Primary spinal syringomyelia

Invited submission from the Joint Section Meeting on Disorders of the Spine and Peripheral Nerves, March 2005

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In the present review the author describes the different types of syringomyelia that originate from abnormalities at the level of the spinal cord rather than at the craniovertebral junction. These include posttraumatic and postinflammatory syringomyelia, as well as syringomyelia associated with arachnoid cysts and spinal cord tumors. The diagnosis and the principles of managing these lesions are discussed, notably resection of the entity restricting cerebrospinal fluid flow. Placement of a shunt into the syrinx cavity is reserved for patients in whom other procedures have failed or who are not candidates for other procedures.

KEY WORDS • syringomyelia • arachnoid cyst • tumor • syrinx • shunt

Primary spinal syringomyelia can be defined as syringomyelia that exists in the absence of an abnormality at the level of the foramen magnum, the most common form of which is CTE. Chiari malformation is the generally used eponym, comprising a small number of anatomical subtypes. There are similarities in the underlying pathophysiology of syringomyelia developing in the presence of CTE and primary spinal syringomyelia. At present our understanding is that in both of these conditions there is a partial obstruction of the subarachnoid space; in the case of CTE, this occurs when the subarachnoid space at the level of the foramen magnum becomes crowded by the descended cerebellar tonsils and the neural elements of the cervicomedullary junction. In primary spinal syringomyelia, partial obstruction of the SAS may also be the result of other causes (Table 1).

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Causes of Partial Obstruction of Subarachnoid Space

In both CTE and primary spinal syringomyelia, the subarachnoid space below the level of the partially obstructed subarachnoid space behaves as a relatively isolated compartment, subject to transmitted pulsations generated by respiration and arterial pulsations. This mechanism has been studied extensively by Oldfield and colleagues, who described the descended tonsils as acting as pistons against the somewhat isolated spinal fluid compartment beneath the tonsils, thereby driving fluid from the subarachnoid space, presumably along the perivascular spaces, into the cord parenchyma. It is not impossible that an area of residual central canal of the spinal cord plays a role in the formation of an expanded syrinx cavity. Accurate analogous studies of primary spinal syringomyelia have not been conducted, but it can be assumed that when pulsatile CSF encounters scar tissue, cyst membrane, or tumor, it fulfills the role of the “piston.” If this, or a somewhat similar mechanism, accounts for the filling of the syringomyelic cavity, it is logical to assume that eliminating the subarachnoid space’s partial obstruction whenever possible would “inactivate the filling mechanism” of the syrinx cavity, to borrow a phrase coined by Sgouros and Williams. It is this concept that forms the basis of the initial surgical approach to primary spinal syringomyelia—that is, surgical resection of scar tissue, arachnoid web, or tumor. Only when such initial procedures fail would one want to consider other treatment options.

Diagnosis of Spinal Syringomyelia

The diagnosis of syringomyelia, including the primary spinal form, is best established using MR imaging. The signal differences between watery CSF and neural tissue clearly outline the syrinx cavity on MR images. Administration of ionic contrast is helpful to define a tumor. Magnetic imaging, however, does not always clearly demonstrate the precise nature, level, or extent of the CSF obstruction. Cardiac-gaited techniques, used to study CSF flow properties at the craniocervical junction, have not been applied as successfully or widely to study other lev-

Abbreviations used in this paper: CSF = cerebrospinal fluid; CTE = cerebellar tonsillar ectopic; MR = magnetic resonance; SAH = subarachnoid hemorrhage; SCI = spinal cord injury.
els of the spine. The use of myelography, particularly in conjunction with computerized tomography scanning, often reveals more precisely the areas of a subarachnoid blockage. From the point of view of the surgeon who must decide whether to perform surgery and at which level(s) of the spine, the accurate delineation of the areas at which the subarachnoid space is narrowed or obstructed is of great importance. Myelography will frequently reveal subtle irregularities in the outline of the subarachnoid space, a feature that will direct the surgeon to an area of interest. Occasionally myelography may demonstrate a complete obstruction to the flow of contrast material. In such cases it is necessary to complete the study by also administering contrast medium at the C1–2 level to delineate the complete extent of subarachnoid scarring.

