The role of lumbar shunts in the management of slit ventricles: does the slit-ventricle syndrome exist?

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Objective. slit-ventricle syndrome (SVS) is a confusing description of presentations in patients with chronic shunt-treated hydrocephalus. These patients are prone to acute deterioration with recurrent malfunction. The authors describe the clinical profile and management outcome of using lumboventricular (LP) shunts in this population of patients.

Methods. Thirty-three patients with slit ventricles and recurrent malfunctions were converted to LP shunts (mean age 12 years). The initial ventricular shunt was placed at a mean age of 16.5 months. Ten patients had failed endoscopic third ventriculostomies prior to placement of their LP shunt. At a previous presentation, in 11 patients suspected to have SVS following revision of the shunt, intracranial pressure normalized after insertion of a contralateral shunt, suggesting that their ventricles were isolated. The rate of infection and malfunction was compared before and after conversion to an LP shunt.

Twenty-seven patients were successfully converted to LP shunts. Four of the 11 patients with isolated ventricles required ventricular shunts in addition to the LP shunt. During a mean follow-up period of 16.7 months, the malfunction rate per patient decreased from 4.81 for ventriculopercutaneous shunts, prior to conversion to 1.48 after conversion to LP shunts, a statistically significant reduction (p < 0.000). No significant difference was found in the rate of shunt infections (7.1% for VP shunts and 9.6% for LP shunts, p = 0.44). No patient presented with acute symptoms following malfunction of an LP shunt or suffered from a Chiari I malformation.

Conclusions. Conversion to an LP shunt is a safe and effective procedure in patients prone to rapid decompensation and recurrent shunt malfunctions from small, slitlike ventricles. The term SVS is confusing. The condition is a manifestation of an unrecognized slitlike isolated ventricle and should be abandoned.

Key Words • hydrocephalus • slit ventricle syndrome • shunt malfunction • lumbar shunt • pediatric neurosurgery

The term SVS was introduced to describe patients with symptoms of increased ICP, small slitlike ventricles, and a slow-filling, clinically functioning shunt.1,2,3 It has evolved into a confusing spectrum of presentations in patients with shunt-treated hydrocephalus. Numerous opinions and recommendations are found in the literature regarding definitions, causes, and treatments of these symptoms. Several authors have attempted to subclassify patients with SVS into those having overdrainage and a low ICP and those having a high ICP. The latter are further categorized into patients with a functioning, intermittently functioning, or malfunctioning shunt.4,5,6,23 A uniform consensus, however, remains elusive.5,6,12,23,25 On one end of the spectrum is an asymptomatic patient with a functioning shunt in whom a CT scan reveals small ventricles.

Abbreviations used in this paper: CSF = cerebrospinal fluid; CT = computed tomography; EVD = external ventricular drain; ICP = intracranial pressure; LP = lumboventricular; SVS = slit-ventricle syndrome; VP = ventriculopercutaneous.

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At the other end of the spectrum is an acutely ill child with a shunt malfunction whose CT scan demonstrates small ventricles and who is at risk of death from elevated ICP.

The treatment recommendations for SVS remain as varied as the definitions and are often dictated by the classification or subclassification used.4,5,7,12,23,24,25,38,29,41 Conservative measures include observation, hydration, diuresis, antimigraine therapy, and use of corticosteroid agents. Surgical management typically ensues, with a shunt exploration and revision. Oftentimes the proximal shunt is changed or a higher-pressure valve and/or antishock device is used. In certain situations, subtemporal decompression or other cranial expansion techniques have been advocated.4,5,12,25 Recent interest in using LP shunts in these patients with increased ICP has met with success.4,5,7,12

To avoid confusion, we herein abandon the term SVS in favor of the pathophysiological description. We present our experience with conversion of ventricular shunts to lumbar shunts in patients with slitlike ventricles, recurrent malfunction, and frequent episodes of acute deterioration.

