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Endoscopic transconjunctival optic nerve sheath fenestration for progressive idiopathic visual field deficit: a case report

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Abstract

Intra-orbital optic nerve sheath fenestration (ONSF) is an effective option in patients with progressive vision loss due to idiopathic intracranial hypertension. Most proposed techniques involve surgical trauma and require disinsertion of the medial rectus muscle; thus, less invasive surgical procedures are needed. Here, a feasible and effective technique of endoscopic intra-orbital ONSF through a conjunctival incision is presented, in a patient with a progressively compromised visual field, papilloedema, and distended subarachnoid space around the optic nerves. The retrobulbar segment of the optic nerve was exposed for incision, avoiding manipulation of the lateral orbital rim bones and irritation of the ciliary microvessels and nerves. The patient regained the entire visual field. ONSF was safely and effectively performed endoscopically through a narrow corridor gained by brushing away the orbital fat with minimal traction on the medial rectus muscle. The small postoperative wound was associated with faster and easier convalescence, and less tissue trauma versus conventional open approaches.

Keywords

Optic nerve sheath fenestration, ocular surgery, endoscopic surgery, endoscopy, intra-orbital, vision loss

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Introduction

Idiopathic intracranial hypertension (IIH) is a rare condition with an annual incidence of 7.8 cases per 100 000 people and an estimated prevalence of 76 cases per 100 000, with an 85% predominance in adult females.[1,2] IIH should be suspected primarily in patients with severe headaches and in obese women of childbearing age, in conjunction with ophthalmoscopic evidence of papilloedema.[3] IIH may also be accompanied by progressive, or occasionally rapid, vision impairment. Conventional treatment options include steroids and diuretics, shunting procedures, or stenting of the intracranial venous sinuses, often with positive results.[4] However, in some cases, the visual field cannot be preserved despite maximum well-tolerated medical therapy, and such patients – according to the relevant literature and also Medical University of Białystok policy – are typically candidates for surgical intervention, including optic nerve sheath fenestration (ONSF).[5] ONSF is considered beneficial for patients with IIH, including those in whom IIH is associated with proliferative diseases, such as leukaemia.[5,6]

Open microscopic approaches (such as the medial transconjunctival approach), approaches through the upper eyelid, or lateral orbitotomy, have all been used in ONSF.[3,4] Most of these techniques involve controlled trauma and tissue disruption through medial rectus muscle disinsertion, eyelid transection, or bone removal. The present authors propose a modified technique of an entirely endoscopic ONSF, in which the only vital tissue to be cut and crossed is the conjunctiva. Herein, the case of a patient with IIH, who was successfully treated with this proposed method, is described. The entirely endoscopic ONSF technique may reduce the risk of complications associated with conventional methods, while effectively lowering the intra-sheath pressure in patients with IIH. The main advantages are decreased surgical time, less tissue traumatisation, and no requirement to suture the eyelid or remove bones.

Case report

A Caucasian male, aged 44 years, was referred to the Department of Neurosurgery, Medical University of Białystok, Poland in September 2017, with a progressive visual field deficit and deteriorating vision in both eyes, accompanied by headache, nausea, and dizziness. Symptoms had started 4 months previously, and the patient was otherwise healthy: body mass index was 25 kg/m^2, and he had no significant medical history or risk factors for IIH development. He had been previously treated with a carbonic anhydrase inhibitor (250 mg acetazolamide [Diuramid], orally, twice daily for 6 weeks) and corticosteroid (80 mg prednisone [Encorton], orally, once daily for 6 weeks) without improvement.

Magnetic resonance imaging (MRI) revealed bilateral widening of the fluid space around the optic nerves with no other intracranial pathology (Figure 1). MRI of the head and spine, including venograms, were normal (data not shown). Lumbar puncture opening pressure was 26 cmH_2O, and cerebrospinal fluid analysis was unremarkable. Inflammatory markers, serum angiotensin-converting enzyme levels, and Lyme disease and syphilis serology were negative.

