Ventricular shunt removal: the ultimate treatment of the slit ventricle syndrome

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Object. The aim of this study was to assess the effectiveness of an algorithm used to evaluate and prescribe treatment for patients having slit ventricle syndrome (SVS).

Methods. All patients included in this protocol underwent fiberoptic intracranial pressure monitoring after removal or externalization of their ventricular shunt systems. A significant number of patients did not need extracranial cerebrospinal fluid (CSF) diversion and tolerated removal of their shunt systems without requiring further intervention. Patients who demonstrated a need for CSF drainage underwent an endoscopic third ventriculostomy, regardless of the putative cause of their hydrocephalus. Sixteen (72.7%) of 22 patients experienced resolution of or significant improvement in their SVS complaints after their inclusion in the protocol. Concomitantly, 14 (64%) of 22 patients were no longer shunt dependent after a mean follow-up period of 21.4 months.

Conclusions. A significant number of patients debilitated by SVS may experience improvement in their symptoms and undergo shunt removal according to this protocol, improving their quality of life and simplifying their medical follow-up.

Key Words • slit ventricle syndrome • endoscopic third ventriculostomy
Management of patients with slit ventricle syndrome

Patients were admitted to the neurosurgical intensive care unit at one of two institutions and underwent a trial of iatrogenically maintained “shunt failure.” Extracranial CSF diversion was prevented by externalizing and occluding the patient’s distal shunt catheters in their peritoneal cavity, but inclusion in the study was not based on distal catheter site location. Enrollment was based only on the preexisting diagnosis of SVS and not on the supposed cause of the patient’s hydrocephalus. However, the majority of patients included in the study had a history that indicated some “obstructive” component underlying their hydrocephalus.

Monitoring of ICP

Patients were admitted to the neurosurgical intensive care unit at one of two institutions and underwent a trial of iatrogenically maintained “shunt failure.” Extracranial CSF diversion was prevented by externalizing and occluding the patient’s distal shunt system or by replacing the system with an external ventricular drain. In either circumstance, ICP was carefully documented in all patients by means of a fiberoptic intraparenchymal monitor (Ca- mino Lalis, San Diego, CA) that was inserted at the time of shunt manipulation. Patients who maintained asymptomatic ICP, whose conditions were stable on clinical examination, and who were found to have stable brain computerized tomography (CT) scans without CSF drainage for at least 72 hours were considered unlikely to need their shunt systems. The systems were removed in these individuals and they were discharged from the hospital with their family prepared to monitor them for evidence of clinical deterioration. Patients who demonstrated symptomatic ICP underwent endoscopic third ventriculostomy to treat their hydrocephalus. In all patients suffering from symptomatic ICP, we were able to dilate the slit ventricles sufficiently to allow intraventricular navigation with a rigid endoscope. This dilation always occurred within 48 hours of CSF outflow obstruction. Predictably, patients became acutely ill in the midst of this maneuver and required prompt surgical attention.

Surgical Technique

Endoscopic third ventriculostomy was performed as follows: a frontal burr hole was made and the right lateral ventricle was cannulated with a No. 14 French peelaway sheath. The foramen of Monro was identified with the aid of a 3-mm rigid endoscope. The third ventricle was entered and its floor was fenestrated by a hydrodissection technique while maintaining direct visualization. If the floor of the third ventricle was thickened and resistant to puncture, an alternative mechanical method was used, such as cautious blunt perforation with a balloon catheter or the tip of the endoscope. One of the senior authors (K.H.M.) preferred a thermal technique in which a monopolar radiofrequency electrode was used (ME2 Micro Endoscopic Electrode; Codman and Shurtleff, Inc., Rayn-
ham, MA), followed by dilation with an inflated No. 2 or 3 French balloon catheter. A third ventriculostomy of at least 5 mm was created to optimize chances for long-term patency. Before the peelaway sheath was withdrawn, an external ventricular drain was placed within the lateral ventricle so that CSF could be drained postoperatively as necessary.

At the completion of the procedure, absolute hemostasis was demonstrated both within the ventricular system and along the transcortical endoscope tract. Typically, the external ventricular drain was removed after 72 hours of monitoring that demonstrated asymptomatic ICP, at which time the third ventriculostomy was regarded as functional. Before the patient was discharged, a patent fenestration was further confirmed radiographically by means of cine magnetic resonance imaging.