Types of Syrinx Cavities: Syringomyelia and Persistent Central Canal

Syringomyelic cavities are often not simple, confluent fluid spaces. Rather, they may be quite complex, with separate parallel channels that may or may not communicate with each other. Such complex channels are perhaps more frequently encountered in patients with posttraumatic spinal syringomyelia. Another important variant of syrinx cavity anatomy was described by Milhorat, et al., in some patients with posttraumatic syringomyelia, the syrinx cavity communicates with the subarachnoid space at the level of the spinal cord injury. It is not clear whether such communications result from secondary rupture of the syringomyelic cavity or are present since the injury occurred and contribute to formation of the syrinx cavity. True syringomyelic cavities need to be distinguished from areas of residual central canal of the spinal cord. The term hydromyelia would seem particularly applicable to the latter imaging finding. The central canal of the cord, present at birth, undergoes progressive involution as an individual ages, as described by Yasui, et al. Although involution of the central canal occurs most rapidly during the first decade of life, it nonetheless continues with every decade. Even in people in their seventh and eighth decades of life and beyond, small focal areas of residual central canal may be present. The characteristic imaging features of the persistent central canal are the following: 1) it is linear or fusiform in appearance on sagittal MR images; 2) most are 2 to 4 mm in maximal width; 3) there may be a single region of persistent central canal of varying rostrocaudal extent, or there may be several discontinuous regions; and 4) axial MR images obtained through these areas reveal an almost perfectly round, centrally placed cavity in the cord (Fig. 1). It is not known whether the presence of an area of residual central canal observed at the time of SCI plays a role in the development of posttraumatic syringomyelia, although such speculation is not unreasonable.

Posttraumatic Syringomyelia

Incidence and Symptoms. The exact incidence of posttraumatic syringomyelia is unknown, but it is very clear that MR imaging has increased the recognition of the condition. Using this modality, current estimates of the incidence are as high as 64% in the pre–MR imaging era; this incidence was frequently cited as 2 to 3%.

Neurological symptoms and pain may develop in patients in whom the initial SCI was not associated with a neurological deficit, or their onset may be sudden and associated with straining or a bout of coughing. In cases involving a history of neurological symptoms the patients may report a sudden ascent of their existing sensory level or a new loss of motor function, often involving their hands or arms. Symptoms may include motor, sensory, autonomic, and sphincter control functions, in any combination. It is presumed that, over time, the progressive thickening of scar tissue accounts for an interval between the injury and the development of symptoms attributable to syringomyelia.

Subtypes. Posttraumatic syringomyelia is presumed to develop when the subarachnoid space has been significantly narrowed or obliterated at the level of injury. As already mentioned, this may occur in the following situations: 1) The lesion may develop in the absence of any recognized trauma to the spine, presumably by arachnoid scarring, which proceeds and progresses after the injury. 2) Severe posttraumatic spinal deformity, such as retropulsed bone or disc or a kyphotic deformity, may contribute to the constriction or obliteration of the subarachnoid space along with scarring. 3) Severe injury to the spinal cord and its coverings is probably the most common underlying form of injury leading to syringomyelia; blood itself may be a contributing factor. The older concept that syringomyelia develops from a focus of traumatic hematoma has not found much support because of more current observations, which benefit from the widespread application of MR imaging (Fig. 2).

Postinflammatory Syringomyelia

Infection and chemical or other sterile inflammations are the most common causes underlying the development of postinflammatory syringomyelia. Granulomatous meningitis—tuberculous and fungal—seems especially likely to be followed by scarring of the subarachnoid space. Postoperative meningitis, particularly after an intradural procedure such as tumor removal, can also lead to postinflammatory syringomyelia. In a relatively small number of patients, syringomyelia may occur after SAH, as from aneurysmal rupture. It is not clear why this complication of SAH develops in some but not in most patients. Foreign chemical substances may also provoke an intense inflammatory response in some individuals; these include io-