On detailed ophthalmological examination, the best corrected visual acuity (BCVA; measured with a Snellen chart and then converted into decimals), with normal light projection, was 0.1 in the right eye and 0.5 in the left eye. Intraocular pressure (IOP), measured with a Goldmann applanation tonometer, was 21 mmHg and 19 mmHg in the right and left eyes, respectively. Biomicroscopic examination of both eyes revealed a normal anterior segment. Fundus

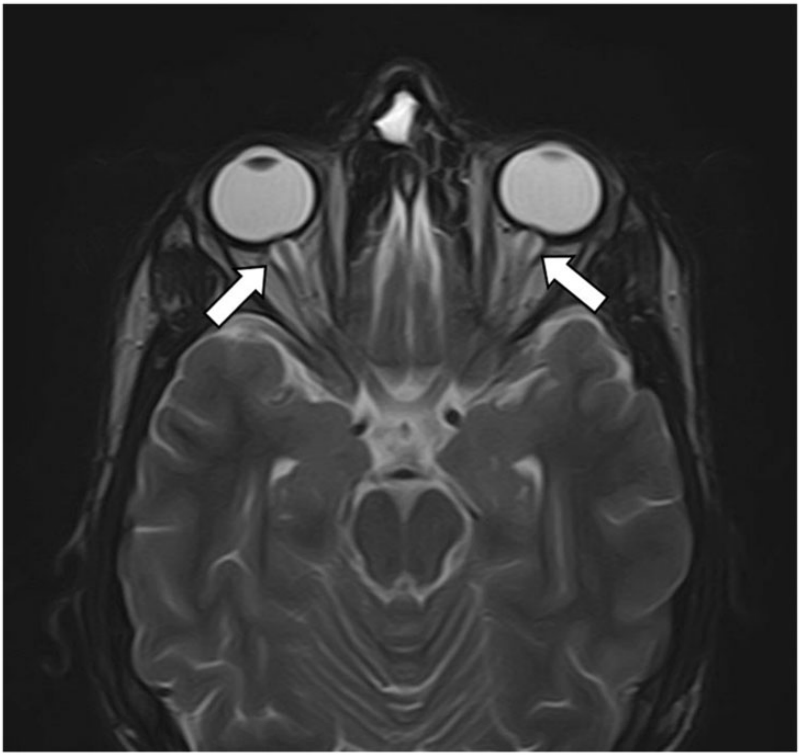

Figure 1. Transverse plane magnetic resonance image showing bilateral widening of the fluid space around the optic nerves in an adult male patient with progressive visual field deficit and deteriorating vision in both eyes.

examination through a dilated pupil showed optic disc oedema with no signs of vasculitis or uveitis. The visual-field test revealed enlarged blind spots (0–30° and 0–60°) and peripheral field defects in both eyes, with more pronounced defects in the right eye (Figure 2 and 3). Optical coherence tomography (OCT) revealed optic nerve oedema with no signs of retinal vasculitis or uveitis (Figure 4).

Considering the rapid progression of signs and symptoms over a few weeks and ineffective pharmacotherapy, surgical fenestration of the right optic nerve sheath was proposed to the patient as a procedure of documented efficacy and safety.[7] A purely endoscopic transconjunctival approach to the optic nerve was selected.[8]

The present report adheres to the ethical principles outlined in the Declaration of Helsinki as amended in 2013 and was approved for publication by the Medical University of Białystok Bioethics Committee. The reporting of this study conforms to CARE guidelines.[9] All patient data were anonymised upon collection, and the patient provided written informed consent for the proposed treatment and publication of the report, images, and surgical video.