Patients who experienced ICP difficulties after the endoscopic procedure were categorized in one of two groups: those who could not maintain a patent third ventriculostomy and those who could but who demonstrated an additional, more distal defect in CSF absorption. In the former group, the persistent hydrocephalus was managed by replacing the ventriculoperitoneal (VP) shunt system. In the latter group, however, conversion of the hydrocephalus to a solely “communicating” variety presented the alternative of placing a lumboperitoneal (LP) shunt for CSF diversion. The decision to replace the VP shunt or to insert an LP shunt was based on the attending physician's preference. All patients included in this protocol were scheduled for neurosurgical follow-up review after discharge and had a strong and well-educated familial support system with members who could monitor them for subsequent clinical difficulties.

### Results

In five (22.7%) of the 22 patients, ICP measurements remained consistently within normal limits despite the absence of CSF drainage. All five of these patients maintained stable clinical and radiographic examinations and

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**TABLE 1**

**Clinical summary of treatment of 22 patients with SVS**

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Cause of Hydrocephalus</th>
<th>Age at 1st Shunt Placement, Sex†</th>
<th>Age at Protocol Inclusion, yrs‡</th>
<th>No. of Shunt Revisions Pre-protocol§</th>
<th>Symptoms of SVS</th>
<th>Monitored ICP W/O CSF Drainage</th>
<th>Improved Symptoms Postprotocol</th>
<th>Surgical Intervention</th>
<th>Shunt-Free Postprotocol</th>
<th>Follow Up (mos)¶</th>
</tr>
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<tbody>
<tr>
<td>1</td>
<td>aqueductal stenosis</td>
<td>27 yrs, M</td>
<td>30</td>
<td>8</td>
<td>headache, nausea, violence, stupor bradycardia, Parinaud’s syndrome, decreased memory loss</td>
<td>increased</td>
<td>ETV</td>
<td>yes</td>
<td>yes</td>
<td>39</td>
</tr>
<tr>
<td>2</td>
<td>tectal astrocytoma</td>
<td>13 yrs, F</td>
<td>19</td>
<td>4</td>
<td>headache, Parinaud’s syndrome, decreased memory loss</td>
<td>increased</td>
<td>ETV</td>
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<td>yes</td>
<td>21</td>
</tr>
<tr>
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<td>1 wk, F</td>
<td>3</td>
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<td>29</td>
</tr>
<tr>
<td>4</td>
<td>IVH of prematurity</td>
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<td>25</td>
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<td>ETV</td>
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<td>yes</td>
<td>6</td>
</tr>
<tr>
<td>5</td>
<td>pineal astrocytoma</td>
<td>9 yrs, F</td>
<td>19</td>
<td>3</td>
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<td>yes</td>
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</tr>
<tr>
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<td>22</td>
<td>6</td>
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<td>increased</td>
<td>ETV</td>
<td>yes</td>
<td>yes</td>
<td>19</td>
</tr>
<tr>
<td>7</td>
<td>IVH of prematurity</td>
<td>1 wk, M</td>
<td>2</td>
<td>5</td>
<td>nausea, vomiting, irritability</td>
<td>increased</td>
<td>ETV</td>
<td>yes</td>
<td>yes</td>
<td>6</td>
</tr>
<tr>
<td>8</td>
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<td>8 wks, M</td>
<td>14</td>
<td>2</td>
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<td>ETW</td>
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<td>no</td>
<td>NA**</td>
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<td>1 wk, M</td>
<td>1.5</td>
<td>25</td>
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<td>ETW</td>
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<td>yes</td>
<td>11</td>
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<tr>
<td>10</td>
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<td>2 yrs, M</td>
<td>8</td>
<td>2</td>
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<td>normal</td>
<td>shunt removal</td>
<td>yes</td>
<td>yes</td>
<td>25</td>
</tr>
<tr>
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<td>15 yrs, F</td>
<td>32</td>
<td>12</td>
<td>headache, visual obsfuscation</td>
<td>normal</td>
<td>shunt removal</td>
<td>yes</td>
<td>no</td>
<td>15</td>
</tr>
<tr>
<td>12</td>
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<td>5 mos, F</td>
<td>12</td>
<td>7</td>
<td>headache, diplopia</td>
<td>increased</td>
<td>LP shunt</td>
<td>yes</td>
<td>no</td>
<td>38‡†</td>
</tr>
<tr>
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<td>aqueductal stenosis</td>
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<td>19</td>
<td>9</td>
<td>headache, lethargy</td>
<td>increased</td>
<td>ETV</td>
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<td>yes</td>
<td>15</td>
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<tr>
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<td>1 wk, F</td>
<td>15</td>
<td>6</td>
<td>fatigue, headache</td>
<td>normal</td>
<td>shunt removal</td>
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<td>yes</td>
<td>27</td>
</tr>
<tr>
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<td>15</td>
<td>2</td>
<td>fatigue, headache</td>
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<td>yes</td>
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<td>tectal glioma</td>
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<td>10</td>
<td>12</td>
<td>headache, visual obsfuscation</td>
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<td>ETV</td>
<td>no</td>
<td>no</td>
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<td>30</td>
<td>11</td>
<td>headache, Parinaud’s syndrome, memory loss</td>
<td>increased</td>
<td>ETV</td>
<td>yes</td>
<td>yes</td>
<td>15</td>
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<td>18</td>
<td>pineal ganglioglioma</td>
<td>1 yr, F</td>
<td>15</td>
<td>1</td>
<td>headache, irritability, lethargy</td>
<td>increased</td>
<td>ETV</td>
<td>no</td>
<td>no</td>
<td>NA**</td>
</tr>
<tr>
<td>19</td>
<td>myelomeningocele</td>
<td>1 wk, M</td>
<td>12</td>
<td>9</td>
<td>headache</td>
<td>increased</td>
<td>ETV</td>
<td>no</td>
<td>no</td>
<td>NA**</td>
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<tr>
<td>20</td>
<td>AVM w/ IVH</td>
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<td>19</td>
<td>4</td>
<td>headache</td>
<td>normal</td>
<td>shunt removal</td>
<td>no</td>
<td>yes</td>
<td>6‡‡</td>
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<tr>
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<td>IVH of prematurity</td>
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<td>headache, blurring vision</td>
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<td>ETV</td>
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<td>no</td>
<td>NA**</td>
</tr>
<tr>
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<td>1 wk, F</td>
<td>24</td>
<td>4</td>
<td>headache, nausea, vomiting, dizziness</td>
<td>increased</td>
<td>ETV</td>
<td>yes</td>
<td>yes</td>
<td>31</td>
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</tbody>
</table>

