OPTIC NERVE SHEATH FENESTRATION FOR TREATMENT OF IDIOPATHIC INTRACRANIAL HYPERTENSION

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The swollen disc attributed to increased intracranial pressure was first described in 1856, shortly after ophthalmoscopy had been developed but 35 years before the first lumbar puncture was performed. Treatment of papilledema was initiated in 1872 by DeWecker.1 The daring and almost intuitive nature of the optic nerve sheath fenestration is all the more remarkable since it was carried out without radiology, without anesthesia, and solely with an instrument that simply was inserted through the conjunctiva and made a bayonet incision without any visualization of the operative field. No details of the visual function of these patients before or after surgery is available. However, almost as an aside, DeWecker notes that headache improved following surgery.1 After a brief flurry of interest in the procedure, it was abandoned and for 65 years or more it was not further reported.2

In the 1960's, Hayreh produced a subdural balloon primate animal model of papilledema. He showed that by opening a window in the sheath of the optic nerve there was communication between the lumbar theca and the orbit through the sheath.3 He also demonstrated that one fenestration would commonly produce bilateral relief of optic disc edema. Subsequently, Australian neuro-ophthalmologists reported success in performing optic nerve sheath fenestrations for papilledema in humans.4,5 Dribs and drabs of case reports were all that was seen of this procedure until the pathological report, Keltner, et al of a patient who had visual loss with papilledema due to an intracranial metastasis. Optic nerve sheath fenestration was done. The patient died some months later and at post mortem examination, pathologic studies suggested two possible mechanisms by which fenestration was effective in reducing papilledema. The first was the possibility of CSF leak into the orbit with absorption of CSF by tissues and the second was that there was a blockade of CSF flow and pressure by scarring. The right or wrong or “yes” to both mechanisms remains incompletely resolved.6,7,8

Subsequently, three major retrospective studies of optic nerve sheath fenestration were published in 1988 from three different centers (Pittsburgh, Iowa City and Philadelphia). Despite the lack of collaboration, the results of the three studies were remarkably similar as regards reduction of papilledema, stability or improvement of vision, the utility of unilateral fenestration for bilateral swelling, relief of headache and the relative paucity of complications.10,11,12

Following these large reports there were descriptions of different surgical approaches and modifications including lateral canthotomy,9 placement of valves14,14a and the application of mitomycin C14,15 to the margins of a fenestration to prevent closure of the window, the use of slits in the sheath as opposed to a window.16

SOME ISSUES REGARDING OPTIC NERVE SHEATH DECOMPRESSION:

What happens to CSF pressure after optic nerve sheath fenestration?

Almost certainly the CSF pressure is reduced to some degree. Evidence of this includes CSF pressure measurement,17,18 decreased caliber of venous vessels,18 a “filtering bleb” is seen on MRI,20,21 headache improves,17,18,19 and the frequency with which papilledema in the unoperated eye improves.10-12, 21

Is the reduction of CSF pressure and improvement or stability of vision a permanent effect? 22,23

This is the big question and gets into territories of opinion and art and not so much science. What is lacking here is a large multicenter follow-up study. Such a study is underway at Mississippi and Utah. As regards pressure, we need a non-invasive way of monitoring pressure since single stick LP measurements don’t tell the whole story.24

Do patients lose vision after the decompression?

Yes, occasionally. Numbers are lacking as are the details of the individual patient circumstances however there are basically three scenarios.

1.) Acute or rapid immediate post-operative visual loss.25 These include AION, branch and central retinal artery occlusions, retrobulbar hemorrhage and a mysterious, transient, 1-3 day long total loss of vision which then completely returns. The most preventable of these is retrobulbar hemorrhage possibly related to the use of large amounts of aspirin or NSAID for headache prior to surgical intervention.

2.) Rapid recurrence of disc swelling or failure of disc swelling to disappear. This usually means that the fenestration or the slits made in the optic nerve sheath were inadequate or sealed rapidly. How
frequently this occurs is difficult to know but should be information that could be obtained in a retrospective study. 26-28

3.) Late visual loss with no disc swelling. This is a terribly frustrating outcome. It usually occurs months to years after fenestration, with no other associated symptoms of TVO, headache or evidence of acutely raised intracranial pressure. Virtually all of these patients had severe visual loss at the time of surgery and while their vision improved, they had persistent defects in color vision, contrast sensitivity acuity, stereoaucity and even Snellen acuity. Discs in these patients are pale and gliotic and visual fields frequently remain constricted. The cause of this gradual visual loss has been attributed variously to axonal swelling damaging the last few axons without detectable swelling, exuberant interstitial gliosis comprising remaining fibers or to apoptosis.

Are there other significant complications? 11,12
Yes, the most common of these is partial or total tonic pupil. Whether this is more common with lateral or medial approach is not clear. Diplopia following medial rectus disconnection is unusual. The appearance of what looks like a filtering bleb is rare and infections are rare.

Does optic nerve sheath fenestration have other effects?
1.) It is associated with reduction or elimination of headache in most patients and this has been observed since DeWecker first did this procedure. How long this headache relief persists has not been studied but in my experience, headache is rarely a persistent problem. For the individuals who have persistent headache but no visual loss, I would be reluctant to recommend ONSF until a seasoned headache clinician had tried to medically control the headaches for a long time. 11,10-12

References
Other References of Interest


