Refractory idiopathic intracranial hypertension treated with stereotactically planned ventriculoperitoneal shunt placement

CORMAC O. MAHER, M.D., JAMES A. GARRITY, M.D., AND FREDRIC B. MEYER, M.D.

Departments of Neurosurgery and Ophthalmology, Mayo Clinic, Rochester, Minnesota

Object. Ventriculoperitoneal (VP) shunts have not been widely used for idiopathic intracranial hypertension (IIH) because of the difficulty of placing a shunt into normal or small-sized ventricles. The authors report their experience with stereotactic placement of VP shunts for IIH.

Methods. The authors reviewed the clinical records of all patients in whom stereotaxis was used to guide the placement of a VP shunt for IIH at their institution. All shunts were placed using stereotactic guidance to target the frontal horn of the lateral ventricle. Patients were contacted at a mean postoperative interval of 15.1 months. No patients were lost to follow up.

The authors identified 13 patients who underwent placement of a stereotactically guided VP shunt for IIH over a 6-year period. A trial of either acetazolamide or steroid therapy had failed in all patients. Prior surgical treatments included optic nerve sheath fenestrations in seven patients and cerebrospinal fluid diversionary procedures, other than stereotactic VP shunt procedures, in nine patients. Twelve patients reported excellent or good durable symptomatic relief at the time of follow up. No patient suffered progression of visual deficits. Four patients experienced persistent headaches following the procedure. Three patients required a revision of the VP shunt for technical failure.

Conclusions. Stereotactically guided VP shunt placement is an effective and durable treatment option in many cases of IIH that are refractory to more traditional medical and surgical approaches.

KEY WORDS • intracranial hypertension • pseudotumor cerebri • ventriculoperitoneal shunt • stereotaxis

Idiopathic intracranial hypertension, also known as pseudotumor cerebri or benign intracranial hypertension, is a disorder of elevated ICP in the absence of any demonstrable disease.^{11,37,47} This disorder may affect children and adults, and women are more frequently affected than men.^{12,23} Typically, patients complain only of headaches. Except for papilledema and sixth cranial nerve paresis, neurological examination is usually normal.⁸ Chronic papilledema may lead to optic atrophy and progressive visual loss. Although subtle visual loss is present in a large majority of patients with IIH, significant loss of central vision is usually a late development.^{47,48} Patients with pseudotumor cerebri must undergo frequent ophthalmological examinations to identify early signs of visual deterioration.

There are several treatment modalities that appear to be effective. Acetazolamide does provide relief to some patients and is often the initial treatment of choice.^{8,18,21,49} Steroid therapy has also been used with some success.⁸ Idiopathic intracranial hypertension has been associated with obesity, and some patients do improve with weight loss.^{21,36} Additionally, special diets and gastric surgery have been suggested as possible treatment modalities.^{1,26,30,44}

Indications for surgical treatment of IIH include significant visual loss, progressive visual loss, or severe headache that persists despite medical intervention. Optic nerve sheath fenestration is a frequently performed surgical procedure for this condition, most often bilaterally in two stages. Optic nerve sheath fenestrations are effective at relieving papilledema and progressive visual loss in most cases.^{4,9,17,25} In contrast to CSF diversionary procedures, however, optic nerve sheath fenestrations are ineffective at relieving headaches. Reported complications of optic nerve sheath fenestrations include peripapillary hemorrhage, new visual field deficits, blindness, cranial nerve paresis, and stroke.^{3,15,32,41–43}

The most frequently performed CSF diversion procedure for IIH has been LP shunt placement.^{7,10,13,22,23,35} Lumboperitoneal shunts result in rapid resolution of symptoms of elevated ICP.²² Although LP shunts are quite effective at relieving intracranial hypertension, shunt failure and low pressure-induced headaches are frequently seen. The difficulty of introducing a catheter into ventricles of normal or small size has led most surgeons to favor LP over VP shunts in these patients. Stereotaxis, however, may be used to overcome this limitation of traditional VP shunt placement.^{16,29,31}

CLINICAL MATERIAL AND METHODS

We reviewed all cases of patients who underwent place-

Abbreviations used in this paper: CSF = cerebrospinal fluid; ICP = intracranial pressure; IIH = idiopathic intracranial hypertension; LP = lumboperitoneal; VP = ventriculoperitoneal.

