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Outcomes of Endoscopic Optic Nerve Decompression in Patients with Idiopathic Intracranial Hypertension

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Abstract

Background—The conventional treatment for idiopathic intracranial hypertension involves weight loss, steroids, diuretics, and/or serial lumbar punctures; however, if the symptoms persist or worsen, surgical intervention is recommended. Surgical options include cerebrospinal fluid diversion procedures, such as ventriculoperitoneal and lumboperitoneal shunts, and optic nerve decompression with nerve sheath fenestration. The latter can be carried out using an endoscopic approach, but the outcomes of this technique have not been firmly established.

Methods—This systematic review examined the outcomes of performing endoscopic optic nerve decompression (EOND) in patients with idiopathic intracranial hypertension (IIH). Six studies were included for a total of 34 patients.

Results—The patients presented with visual field disturbances [32 of 32 (100%)], visual acuity disruptions [33 of 34 (97.1%)], papilledema [26 of 34 (76.5%)], and persistent headache [30 of 33 (90.1%)]. The mean duration of the symptoms ranged from 7 to 32 months. Overall, the patients showed post-EOND improvement of the signs and symptoms associated with IIH, specifically visual field deficits (93.8%), visual acuity (85.3%), papilledema (81.4%), and headaches (81.8%). Interestingly, 11 cases showed postoperative improvement in their symptoms with bony decompression of the optic canal alone, without nerve sheath fenestration. The authors did not report any major adverse events or complications with this approach.

Conclusion—EOND appears to be a promising and safe surgical alternative for patients with IIH who fail to respond to medical treatment. Further studies are needed before we can attest to the clinical validity of this procedure.

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Keywords

idiopathic intracranial hypertension; pseudotumor cerebri; papilledema; endoscopic optic nerve decompression; nerve sheath fenestration

Introduction

Idiopathic intracranial hypertension (IIH) is a syndrome of unknown pathogenesis characterized by elevated intracranial pressure (ICP).^{1–5} Epidemiological studies of IIH indicate that this condition affects predominantly obese, middle aged women.^{1,4–7} As a consequence of frequent hospitalizations and decreased productivity, the healthcare costs for IIH in the US have been estimated at over 444 million dollars annually.⁸ The patient symptoms include visual disturbances, headaches, nausea, and vomiting.^{1–5,7}

The conventional treatment for IIH involves weight loss, steroids, diuretics, and serial lumbar punctures.^{1,3–5,7} However, if the symptoms persist or worsen, surgical intervention is recommended, including cerebrospinal fluid (CSF) diversion procedures like ventriculoperitoneal and lumboperitoneal shunts. Another surgical option is optic nerve decompression (OND), which can be carried out using different surgical approaches such as a medial orbitotomy, lateral orbitotomy, a transconjunctival approach, and most recently, endonasal endoscopic optic nerve decompression (EOND).^{1,2,4,5,7,9} Unfortunately, there is a paucity of information in the medical literature regarding the outcomes of EOND in patients with IIH.^{2,3,5} This surgical technique has shown promising results in the setting of traumatic orbital neuropathy (TON) and fibro-osseous tumors involving the optic canal.¹⁰ Therefore, the purpose of this review was to examine the published EOND outcomes in patients with IIH. This study also focused on the clinical implications of nerve sheath fenestration as part of the procedure to address the pathophysiology of IIH.^{11–13}

Methods

Study selection

This study was approved by the University of Puerto Rico Institutional Review Board, and the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) guidelines were used for this review. We performed an online medical literature search using PubMed, EMBASE, and the Cochrane Library, which was limited to the following keywords: IIH, pseudotumor cerebri, endoscopic endonasal, and OND. English publications of any type from all years were eligible, and the search provided a total of 150 medical articles. A screening of the titles was conducted to exclude any duplicates. After screening, full-text reviews of the remaining articles were performed to eliminate those studies not related to IIH, as well as publications assessing surgical techniques other than EOND as the treatment modality. Only six articles met the required inclusion criteria and quantitative data for our review (Figure 1).

Data extraction

An individual code was assigned to each eligible patient, and specific data was retrieved and organized into a personalized Excel table, including the sex, age, duration of symptoms, and number of cases. We extracted surgical information regarding the eye selected for surgery, degree of decompression, management of the optic nerve sheath, and study design. The visual acuities, visual fields, and funduscopy exam findings were acquired and compared to the postoperative outcomes. In addition, the studies were scrutinized for any reported complications; however, information about the previous use of medications for IIH or CSF diversion procedures was not quantitatively available for the analyses.

