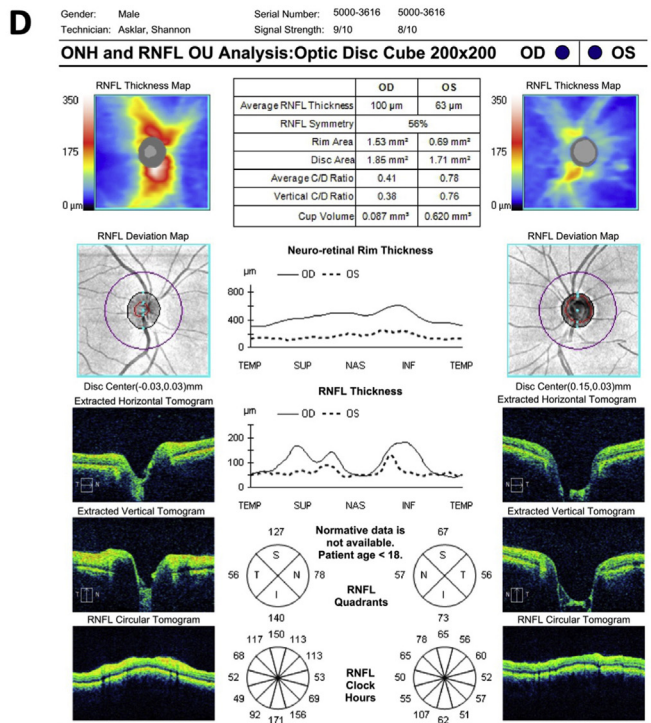
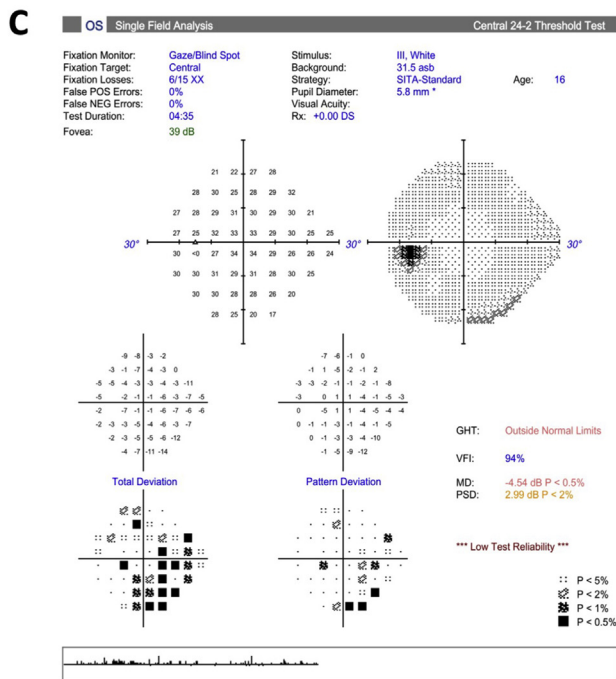


Fig. 3. A) Humphrey Visual Field (HVF) of left eye (OS) with early superior and inferior arcuate scotomas at presentation. B) Optical coherence tomography (OCT) of the retinal nerve fiber layer (RNFL) showing superior and inferior thinning OS at presentation. C) HVF OS with progressive inferior arcuate scotoma after 1.5 years. D) OCT RNFL showing progressive thinning OS after 1.5 years.

Table 1
Surgical interventions for IEEVP reported in the English literature.

