Effect of Optic Nerve Sheath Fenestration on Papilledema of the Operated and the Contralateral Nonoperated Eyes in Idiopathic Intracranial Hypertension

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Purpose: To evaluate the effect of optic nerve sheath fenestration (ONSF) on papilledema grade in the operated eyes and the contralateral nonoperated fellow eyes in patients with idiopathic intracranial hypertension (IIH).

Design: Retrospective review.

Participants: A total of 78 patients underwent ONSF, and 20 patients served as controls.

Methods: Charts of patients with IIH who had ONSF at the University of Iowa Hospital and Clinics were reviewed for age, gender, body mass index, and clinical findings. Optic disc photographs were graded by a masked observer using the Frisén papilledema grading scale at preoperative baseline and postoperatively at 2 weeks, 3 months, 6 months, and 12 months follow-up. Wilcoxon signed-rank test was used to examine the change in papilledema grade in both operated and nonoperated eyes at each time point.

Main Outcome Measures: Grade of papilledema.

Results: Sixty-two patients (52 women and 10 men) with a mean age of 32 years (range, 13–57 years) underwent unilateral ONSF. The median grade of papilledema for operated and nonoperated eyes was 3 and 2, respectively, at preoperative baseline. Postoperatively the grade was 2 in each eye at 2 weeks (\(P<0.0001\) and \(P<0.0002\) for operated and nonoperated eyes, respectively), 1 in each eye at 3 months (\(P<0.0001\) for both operated and nonoperated eyes), and 0.5 and 1 for operated and nonoperated eyes, respectively, at 12 months follow-up (\(P<0.0001\) for both operated and nonoperated eyes). There was no significant difference in grade of disc edema or reduction of disc edema on the basis of age, gender, or body mass index.

Conclusions: Unilateral ONSF significantly decreases the grade of papilledema in both ipsilateral (operated) and contralateral (unoperated) eyes. The reduction of the papilledema and the stability of visual field in the contralateral (nonoperated) eyes suggest that bilateral ONSF may not always be necessary in patients with bilateral visual loss and papilledema due to IIH.

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Idiopathic intracranial hypertension (IIH) is a condition of increased intracranial pressure of unknown cause. The most common symptoms are headache, visual loss, transient visual obscuration, and diplopia. Visual loss is one of the major morbidities, with blindness reported in up to 10% of patients and visual field defects of some type in up to 90% of patients.³ Optic nerve sheath fenestration (ONSF) is an effective treatment in patients with IIH who have progressive visual loss from chronic papilledema. Although prior reports have shown reduction of papilledema in both operated and unoperated eyes,¹ ² ³ ⁴ we hypothesized that the reduction in the grade of papilledema and visual field loss might be sufficient in the contralateral, unoperated eye to obviate a contralateral ONSF in a majority of patients. To our knowledge, this is the first study in the English language ophthalmic literature to test this hypothesis by systematically quantifying the reduction of papilledema in the fellow unoperated eyes in patients with IIH requiring ONSF.

Materials and Methods

After approval was obtained from the institutional review board of the University of Iowa Hospital and Clinics, the medical records of patients who had ONSF for IIH at the University of Iowa Hospital and Clinics from January 1988 to January 2007 were retrospectively reviewed. In this study, only patients who met the modified Dandy criteria were included. Patients who did not fulfill the modified Dandy criteria for IIH; patients with incomplete documentation, inadequate or incomplete fundus photographs, or inadequate follow-up; or patients with history of chronic systemic steroid use were excluded. Patients who had ONSF or cerebrospi-
nal fluid (CSF) shunt procedures before presentation to the University of Iowa Hospital and Clinics were excluded. All patients were evaluated individually with a full neuro-ophthalmologic examination, visual field testing, and fundus photographs preoperatively and in the postoperative visits.

