The role of lumboperitoneal shunts in the treatment of syringomyelia

Clinical article

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Object. The role of thecoperitoneal shunts in the management of syringomyelia is not well defined. In this study, the authors analyze the outcome of lumboperitoneal shunt procedures carried out to treat syringomyelia in their institution.

Methods. The authors retrospectively reviewed the medical records of 19 patients who underwent lumboperitoneal shunt procedures for syringomyelia.

Results. The mean follow-up duration was 25 months (range 3–51 months). Of 16 cases followed up, only 5 patients reported clinical improvement in their preoperative symptoms, but of these, 2 had clear radiological evidence of improvement. Three of 6 patients with syringomyelia due to spinal arachnoiditis improved.

Conclusions. Lumboperitoneal shunts may lead to useful improvement in the symptoms of a patient with syringomyelia while avoiding the risk of neurological deterioration inherent in myelotomies required for syrinx shunting procedures. (DOI: 10.3171/2010.3.SPINE0964)

Key Words • syringomyelia • lumboperitoneal shunt • shunting

Syringomyelia can be caused by a variety of underlying pathological entities, but the common feature, in most cases, is a blockage in the spinal subarachnoid channels, which causes a pressure differential to develop across the 2 sides of the obstruction. The resulting abnormal hydrodynamic forces then act through the perivascular spaces of the cord parenchyma to create the syrinx.1,2,10 The rational method of treatment, therefore, is to unblock the CSF channels whenever feasible. Craniocervical decompression for hindbrain hernia is the best example of this approach and results are usually satisfactory. With posttraumatic syringomyelia, laminectomy combined with creation of a conduit for CSF flow represent the equivalent approach, although success rates with this form of surgery are less than with hindbrain related syringomyelia.

Sometimes surgery to create free flow of CSF proves unsuccessful. On other occasions fibrosis may be too extensive for any realistic attempts to be made. At times it proves impossible to identify a focal point of obstruction to CSF flow. Thus, when normal CSF circulation cannot be reestablished, direct drainage of syringomyelia cavities, using syringosubarachnoid, syringopleural, and syringoperitoneal shunts, is a well-defined and accepted treatment modality.3,11,14,15,21 The main disadvantage of such procedures is that they carry the risk of worsening existing neurological deficits because of the myelotomy required to place the shunt tubing into the syrinx cavity.16

The role of thecoperitoneal shunts in the management of syringomyelia is less well defined. The rationale for using such systems depends on whether CSF is drained from above or below the intraspinal block. On the one hand, a shunt placed above aims to reduce the filling pressure, thus preventing syrinx progression. On the other hand, drainage from below may encourage bulk flow of water through the cord, thereby reducing its accumulation within the syrinx cavity. Thecoperitoneal shunts have the particular advantage of avoiding the risks associated with a myelotomy, and LP shunts have a low risk of producing a neurological deficit.

Abbreviation used in this paper: LP = lumboperitoneal.
In this study, we analyze the results of LP shunting procedures carried out to treat syringomyelia, in our unit.

Methods

Study Method

We reviewed the medical records and radiological findings of 19 patients who had undergone LP shunt procedures for syringomyelia, in our unit, between October 2001 and May 2006. Data extracted included age, sex, and underlying etiology, as well as clinical and radiological outcome.

Surgical Technique

The senior author (G.F.) performed or supervised all the surgical procedures. For all LP shunt placement procedures, the patients were placed in a lateral decubitus position. Following cleaning and draping, a midline lumbar incision centered over L3–4 interspinous space was made. A Tuohy needle was then inserted into the L3–4 or L4–5 interspinous space and the LP shunt catheter inserted. In all cases a Medtronic LP shunt catheter was used. This system relies on terminal slit valves to provide a fixed resistance to CSF flow. In cases in which the lumbar theca was judged to communicate with the spinal CSF channels higher up, these valves were left intact. In cases in which there was a complete obstruction to CSF flow in the spinal canal, the terminal slit valves were cut off so as to maximize the drainage effect of the shunt. The distal shunt catheter was then placed into the peritoneal cavity.