Fig. 1. A frame-based stereotactic system is used for planning the ventricular catheter placement. The target (*left*) as well as entry (*right*) points are selected using stereotactic planning software.

ment of a VP shunt for IIH in which stereotaxis was used at our institution. We identified 13 patients in whom this procedure was performed between 1993 and 1999. There were 10 females and three males; their mean age was 31.5 years (range 6–54 years). The diagnosis of IIH was made by a neurologist based on history, examination, imaging studies, and ICP measurements when available. Formal ophthalmological testing as well as a standardized neurological examination (by an independent examiner) was undertaken preoperatively in each case.

We favor an occipital burr hole, as this allows for placement of the entire shunt system in a single sterile field, obviating the need for temporary closure and removal of the head frame. Patients undergo application of a stereotactic head frame followed by stereotactic computerized tomography scanning. Following data acquisition, patients are positioned supine on the operating table with the head turned 90° relative to the body and parallel to the floor. The shoulder may be raised slightly to facilitate head rotation. An entry point is chosen that is approximately 6 cm superior to the external occipital protuberance and 3 cm lateral to the midline. For posterior burr holes, a target is chosen in the anterior body of the lateral ventricle (Fig. 1). The catheter is advanced according to these coordinates. The peritoneal catheter is passed subcutaneously and placed in the abdomen. In a single case, a magnetic resonance imaging–based frameless stereotactic system was used (Fig. 2).

Patients attended a follow-up visit at a mean postoperative interval of 15.2 months (range 1–38 months). No patient was lost to follow up. Patients were asked about changes in vision, headaches, medication use, and any additional required surgical interventions.

RESULTS

Prior to the placement of a VP shunt, 11 patients suffered a loss of visual acuity and two had normal vision (Table 1). The mean interval between the onset of diagnostic symptoms and placement of a stereotactic VP shunt was 2.8 years (range 2 months–9 years). In all patients a trial of either acetazolamide or steroid medication had failed. Prior surgical treatments included optic nerve sheath fenestrations in seven patients and CSF diversion-

Fig. 2. Photograph showing placement of a ventricular catheter in which frameless stereotaxis is used.

ary procedures other than a stereotactic VP shunt placement in nine patients.

Of the 13 patients included in the analysis, 12 reported excellent or good durable symptomatic relief at the time of follow up (Table 2). No patients suffered progression of visual deficits. Four patients who presented with frequent headaches continued to experience persistent headaches following the procedure. Three patients required revision of the VP shunt for system failure: for proximal obstruction and distal obstruction in one case each; in the third case, the site of the obstruction was not recorded. There were no instances of infection or overdrainage.

DISCUSSION

There are many effective treatment modalities for IIH. Optic nerve sheath fenestration is the most frequently performed surgical treatment for IIH at our institution. Although generally considered to be a safe procedure, complication rates as high as 40% have been reported.³² The most frequently reported procedure-related complications are strabismus, peripapillary hemorrhage, and pupillary dysfunction. Rarely reported complications associated with optic nerve sheath fenestration include death, worsening of visual field deficits, permanent loss of vision and stroke.^{28,32,34,42,43} Furthermore, repeating an optic nerve sheath fenestration is usually not indicated. Secondary operations have a lower rate of symptomatic improvement and a higher complication rate compared with the primary procedure.^{32,42,43}

In contrast to optic nerve sheath fenestrations, CSF shunt procedures lower ICP. For this reason, shunts are

Stereotactic VP shunt for IIH

TABLE 1
Summary of characteristics in patients with IIH

Case No.	Age (yrs), Sex	Duration of Symptoms (mos)	Prior Surgery	Visual Symptoms
1	40, F	84	bilat optic nerve fenestrations & VP shunt	blurred vision & decreased visual fields
2	26, F	16	foramen magnum-to-atrial shunt	none
3	23, F	72	bilat optic nerve fenestrations	blurred vision
4	38, F	7	bilat optic nerve fenestrations	progressive visual loss
5	36, M	24	LP shunt	none
6	45, F	12	LP shunt	blurred vision
7	30, F	65	LP shunt	blurred vision
8	54, F	2	bilat optic nerve sheath fenestrations	visual obscurations
9	6, M	19	LP shunt	blurred vision
10	37, F	10	LP shunt	blurred vision
11	37, M	24	bilat optic nerve sheath fenestrations & LP shunt	blurred vision
12	27, F	108	bilat optic nerve fenestrations & multiple LP shunts	severe acuity loss
13	16, F	12	bilat optic nerve sheath fenestrations	severe acuity loss