Statistical analysis

The frequencies, percentages, means ($\pm$ standard deviation), and medians (interquartile range) were included in the descriptive tables. In addition, the overall results from the postsurgery outcomes of the studies, according to the optic sheath management, were graphed. The power sample limitations of both groups did not allow for a statistical comparison between the groups and interventions.

Results

Six studies were included in our review, for a total of 34 patients, and most of the patients were adult females (85%). The surgical procedure was predominantly performed unilaterally (88%), irrespective of bilateral papilledema. The degree of optic nerve decompression extended to at least 180 degrees in most of the studies, but the data needed to analyze this variable was incomplete (Table 1). Across the articles, there was not enough specific information regarding previous medical treatments or prior surgical management, but all of the authors stated that the patients failed conventional therapy and had persistent symptoms prior to EOND. The mean duration of symptoms ranged from 7 to 32 months (Table 2).

In those patients in whom an optic nerve sheath fistula was created ($n=23$), all of the measured variables improved: perimetry (100%), visual acuity (91.3%), papilledema (87.5%), and headaches (100%). In those cases in which the optic sheath remained intact ($n=11$), the measured variables improved as follows: perimetry (81.8%), visual acuity (72.7%), and papilledema (72.7%). The symptoms of those with headaches were ameliorated in 45.5% of the cases (Figure 2). Ophthalmological improvement was defined as objective evidence of recovery or the complete resolution of visual deficits. Headache improvement was characterized by the patient's subjective self-reporting of their pre and postsurgical symptoms. Postoperatively, two of the patients in the fenestration group showed worsening papilledema, while three patients in the non-fenestration group showed deterioration of their visual parameters. The authors of all of the studies specifically mentioned the absence of any major complications (Table 3).

Discussion

Surgery for the treatment of IIH is usually considered once a patient develops progressive loss of vision or severe papilledema, despite maximal medical therapy. CSF shunting procedures have demonstrated a 96% resolution of papilledema,³¹ and there are numerous

reports showing significant visual improvement following transconjunctival optic nerve sheath fenestration (ONSF).³² However, due to the potential complications from these surgical interventions, a conservative approach is advocated initially. As a surgical alternative, EOND has not been considered in previous reviews.^{1,8} Multiple authors have demonstrated the advantages of the endoscopic approach to the optic nerve, but few studies have been performed evaluating its efficacy and safety in treating the symptoms of IIH.^{15–18}

For patients with IIH who fail to respond to medical management, a CSF diversion procedure is the standard surgical treatment.^{9,19,20} Previous studies have demonstrated overall improvement in visual obscurations and headaches after CSF shunting. Moreover, proponents of this approach claim that this procedure directly addresses ICP, which is the underlying pathology of IIH, while avoiding any direct risk to vision. However, cases of visual loss with functioning shunts have been described. Shunt obstructions are frequent, with articles reporting a 56% complication rate, 51% revision rate, and 0.5% inpatient mortality rate.^{19,21,26,27}

The supporters of ONSF assert that this procedure directly treats the problem for which the surgery is indicated, namely, severe papilledema with visual loss. Unilateral ONSF may not only result in ipsilateral improvement, but also lead to recovery in the contralateral eye. There are various external transorbital approaches for this procedure, but medial transconjunctival access is the safest and most commonly used today.²² Medial transconjunctival access allows for an easier approach to the optic nerve, avoids removal of the lateral bony orbital wall, and decreases the risk of damage to the ciliary ganglion. Chandrasekaran et al.²³ evaluated the complication rate (15.6%) of this approach, and found that 3/52 patients had transient postoperative diplopia, 2/52 had transient anisocoria, and one patient had an optic disc hemorrhage. Overall, there were five self-limiting complications reported in this series of 52 patients. In a large retrospective study, Moreau et al.²⁴ found that complications were seen in 17 of 236 patients treated for IIH by an orbital approach to the optic nerve (7.2%), with 15 patients developing ocular dysmotility and two patients having corneal dellen. Six patients eventually required strabismus surgery. The complication rates with this approach vary from 4.8% to 45% in the literature, and include orbital hemorrhage, retinal artery occlusion, sudden intraocular pressure increase, iris sphincter paresis, accommodation failure, and transient diplopia.^{5,28} A superomedial lid crease approach has been proposed as a safer technique to access the optic nerve.³⁶ Still, in the case series published by Pelton et al. a 22% complication rate was described including one case of tonic pupil, one case of transient vertical diplopia, and two cases of transient medial ptosis.³⁷