Operative technique

The patient was placed in the supine position, and the procedure was conducted under general anaesthesia. The eyelids

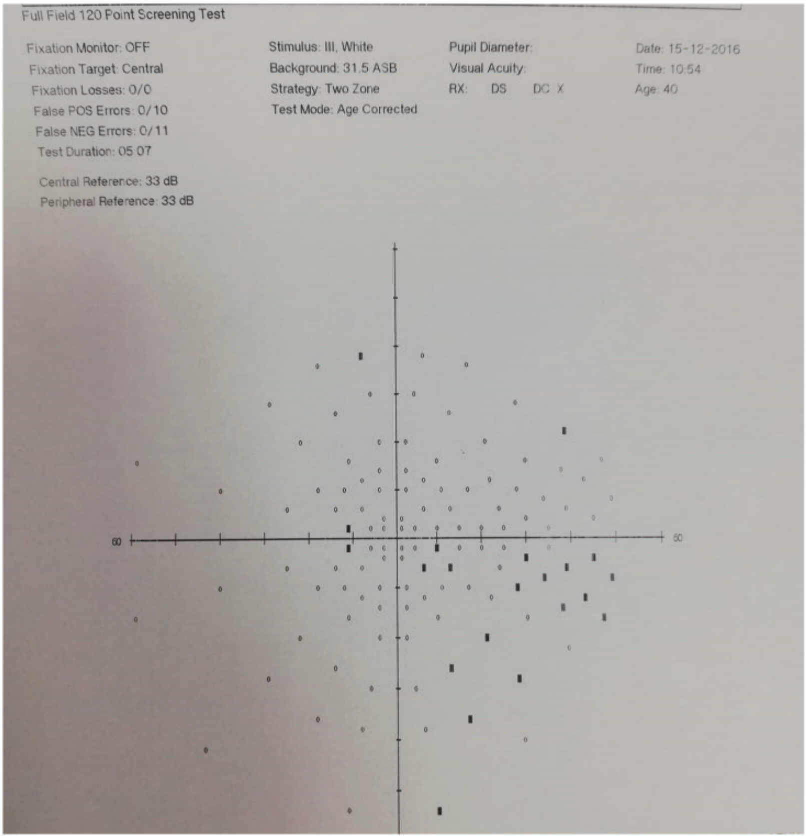

Figure 2. Preoperative left eye visual field test results in an adult male patient, showing enlarged blind spots and peripheral field defects.

were retracted with a lid speculum, and a 180° medial conjunctival incision was made close to the right-eye corneal limbus. The attachments of the superior, inferior, and medial rectus muscles to the eyeball were visualised and spared. Stay sutures were placed at each muscle to enable gentle traction of the globe in the superolateral direction. Through the conjunctival incision, two neurosurgical cotton swabs were gently forced deep into the intraconal space below the medial rectus muscle. A 10-mm wide spatula was inserted into the gap between the two cotton swabs close to the eyeball surface. The orbital content was pushed away towards the nose and into the resulting narrow space. A 0° telescope (Hopkins II, diameter 4 mm, length 18 cm; Karl Storz, Tuttlingen, Germany) was introduced inferiorly to the medial rectus muscle. Traction on the stay sutures was then released and replaced by pushing the globe away from the side of the telescope, while its focus was directed to the retrobulbar segment of the optic nerve. With awareness of the network of fragile ciliary microvessels entwining the retrobulbar segment, and the presence of short ciliary nerves heading to the pupillary sphincter, all preparations were performed with continuous monitoring of the trade-off between the need for creating space and the degree of unavoidable pressure exerted on the ocular content. The procedure was