Preoperative and Postoperative Evaluation and Diagnostic Imaging

In our series, all patients with Chiari malformation had previously undergone craniovertebral decompression, prior to consideration for insertion of LP shunts. Also, in patients with posttraumatic syringomyelia, our preference is to perform, initially, a laminectomy and creation of a spinal CSF flow and to offer LP shunting if this operation proves ineffective or impractical.

Preoperatively, Gd-enhanced MR imaging of the entire spine was performed in all cases. Diagnostic CT myelography or CSF infusion studies were performed in selected cases in which an evident underlying cause for the syringomyelia could not be identified.

Routine postoperative spinal MR imaging was performed 3–6 months after surgery. Further management decisions, including the need for further MR imaging, was based on patients’ clinical symptoms such as neurological deterioration.

Results

Patient Population

The mean age of our 19 patients was 48 years (range 24–73 years). There were 14 men and 5 women. The underlying cause of syringomyelia was hindbrain herniation in 5 patients, posttraumatic spinal arachnoiditis in 5, postmeningitic spinal arachnoiditis in 1, and intramedullary spinal hemangioblastomas in 2 patients. The cause was not apparent in 6 patients. The 5 patients with hindbrain herniations had previously undergone craniovertebral decompression.

Clinical and Radiological Outcome

The mean follow-up duration was 25 months (range 3–51 months). Three patients were lost to follow-up. Of the remaining 16 cases, only 5 patients (1 in 3) reported clinical improvement in their preoperative symptoms (Table 1). In 2 of these 5 patients, MR imaging demonstrated a reduction in the size of their syrinx cavities, although none of the other 14 cases had any radiological improvement (Fig. 1). Table 2 provides a summary of the patients who did not have any clinical improvement as well as those lost to follow-up.

Of the 5 patients who reported improvement, spinal arachnoiditis was the underlying cause in 3 of them. Looked at another way, half (3 of 6) of the arachnoiditis group improved.

Discussion

Historically, the clinical management of syringomyelia has been influenced by the prevailing concepts of the underlying filling mechanism. A well-known, early hypothesis, applied to hindbrain-related syringomyelia, was that the cavity forms when a differential pressure between the cranial and the spinal compartments creates a caudally directed pulse wave, driving CSF through the fourth ventricle and into the central canal of the spinal cord. Although subsequently discredited, this theory led to the development of an effective surgical procedure, in the form of craniovertebral decompression.

With the advent of MR imaging, it became clear that this hypothesis could not explain the development of all forms of syringomyelia, particularly those seen in the absence of hindbrain abnormalities.9 Direct drainage of syringomyelia cavities was commonly practiced in such cases as a last resort.

The use of thecoperitoneal shunts in the management of syringomyelia is less well established. Its application is, however, consistent with current concepts of the pathogenesis of syringomyelia. Obstruction in the subarachnoid space causes a pressure differential to exist between the 2 sides of the blockage.12 In animal models of posttraumatic syringomyelia, CSF has been shown to enter the syrinx cavities from the subarachnoid space, via perivascular spaces. Abnormal hydrodynamic forces then act within the cord parenchyma to propagate the syrinx.4 Human studies have also revealed altered CSF hydrodynamics in the form of increased CSF pulse pressure and CSF velocities in patients with syringomyelia.6,13 It is reasonable to suggest that an increase in the spinal CSF compliance, brought about following a LP shunt insertion, could improve CSF hydrodynamics and prevent syrinx progression.

The particular advantage of thecoperitoneal shunts is that the risks of neurological deterioration, caused by the myelotomy required for shunt insertion into the syr-
Lumboperitoneal shunts in the treatment of syringomyelia

TABLE 1: Profile of patients with clinical improvement following LP shunt placement*