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* AS = aqueductal stent; AVM = arteriovenous malformation; ETV = endoscopic third ventriculostomy; NA = not applicable.
† Age at first shunt placement: mean age 6.5 years; median 8.5 months; range 1 week to 27 years.
‡ Age at protocol inclusion: mean age 17.3 years; median 15 years; range 1.5 to 49 years.
§ Number of shunt revisions before protocol: mean 8.0; median 6; range 1 to 25.
¶ Follow up after shunt independence: median 15.5 months; mean 21.4 months; range 6 to 45 months.
** VP shunt replaced after failed ETV.
†† LP exchanged for VP shunt.
‡‡ Repeat AVM hemorrhage with VP shunt reintension after 6 months without a shunt.
Management of patients with slit ventricle syndrome

tolerated discharge from the hospital without shunt systems. In three of these patients, their presenting headache complaints either improved substantially or resolved completely after CSF drainage was discontinued. The other two patients in this subgroup with normal ICP reported minimal changes in their headaches. One of these two patients later experienced improvement after receiving pharmacological treatment that had previously been ineffective. This patient did not have a family history of migraine but is thought to suffer from a form of “acquired” migraine as has been described previously in shunt-dependent children. Six months after discharge, the other patient suffered a severe intraventricular hemorrhage (IVH) related to an arteriovenous malformation and required reinsertion of a shunt for posthemorrhagic hydrocephalus. Four of these five patients have remained shunt free with stable clinical examinations and CT studies after a mean follow-up period of 28 months.