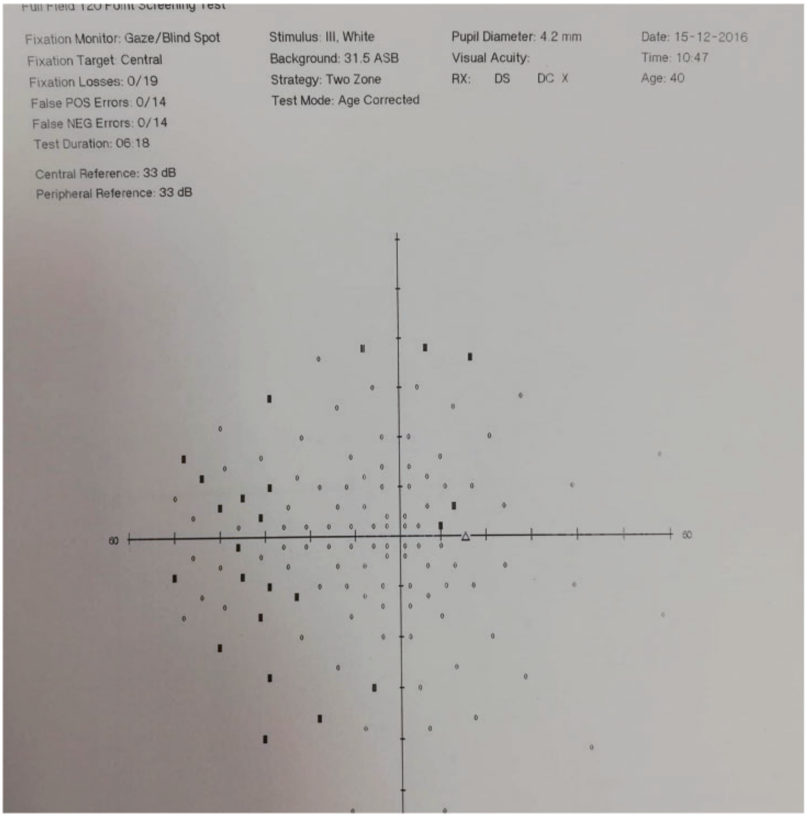

Figure 3. Preoperative right eye visual field test results in an adult male patient, showing enlarged blind spots and peripheral field defects.

performed expeditiously due to: (1) the danger of damage from the heat generated at the tip of the telescope; and (2) orbital fat, which tends to change consistency with time, collapses into the surgical canal, and eventually obscures the view. Of important note, the procedure must immediately stop upon any sign of pupil malfunction and resume after normalisation of the pupil size and shape.

To help maintain surgical-canal patency, the fat was retracted with two neurosurgical cotton swabs, using swinging/rotating movements of this provisional tool, so that the fat stuck lightly to the cotton surface and could be pushed away. Keeping the fat out of the surgical corridor using a tool with a slippery surface would be more challenging, therefore, the cotton swabs were exchanged several times during the procedure because their tips tended to fall apart after prolonged contact with orbital fat. With patency maintained, it was possible to identify the proximal retrobulbar portion of the optic nerve surrounded by the posterior ciliary vessels. Careful intraorbital retraction of the optic nerve was achieved with vectors directed perpendicular to the nerve rostrally, to avoid additional nerve stress or dysfunction manifested by pupillary dilation. During the procedure, the endoscope was attached to a mechanical holder (Karl Storz), but not fixed, to enable free-hand manipulation during most

