Outcomes of medical and surgical management in eyes with idiopathic elevated episcleral venous pressure

Nagalekshmi Ganesh, Md Shahid Alam, Ronnie J George, Shantha Balekudaru, Lingam Vijaya

**Purpose:** Idiopathic elevated episcleral venous pressure (IEEVP) is a rare cause of secondary glaucoma and is a diagnosis of exclusion. The aim of this study was to describe the clinical presentation and analyze the outcomes of medical and surgical management in eyes diagnosed with idiopathic elevated episcleral venous pressure. **Methods:** A retrospective analysis of eyes diagnosed with IEEVP over a 5-year period between April 2012 and March 2016 was performed. The demographic details, medical history, and clinical course of the cases were obtained from the medical records. Data pertaining to the severity of glaucomatous damage, response to medical management, need for surgical intervention, and their outcomes were analyzed. **Results:** Fifteen eyes of 13 patients were included. Thirteen eyes (86.6%) had open angle configuration. Among the 13 eyes that had glaucoma, eight eyes (61.5%) had severe glaucoma, four eyes (30.7%) had moderate glaucoma, and one eye (7.6%) had mild glaucoma. The median follow-up was 210 days. Seven of the 15 eyes (46.6%) required a glaucoma filtration procedure, and three underwent prophylactic sclerotomy. 71.4% of these eyes had complete success. One out of the seven operated eyes required choroidal drainage post-operatively. **Conclusion:** IEEVP is an extremely rare condition and presents with raised intra-ocular pressure and tortuous episcleral vessels. The management of IEEVP is similar to that of primary open angle glaucoma. Uveal effusion is to be anticipated, and hence, combining trabeculectomy with prophylactic sclerotomy is advisable.

**Key words:** Carotid cavernous fistula, episcleral venous pressure, idiopathic, intra-ocular pressure, orbital pressure

The aqueous outflow is facilitated by the difference in pressure between the anterior chamber and the episcleral veins. The normal episcleral venous pressure (EVP) is 8–10 mm of Hg.\(^1\) According to the Goldman equation, “Intra-ocular pressure (IOP) = Aqueous flow rate (F)/Coefficient of outflow facility (C) + Episcleral venous pressure (EVP),” which implies that an increase in EVP would increase the IOP.\(^2\) The exact relationship however is not well understood. Causes of the increase in EVP include arteriovenous anomalies and venous obstruction, although a significant proportion is idiopathic.\(^3\)

Idiopathic elevated episcleral venous pressure (IEEVP) leading to secondary glaucoma was first described by Minas and Podos in 1968.\(^4\) The evanescent venous congestion and secondary glaucoma are probably because of poor episcleral venous drainage, the cause remaining unknown. It is a diagnosis of exclusion presenting with arterialized episcleral vessels and secondary glaucoma.\(^5\) This is a rare cause for glaucoma with most of the literature being limited to isolated case reports, the largest case series being that of six cases.\(^6\) We report the presentation and management outcome of a series of 13 cases of IEEVP. To the best of authors’ knowledge, this is by far the largest case series on this rare entity.

**Methods**

A retrospective chart review was performed for all cases diagnosed with IEEVP over a period of 5 years, between March 2012 and April 2016. All patients with dilated episcleral veins and a raised IOP (≥21 mmHg), without any other cause for elevation in IOP and any other contributory factors on neuroimaging [magnetic resonance imaging (MRI) brain and orbit] causing a secondary rise in EVP, were diagnosed with IEEVP and included in the study [Fig. 1]. Cases with orbital pathologies such as thyroid eye diseases, carotid cavernous fistula, phakomatosis, idiopathic orbital inflammation, or orbital neoplasms which could cause an elevation of the EVP were excluded. Details pertaining to the patient demographics, medical history, and clinical course were extracted from the records. Eyes with structural or functional changes characteristic of glaucomatous optic neuropathy were considered to have glaucoma based on the International Society of Geographical and Epidemiological Ophthalmology (ISGEO) criteria.\(^7\) The angle was graded by modified Shaffer’s grading and classified as open and closed angles as per ISGEO criteria.\(^1,7\) Based on the Hoddap...
Anderson and Parish criteria for Humphrey visual fields, glaucoma was classified as mild, moderate, or severe.[9] For those with IOP higher than 21 mm of Hg, a normal optic nerve and normal visual field were classified as ocular hypertension.[10] The fellow eye status, need for surgical intervention, type and outcomes of surgery in terms of complications (intra-operative and post-operative), post-operative IOP control, and need for anti-glaucoma medication at final follow-up were analyzed. Data were entered and analyzed in a Microsoft Excel spreadsheet.