Of the remaining 17 patients, 16 underwent endoscopic third ventriculostomy for treatment of their hydrocephalus. Of these 17 patients, 10 maintained asymptomatic ICP. Their symptoms improved and they have sustained their shunt-free status after a mean follow-up period of 18.8 months. Endoscopic third ventriculostomy was unsuccessful in managing ICP in six patients, and in four of these patients failure of the endoscopic procedure was apparent before they were discharged from the intensive care unit. Of the two remaining patients, one presented with symptomatic hydrocephalus 2 weeks after undergoing third ventriculostomy and the other presented similarly 14 months postoperatively. In both of these “late” failures, repeated ventriculoscopy revealed a scarred-over ventriculostomy defect, and the endoscopic third ventriculostomy was repeated, resulting in subsequent 2-week and 3-month intervals of success, respectively. Five of the six patients with obstructed or otherwise unsuccessful ventriculostomies underwent reinsertion of a VP shunt system that incorporated either a high-pressure valve with an antisiphon device or a flow-controlled valve. The sixth patient was an infant with Apert’s syndrome and aqueductal stenosis. A secondary effort to treat his hydrocephalus by endoscopically guided aqueductal stent placement was unsuccessful by itself, but later placement of an LP shunt resolved his hydrocephalus and SVS complaints.

The remaining patient had been referred to us after a primary diagnosis of congenital aqueductal stenosis. After initiation of the protocol, however, findings on her CT scan appeared to be consistent with a communicating type of hydrocephalus, and aqueductal patency was demonstrated by means of intraventricular injection of contrast material. The patient and her family declined the endoscopic procedure, and an LP shunt was placed, which resulted in resolution of the patient’s hydrocephalus and SVS complaints.

No patient died during this protocol, and morbidity was limited to transient short-term memory loss in two patients who underwent the endoscopic procedure. In both cases, the cognitive deficits resolved within 6 months.

Discussion

Although the development of valve-regulated shunt systems in the 1950s represented a profound advance in the surgical management of hydrocephalus, SVS has been a troublesome complication associated with their use. This clinical syndrome is not to be confused with the radiographic finding of slit ventricles, the prevalence of which has been estimated in one review to be as high as 80% of children undergoing shunt placement and which was found in that study to be symptomatic in only 31 (11.5%) of 270 patients. Although other investigators have reported a correlation as high as 69% between the presence of SVS complaints and slit ventricles on CT scans in 75 patients, many individuals clearly tolerate the presence of slit ventricles with no apparent consequences. Within the subgroup of patients with shunts in place who display clinically significant manifestations of SVS, a substantial percentage improve with conservative treatment. Unfortunately, the patients whose symptoms are refractory to medical therapy typically present a management dilemma that has been disproportionately costly, challenging, and vexing for both patients and their physicians.

Treatment of SVS

Although various medical and surgical strategies have been advocated to treat patients with SVS, no single therapy has demonstrated uniform or long-standing success. In patients who have functioning shunts and who experience mild symptoms of SVS that minimally interfere with daily activities, nonsurgical options include nonnarcotic analgesic drugs, diuretic therapy, and antimigraine medications. In some circumstances, the efficacy of antimigraine therapy in patients with SVS is thought to be mediated through a primary migraine pathway, given the recognized association of migraine attacks with indwelling CSF shunts in young patients. Antimigraine medications are also thought to impart a vasomotor stability that limits vascular congestion and attendant intracranial hypertension. This point is particularly relevant to the management of patients with SVS, given their relatively noncompliant brains and severely limited capacity to buffer the increases in intracerebral blood volume that accompany vascular dilation. For patients with more severe symptoms that persist despite pharmacological treatment, surgical intervention is considered.

The most common surgical treatment for SVS has been shunt revision, the goal of which is to minimize symptomatic overdrainage of CSF. The preferred method of shunt modification varies among institutions but typically involves increasing the valve resistance of the apparatus, incorporating an antisiphon device within the shunt system, a combination of these maneuvers, or placement of a flow-control valve. Obviously, patency of the proximal and distal shunt components should be ensured at the time of revision.