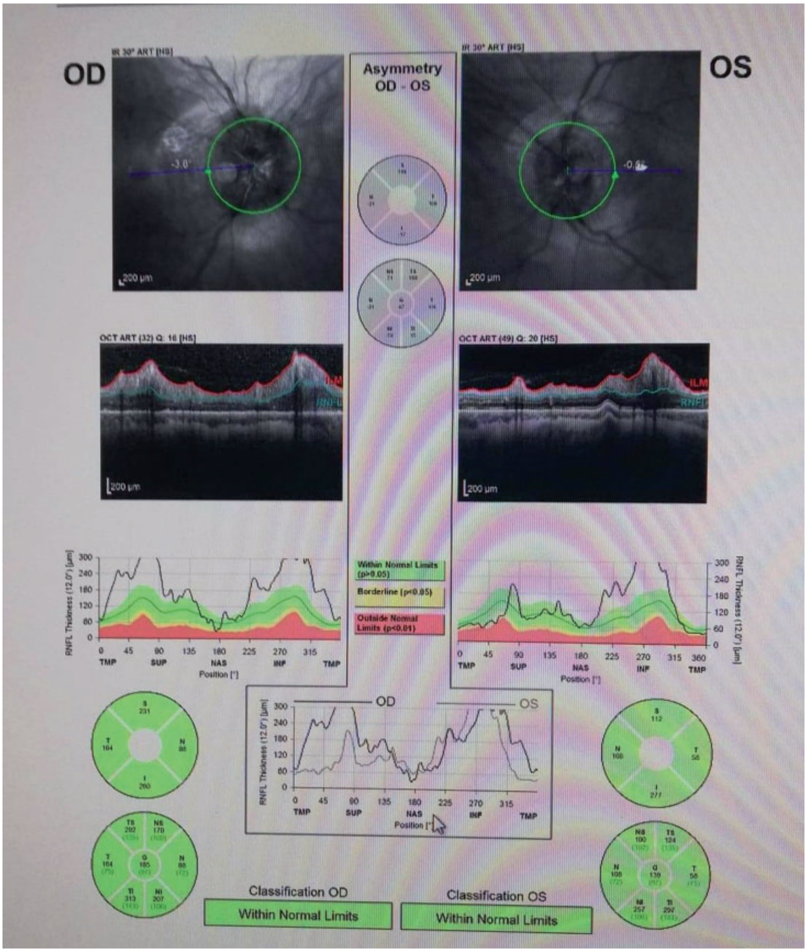

Figure 4. Preoperative optical coherence tomography (OCT) revealing optic nerve oedema with no signs of retinal vasculitis or uveitis in an adult male patient with progressive visual field deficit and deteriorating vision in both eyes. OD, right eye; OS, left eye; RNFL, retinal nerve fibre layer; TMP, temporal; SUP, superior; NAS, nasal; INF, inferior.

manoeuvres. The endoscope was fixed with a pneumatic holder (Point Setter; Matika Kohki, Tokyo, Japan) after achieving a satisfactory view of the optic nerve sheath. Incision of the dural sheath and arachnoid layer was performed with a diamond microknife (Evonos GmbH, Tittlingen, Germany), and was followed by an abundant gush of cerebrospinal fluid, marking the attainment of the procedural goal. The incision was stretched with careful blunt dissection to protect against spontaneous sealing, and the procedure was completed using absorbable Vicryl 8-0 conjunctival sutures (Ethicon, Lidingo, Sweden). The endoscopic intra-orbital ONSF technique performed in the present case is available to view online in Supplemental Video 1.

The patient was discharged from hospital on postoperative day 3 with recommendation of ophthalmological and neurosurgical follow-up. At 3 weeks following hospital discharge, the BCVA in the right eye was 0.9 with normal light projection, and the left eye remained at 0.5. The IOP was 13 mmHg and 19 mmHg in the right and left eyes, respectively. Biomicroscopic examination of the right eye revealed minor signs of conjunctival irritation, such as enlarged conjunctival vessels and chemosis of the conjunctival sac. Fundus examination showed a normal optic disc, macula with no reflex, and improvement in the OCT image (Figure 5) with full restoration of the visual field (Figure 6), and no impairment in ocular motility. MRI revealed no intra-orbital fluid collection (data not shown).

Discussion

Optic nerve sheath fenestration essentially differs from 'external' decompression of the nerve in its optic canal, and the concept