more effective than optic nerve sheath fenestrations at relieving headaches.⁴⁰ In treating patients with IIH, shunts may be effectively placed in the lumbar cistern, the cisterna magna, or the ventricles. The use of LP shunts for this condition has been well described.^{5,13,19,20,22} Eggenberger, et al.,¹³ have reported that LP shunt therapy has relieved symptoms in all 27 patients who were studied. Shunt revision was required in 56% of these patients at a median follow-up period of 47 months, most often because the shunt became obstructed. This high rate of obstruction-related shunt failure has also been found in other series.^{7,35} There are reports of rapid progression of visual loss following LP shunt failure or removal of the system.^{27,33} Another frequent LP shunt-related complication is intracranial hypotension resulting from excessive drainage of CSF.^{7,13,22,35,39} Lumbar radiculopathy is a relatively frequent complaint following LP shunt placement.^{14,39} Eggenberger, et al., reported that five of their 27 patients who underwent LP shunt placement complained of radicular pain and three of the five required shunt revision for this reason. Tonsillar herniation has been reported following LP shunt placement.^{6,45,50} Chumas, et al.,⁶ have reported that 4.2% of 143 children treated with LP shunts, most of whom were being treated for communicating hydrocephalus, suffered symptomatic tonsillar herniation. Five of their patients required surgical decompression for the herniation and one child died as a result of the herniation. Because only 10 of the patients reported by Chumas, et al., were being treated for IIH, it is not clear if tonsillar herniation occurs with significant frequency in this subgroup of patients. Furthermore, the use of an adjustable-valve LP shunt may decrease the incidence of tonsillar herniation and posture-related headache.

TABLE 2
Summary of outcome data after VP shunt placement

Case No.	Follow-Up Visit			Shunt Revision or Failure
	Head-ache	Vision	Length (mos)	
1	no	stable	6	no
2	no	stable	6	no
3	no	improved	4	no
4	yes	improved	10	no
5	yes	stable	4	no
6	no	improved	6	no
7	no	improved	23	no
8	yes	stable	31	distal revision
9	yes	stable	29	total revision
10	no	stable	1	no
11	no	stable	2	no
12	no	slight improvement	1	no
13	no	stable	38	proximal revision

Ventricular shunts have been infrequently used in IIH because of the difficulty associated with cannulating small-sized ventricles. As has been described by Tulipan, et al.,⁴⁶ in a series of seven patients, current stereotactic techniques make placement of the ventricular catheter straightforward. Both LP and VP shunts are associated with a relatively low infection rate,^{20,39} although the infection rate of VP shunts may be slightly higher.² The rate of distal (that is, peritoneal) obstruction is low and probably equivalent to that seen in LP shunts.^{7,13,14,38} Subdural hematomas may be caused by treatment with both VP and LP shunts.²⁴ However, one of the most frequent and dangerous complications that follows LP shunt placement, tonsillar herniation, is not associated with VP shunt therapy. In one reported case of IIH, a ventricular shunt was successfully used to treat tonsillar herniation that resulted from the use of an LP shunt.⁴⁵

Our initial results support stereotactically guided VP shunt placement as an effective and durable treatment option in cases of refractory IIH. Stereotactic VP shunt has a long patency rate and a low incidence of the low-pressure headache syndrome. We have not attempted a direct comparison between the effectiveness of VP shunt procedures and other modalities, such as optic nerve fenestration or LP shunting, because our inclusion criteria selected those patients for whom optic nerve fenestration, LP shunts, acetazolamide, or steroid therapy were not effective.