In eyes that underwent surgery, complete success and qualified success were defined as IOP of <20 mmHg without and with one or more topical anti-glaucoma medications, respectively, at the final follow-up. Eyes which required minor interventions such as bleb needling or choroidal drainage to manage post-operative complications were also considered to have qualified success. Failure was defined as the need for a repeat glaucoma filtration procedure or glaucoma implants for IOP control after a failed trabeculectomy in these cases.

**Results**

Fifteen eyes of 13 patients were included. Two cases had bilateral involvement. The median age at presentation was 57 years (range: 29–72 years). There was no gender predilection or significant difference in laterality [Table 1]. There was no association with any systemic diseases. Eleven (73.3%) out of 15 eyes had a best corrected visual acuity of 0.3 log MAR or better at presentation. Eight eyes (54%) had a clear lens at presentation, whereas the rest had early cataractous changes.

The mean IOP at presentation was 27.3 ± 8.5 mm of Hg (16–50 mm of Hg). Three eyes with IOP less than 21 mm of Hg at presentation were also included as their baseline IOP was not available but were already on anti-glaucoma medications and had glaucomatous cupping with the corresponding field defects.

Thirteen eyes (86.6%) had open angle configuration. Among them, nine eyes had blood in Schlemm’s canal on visualization with gonioscopy. Two eyes had closed angles. Although angle closure was bilateral and clinically consistent with primary angle closure, dilated episcleral veins and elevated pressure were seen in only one of the eyes in both the cases.

Out of the 15 eyes included in the study, all 13 eyes (86.6%) with open angles had glaucoma; two eyes with closed angles had normal discs and visual fields. Among the 13 eyes, eight eyes (61.5%) had severe glaucoma, four eyes (30.7%) had moderate glaucoma, and one (7.6%) had mild glaucoma. Three eyes with advanced disc changes had vision less than counting fingers at 1 meter precluding Humphrey visual field testing. The average pachymetry was 524 ± 37.1 microns (465–598 microns). Two cases had bilateral IEEVP; one had ocular hypertension in the contra-lateral eye, and in the two eyes with closed angle configuration on gonioscopy, a laser peripheral iridotomy was performed.

Fourteen eyes were on topical anti-glaucoma medication at presentation. The mean number of agms used at presentation was 2.13. However, all 15 study eyes required topical anti-glaucoma medications during subsequent follow-up. The median follow-up was 210 days. Five eyes had follow-up of less than a month. Of the 15 study eyes, seven had undergone trabeculectomy augmented with Mitomycin-C, and three eyes had prophylactic sclerostomies with trabeculectomy to prevent choroidal effusion. All three cases had advanced glaucoma

**Table 1: Demographic details and clinical features**

| Parameters                  | Numbers and Percentage |
|-----------------------------|------------------------|
| Age (Years)                 | Median Age: 57 years (29-72 years) |
|                             | 20-40: 2               |
|                             | 41-60: 8               |
|                             | 61-80: 3               |
| Sex                         | Males: 7               |
|                             | Females: 6             |
| Laterality                  | Right Eye: 5           |
|                             | Left Eye: 6            |
|                             | Both Eyes: 2           |
| Best corrected visual acuity| Mean log Mar: 0.5      |
|                             | Better than log Mar 0.3: 11 eyes |
|                             | Less than Log Mar 1.3: 4 eyes |
| Gonioscopy                  | Open angle: 13         |
|                             | Closed angle: 2        |
|                             | Blood in Schlemm’s canal: 9 |
| Pachymetry                  | Mean: 524±37.1 microns (465-598 microns) |
|                             | >520 micrometers: 7    |
|                             | <520 micrometers: 6    |
|                             | Not available: 2       |
| Glaucoma staging            | No glaucoma: 2 eyes    |
|                             | Mild glaucoma: 1 eye   |
|                             | Moderate glaucoma: 4 eyes |
|                             | Severe glaucoma: 8 eyes |