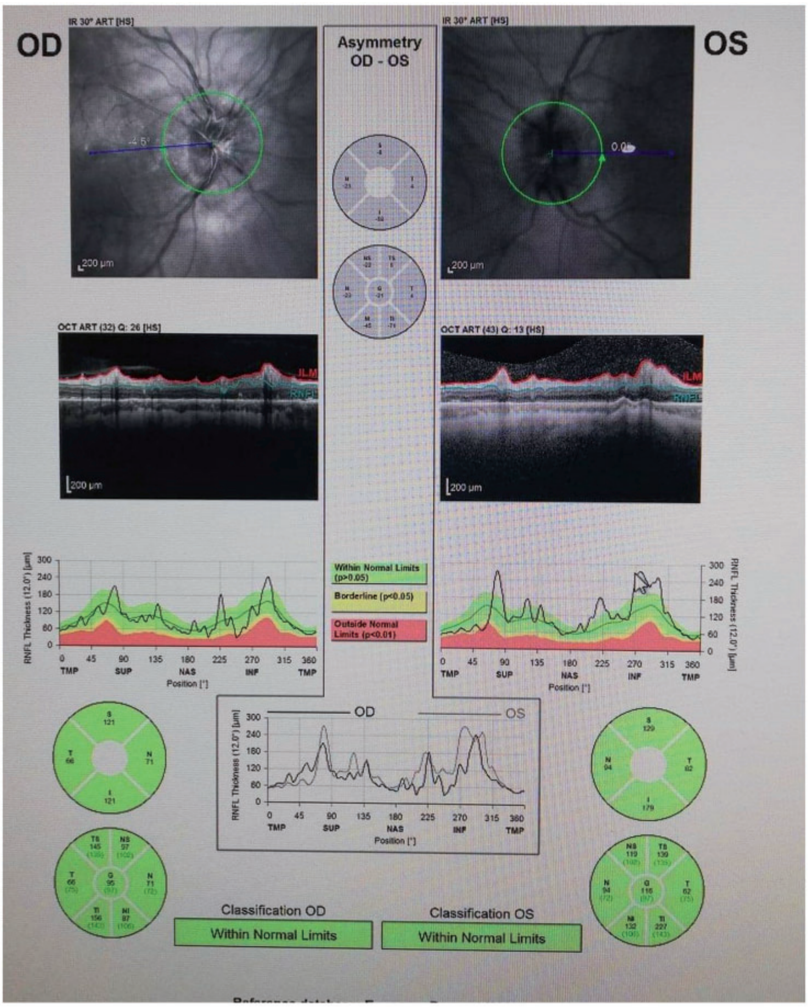

Figure 5. Postoperative optical coherence tomography (OCT) revealing increase in optical nerve thickness. OD, right eye; OS, left eye; RNFL, retinal nerve fibre layer; TMP, temporal; SUP, superior; NAS, nasal; INF, inferior.

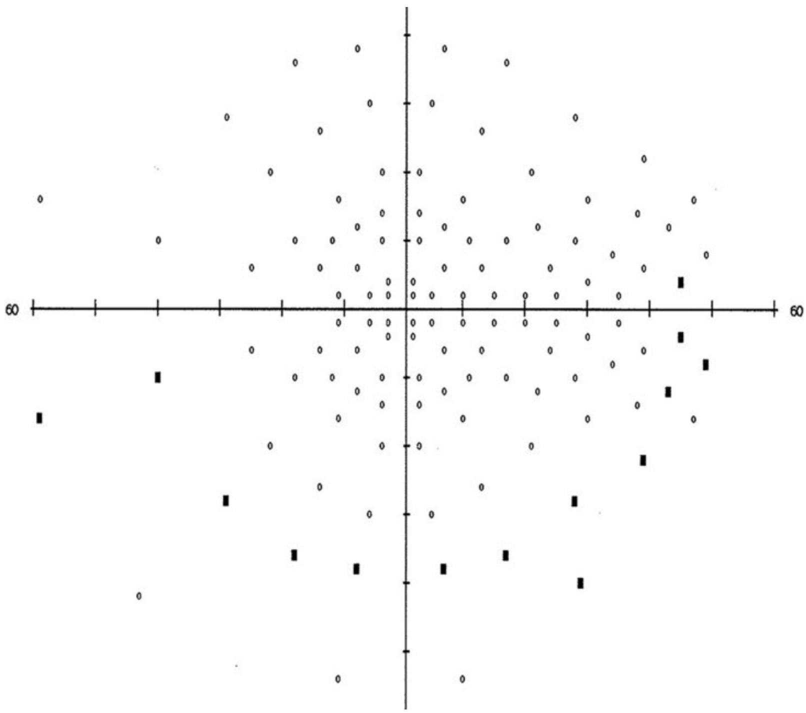

Figure 6. Postoperative right eye visual field test results in an adult male patient, showing improved visual field.