Transnasal endoscopic surgery offers minimally invasive access to the optic nerve, while avoiding the medial rectus disinsertion required for the transconjunctival approach. EOND does not have a limitation with regard to eyes set deeply into the orbit,²¹ and provides better visualization of the optic nerve. No major surgical complications were reported in the 34 patients in whom EOND was performed. However, endoscopic procedures come with their own set of potential complications, including but not limited to: CSF leak, meningitis, epistaxis, subcutaneous orbital emphysema, and visual deterioration secondary to thermal injury from drilling the optic canal. Another concern is that by creating a direct opening into the optic nerve through a nonsterile nasal cavity, an infectious process may gain access to

the central nervous system.¹⁰ None of these complications were reported in our systematic review of the literature, but this was limited by the small sample size and a lack of randomized control trials. Papilledema has three pathological components: increased and fluctuating pressure in the distal optic nerve sheath, elevation of the central retinal venous pressure, and impaired perfusion of the nerve fibers traversing the lamina cribrosa.³⁵ However, it only occurs if the subarachnoid space surrounding the optic nerve is patent. The transmitted CSF pressure toward the optic nerve is lowered when a nerve sheath fenestration is performed. The mechanism by which this occurs is thought to be either via the formation of a permanent CSF fistula or by the obliteration of the subarachnoid space surrounding the optic nerve via fibroblast proliferation, preventing the transmission of the pressure distal to the operative site.²⁹ Supporting this argument, electron microscopy studies have shown intra-axonal swelling without evidence of extracellular edema in IIH patients.

Performing an ONSF has potential complications, including inadvertent damage to the ophthalmic artery² and a higher risk of meningitis.¹⁰ What was puzzling to us was that in our review, bony decompression alone, without nerve sheath fenestration, resulted in an improvement in the signs and symptoms of IIH. Unfortunately, the authors did not provide plausible theories that might explain these findings. It may be possible that decompression of the optic nerve at the level of the optic canal allows for expansion of the nerve, but this hypothesis has not been proven. Gupta et al.²⁵ reported that if only the bone overlying the canalicular segment is removed, the pressure on the optic nerve would not decrease, since the pressure comes from within the nerve and not from the outside. Prior studies have shown that to improve the visual symptoms, a nerve fenestration is necessary to decrease the CSF pressure on the optic nerve.²⁹ One recent study analyzing the size of the optic canal in patients with IIH showed an association between larger optic canals and more severe papilledema,³⁰ which makes the results of Sencer et al.⁴ and Patrocinio et al.³ even more intriguing.

Headaches are the most common presenting symptom in patients with IIH, which can be debilitating and significantly affect the quality of life.¹⁴ While CSF diversion procedures have been documented to improve headaches in up to 82% of patients,³¹ the literature reports that only 50% of patients who undergo a transorbital approach to ONSF will experience relief from headaches.^{32,33} The mechanism of action is not well established, and hypotheses include reduced ICP secondary to CSF drainage through the fenestration site, decreased stretching of the meninges, or even a placebo effect. Our study showed a marked improvement in headaches in all of the patients who underwent EOND with sheath fenestration while only 45% of patients reported similar outcomes after EOND without fenestration.

Conclusion

EOND appears to be a reasonable surgical alternative for patients with IIH with visual complaints and headaches that failed to respond to medical treatment. Based on our limited review, EOND is a safe procedure for treating symptomatic IIH with positive outcomes similar to those of other orbital approaches to the optic nerve. However, further studies are

needed before we can attest to the clinical validity of this procedure in patients with IHH. In addition, we should better define the selection criteria for this procedure.

Limitations

Systematic review limitations include reporting and selection biases by the original authors, which contribute to a degree of error. In terms of data extraction, this study was limited by the small number of studies available in the literature, the absence of randomized control trials, and no long-term studies. Most of the articles available for this topic were retrospective, with a low number of patients per study. Another perhaps more important limitation was that the results of the study were dependent upon the experience of each surgeon and their practice patterns, the limited amount of data reported, and the lack of detail about previous treatments.

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Figure 1.
Review of literature flow chart.

Figure 2.
Signs and symptoms improvement after endoscopic optic nerve decompression according to the nerve sheath management.

Table 1

Articles on endoscopic optic nerve decompression in patients with IIH.