First proposed by Epstein, et al., cranial expansion (that is, subtemporal decompression) for the treatment of SVS has successfully relieved symptoms and diminished the number of hospitalizations for shunt revision. The precise mechanism by which subtemporal decompression benefits patients with SVS remains controversial but is thought to involve a combination of prolonged proximal catheter patency and amelioration of ICP spikes. Calvarial and dural openings created during subtemporal...
decompression may induce an ipsilateral ventricular dilatation that improves the longevity of the proximal catheter.\textsuperscript{6,9} The surgical fontanelle created in these patients is also thought to dampen ICP waves that could otherwise contribute to symptoms of the SVS complex.

In more recent studies, third ventriculostomy has been advanced as a treatment option for SVS. Although typically regarded as a salvage procedure for patients in whom more conventional management efforts have failed, successful outcomes have been reported after this procedure was performed by open, stereotactic, and endoscopic routes.\textsuperscript{16,23,28}

**Outcomes of Third Ventriculostomy**

Reddy, et al.,\textsuperscript{21} reported encouraging outcomes after performing third ventriculostomy via a frontotemporal craniotomy in five patients with SVS in whom conventional medical and surgical interventions had failed. Walker, et al.,\textsuperscript{28} performed endoscopic third ventriculostomy to treat seven patients with SVS. Two patients from the latter study attained independence from shunts, and the SVS improved in three others. Unfortunately, the origins of the hydrocephalus in individual patients and the duration of postoperative follow-up review were not specified in that report. Walker, et al., prefer to reserve endoscopic third ventriculostomy for patients in whom all other available therapies, including subtemporal decompression, have failed. In our experience as reported here, 10 (62.5\%) of 16 patients no longer needed shunts and were cured of their SVS complaints by means of endoscopic third ventriculostomy at a mean follow-up period of 18.8 months.

Although the most promising candidates for successful endoscopic third ventriculostomy may be young adults suffering from an acquired nonneoplastic aqueductal stenosis,\textsuperscript{16} significant rates of shunt independence have been achieved both in younger patients and in those with other causes of hydrocephalus including congenital aqueductal stenosis, tumors, and myelomeningocele. These outcomes are well summarized in a review by Drake.\textsuperscript{4} Contrasting the low morbidity accompanying endoscopic third ventriculostomy with the profound advantages of independence from shunts, we elected to offer this procedure to all patients presenting with SVS (Table 2). The success of endoscopic third ventriculostomy in two patients who were shunt dependent as a consequence of previous IVHs illustrates the limitations of systematically excluding patients as candidates for this procedure based on the outdated and arbitrary designations of their hydro-}

**TABLE 2**

<table>
<thead>
<tr>
<th>Cause of hydrocephalus</th>
<th>Successful</th>
<th>Failed</th>
</tr>
</thead>
<tbody>
<tr>
<td>acquired aqueductal stenosis</td>
<td>3</td>
<td>0</td>
</tr>
<tr>
<td>congenital aqueductal stenosis</td>
<td>3</td>
<td>2</td>
</tr>
<tr>
<td>tectal/pineal/cerebellar tumor</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>IVH</td>
<td>2</td>
<td>1</td>
</tr>
<tr>
<td>myelomeningocele</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>total</td>
<td>10</td>
<td>6</td>
</tr>
</tbody>
</table>

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cephalus as either “obstructive” or “communicating.” By definition, all patients with hydrocephalus have an obstruction to CSF absorption at some point along their CSF circulatory pathway and the more accurate descriptors previously offered by Ransohoff, et al.,\textsuperscript{22} of hydrocephalus as either intra- or extraventricular remain most meaningful. One would expect endoscopic third ventriculostomy to be successful whenever the ventricular fenestration bypasses the obstruction, whether it exists within the ventricular system or at the level of the basal cisterns. Furthermore, if symptomatic hydrocephalus is related to a series of partial circulatory obstructions, circumventing some of these points by means of endoscopic third ventriculostomy might be sufficient to enable patients to compensate for their remaining disease and to avoid dependence on a shunt system. In this study, the SVS complaints of 16 (72.7\%) of 22 patients resolved or improved markedly after either VP shunt removal (four patients), VP shunt removal followed by endoscopic third ventriculostomy (10 patients), or conversion of an existing VP to an LP system (two patients).