Paper	Age (years)	Sex	Affected eye(s) with IEEVP	Surgical Intervention	Last reported IOP after surgery (mmHg)	Presence of choroidal effusion after intervention
Radius and Maumenee, 1978 ²	42	M	OD > OS	Filtration surgery OD	OD 15	No
Radius and Maumenee, 1978 ²	48	F	OD > OS	Filtration surgery OD	OD 19	No
Radius and Maumenee, 1978 ²	63	F	OD > OS	Penetrating cyclodiathermy × 2 OD	OU < 20	No
Minas and Podos, 1968 ³	60	F	OU	Sclerectomy OD	OD 4	No
Bellows et al., 1979 ²²	27	F	OU	Sclerectomy + posterior sclerostomy OD	OD 7	Yes OD
Bellows et al., 1979 ²²	25	F	OU	Sclerectomy + posterior sclerostomy × 2 OD	OD 10, OS 11	Yes OU
Güven et al., 2002 ⁹	32	M	OD	Sclerectomy + posterior sclerostomy OS	OD 16-19	No
Parikh et al., 2011 ¹⁵	65	F	OD > OS	Non-penetrating deep sclerectomy OD	OD 22	Yes OD
Talusan et al., 1983 ²⁰	68	F	OD	Sclerectomy with sclerostomy inferotemporally + trabeculectomy superiorly OD		
Bigger, 1975 ²³	50	F	OD	Trabecular trephine OD	OD 8	No
Lanzl et al., 1996 ¹³	34	M	OS	Trabeculectomy OD	OD 12	No
Griehaber et al., 2007 ⁷	39	M	OS	Trabeculectomy OS	OS < 20	No
Rhee et al., 2009 ¹⁷	39	M	OS	Trabeculectomy + MMC OS	OS 9	No
Rhee et al., 2009 ¹⁷	50	M	OD	Trabeculectomy with MMC OD	OD 11	No
Rhee et al., 2009 ¹⁷	40	F	OS > OD	Trabeculectomy with MMC OS	OS 16	Yes OS
Rhee et al., 2009 ¹⁷	60	M	OS > OD	Trabeculectomy with MMC OS	OD 10	No
Cymbor et al., 2013 ⁵	15	M	OD	Trabeculectomy OD	OD 5	No
Stock et al., 2013 ¹⁹	69	M	OS	Trabeculectomy OS	OS 12	Yes OS
Pradhan et al., 2015 ¹⁶	40	F	OU	Trabeculectomy with MMC OD, followed by sclerostomy for choroidal effusion on POD4	OD 13, OS 8	Yes OU
Rong and Li, 2018 ¹⁸	50	F	OU	Trabeculectomy with MMC OS		
				Trabeculectomy with 5FU OU	OU 8-11	No

Abbreviations: IEEVP, Idiopathic elevated episcleral venous pressure; IOP, Intraocular pressure; M, Male; F, Female; OD, Right eye; OS, Left eye; OU, Both eyes; MMC, Mitomycin C; 5FU, Fluorouracil; POD4, Post-operative day 4.

distal to the trabecular meshwork.

In our patient, we elected to perform a non-penetrating deep sclerectomy and viscocanalostomy, which theoretically reduces the risk of hypotony and choroidal detachment compared to a trabeculectomy. Mechanistically, the viscocanalostomy dilated the Schlemm canal, which may improve outflow by recruiting collector channels without elevated venous pressure. The deep sclerectomy left a patent space behind the anterior trabeculum and Descemet membrane, which allows aqueous to efflux into a proposed intrascleral lake. The surgery and post-operative course were uncomplicated and he has maintained IOP of 16 mmHg on no drops at post-operative month 3.

3. Conclusions

In young patients presenting with elevated IOP and blood in Schlemm canal, it is important to rule out vision and life-threatening causes of increased EVP, prior to arriving at the diagnosis of IEEVP. Dilated episcleral vessels may not be obvious in young patients due to a thick Tenon's capsule. In these patients, glaucoma evaluation should be performed and patients should be managed similarly to primary open-angle glaucoma with medications as first line therapy. If surgery is needed, nonpenetrating procedures such as deep sclerectomy with viscocanalostomy may be a safe and efficacious alternative to bleb-forming or cyclodestructive procedures. We may be able to have better diagnostic criteria for IEEVP if more objective techniques for EVP measurement are developed for routine use. Lastly, the creation of an international registry-style database may be useful in gathering long-term outcomes data on this uncommon disease.

Patient consent

The patient's legal guardian consented to publication of the case in writing.

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Authorship

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Declaration of competing interest

The following authors have no financial disclosures: CQS, CMM, TCC.

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