Authors & Year	Study design	No. of Pts	No. of Eyes	Males	Females	Surgical Site	Degree Decompression	ONSF
Patrocínio et al., 2005	Case Report	1	1	0	1	UWV	180	N
Gupta et al., 2007	Prospective	18	18	4	14	UWV	NA	Y
Koc et al., 2008	Case Report	2	4	0	2	BL	> 180	Y
Berhouma et al., 2014 ^a	Retrospective	1	1	0	1	UNW	> 180	Y
Sencer et al., 2014 ^b	Retrospective	10	10	1	9	UNW	NA	N
Yildirim et al., 2015	Retrospective	2	4	0	2	BL	> 180	Y
OVERALL		34	38	5 (15%)	29 (85%)			

Abbreviations: IIH, Idiopathic Intracranial Hypertension; Pts, patients; UWV, Unilateral with worst vision; BL, Bilateral; ONSF, Optic Nerve Sheath Fenestration; NA, Not Available

Note: Patients were adults unless otherwise specified;

^aOnly 1 out of 11 patients included in the study had IIH;

^bTwo of the patients were children.

Table 2

Summary of articles on IIH symptoms before and after surgery by optic nerve sheath fenestration.

Authors & Year	Mean ^a and Median ^b Duration of Symptoms (in months)	Range of Follow-Up time (in months)	Optic Nerve Sheath Fenestration							
			Pre-Surgery Symptoms (No. cases/N)				Post-Surgery Symptoms			
			Visual Fields	Visual Acuity	Papilledema	Headache	Visual Fields	Visual Acuity	Papilledema	Headache
Gupta et al., 2007	17.8 ± 10.3 15 (9 – 24)	3 – 45	18/18	18/18	11/18 ^c	18/18	I-18	I-17 NC-1	I-10 W-1	R-7 I-11
Koc et al., 2008	7 ± 1.4 7 (6 – 8)	15 – 17	NA	2/2	2/2	2/2	NA	I-1 NC-1	I-1 W-1	R-2
Berhouma et al., 2014 ^d	NA	NA	1/1	1/1	1/1	NA	R-1	I-1	R-1	NA
Yildirim et al., 2015	16 ± 11.3 16 (8 – 24)	12	2/2	2/2	2/2	2/2	I-2	I-2	I-2	R-1 I-1
SUMMARY	16.6 ± 10.1 13 (8 – 24)	3 – 45	21/21	23/23	16/23	22/22	R-1 I-20	I-21 NC-2	R-1 I-13 W-2	R-10 I-12

Authors & Year	Mean ^a and Median ^b Duration of Symptoms (in months)	Range of Follow-Up time (in months)	Optic Nerve Sheath Intact							
			Pre-Surgery Symptoms (No. cases/N)				Post-Surgery Symptoms			
			Visual Fields	Visual Acuity	Papilledema	Headache	Visual Fields	Visual Acuity	Papilledema	Headache
Patrocinio et al., 2005	12	6	1/1	1/1	1/1	1/1	I-1	I-1	I-1	R-1
Sencer et al., 2014 ^e	28.6 ± 32.0 12 (6 – 36)	8 – 55	10/10	9/10	9/10	7/10	I-8 NC-1 W-1	I-7 NC-1 W-2	R-4 I-3 NC-3	I-4 NC-6
SUMMARY	27.1 ± 30.8 12 (6 – 36)		11/11	10/11	10/11	8/11	I-9 NC-1 W-1	I-8 NC-1 W-2	R-4 I-4 NC-3	R-1 I-4 NC-6

Abbreviations: IIH, Idiopathic Intracranial Hypertension; NA, Not Available; I, Improved; NC, No change; R, Resolved; W, Worse

^aStandard deviation was included;

^bInterquartile range was included;

^cSeven patients had disc atrophy.

Table 3

Pre- and post-surgery description of visual parameters and headaches complains.