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (yrs), Sex</th>
<th>Diagnosis/Etiology</th>
<th>Surgical History</th>
<th>Preop Presentation</th>
<th>Clinical Improvement</th>
<th>Radiological Improvement</th>
<th>Subsequent Procedures</th>
<th>FU (mos)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>39, M</td>
<td>posttraumatic syringomyelia, C6–T12 syrinx, T-6 complete paraplegia</td>
<td>decompressive laminectomy &amp; pedicle screw fixation</td>
<td>rt hand numbness &amp; weakness</td>
<td>yes</td>
<td>yes</td>
<td>revision of LP shunt 4 yrs after initial insertion</td>
<td>51</td>
</tr>
<tr>
<td>2</td>
<td>50, M</td>
<td>posttraumatic C5–6 fracture-dislocation</td>
<td>none</td>
<td>progressive numbness involving both UE</td>
<td>yes</td>
<td>yes</td>
<td>none</td>
<td>20</td>
</tr>
<tr>
<td>3</td>
<td>29, M</td>
<td>idiopathic, C1–T11 syrinx</td>
<td>none</td>
<td>rt-sided hyperhidrosis, numbness in both hands, gait disturbance</td>
<td>yes</td>
<td>no</td>
<td>none</td>
<td>16</td>
</tr>
<tr>
<td>4</td>
<td>58, M</td>
<td>Chiari, C2–6 syrinx</td>
<td>craniovertebral decompression</td>
<td>gait disturbances, rt hand wasting &amp; weakness, numbness in both hands, neurogenic bladder</td>
<td>yes</td>
<td>no</td>
<td>LP shunt revised</td>
<td>48</td>
</tr>
<tr>
<td>5</td>
<td>68, M</td>
<td>listeria meningitis, C5–T12 syrinx</td>
<td>previous VP shunt for associated hydrocephalus following meningitis</td>
<td>gait disturbance, bilateral LE spasticity, neurogenic bladder</td>
<td>yes</td>
<td>no</td>
<td>none</td>
<td>3</td>
</tr>
</tbody>
</table>

* FU = follow-up; LE = lower-extremity; UE = upper-extremity.

Inx cavity, are avoided. There have, however, been very few reports on the use of thecoperitoneal shunts for syringomyelia and most of these have been limited to case reports. Lumboperitoneal shunting, in combination with myelotomy, was first used by Park et al.\textsuperscript{10} in cases of syringomyelia associated with Chiari malformation or myelomeningoceles. Vengsarkar et al.\textsuperscript{18} reported an excellent clinical and radiological response in 3 patients with syringomyelia associated with Chiari malformation, managed only with LP shunts. Vassilouthis et al.\textsuperscript{17} reported 3 cases in which LP shunt treatment was followed by clinical improvement in all patients. Most recently, Lam and associates\textsuperscript{8} presented their results on thecal shunt placement for treatment of obstructive primary syringomyelia. They reported clinical improvement in 6 of 7 cases of posttraumatic, postsurgical, or postinflammatory syringomyelia in patients who had undergone thecoperitoneal, thecopleural, or thecoatrial shunt placement. Table 3