References

1. Amaral JF, Tsiaris W, Morgan T, et al: Reversal of benign intracranial hypertension by surgically induced weight loss. **Arch Surg** 122:946-949, 1987

2. Aoki N: Lumboperitoneal shunt: clinical applications, complications, and comparison with ventriculoperitoneal shunt. **Neurosurgery** 26:998-1004, 1990

3. Brodsky MC, Rettele GA: Protracted postsurgical blindness with visual recovery following optic nerve sheath fenestration. **Arch Ophthalmol** 115:1473-1474, 1997

4. Brouman ND, Spoor TC, Ramocki JM: Optic nerve sheath decompression for pseudotumor cerebri. **Arch Ophthalmol** 106:1378-1383, 1988

5. Burgett RA, Purvin VA, Kawasaki A: Lumboperitoneal shunting for pseudotumor cerebri. **Neurology** 49:734–739, 1997
6. Chumas PD, Armstrong DC, Drake JM, et al: Tonsillar herniation: the rule rather than the exception after lumboperitoneal shunting in the pediatric population. **J Neurosurg** 78:568–573, 1993
7. Chumas PD, Kulkarni AV, Drake JM, et al: Lumboperitoneal shunting: a retrospective study in the pediatric population. **Neurosurgery** 32:376–383, 1993
8. Cinciripini GS, Donahue S, Borchert MS: Idiopathic intracranial hypertension in prepubertal pediatric patients: characteristics, treatment, and outcome. **Am J Ophthalmol** 127:178–182, 1999
9. Corbett JJ, Nerad JA, Tse DT, et al: Results of optic nerve sheath fenestration for pseudotumor cerebri. The lateral orbitotomy approach. **Arch Ophthalmol** 106:1391–1397, 1988
10. Cornblath WT, Miller NR: Pseudotumor cerebri treated with lumboperitoneal shunt. **Ann Neurol** 26:183, 1989 (Abstract)
11. Dandy WE: Intracranial pressure without brain tumor: diagnosis and treatment. **Ann Surg** 106:492–513, 1937
12. Durcan FJ, Corbett JJ, Wall M: The incidence of pseudotumor cerebri. Population studies in Iowa and Louisiana. **Arch Neurol** 45:875–877, 1988
13. Eggenberger ER, Miller NR, Vitale S: Lumboperitoneal shunt for the treatment of pseudotumor cerebri. **Neurology** 46:1524–1530, 1996
14. Eisenberg HM, Davidson RI, Shillito J Jr: Lumboperitoneal shunts. Review of 34 cases. **J Neurosurg** 35:427–431, 1971
15. Flynn WJ, Westfall CT, Weisman JS: Transient blindness after optic nerve sheath fenestration. **Am J Ophthalmol** 117:678–679, 1994
16. Frank EH, Quattrocchi KB, Hein L: An adaptor for stereotactic insertion of unitized shunt systems. **Surg Neurol** 35:400–402, 1991
17. Goh KY, Schatz NJ, Glaser JS: Optic nerve sheath fenestration for pseudotumor cerebri. **J Neuroophthalmol** 17:86–91, 1997
18. Gucer G, Viernstein L: Long-term intracranial pressure recording in the management of pseudotumor cerebri. **J Neurosurg** 49:256–263, 1978
19. Guy J, Johnston PK, Corbett JJ, et al: Treatment of visual loss in pseudotumor cerebri associated with uremia. **Neurology** 40:28–32, 1990
20. James HE, Tibbs PA: Diverse clinical applications of percutaneous lumboperitoneal shunts. **Neurosurgery** 8:39–42, 1981
21. Johnson LN, Krohel BG, Madsen RW, et al: The role of weight loss and acetazolamide in the treatment of idiopathic intracranial hypertension. **Ophthalmology** 105:2313–1317, 1998
22. Johnston I, Besser M, Morgan MK: Cerebrospinal fluid diversion in the treatment of benign intracranial hypertension. **J Neurosurg** 69:195–202, 1988
23. Johnston I, Paterson A, Besser M: The treatment of benign intracranial hypertension: a review of 134 cases. **Surg Neurol** 16:218–224, 1981
24. Jones RF: Long-term results in various treatments of hydrocephalus. **J Neurosurg** 26:313–315, 1967
25. Kelman SE, Heaps R, Wolf A, et al: Optic nerve decompression surgery improves visual function in patients with pseudotumor cerebri. **Neurosurgery** 30:391–395, 1992
26. Kupersmith MJ, Gamell L, Turbin R, et al: Effects of weight loss on the course of idiopathic intracranial hypertension in women. **Neurology** 50:1094–1098, 1998