**Significance of ICP Monitoring**

Monitoring of ICP can clarify the origin of headaches in patients with shunts and is valuable in the diagnostic evaluation of untreated ventriculomegaly.\textsuperscript{1,11,24} Patients with symptomatic slit ventricles may be habituated to significantly low ICP in the upright position (mean \(-239 \text{ mm H}_2\text{O}\)), and clinical sequelae have resulted from attempts to raise ICP abruptly to a more normal range.\textsuperscript{10} Consequently, “normal” ICP readings by conventional standards can actually reflect a relatively elevated and pathological ICP for patients with SVS.

To assess the impact of obstructing CSF drainage or the efficacy of endoscopic third ventriculostomy during this study, we therefore focused on “asymptomatic” as opposed to “normal” ICP values. Through our protocol, we identified five patients who had previously undergone shunt placement for treatment of hydrocephalus from disparate causes and who had regained the intrinsic capacity to manage their CSF dynamics and no longer required their shunt systems. This finding illustrates the importance of rigorously reevaluating patients deemed shunt dependent because of their history or by classic criteria when they present with symptoms suggesting shunt overdrainage.

**Advantages and Disadvantages of Endoscopic Third Ventriculostomy**

In managing patients with hydrocephalus, the opportunity to avoid placement of a shunt or to remove a preexisting shunt can be considered the ultimate triumph. The benefit of achieving independence from shunts is multifaceted and cannot be overstated. In addition to avoiding the long-term complications that accompany shunt dependence,\textsuperscript{25} including a 2.9\% mortality rate (reported over 17 years for 70 patients who received the devices during their first 2 years of life),\textsuperscript{27} shunt removal lifts an emotional burden and social stigma that many patients ascribe directly to the physical presence of their shunt system. Furthermore, when shunt dysfunction is not a diagnostic consideration for families and general medical practition-
ers who encounter patients with a history of hydrocephalus, anxiety is alleviated, the threshold for which a neurosurgical referral would otherwise have been pursued is raised, and the overall care of these patients is simplified.

In experienced hands, the risks associated with endoscopic third ventriculostomy are low. The most likely sources of morbidity include infection, injury to adjacent neural structures, and acute or delayed hemorrhage. In a single case report, cardiac asystole occurred after fenestration of the third ventricular floor. Aside from arterial catastrophe, most postoperative complications have been reported as transient. Our experience, which included only two patients who suffered short-term memory losses that corrected within 6 months of their procedures, mirrors this finding.

However, patients with SVS pose an inherently greater challenge to surgeons contemplating endoscopic ventriculoscopy and ventriculostomy. The anatomical spaces available for maneuvering an endoscope are relatively small in these patients, and obscured intraventricular landmarks typically accompany a history of multiple previous shunt placement procedures, which is almost ubiquitous in the population of patients with SVS. These impediments to endoscopic surgery are obviously intensified when accompanied by intercurrent bouts of ventriculitis or shunt infection. Neurosurgeons planning to perform endoscopic third ventriculostomy in patients with SVS should have experience with this procedure under more favorable circumstances.

It is also worth repeating that patients with SVS typically are made very ill during the course of dilating their ventricles as described in this protocol. Preparations must be made to facilitate surgical intervention on an emergency basis as soon as sufficient ventricular size is achieved. Given our experience with the one patient who developed an occlusion of his third ventriculostomy 14 months postsurgery, our current opinion is that all patients warrant long-term scheduled neurosurgical follow-up review after endoscopic third ventriculostomy.

Conclusions

Patients with SVS can be safely evaluated for their continued need for extracranial CSF diversion. The fact that the intrinsic CSF absorptive or circulatory mechanisms normalized over time in several patients underscores how the designation of patients as “shunt dependent” can become a self-fulfilling prophecy when based only on historical and untested criteria. For most patients who demonstrate persistent hydrocephalus, we advocate endoscopic third ventriculostomy as a means to treat their SVS complaints and to obviate the need for shunt hardware. Finally, eradication of SVS can be advanced as further justification for advocating endoscopic third ventriculostomy as the initial therapeutic option for patients newly presenting with symptomatic hydrocephalus, particularly in the age and etiological subgroups that have experienced the greatest statistical success with this procedure.

References


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