![Fig. 1. Sagittal T2-weighted MR image of posttraumatic syrinx cavity preoperatively (left) and the appearance 12 months after insertion of LP shunt (right).](image-url)
<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (yrs), Sex</th>
<th>Diagnosis/Etiology</th>
<th>Surgical History</th>
<th>Preop Presentation</th>
<th>Clinical Improvement</th>
<th>Radiological Improvement</th>
<th>Subsequent Procedures</th>
<th>FU (mos)</th>
</tr>
</thead>
<tbody>
<tr>
<td>2</td>
<td>69, F</td>
<td>idiopathic</td>
<td>none</td>
<td>rt hand pain &amp; clawlike attitude</td>
<td>no</td>
<td>no</td>
<td>none (died 27 mos later due to pneumonia)</td>
<td>27</td>
</tr>
<tr>
<td>4</td>
<td>60, F</td>
<td>Chiari malformation</td>
<td>craniovertebral decompression</td>
<td>rt hand clawing, dysesthetic pain in both UE</td>
<td>no</td>
<td>no</td>
<td>none</td>
<td>10</td>
</tr>
<tr>
<td>5</td>
<td>44, M</td>
<td>idiopathic, C2–T4 syrinx</td>
<td>none</td>
<td>gait disturbance, numbness in trunk &amp; bilat LE</td>
<td>no</td>
<td>no</td>
<td>syringopleural shunt</td>
<td>10</td>
</tr>
<tr>
<td>6</td>
<td>45, M</td>
<td>Chiari, C3–T4 syrinx</td>
<td>craniovertebral decompression</td>
<td>altered temperature sensation in both UE &amp; Lt LE</td>
<td>no FU</td>
<td>no FU</td>
<td>no FU</td>
<td>no FU</td>
</tr>
<tr>
<td>7</td>
<td>60, F</td>
<td>Chiari, C2–7 syrinx</td>
<td>craniovertebral decompression</td>
<td>gait disturbance, neurogenic bladder, bilat UE numbness</td>
<td>no</td>
<td>no</td>
<td>none</td>
<td>40</td>
</tr>
<tr>
<td>8</td>
<td>34, M</td>
<td>idiopathic, normal CSF infusion study &amp; CT myelography</td>
<td>none</td>
<td>bilat LE spasticity, gait disturbance, altered temperature sensation in rt hand</td>
<td>FU</td>
<td>no FU</td>
<td>no FU</td>
<td>no FU</td>
</tr>
<tr>
<td>9</td>
<td>73, M</td>
<td>idiopathic, T7–11 syrinx, normal CSF infusion study</td>
<td>none</td>
<td>bilat LE weakness &amp; spasticity, neurogenic bladder</td>
<td>no (worse)</td>
<td>no</td>
<td>none</td>
<td>11</td>
</tr>
<tr>
<td>10</td>
<td>31, M</td>
<td>von Hippel-Lindau, multiple spinal hemangioblastomas, C2–T6 syrinx</td>
<td>2 previous cervical laminectomies for tumor excision</td>
<td>dysesthetic pain on rt side of trunk &amp; rt LE, numbness in rt hand &amp; trunk to T-4 dermatome, Lt hand weakness</td>
<td>no FU</td>
<td>no FU</td>
<td>none (died 3 mos later)</td>
<td>no FU</td>
</tr>
<tr>
<td>12</td>
<td>24, M</td>
<td>von Hippel-Lindau, spinal tumors, C2–T11 syrinx</td>
<td>none</td>
<td>numbness in Lt LE</td>
<td>no</td>
<td>no</td>
<td>had syringopleural shunt later</td>
<td>7</td>
</tr>
<tr>
<td>13</td>
<td>50, M</td>
<td>posttraumatic syringomyelia, C6–7 fracture-dislocation, C-7 paraplegia, C5–7 syrinx</td>
<td>cervical laminectomy &amp; adhesiodysis</td>
<td>paresthesia in both hands, worsening LE spasticity &amp; dysesthetic pains in LE</td>
<td>no</td>
<td>no</td>
<td>had syringopleural shunt later but still no improvement</td>
<td>25</td>
</tr>
<tr>
<td>14</td>
<td>44, F</td>
<td>idiopathic, C4–T1 syrinx, normal CT myelography</td>
<td>none</td>
<td>Lt UE paresthesia &amp; dysesthetic pain</td>
<td>no</td>
<td>no</td>
<td>none</td>
<td>34</td>
</tr>
<tr>
<td>15</td>
<td>45, M</td>
<td>posttraumatic, T-6 paraplegia, C7–6 syrinx</td>
<td>thoracic laminectomy &amp; adhesiodysis</td>
<td>upward extension of numbness in trunk</td>
<td>no</td>
<td>no</td>
<td>syringopleural shunt (radiological improvement)</td>
<td>30</td>
</tr>
<tr>
<td>16</td>
<td>37, F</td>
<td>Chiari malformation</td>
<td>craniovertebral decompression</td>
<td>numbness of Lt side of body</td>
<td>no</td>
<td>no</td>
<td>LP shunt changed to VP shunt</td>
<td>32</td>
</tr>
<tr>
<td>17</td>
<td>54, M</td>
<td>posttraumatic, T7–8 fracture-dislocation, T4–7 syrinx, incomplete T-6 paraparesis</td>
<td>thoracic laminectomy &amp; adhesiodysis; syringopleural shunt, ligated for pressure dissociation headaches</td>
<td>worsening numbness in Lt LE</td>
<td>no</td>
<td>no</td>
<td>revision of thoracic laminectomy &amp; adhesiodysis, insertion of syringosubarachnoid shunt</td>
<td>34</td>
</tr>
</tbody>
</table>

* VP = ventriculoperitoneal.
Lumboperitoneal shunts in the treatment of syringomyelia