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Clinical Material and Methods
Thirty-three patients who had presented between 1998 and 2003 at the Children’s Hospital of Michigan with recurrent shunt malfunctions, slit ventricles, and documented elevated ICP were considered for placement of LP shunts. All patients had episodes of acute clinical deterioration from increased ICP despite slit ventricles visible on a CT scan. There were 23 male and 10 female patients (mean age 12.7 ± 7.9 years; range 4–42 years). Medical had ventriculomegaly (15 patients) or posthemorrhagic hydrocephalus (eight patients). The mean age at first shunt placement was 16.5 months (range 1–108 months). The mean duration for which the patients had a shunt was 112 months (range 36–264 months), with a mean of 26 revisions. Eleven patients had more than one ventricular catheter placed during a previous surgery. These patients had presented with slit-like ventricles and a high ICP, measured at surgery by using a manometer. The high pressure did not come down even after replacement of the obstructed proximal catheter. A second catheter was inserted on the contralateral side at the same surgery that was successful in normalizing the pressure as measured by a manometer.

Patients were evaluated in the standard fashion, which included a shunt tap, shunt injection, and/or a CT scan. At surgery, the obstructed proximal catheter was removed and an EVD was inserted. A Codman ICP monitor (Codman, Inc., Raynham, MA) and a percutaneous lumbar drain were placed. The patient then underwent CT scanning with intra-ventricular contrast as well as magnetic resonance imaging. A fracture guide was used to guide endoscopic third ventriculostomy was performed in patients who had obstructive hydrocephalus. This procedure was accomplished without difficulty despite the small size of the third ventricle. An endoscopic fenestration of the septum pellicudum was performed in patients suffering from obstruction at the level of the foramen of Monro. An endoscopic fenestration at the level of the atrium was attempted in patients with an isolated temporal horn. Once the diagnosis of a communicating ventricle was confirmed on a repeated CT ventriculogram, the EVD was clamped for a mean of 2.5 days. If this procedure was well tolerated, the lumbar drain was then converted to an LP shunt. A horizontal–vertical valve appropriate in vertical pressure to the height of the patient was inserted in the lumbar shunt to minimize overdrainage in all patients. The EVD and ICP monitors were removed and a ventricular reservoir was left for future access. In addition to the LP shunt, patients who continued to have isolated ventricles underwent either replacement of their VP shunt to drain the isolated ventricle or insertion of a ventriculocerebral shunt from the isolated ventricle to the communicating ventricle. Procedures in patients who failed to tolerate EVD clamping were considered failures, so their VP shunts were reinstituted and their LP drains removed.

Results
In 11 patients who had required a second ventricular catheter during a previous surgery for a suspected isolated slit ventricle, a CT ventriculogram confirmed an isolated ventricle on the side of the initial single shunt: five at the level of the foramen of Monro and six at the level of the atrium resulting in an isolated temporal horn. Endoscopic fenestration of the septum pellicudum was successful in all five patients who had obstruction at the level of foramen of Monro, but none of the six with endoscopic patients after the level of the atrium had successful outcomes. One of these six patients underwent a ventriculocerebral shunt and three required a VP shunt in addition to the LP shunt. Two of the six patients’ conversions to an LP shunt failed due to complication of VP shunt.

Ten patients had obstructive hydrocephalus diagnosed using CT ventriculography and had failed to respond to an endoscopic third ventriculostomy. After confirming patency of the ventriculostomy, six patients underwent placement of an LP shunt. Four patients failed to tolerate a lumbar drain and were switched back to a ventricular shunt. In 12 patients good communication to the spinal subarachnoid space was evident and all 12 were successfully converted to LP shunts.

