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Optic nerve sheath fenestration for idiopathic intracranial hypertension: A seven year review of visual outcomes in a tertiary centre☆

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ABSTRACT

Purpose: To determine visual outcomes of patients with Idiopathic intracranial hypertension (IIH), who underwent optic nerve sheath fenestration (ONSF), utilising the two most commonly used tools for monitoring visual function (visual acuity and visual fields) and a third less commonly used tool, colour vision.
Methods: A retrospective study of ONSF patients from 2004 to 2011. Patients' symptoms, body mass index, CSF opening pressure, and visual outcomes were analysed.
Results: ONSF's were carried out on 31 eyes of 14 patients. 64% were female and 36% were male. The most predominant symptom was a headache (93%). 71% of patients had a BMI > 30. The average CSF opening pressure was 36 mmHg (range 22–64). Post ONSF, visual acuity (VA) improved in 24.1%, remained stable in 62.1% and worsened in 13.8% of operated eyes. 6% were lost to follow up. Visual fields (VF) were reliable in 48% of operated eyes. Of these 33.4% improved, 53.3% remained the same and 13.3% worsened. Colour vision (CV) improved or remained stable in 87%, and worsened in 13% of operated eyes. 4 patients had tertiary procedures (LP or VP shunts). ONSF resulted in statistically significant improvement/stabilisation in visual acuity, visual fields and colour vision. Most importantly, this was not dependent on the body mass index.
Conclusion: ONSF is a safe procedure in experienced hands. It predominantly stabilises visual function in majority of maximally medicated patients but also offers improved visual function to some patients. Colour vision monitoring is a useful adjunct in patient with unreliable visual fields. Unfortunately patients whose visual function deteriorated despite maximal medical and surgical treatment were often those who presented late or had a delay in their clinical diagnosis.

1. Introduction

Idiopathic intracranial hypertension (IIH) is the syndrome of raised intracranial pressure without clinical, laboratory or radiological evidence of intracranial pathology. IIH is a relatively rare disease but rapidly increasing incidence is reported due to a global increasing incidence of obesity. In patients who have been diagnosed with idiopathic intracranial hypertension, when medical therapy fails or when visual dysfunction deteriorates, surgical therapies should be considered. The main procedures performed include lumboperitoneal shunt (LPS) [1–3], ventriculoperitoneal shunt (VPS) [3–7], optic nerve sheath fenestration (ONSF) [8–11] and dural venous sinus stenting (DVSS) [12]. With medical intervention, disease course is generally self-limiting. However, some patients experience a disabling condition of chronic severe headache and visual disturbances that can limit their capacity to work. Permanent severe visual defects are infrequent but serious complications, which can have physical, psychological and social consequences. While most studies on ONSF have rightly focused on visual acuity and visual fields for monitoring [10,13–17], visual field test results can at times be unreliable. We believe a third parameter, colour vision, is very relevant in monitoring visual function and assessing outcomes especially when visual field test results are persistently unreliable. We report a seven-year review

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of visual outcomes, utilising these three visual function tools, following optic nerve sheath fenestration in a tertiary centre in the United Kingdom.

2. Methods

A retrospective study of ONSF patients from 2004 to 2011.

2.1. Patient selection and characteristics

Patients from the neurology department of various units who were diagnosed with idiopathic intracranial hypertension, papilloedema (Frisen scale grade 2–4) and deteriorating visual symptoms (visual acuity and visual fields) despite maximal medical therapy, requiring urgent intervention, were referred to our tertiary centre for consideration of an optic nerve sheath fenestration. Demographic and clinical features gathered included age, gender, cerebrospinal fluid (CSF) opening pressure, body mass index, visual symptoms, non-visual symptoms, visual acuity, visual fields, colour vision and the presence or absence of a relative afferent pupillary defect (RAPD).

2.2. Procedure

Surgery was carried out under general anaesthesia. A single dose of cefuroxime was given intravenously just before commencing surgery. A medial approach was utilised to gain access to the optic nerve. An eye speculum was inserted to maintain an adequately sized palpebral aperture. A 270° conjunctival peritomy was performed incorporating the inferior, medial and superior limbus. Relaxing incisions are made at the end of the peritomy. Conjunctiva and Tenon's capsule are dissected and reflected off the sclera. The medial rectus muscle was exposed, secured with a double ended 6.0 Vicryl suture and dis-inserted. The globe was rotated laterally using traction sutures placed at the site of the medial rectus disinsertion.

Gentle blunt dissection was carried out to reveal the proximal medial aspect of the optic nerve. The optic nerve sheath (dura) is identified and a 2–3 mm window cut open, in the avascular portion of the dura with a MVR blade 2 mm posterior to the junction of the globe and optic nerve. A gush of cerebrospinal fluid is seen emitting out of the window. The overlying dura was then excised to ensure long-term patency of the window. The medial rectus was reattached at its insertion using a 6.0 Vicryl suture. The conjunctiva was closed using 8.0 Vicryl sutures. Chloramphenicol eye ointment was applied topically.