The decision for performing ONSF was made by the evaluating surgeon and neuro-ophthalmologist on the basis of progression, acuteness, or severity of visual loss despite maximally tolerated medical therapy. Optic nerve sheath fenestration was performed with the patient under general anesthesia. A lateral orbitotomy without mitomycin C approach was used in all cases. A lateral canthotomy surgical approach without the need for bone removal was used in 95% of the patients, and 5% patients had the bone removed. Expression of CSF was observed when the initial incision into the dural sheath was made and a window of dural sheath approximately 3×4 mm in size was opened in all patients. Standard follow-up occurred at 1 week, 3 months, and 6 months thereafter by full neuro-ophthalmologic examination, visual field testing, and documentation of optic disc edema with fundus photography. Patients were asked to continue on medical treatment for IIH postoperatively until complete or near complete resolution of papilledema.

A neuro-ophthalmologist (AGL) served as a masked grader and evaluated the optic disc photographs taken at the immediate preoperative visit and at the 1-week, 3-month, 6-month, and 12-month postoperative visits for grading papilledema using the Frisén scale (Table 1, available at http://aaojournal.org). Patients included in this study had visual field done at the preoperative visit and compared it with the last follow-up (Table 2, available at http://aaojournal.org). The baseline grade of disc edema was 2 in each eye at 6 months follow-up (P=0.0001 and 0.0026, respectively). The field of vision improved significantly after unilateral ONSF for both operated and contralateral eyes (P=0.0017 and 0.0009, respectively) with a follow-up of 12 months postoperatively. More descriptive statistics on the visual field changes for patients who underwent unilateral ONSF are shown in Table 6A and B (available at http://aaojournal.org).

Discussion

Papilledema is the ophthalmologic hallmark of increased intracranial pressure. Papilledema can lead to permanent visual loss if left untreated and is the major morbidity in IIH. Optical nerve sheath fenestration provides an effective surgical treatment for patients in whom medical therapy fails. In our study, unilateral ONSF decreased the grade of papilledema in both ipsilateral (operated) and contralateral (nonoperated) eyes. The median grade of disc edema in operated eyes was higher than in nonoperated eyes at baseline, probably reflecting a bias in our center to operate on the worse eye first. The reduction of grade of papilledema in operated eyes was greater than in nonoperated eyes, but nonoperated eyes also experienced significant reduction in the disc edema grade. At 3 and 6 months, both operated and nonoperated eyes had the same grade reduction of edema, but at last follow-up the final grade of edema was less in the operated eye compared with the nonoperated eye. After unilateral ONSF, the majority of our study patients experienced improvement in the visual function in both operated and nonoperated eyes. The reduction in disc edema and visual stabilization was clinically sufficient to obviate the need to proceed with a contralateral ONSF in the majority of our patients.

We acknowledge that the mechanism for this contralateral surgical effect is not certain and could be related to decreased intrasheath CSF in both optic nerves after unilateral ONSF, regression toward the mean, spontaneous improvement, or simply better patient compliance with maximum medical therapy after unilateral surgery. Many histopathologic studies demonstrated soft-tissue proliferation at the fenestration site in the optic nerve. Hayreh found proliferation of connective tissue with adherence to the pia at the site of surgery in 9 monkeys with induced papilledema that had been treated with ONSF. Two separate studies observed occlusion of the dural fenestration sites with granulation tissue in 3 patients who died 10, 14, and 21 days after ONSF. On the other hand, a histopathologic report of a patient who died 39 days after ONSF showed patent subarachnoid space around the optic nerve and intact fistula. Magnetic resonance imaging of the optic nerve after ONSF demonstrated a periorbit fluid collection indicating CSF filtration as the dominant finding during the early postoperative period. This cyst-like structure disappeared in almost all eyes during the late period. Two mechanisms have been proposed that could explain this effect. The first is the closure of the dural sheath around the optic nerve, which may result in CSF accumulation along the dura and stimulation of the optic nerve sheath. The second is the direct drainage of CSF through the fenestration, which may lead to the reduction of disc edema.