**Figure 1:** (a): Bilateral tortuous episcleral vessels in a patient with bilateral IEEVP. (b and c): Right-sided tortuous episcleral vessels in a patient with right IEEVP.
and high pre-operative IOP. The average duration between diagnosis and surgery was 276 days. The mean pre-operative IOP was 30.0±10.50 mmHg (20–52 mm of Hg), whereas the mean post-operative IOP was 15±5.5 mmHg (6–24 mm of Hg).

None of the eyes had intra-operative complications. The mean follow-up for the eyes that underwent trabeculectomy was 334.5 days. Among the four eyes that underwent trabeculectomy without prophylactic scleromies, two had vascularized blebs which eventually responded to sub-conjunctival 5-fluorouracil injections, one had a shallow anterior chamber without signs of choroidal effusion and was conservatively managed with cycloplegics, and one had an uneventful post-operative course. Among the three eyes that underwent trabeculectomy with prophylactic scleromies, one had an uneventful course, one had a failing bleb which was treated by bleb massage, and one had a developed shallow anterior chamber with choroidal detachment. They all had open angle glaucoma pre-operatively. This was managed with oral steroids and choroidal drainage.

The visual acuity improved by at least one line in four (57.1%) out of seven patients who underwent trabeculectomy, remained stable in two patients, and decreased by one line in one of the eyes over a follow-up of 20 months because of the development of cataract. The details have been provided in Table 2.

None of the seven eyes that underwent the glaucoma filtration procedure needed anti-glaucoma medications at final follow-up, whereas in eyes that did not undergo the glaucoma filtration procedure, the need for anti-glaucoma medications increased by an average of 0.9. Post-operatively, one case with an IOP of 24 mm of Hg was lost to follow-up within a month and could not be classified as success or failure. Thus, there was complete success in five (71.4%) out of seven eyes that underwent trabeculectomy, and one eye that required choroidal drainage was considered as qualified success.

### Discussion

All the 15 eyes in the present study had dilated and tortuous episcleral veins with an IOP of more than 21 mm of Hg or glaucomatous cupping. There have been two major case series on IEEVF so far, one by Rhee et al.[11] included six bilateral cases and one by Talusan et al.[11] had six unilateral cases and one bilateral case. The present study includes 11 unilateral and two bilateral cases. The median age at presentation was 57 years with a wide range between 29 and 72 years. This varied age of onset has been reported in other similar studies.[12] Although a few earlier studies reported a preponderance of the condition in the right eye, and no logical conclusion could be drawn from the observation.[11,12] However, a similar pattern was not observed in the later studies and in our series too.[13] Although familial association had been reported in one of the very early studies by Minas and Podos, none of our patients had a positive family history.[13]

Two of the 15 study eyes had a closed angle, and both had unilateral disease. We included them as they fulfilled the inclusion criteria of arterialized episcleral veins with a raised IOP of 21 mm of Hg or more. Also, the difference in the IOP between the contra lateral eyes was not attributable to the asymmetry in the angle configuration. Rhee et al.[14] had reported the condition in eyes with plateau iris syndrome and

---

**Table 2**: Comparison of pre-operative and post-operative data of eyes that underwent the glaucoma filtration procedure