Overall, 27 of the 33 patients were successfully converted to LP shunts. Each patient was reviewed individually. The number of LP shunt malfunctions and infections during the follow-up period were compared with the number of VP shunt malfunctions and infections over the same number of months prior to LP shunt conversion. The mean follow-up period was 16.7 months (range 3–63 months). A mean of 4.81 shunt malfunctions per patient occurred for the 27 patients with VP shunts, during a mean of 16.7 months prior to conversion to LP shunts. This rate compared with a mean of 1.48 malfunctions per patient for the 5-month period of the LP shunts during a mean follow-up period of 16.7 months. Using the paired Student t-test, a statistically significant (p < 0.0001) decrease in the revision rate for malfunction was seen following conversion to LP shunts. Proximal malfunction was significantly less with lumbar than with ventricular shunts (Table 1).

In the six patients whose conversions to LP shunts failed, the mean number of revisions of their VP shunts during a mean of 14.3 months prior to attempted conversion was not significantly different from the revision rate for VP shunts during a mean 14.3-month follow-up period (5.33 compared with 4.33, p = 0.34).

The rate of infection per procedure per patient was 7.1 ± 2% (standard error of mean) for VP shunts and 9.6 ± 3% (standard error of mean) for LP shunts. This difference was not statistically significant (p = 0.44).

Seven patients had the same patient swelling at the site of the LP shunt. In none of them was a discontinuity in the system seen on preoperative x-ray films, nor did they experience malfunctioning shunts during surgery. The condition was effectively resolved with a single blood patch placed at a

<table>
<thead>
<tr>
<th>Malfunction Rate and Type</th>
<th>VP (mean ± SD)</th>
<th>LP (mean ± SD)</th>
<th>Significance</th>
</tr>
</thead>
<tbody>
<tr>
<td>overall†</td>
<td>4.81 ± 4.00</td>
<td>1.48 ± 2.30</td>
<td>p &lt; 0.0000</td>
</tr>
<tr>
<td>proximal</td>
<td>4.04 ± 3.70</td>
<td>0.11 ± 0.30</td>
<td>p &lt; 0.0000</td>
</tr>
<tr>
<td>valve</td>
<td>0.56 ± 0.90</td>
<td>0.70 ± 1.30</td>
<td>NS</td>
</tr>
<tr>
<td>distal</td>
<td>0.22 ± 0.40</td>
<td>0.37 ± 0.90</td>
<td>NS</td>
</tr>
<tr>
<td>shunt site swelling</td>
<td>0</td>
<td>0.30 ± 0.50</td>
<td>p &lt; 0.0000</td>
</tr>
<tr>
<td>infection</td>
<td>0.44 ± 0.70</td>
<td>0.56 ± 1.00</td>
<td>NS</td>
</tr>
</tbody>
</table>

* NS = not significant; SD = standard deviation.
† Mean number of malfunctions per patient during the follow-up period.

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level above the LP shunt in five patients and with multiple patches in two patients.

During the follow-up period, no patient presented with acute deterioration or bradycardia following shunt malfunction. Additionally, in no patient was there radiographic or clinical evidence to suggest development of acquired Chiari I malformation.

Discussion

Increased ICP With a Functioning Shunt: Isolated Silt Ventricles

Eleven patients who had previously presented with slit ventricles and acute clinical deterioration had maintained a high ICP despite drainage of CSF after adjustment of the proximal catheter. These patients had required placement of a ventricular catheter on the contralateral side. When these patients were put through the protocol of lumbar drain and EVD placement, the CT ventriculogram confirmed that the ventricle on the side of the initially functioning shunt was isolated and was not draining the rest of the system. Because altered pathophysiology prevents ventricular dilation in chronically shunt-treated patients, it becomes impossible to recognize this condition on CT scans or MR images unless intraventricular contrast is used. We believe that patients reported in the literature as having SVS with high ICP and a functioning shunt are truly patients with unrecognized siltate isolated ventricles. The functioning shunt drains the isolated ventricle, whereas the undrained part of the ventricular system maintains the high ICP.