Results

In our series, 62 patients had unilateral ONSF, 10 patients had bilateral ONSF, and 6 patients had ONSF followed by CSF shunt procedure. The mean age of the patients was 32 years (range, 13–57 years) with a male to female ratio of 1:5.5. The median grade of papilledema for patients who had unilateral ONSF was grade 3 for the operated eye and grade 2 for nonoperated eyes at preoperative baseline. Postoperatively, the grade was 2 in each eye at 2 weeks (P<0.0001 and <0.0002 for operated and nonoperated eyes, respectively), 1 in each eye at 3 months (P<0.0001 for both operated and nonoperated eyes), 1 in each eye at 6 months (P<0.0001 for both operated and nonoperated eyes), and 0.5 and 1 for operated and nonoperated eyes, respectively, at 12 months follow-up (P<0.0001 for both operated and nonoperated eyes) (Table 2, available at http://aaojournal.org). The baseline grade of papilledema and the change in edema grade for patients who had bilateral ONSF and ONSF are shown in Tables 3 and 4 (available at http://aaojournal.org).

The “no surgery” control group also showed significant changes in edema score at follow-up in both eyes, but there was no significant difference in the change in edema score between the 2 eyes (Table 5, available at http://aaojournal.org). The change in edema score at follow-up observed in the unilateral ONSF group did not differ significantly from the change in edema score in the no surgery group (P>0.20). There was no significant difference in the grade of disc edema or reduction of disc edema on the basis of age or body mass index, with P values of 0.191 and 0.889 for operated eyes and 0.478 and 0.646 for contralateral eyes, respectively.

Of those patients with baseline edema of 2, 3, or 4, there was no significant difference in the distribution of level of edema at 12 months follow-up in both the operated and contralateral eyes (P=0.223 and 0.715, respectively).

The visual acuity improved significantly after unilateral ONSF in both operated and nonoperated eyes in the study group with P values of 0.005 and 0.026, respectively. The field of vision improved significantly after unilateral ONSF for both operated and contralateral eyes (P=0.0017 and 0.0009, respectively) with a follow-up of 12 months postoperatively.
in the literature to explain how ONSF works. Cerebrospinal fluid filters through the dural opening into the orbit with a subsequent decrease in subarachnoidal CSF pressure around the optic nerve. Another potential explanation is that the fibrous tissue obliterates the operation site and prevents the direct transmission of elevated CSF pressure to the optic nerve head. One study suggested that transient drainage of the CSF through a dural window for a few weeks may be sufficient for papilledema to resolve and the intracranial CSF hemostasis to become reestablished. After ONSF, we asked our patients to continue on medical treatment until complete or near-complete resolution of papilledema.

Curry et al reported an increasing incidence of CSF shunting for IIH in the population of the United States and a concomitant flat or declining rate of ONSF. In Curry et al’s study, there were 2779 admissions for CSF shunting procedures (new or revision) in IIH. The in-hospital mortality rate for new shunts was 0.5% (0.9% for ventriculotubal shunts and 0.2% for lumbar shunts), and the estimated total US caseload of CSF shunting procedures for IIH increased 350% between 1988 and 2002 (P<0.001).

Optic nerve sheath fenestration, “in contrast to shunting procedures,” is a safer procedure, has a shorter surgical time, has a faster patient recovery time, and has essentially no mortality from the procedure itself. Optic nerve sheath fenestration is our first-line surgical intervention in patients with IIH and visual loss due to papilledema that fails maximum medical therapy.

We recognize the limitations of our study, however, including the retrospective nature of the design, relatively small sample size, associated variability in visual fields assessment and photographic technique over the years, selection, and ascertainment bias. Despite these limitations, we believe our study shows that contralateral improvement in the grade of the papilledema and visual field after unilateral ONSF may be sufficient to obviate the need for contralateral surgery.

In conclusion, it has been argued in the past that a patient with vision loss and papilledema can be spared 2 surgical procedures if shunting is performed rather than bilateral ONSF. On the basis of our results, we believe that this argument against ONSF is theoretic only and that most of our patients did well bilaterally after unilateral surgery.

References


Footnotes and Financial Disclosures

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