Authors & Year	No. Patient	Pre-Surgery				Post-Surgery				Complications
		Visual Field	Visual Acuity	Fundoscopic Evaluation	Headaches	Visual Field	Visual Acuity	Fundoscopic Evaluation	Headaches	
Patrocinio et al., 2005 ^{a,b}	1	L Moderate Superior Tempo-ral R Moderate Superior Inferior Temporal	L 20/20 R 20/50	Bilateral High Grade Papilledema	HA	L Superior Enlarged Blind Spot R Mild Superior Inferior Temporal	L 20/20 R 20/25	L mild R resolved	resolved	none
Gupta et al, 2007 ^a	Overall	9 inferonasal scotomas 3 constriction of visual fields 2 arcuate scotomas 2 altitudinal loss 1 temporal wedge defects 1 total loss of vision				The perimetric findings improved in all 18 patients (p < 0.005)				2 cases of nasal synechiae
	1		L 6/6 R 6/9	+	HA		L 6/6 R 6/6	reduced	resolved	
	2		L 6/24 R 6/36	+	HA		L 6/6 R 6/9	reduced	reduced	
	3		L 6/24 R 6/60	+	HA		L 6/9 R 6/24	reduced	reduced	
	4		L 6/24 R PL –	DA	HA		L 6/9 R PL –	DA	resolved	
	5		L 6/24 R HMCF	DP	HA		L 6/24 R 3/60	DP	reduced	
	6		L 6/36 R 3/60	DP	HA		L 6/18 R 6/36	DP	reduced	
	7		L 6/24 R 6/36	+	HA		L 6/6 R 6/9	reduced	reduced	
	8		L 6/60 R 6/24	+	HA		L 6/24 R 6/6	reduced	resolved	
	9		L HMCF R 6/36	DP	HA		L 6/36 R 6/9	DP	resolved	
	10		L PL + R 6/36	DP	HA		L 6/60 R 6/24	DP	reduced	
	11		L 6/18 R 6/60	+	HA		L 6/6 R 6/18	reduced	resolved	
	12		L 1/60 R 6/18	DP	HA		L 6/24 R 6/6	DP	reduced	

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Authors & Year	No. Patient	Pre-Surgery					Post-Surgery				Complications
		Visual Field	Visual Acuity	Fundoscopic Evaluation	Headaches	Visual Field	Visual Acuity	Fundoscopic Evaluation	Headaches		
	13		L 6/6 R 6/24	+	HA		L 6/6 R 6/6	reduced	reduced		
	14		L 6/36 R PL +	DP	HA		L 6/36 R 6/60	DP	reduced		
	15		L 6/24 R 6/60	+	HA		L 6/9 R 6/36	reduced	resolved		
	16		L 6/60 R 6/18	+	HA		L 6/36 R 6/9	reduced	reduced		
	17		L 6/36 R 6/6	+	HA		L 6/18 R 6/6	reduced	resolved		
	18		L 6/60 R 6/9	+	HA		L 6/36 R 6/9	DA	reduced		
Koc et al., 2008 ^c	1	NA	L 7/10 R 3/10	L + R +	HA	NA	L 7/10 R 3/10	L resolved R resolved	resolved	none	
	2	NA	L 5/10 R 9/10	L + R +	HA	NA	L 7/10 R 10/10	OA OA	R	none	
Berhouma et al., 2014 ^a	1	Normal	CF 10	Grade I	NA	normal	8/10	resolved	NA	none	
Sencer et al., 2014 ^{a,b,d,e}	Overall				7 pts w/HA				4 pts resolved HA 1 pt improved HA	1 case serous retinopathy	
	1	NA	NA	L + R +		improved	worsened	L improved R resolved			
	2	NA	NA	L + R +		improved	improved	L NC R NC			
	3	NA	NA	L + R +		improved	improved	L resolved R NC			
	4	NA	NA	L + R +		worsened	worsened	L improved R resolved			
	5	NA	NA	L - R DA		NC	NC	L improved R NC			
	6	NA	NA	L + R +		improved	improved	L NC R resolved			
	7	NA	NA	L + R +		improved	improved	L improved R NC			
	8	NA	NA	L + R +		improved	improved	L improved R NC			

Authors & Year	No. Patient	Pre-Surgery				Post-Surgery				Complications
		Visual Field	Visual Acuity	Fundoscopic Evaluation	Headaches	Visual Field	Visual Acuity	Fundoscopic Evaluation	Headaches	
	9	NA	NA	L + R +		improved	improved	L improved R improved		
	10	NA	NA	L + R +		improved	improved	L NC R improved		
Yildirim et al., 2015 ^c	1	L Superior Hemianopsia	L 2/10 R 7/10	+	HA	improved	L 4/10 R 7/10	Markedly regressed	improved	none
	2	L Both Superior Quadrants	L 4/10 R 9/10	+	HA	improved	L 5/10 R 9/10	Markedly regressed	resolved	none

Definitions: HMcF: The perception of hand motion at 2 feet; CF 10: counting fingers at 10cm; PL: perception of light (positive or negative); DP: Disc Pallor; DA: Disc Atrophy; OA: Optic Atrophy; +/- papilledema; HA: headaches reported; L:left; R right, NA: not available; NC: no change

^aUnilateral surgery on worst vision eye.

^bDid not opened Optic Nerve Sheath

^cBilateral surgery

^dVisual field improvement defined as more than 5 dB change in mean deviation values in Humphrey automated visual field analysis.

^eVisual acuity improvement defined as improvement of two or more lines on Snellen chart.