The report by Le, et al., on seven patients in whom the addition of an LP shunt to the existing functioning VP shunt resolved the symptoms of "SVS" appears to represent an unrecognized isolated slit ventricle on the side of the functioning shunt rather than an improvement in the buffering capacity, as conjectured by the authors. This situation may well be true for patients of so-called SVS, who improve after a third ventriculostomy while retaining a shunt, undergoing upgrading of the valve or experiencing subtemporal decompression (as discussed later).

Prolonged ventricular shunt treatment is known to result in siltate ventricular isolation.22 One explanation may be that the normal flow of CSF from the lateral ventricles toward the third ventricle maintains the patency of the foramen of Monro. Rekate, et al., have shown that a reversal of flow from the third ventricle to the lateral ventricle results in the shifting of the septum pellucidum to the side of the shunt and produces a functional valvular obstruction of the ipsilateral foramen of Monro that may over time become permanent (Fig. 1A). Likewise, in patients who have a parietal shunt, reversal of flow augments the narrowing at the body of the lateral ventricle and results in the isolation of the posterior part of the lateral ventricle and the temporal horn (Fig. 1B). In some of these patients who have a functional closure, dilation of the ipsilateral ventricle induced by upgrading the valve,21,23,26 by performing ipsilateral subtemporal decompression,12,13 or by cranial expansion may relieve the functional obstruction and provide symptomatic—although usually temporary—relief.5,22 Furthermore, Buxton, et al., have shown that, in fact, patients who underwent cranial decompression had a 68% increase in the rate of revision during the first 3 years of follow up.

Endoscopic fenestration of the septum was uniformly successful in patients with obstruction at the level of the foramen of Monro (Fig. 2A-C). We did not have success with endoscopic procedures in posterior isolation (Fig. 2D and E). The larger surface area created by fenestration of the ventricular atrium has a propensity to reclose in comparison with fenestration of a thin septum. In addition to LP shunts, retention of VP shunts on the side of the isolated ventricle is an effective strategy suggested by a good outcome in three of our patients and in seven patients reported on by Le, et al., although we believe that a ventriculoventricular shunt (Fig. 2E) may be a better alternative. Because no siphoning force exists in a ventriculoventricular shunt, it may eliminate the problem of overdrainage that causes subsequent coaptation of the ventricle and leads to proximal malfunction from catheter occlusion. Furthermore, the bidirectional flow through the ventriculoventricular shunt may prevent future obstruction. A larger series is required to substantiate this hypothesis.

Prolonged shunt treatment can also lead to secondary aqueductal obstruction.11,12 The entrapped cortical subarachnoid CSF that is unable to exit through the arachnoid villi or to drain through the ventricular shunt because of development of acquired aqueductal obstruction may maintain a high ICP. The drainage of this fluid may be best accomplished by LP shunt treatment or by a third ventriculostomy, as reported previously.22

Increased ICP With a Functioning Shunt: Shunt Pseudotumor

We did not encounter any patient with a functioning shunt, slit ventricles, and a communicating ventricular system, although such an entity has been described as a combination of pseudotumor cerebri and shunted hydrocephalus.2 These patients have hydrocephalus, secondary to increased venous pressure from venous outlet obstruction related to craniostenosis.23 Presumably, after shunt placement, slitlike ventricles develop in these patients, but the high venous pressure maintains high ICP. The rarity of the entity demands that every effort be made to rule out isolat-
ed slit ventricles in a patient presenting with high ICP, slit ventricles, and a functioning shunt before labeling the condition a "shunt pseudotumor."

**Outcome After Receiving LP Shunts**

All patients experienced resolution of their symptoms following placement of an LP shunt. Most importantly, none of them subsequently presented with acute deterioration or bradycardia. This type of acute deterioration can result from changes in intracranial venous compliance induced by the chronic overdrainage from a ventricular shunt. At the time of shunt malfunction a rapid collapse of bridging veins occurs, followed by an acute rise in ICP. Presumably, the lumbar shunt has a greater influence on the compliance of the epidural venous plexus; therefore, the intracranial venous system is relatively spared at malfunction, preventing acute deterioration. Alternatively, McLaurin and Olivi believe that LP shunts, being distant from the ventricles, do not dampen the intraventricular pulse pressure and therefore are unlikely to promote formation of slit ventricles.