## TABLE 3: Reports on the use of thecoperitoneal or LP shunts in the treatment of syringomyelia

<table>
<thead>
<tr>
<th>Author &amp; Year</th>
<th>No. of Patients</th>
<th>Mean FU</th>
<th>Percentage (no. of patients)</th>
<th>Remarks/Use of Shunt</th>
</tr>
</thead>
<tbody>
<tr>
<td>Vengsarkar et al., 1991</td>
<td>3</td>
<td>7 days</td>
<td>100 (3) 100 (3)</td>
<td>all had Chiari-associated syrinx; used as primary treatment in 1 patient</td>
</tr>
<tr>
<td>Vassilouthis et al., 1993</td>
<td>3</td>
<td>4 mos</td>
<td>100 (3) 100 (3)</td>
<td>varied etiology: idiopathic, Chiari Type I, &amp; posttraumatic; used as primary treatment for Chiari-associated syrinx</td>
</tr>
<tr>
<td>Lam et al., 2008</td>
<td>7</td>
<td>33 mos</td>
<td>86 (6) 86 (only 6 had postop images)</td>
<td>all had postarachnoiditis-related syringomyelia</td>
</tr>
</tbody>
</table>

provides a summary of all relevant previously published reports.

Here we have presented our experience with 16 of 19 patients with syringomyelia, managed wholly or in part with LP shunts. One in 3 patients enjoyed definite improvement in their symptoms. Clearly, this could represent a placebo effect. The radiological improvement seen in 2 of these 5 cases, however, is a more objective measure of success and would suggest that the response is, after all, physiologically based. On one hand, a surgical success rate of 1 in 3 is not, on the face of it, a very good result. On the other hand, such a relatively minor procedure may be worth trying before subjecting patients to a myelotomy, particularly if they are at least partly intact neurologically. It is also a useful procedure to consider in somebody who may have significant risk factors associated with anesthesia.

With our small patient numbers, we have not been able to relate the outcome to the underlying cause of the syrinx (Table 1). However, it is noteworthy that half (3 of 6) of the patients with postarachnoiditis syringomyelia improved. Lam et al. reported clinical improvement in 6 (86%) of 7 patients with postarachnoiditis-related syringomyelia treated with thecal shunt placement. These results may suggest better outcomes in patients with postarachnoiditis-related syringomyelia compared with syringomyelia-related to other etiologies.

In our series, all patients with Chiari malformation had previously undergone craniovertebral decompression, prior to insertion of LP shunts. This is in contrast to the patients in the Vengsarkar et al. and Vassilouthis et al. reports, in which LP shunts were used as primary treatment for hindbrain-associated syringomyelia. The use of LP shunts for syringomyelia treatment, in the presence of Chiari malformation, has been criticized in the past as it may aggravate cerebellar herniation.

In patients with posttraumatic syringomyelia, our preference is to perform, initially, a laminectomy and creation of a spinal CSF flow, but to offer LP shunting if this operation proves ineffective.

Although not all of our responsive patients had radiological evidence of improvement, Goel and Desai have observed that, with regard to different surgical procedures for syringomyelia, clinical outcome, rather than radiological improvement, is the more appropriate indicator of the surgical result. Pillay also observed that clinical improvement need not be concomitant with a decrease in syrinx size.

## Conclusions

The use of LP shunts in the treatment of syringomyelia is, we believe, a valid concept with a rational basis. It has particular application in cases of syringomyelia in which no site of obstruction to CSF flow can be demonstrated. In cases that do not respond to other procedures, such as suboccipital decompression for hindbrain-related syringomyelia or spinal arachnoid scar resection for posttraumatic syringomyelia, an LP shunt may lead to useful improvement in a patient’s symptoms. At present we cannot readily identify those patients who might benefit from this procedure, although a trial lumbar puncture may be a useful preliminary to the operation.

## Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: Oluigbo, G. Flint. Acquisition of data: Oluigbo, Thacker. Analysis and interpretation of data: Oluigbo, Thacker, Flint. Drafting the article: Oluigbo, Flint. Critically revising the article: Oluigbo, Thacker, Flint. Reviewed final version of the manuscript and approved it for submission: Oluigbo, Flint. Statistical analysis: Oluigbo, Thacker. Administrative/technical/material support: Oluigbo, Thacker, Flint. Study supervision: Flint.

## References


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