| Case | Surgical procedure | Pre-operative IOP (mmHg) | Revised IOP at final follow-up (mmHg) | BCVA at final follow-up in Log Mar | AGMs used at final follow-up | Final follow-up (Months) | Hypotony and choroidal effusion | Vascularized bleb | Hypotony and shallow anterior chamber | Nil | Nil | Vascularized bleb | Nil | Vascularized bleb | Bleb failure |
|------|--------------------|---------------------------|----------------------------------------|-----------------------------------|------------------------------|----------------------------|--------------------------|-----------------|-----------------------------------|-----|-----|-----------------|-----|-----------------|-------------|
| 1.   | Trabeculectomy with sclerotomy | 29                        | 16                                     | 0.0                              | 0.0                          | 0                          | 0                        | 0               | 0                                 | 0.0 | 0.0 | 0                | 0   | 0                | 0           |
| 2.   | Trabeculectomy          | 22                        | 14                                     | 0.0                              | 0.0                          | 0                          | 0                        | 0               | 0                                 | 0.0 | 0.0 | 0                | 0   | 0                | 0           |
| 3.   | Trabeculectomy          | 20                        | 14                                     | 0.0                              | 0.0                          | 0                          | 0                        | 0               | 0                                 | 0.0 | 0.0 | 0                | 0   | 0                | 0           |
| 4.   | Trabeculectomy with sclerotomy | 28                        | 19                                     | 0.0                              | 0.0                          | 0                          | 0                        | 0               | 0                                 | 0.0 | 0.0 | 0                | 0   | 0                | 0           |
| 5.   | Trabeculectomy          | 28                        | 19                                     | 0.0                              | 0.0                          | 0                          | 0                        | 0               | 0                                 | 0.0 | 0.0 | 0                | 0   | 0                | 0           |
| 6.   | Trabeculectomy          | 28                        | 19                                     | 0.0                              | 0.0                          | 0                          | 0                        | 0               | 0                                 | 0.0 | 0.0 | 0                | 0   | 0                | 0           |
| 7.   | Trabeculectomy          | 32                        | 24                                     | 0.0                              | 0.0                          | 0                          | 0                        | 0               | 0                                 | 0.0 | 0.0 | 0                | 0   | 0                | 0           |

**Notes**: IOP: Intra-ocular pressure, BCVA: Best corrected visual acuity, AGM: Anti-glaucoma medication.
suggested that the association is likely to be genetic. Nine out of the 13 eyes with an open angle had blood in Schlemm’s canal, seven eyes had in all quadrants, with one each in one and two quadrants. No significant association was found between the presence of blood in Schlemm’s canal and disease severity or post-operative complications. The inconsistency of the finding of blood in Schlemm’s canal has been attributed to the valve mechanism between the anterior chamber and Schlemm’s canal at higher IOP.

Although all cases underwent MRI of brain and orbit and were found to be normal, six cases also underwent vascular imaging in the form of either digital subtraction angiography (DSA) or magnetic resonance angiogram (MRA). However, all the patients had detailed clinical evaluation by the oculoplastic surgeons to rule out a carotid cavernous fistula (CCF) or other known contributory factors for elevated EVP, and vascular imaging was ordered based on clinical examination. Thyroid orbitopathy and CCF are two common orbital conditions that can have raised IOP secondary to an elevated EVP. It is necessary to rule out any sort of underlying orbital pathology before labeling the patient as a case of IEEVP.

The management of IEEVP is similar to that of primary open angle glaucoma. Because IEEVP is primarily an aqueous outflow pathology, aqueous suppressants are more effective than aqueous outflow enhancers. Apraclonidine in particular has been shown to be effective for lowering IOP in cases of elevated EVP because the drug causes arterial vasodilatation, which decreases the amount of blood flow to the eye.[14] Glaucoma filtration surgery is indicated in cases where medical management fails. Several case reports point out that trabeculectomy in these eyes can be complicated by intra-operative or post-operative uveal effusions.[15‑17] One case report recommends performing a “tight” trabeculectomy with multiple releasable sutures to titrate the IOP gradually and prevent anterior chamber shallow during surgery.[18] In our study, seven eyes required trabeculectomy and prophylactic sclerotomies were performed in three. Pre-placed sutures were used to prevent intra-operative shallowing of the anterior chamber in cases who underwent trabeculectomy without sclerotomy. Releasable sutures were used in all eyes that underwent trabeculectomy; however, only one required the release of the suture on post-operative day 5. Five (71.4%) out of our seven cases qualified for complete success. One of these developed post-operative choroidal effusion and needed choroidal drainage because of a persistent flat anterior chamber. The effusion was noted at an IOP of 14 mm of Hg. Similar cases of post-operative choroidal effusions at normal IOP have been reported earlier.[19]

**Conclusion**

IEEVP, although a rare condition, should be considered as one of the differential diagnoses in congested eyes with glaucomatous cupping or raised IOP. It is imperative to rule out other possible associations by meticulous clinical examination and neuroimaging. The management is essentially similar to that of primary open angle glaucoma. According to our study, trabeculectomy with or without prophylactic sclerotomies offers favorable outcomes; however, long-term prospective studies are needed to confirm these observations. It is pertinent to look for post-operative choroidal effusion, even at normal IOP post-operatively in these patients.