Some evidence in the literature supports the use of LP shunts in patients with slit ventricles. Ide, et al. described one patient with slit ventricles secondary to shunt treatment for posttraumatic hydrocephalus. A CT cisternogram confirmed communicating hydrocephalus, and the symptoms completely resolved after placement of an LP shunt. Rakete and Wallace performed a retrospective review of 25 patients with LP shunts and described nine patients with a severe form of slit ventricles, incapacitating headaches, and recurrent proximal shunt failure. They did not, however, describe the effect of LP shunts on the rate of subsequent malfunctions. Finally, Le, et al. retrospectively reviewed seven patients with a diagnosis of SVS who had presented with intermittent symptoms of apparent shunt malfunction, small ventricles visible on CT scans, and a functioning shunt demonstrated either by shunt tap or seen at exploration. All seven patients received an LP shunt while maintaining their VP shunts. No patient required a revision during the follow-up period of 2 to 25 months.

Our study shows that the LP shunt clearly had a favorable influence on the rate of malfunction. The high rate of shunt malfunction in patients with small ventricles is related to collapse of the ventricles, bringing the choroid plexus into proximity with the proximal catheter and thereby allowing the plexus to grow into the catheter. Moving the catheter into the lumbar spine understandably reduced the rate of proximal malfunction significantly (Table 1). Furthermore, despite the prolonged period of externalization that some patients had to undergo during the evaluation for conversion to an LP shunt, we found no significant difference in the rate of infection. Aoki reported a 40% lower incidence of LP shunt malfunction and a lower rate of infection in pediatric patients compared with those with a VP shunt.

**Complications With LP Shunts**

A unique complication rarely discussed in the adult literature is the type of recurrent swelling around the LP shunt site that developed in seven of our patients and that probably arose from a leak around the proximal catheter. In children, the smaller bulk of the spinal musculature may fail to contain the minor leakage that otherwise is expected around the catheter. The condition was effectively managed with a single blood patch in five patients and multiple blood patches in the remaining two patients.

Arguments have been made that the placement of LP shunts...
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shunts increases the risk of hindbrain herniation (Chiari I malformation), Authors of several articles in the early 1990s reported a large percentage of patients with LP shunts demonstrated radiographic evidence of hindbrain herniation, with a smaller percentage becoming symptomatic. None of these authors reports the use of the horizontal–vertical valve system (Integra Neurosciences Implants, Sophia Antipolis Cedex, France), which offers higher resistance in the upright position to minimize overdrainage with LP shunts. Instead, a straight catheter or T shunt with a distal slit valve was used in these series. It is likely that the use of the horizontal–vertical valve in our patients prevents the long-term consequences of LP shunt treatment. This theory corroborates the observations made by Rekate and Wallace in their review of 25 patients who received LP shunts for several conditions. In none of the patients was there symptomatic or radiographic evidence of Chiari I malformation. Their explanation for discordance with prior data rests on three points. First, the LP shunts were placed in an older patient population; second, they were placed percutaneously; and last, the horizontal–vertical high-resistance valves were used. Our patient population appears similar to that of Rekate and Wallace in this regard. In their 9.6-year follow-up study, Rekate and Wallace suggest that LP shunts in older children can be used without excessive risk of hindbrain herniation.

Conclusions

Slit-ventricle syndrome is a confusing term and should be abandoned in favor of a pathophysiological description. Patients with slitlike ventricles, increased ICP, and a functioning shunt likely have an unrecognized isolated ventricle. These patients and those with recurrent proximal malfunctions are best treated using a combination of LP shunts, third ventriculostomy, fenestration of the isolated ventricle, or placement of a ventriculovenous shunt. In this patient population, placement of an LP shunt appears to be a safe and effective intervention that significantly reduces the rate and severity of subsequent malfunctions.

References


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