Refractory idiopathic intracranial hypertension treated with stereotactically planned ventriculoperitoneal shunt placement

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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. This disorder may affect children and adults, and women are more frequently affected than men. Typically, patients complain only of headaches. Except for papilledema and sixth cranial nerve paresis, neurological examination is usually normal. 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. 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. Steroid therapy has also been used with some success. Idiopathic intracranial hypertension has been associated with obesity, and some patients do improve with weight loss. Additionally, special diets and gastric surgery have been suggested as possible treatment modalities.

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. 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.

The most frequently performed CSF diversion procedure for IIH has been LP shunt placement. Lumboperitoneal shunts result in rapid resolution of symptoms of elevated ICP. 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.

CLINICAL MATERIAL AND METHODS
We reviewed all cases of patients who underwent place-
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 diversionary 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. 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. 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.

In contrast to optic nerve sheath fenestrations, CSF shunt procedures lower ICP. For this reason, shunts are
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.5,13,18,20,22 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 hypertension resulting from excessive drainage of CSF.7,13,22,35,39

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (yrs), Sex</th>
<th>Duration of Follow-Up Visit</th>
<th>Shunt Revision</th>
<th>Visual Symptoms</th>
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<tbody>
<tr>
<td>1</td>
<td>40, F</td>
<td>84</td>
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<tr>
<td>2</td>
<td>26, F</td>
<td>16</td>
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<td>3</td>
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<td>4</td>
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<td>7</td>
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<td>24</td>
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<td>6</td>
</tr>
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</table>

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. 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 latency 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


47. Unpublished data.