**Financial support and sponsorship**
Nil.

**Conflicts of interest**
There are no conflicts of interest.

**References**

1. Brubaker RF. Determination of episcleral venous pressure in the eye. A comparison of three methods. Arch Ophthalmol 1967;77:110-4.

2. Moster M, Ichpuliani P. Episcleral venous pressure and glaucoma. J Curr Glaucoma Pract 1996;3:1143-55.

3. Weinreb RN, Jeng S, Goldstick BJ. Glaucoma secondary to elevated episcleral venous pressure. In: Ritch R, Shields MB, Krupin T, editors. The Glaucomas. St. Louis: CV Mosby; 1989. p. 1130.

4. Podos SM, Minas TF, Macri FJ. A new instrument to measure episcleral venous pressure: Comparison of normal eyes and eyes with open-angle glaucoma. Arch Ophthalmol 1968;80:209-13.

5. RADIUS RL, Maumenee AE. Dilated episcleral vessels and open-angle glaucoma. Am J Ophthalmol 1978;86:31-5.

6. Rhee DJ, Gupta M, Moncavage MB, Moster ML, Moster MR. Idiopathic elevated episcleral venous pressure and open-angle glaucoma. Br J Ophthalmol 2009;93:231-3.

7. Foster PJ, Buhrmann R, Quigley HA, Johnson GJ. The definition and classification of glaucoma in prevalence surveys. Br J Ophthalmol 2002;86:238-42.

8. Shaffer RN. Symposium: primary glaucomas. III. Gonioscopy, ophthalmoscopy and perimetry. Trans Am Acad Ophthalmol Otol 1960;62:112-27.

9. Kass MA, Heuer DK, Higginbotham EJ, Johnson CA, Keltner JL, et al. The ocular hypertension treatment study: A randomized trial determines that topical ocular hypotensive medication delays or prevents the onset of primary open-angle glaucoma. Arch Ophthalmol 2002;120:701-13.

10. Hodapp E, Parrish RK II, Anderson DR. Clinical Decisions in Glaucoma. St. Louis: The CV Mosby Co; 1993. p. 52-61.

11. Talusan ED, Fishbein SL, SchwartzB. Increased pressure of dilated episcleral veins with open-angle glaucoma without exophthalmos. Ophthalmology 1983;90:257-65.

12. Foroozand R, Buono LM, Savino PJ, Sergott RC. Idiopathic dilated episcleral veins and increased intraocular pressure. Br J Ophthalmol 2003;87:652-4.

13. Minas TF, Podos SM. Familial glaucoma associated with elevated episcleral venous pressure. Arch Ophthalmol 1968;80:202-8.

14. Mantziosinos N, Weinreb R. Apraclonidine reduces intraocular pressure in eyes with increased episcleral venous pressure. J Glaucoma 1992;1:42-3.

15. Bellows AR, Chylack LT Jr, Epstein DL, Hutchinson BT. Choroidal effusion during glaucoma surgery in patients with prominent episcleral vessels. Arch Ophthalmol 1979;97:493-7.

16. Parikh RS, Desai S, Kothari K. Dilated episcleral veins with secondary open-angle glaucoma. Indian J Ophthalmol 2011;59:153-5.

17. Bhagat N, Lim JJ, Minckler DS, Green RL. Posterior uveal effusion syndrome after trabeculectomy in an eye with ocular venous congestion. Br J Ophthalmol 2004;88:153-4.

18. Pradhan ZS, Kuruvilla A, Jacob P. Surgical management of glaucoma secondary to idiopathic elevated episcleral venous pressure. Oman J Ophthalmol 2015;8:120.