of alleviating intrathecal pressure that is affecting optic nerve function is not new in itself.[3] Nevertheless the decision to apply the procedure at a sufficiently early stage has long been problematic, in view of the fact that optic nerve sheath incision just behind the globe has historically required extensive surgery with uncertain results.[1] In 2008, Pillai et al.[10] published a cadaver study of a minimally invasive (i.e. avoiding transection of the medial rectus) endoscopic transconjunctival approach to the optic nerve in the medial intraconal space – an approach that, to the best of the present authors' knowledge, has not been applied in clinical practice to date. Here, the case of a patient who underwent a fully endoscopic ONSF according to the technique described by Pillai et al.[10] is described.

The presented technique appears to minimize invasiveness almost entirely, and the most essential aspect is sparing of the medial rectus muscle. In contrast to the present endoscopic approach, all other microsurgical techniques require transection and consequent suturing of this muscle, which significantly increases the risk of postoperative diplopia.[3]

A frequently raised concern regarding the fully endoscopic approach is that the procedure is technically challenging because the medial orbit is a small, confined space, and the endoscope is relatively large compared with the available surgical corridor. Furthermore, there is only limited bimanual instrumentation that fits in the medial orbit, and some surgeons might find visualisation adequate via surgical loupes or a surgical microscope, negating the need for an endoscope, which may additionally limit the mobility of other instruments. In addition, when a complication occurs, such as intra-orbital bleeding, an endoscopic view would be suboptimal to address the bleeding vessel.

To counteract these concerns, it must first be noted that high proficiency in endoscopic techniques is a prerequisite for such advanced surgery. Working in narrow anatomical spaces is an inherent feature of endoscopy and all of the aforementioned manoeuvres should be viable for a surgeon experienced in endoscopy, including haemostasis. In the medial orbit, the only sources of excessive bleeding are the ciliary arteries, and damage to these arteries must be avoided at all costs. Potential problems with bimanuality were resolved in the present case by fixing the endoscope with a Point Setter pneumatic holder. A microscope allows for good visualisation of the orbital content, but apparently at the cost of transection of the medial rectus muscle, which is required for rotation of the globe to bring the optic nerve into the view of the microscope.[3] With the present endoscopic technique, there is no need for either muscle transection or any significant eyeball rotation.

Optical coherence tomography, a non-invasive tool for imaging of the optic disc, is also used to follow patients after ONSF.[11,12] Nevertheless, differentiating between the resolution of papilloedema and secondary optic nerve atrophy may be difficult with this method. Therefore, an additional assessment of visual evoked potentials is recommended.[3]

As this is a report of only one clinical case, definite conclusions may not be drawn. However, the immediate vision recovery and favourable clinical outcome in the present patient, obtained with a short hospital stay, ought to encourage ophthalmologists to pursue surgical intervention without unnecessary delay, at least if an option of transconjunctival, minimally invasive endoscopic decompression of the optic nerve is available. Further studies should address whether the endoscopic procedure yields better results than the microscopic technique. Future directions for IIH patient care may also involve endovascular stenting or cerebral shunting procedures. Randomised controlled trials are needed to better define the role of these modalities and provide more information about its safety and effectiveness.

In conclusion, surgery is moving towards the minimisation of injury and shortening of surgery duration. In addition, sparing the medial rectus muscle must be emphasised as an important prerequisite for reducing the risk of postoperative ocular misalignment and strabismus.[3] In our opinion, the endoscopic ONSF procedure described herein may minimise this risk, require less surgical time, and enable quicker recovery following surgery due to less trauma compared with conventional ONSF techniques.

Declaration of conflicting interest

The authors declare that there is no conflict of interest.

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Supplemental material

Supplemental material for this article is available online.

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