27. Liu GT, Volpe NJ, Schatz NJ, et al: Severe sudden visual loss caused by pseudotumor cerebri and lumboperitoneal shunt failure. **Am J Ophthalmol** 122:129–131, 1996
28. Mauriello JA Jr, Shaderowsky P, Gizzi M, et al: Management of visual loss after optic nerve sheath decompression in patients with pseudotumor cerebri. **Ophthalmology** 102:441–445, 1995
29. McCallum J: Combined frameless stereotaxy and neuroendoscopy in placement of intracranial shunt catheters. **Pediatr Neurosurg** 26:127–129, 1997
30. Newborg B: Pseudotumor cerebri treated by rice reduction diet. **Arch Intern Med** 133:802–807, 1974
31. Pang D, Grabb PA: Accurate placement of coronal ventricular catheter using stereotactic coordinate-guided free-hand passage. Technical note. **J Neurosurg** 80:750–755, 1994
32. Plotnik JL, Kosmorsky GS: Operative complications of optic nerve sheath decompression. **Ophthalmology** 100:683–690, 1993
33. Repka MX, Miller NR, Savino PJ: Pseudotumor cerebri. **Am J Ophthalmol** 98:741–746, 1984
34. Rizzo JF III, Lessell S: Choroidal infarction after optic nerve sheath fenestration. **Ophthalmology** 101:1622–1626, 1994
35. Rosenberg ML, Corbett JJ, Smith C, et al: Cerebrospinal fluid diversion procedures in pseudotumor cerebri. **Neurology** 43:1071–1072, 1993
36. Rowe FJ, Sarkies NJ: The relationship between obesity and idiopathic intracranial hypertension. **Int J Obes Relat Metab Disord** 23:54–59, 1999
37. Rush JA: Pseudotumor cerebri: clinical profile and visual outcome in 63 patients. **Mayo Clin Proc** 55:541–546, 1980
38. Sainte-Rose C, Piatt JH, Renier D, et al: Mechanical complications in shunts. **Pediatr Neurosurg** 17:2–9, 1991
39. Selman WR, Spetzler RF, Wilson CB, et al: Percutaneous lumboperitoneal shunt: review of 130 cases. **Neurosurgery** 6:255–257, 1980
40. Sergott RC, Savino PJ, Bosley TM: Modified optic nerve sheath decompression provides long-term visual improvement for pseudotumor cerebri. **Arch Ophthalmol** 106:1384–1390, 1988
41. Smith KH, Wilkinson JT, Brindley GO: Combined third and sixth nerve paresis following optic nerve sheath fenestration. **J Clin Neuroophthalmol** 12:85–87, 1992
42. Spoor TC, McHenry JG: Complications of optic nerve sheath decompression. **Ophthalmology** 100:1432–1433, 1993 (Letter)
43. Spoor TC, McHenry JG: Long-term effectiveness of optic nerve sheath decompression for pseudotumor cerebri. **Arch Ophthalmol** 111:632–635, 1993
44. Sugerman HJ, Felton WL III, Salvant JB Jr, et al: Effects of surgically induced weight loss on idiopathic intracranial hypertension in morbid obesity. **Neurology** 45:1655–1659, 1995
45. Sullivan LP, Stears JC, Ringel SP: Resolution of syringomyelia and Chiari I malformation by ventriculoatrial shunting in a patient with pseudotumor cerebri and a lumboperitoneal shunt. **Neurosurgery** 22:744–747, 1988
46. Tulipan N, Lavin PJ, Copeland M: Stereotactic ventriculoperitoneal shunt for idiopathic intracranial hypertension: technical note. **Neurosurgery** 43:175–176, 1998
47. Wall M: Idiopathic intracranial hypertension. **Neurol Clin** 9:73–95, 1991
48. Wall M, George D: Visual loss in pseudotumor cerebri. Incidence and defects related to visual field strategy. **Arch Neurol** 44:170–175, 1987
49. Wandstrat TL, Phillips J: Pseudotumor cerebri responsive to acetazolamide. **Ann Pharmacother** 29:318, 1995 (Letter)
50. Welch K, Shillito J, Strand R, et al: Chiari I “malformations”—an acquired disorder? **J Neurosurg** 55:604–609, 1981

Manuscript received December 18, 2000.

Accepted in final form January 26, 2001.

Address reprint requests to: Cormac O. Maher, M.D., Mayo Clinic, 200 First Street, SW, Rochester, Minnesota 55905. email: maher.cormac@mayo.edu.