In the immediate postoperative period the patient's pupillary reactions are assessed, checking for a relative afferent pupillary defect. The patient's gross visual acuity is also assessed. The next day the patient has a formal clinical assessment documenting Snellen visual acuity as well as Humphrey visual field testing. Following this, the patient is seen and clinically assessed on a 1–2 weekly basis for the first 2–4 weeks and then 1–2 monthly, until full visual stabilisation has been clearly documented. Visual stabilisation was defined as no change in visual acuity of greater or less than 2 snellen acuity lines after 2–3 successive monthly visits. Failure was defined as any of the following: A worsening visual acuity of greater than 2 snellen acuity lines from the pre-operative baseline, a worsening of the visual field with a change in mean deviation (MD) of greater than −2 Decibels (dB) from the pre-operative baseline or progressive, persistent or worsening symptoms. All patients were also under the joint care of the neurologists. Any patient who met the failure criteria at any point in time during their post operative follow up, was immediately referred back to the neurologists and neurosurgeons.

2.3. Clinical assessment and parameters

Visual acuity was carried out using a Snellen acuity chart. An improvement in Visual acuity was defined as an increase of two or more Snellen acuity lines i.e. more lines read, compared to that measured at the preoperative baseline. Conversely a reduction in visual acuity was defined as a decrease of two or more Snellen acuity lines i.e. less lines read, compared to that measured at the preoperative baseline. All patients had biomicroscopic stereoscopic fundoscopy.

Visual fields were carried out on all patients at their first visit and subsequently, using the Humphrey visual field analyser, utilising a 30-2 or 24-2 SITA program. Visual field improvement was defined as a decrease in the mean deviation of ≥2 dB from that measured at the preoperative baseline, giving rise to an increasingly positive mean deviation. Visual field deterioration was defined as an increase in the mean deviation of ≥−2 dB from that measured at the preoperative baseline, giving rise to an increasingly negative mean deviation. Unreliable visual fields were classified as visual fields with either one or all of the following criteria – greater than 3 fixation losses, greater than 33% of false positives or greater than 33% of false negatives.

Colour vision was assessed using Ishihara colour test plates. Colour vision improvement was defined as an increase in test plates read of >2 tests plates, compared to test plates measured at the pre-operative baseline. Conversely deterioration was defined as a decrease in test plates read of >2 tests plates, compared to test plates measured at the pre-operative baseline.

Patient's results were classified from baseline as, stable, improved or worse. A Wilcoxon signed ranked test was used for statistical analysis.

3. Results

Of the 15 patients referred over 7 years, One was unsuitable for surgery, because his optic disc swelling was attributable to other causes following further investigation.

ONSFs were carried out on 31 eyes of 14 patients. 3 patients had unilateral ONSF. 11 patients had non-simultaneous bilateral ONSF, of these, 6 eyes (19%), of 4 patients later had repeat procedures. The average time that elapsed before a repeat procedure was 20 months (range 6–32 months). 64% ($n = 9$) were female and 36% ($n = 5$) were male. The mean age at presentation was 36 years (range 17–61 years). Predominant symptoms were headache 93%, ($n = 13$), diplopia – secondary to a 6th nerve palsy 43%, ($n = 6$) and transient visual obscurations 43%, ($n = 6$). 71% of patients had a BMI > 30. All patients presented with papilloedema (Frisen scale grades 2–4). The average CSF opening pressure was 36 mmHg (range 22–64 mmHg). There were no intra-operative complications in any of the patients. Post operatively two patients experienced transient diplopia and one patient transient ocular discomfort. One patient (2 eyes) was lost to follow up. Post ONSF, during the intervening follow up period, papilloedema improved or resolved in all patients, Fig. 1a. Visual acuity improved in 24.1% (BMI range <25 or >40), remained stable in 62.1% (BMI range >25 to <40) and worsened in 13.8% (BMI >30 to <40) of operated eyes. Visual fields were reliable in 48% of operated eyes. Of these 33.4% improved (average mean deviation (MD) 7.7 dB, range 4–16 dB), Fig. 1b. 53.3% remained the same and 13.3% worsened (average MD −13 dB, range −2 to −23 dB). Colour vision improved or remained stable in 87%, and worsened in 13% of operated eyes. In 2 of the 3 patients who had unilateral ONSF, vision significantly improved in their non-operated eyes (≥2 snellen acuity lines) at initial and subsequent post-operative follow up. In the 3rd patient who had a unilateral ONSF vision remained stable in the non-operated eye (snellen