**TABLE 1**

<table>
<thead>
<tr>
<th>Cause of Partial Obstruction</th>
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<tbody>
<tr>
<td>traumatic scar due to SCI or op</td>
</tr>
<tr>
<td>postinflammatory scar due to infection, blood, or foreign substance</td>
</tr>
<tr>
<td>bone fragments or spinal deformity</td>
</tr>
<tr>
<td>disc herniation</td>
</tr>
<tr>
<td>arachnoid cyst</td>
</tr>
<tr>
<td>intradural or extradural tumors</td>
</tr>
</tbody>
</table>
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phenylate (Pantopaque), an oil-based, radiopaque contrast material that was used many years ago.26

Because of the generally diffuse nature of meningitis, SAH, or chemical spread within the subarachnoid space, scarring of the arachnoid in these situations often extends over many spinal segments or is even multifocal (Fig. 2 center and right).

Postsurgical scarring tends to be more focal. Even when postoperative infection is absent, syringomyelia in this case might be regarded as postinflammatory. It will be treated separately in this review.

**Postsurgical Syringomyelia**

I have treated patients who presented with syringomyelia many years after an apparently uncomplicated and other-

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Fig. 1. Representative T₂-weighted MR images.  
**Left:** Sagittal cervical image demonstrating a typical fusiform cavity, a residual focus of central canal, at C6-7. Subsequent C6-7 anterior discectomy and interbody fusion did not alter the appearance of this cavity during a 5-year follow-up period.  
**Right:** Axial thoracic image revealing the typical central, round appearance of such central canal cavities.

Fig. 2.  
**Left:** Sagittal T₁-weighted MR image revealing posttraumatic syringomyelia in a 20-year-old man who became paraplegic following a motor vehicle accident; associated compression fractures of the thoracic spine from T-4 to T-7 are present.  
**Center and Right:** Sagittal T₁-weighted MR images revealing postinflammatory syringomyelia in a 40-year-old woman with a 3-year history of meningitis before presenting with syringomyelia.
wise uneventful benign intradural neoplasm resection (one patient after 24 years, the other 41 years later). Both tumors were reported to be neurofibromas. It is not known whether intra- or postoperative bleeding into the subarachnoid space contributed to the development of syringomyelia or whether the development of syringomyelia reflects the propensity to develop scar tissue in a given individual. In another case, syringomyelia developed following incomplete resection of a thoracic meningioma.

Arachnoid Cysts and Diverticula

Partial obstruction of the subarachnoid space may occur in the presence of a web or pouch of arachnoid that, in itself, is benign in appearance, although sometimes thickened by exposure to the continuous pulsatile motion of CSF during a long period.

Such arachnoid webs or pouches are very discrete. Thus, on imaging studies, they may appear as abrupt changes in the caliber of the cord’s outline. In other instances they can only be identified as subtle irregularities on myelograms. Widening of the subarachnoid space rostral to the web is often seen (Fig. 3).

In my experience arachnoid cysts and diverticuli were encountered in the thoracic region and most frequently manifested at presentation as disturbances of gait, although other neurological signs and symptoms have been reported.

True Syringomyelia in Association With Spinal Tumors and Disc Herniations

True syringomyelic cavities may develop in association with intradural spinal tumors. Theoretically, compression of the subarachnoid space can also lead to syringo-

Fig. 3. Computerized tomography myelogram of thoracic spine demonstrating the abrupt change in cord diameter at the level of an arachnoid cyst.

Fig. 4. Contrast-enhanced T₁-weighted MR image revealing an intramedullary tumor at C₄–₆ with an associated syrinx cavity extending both rostrally and caudally. The cavity collapsed completely after gross-total excision of this ependymoma.
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**TABLE 2**

**Pathological entity and procedures in 64 surgically treated patients with primary spinal syringomyelia**

<table>
<thead>
<tr>
<th>Cause of Syringomyelia</th>
<th>No. of Cases</th>
<th>No. of Ops</th>
</tr>
</thead>
<tbody>
<tr>
<td>traumatic scar/bone</td>
<td>26</td>
<td>58</td>
</tr>
<tr>
<td>postinflammatory</td>
<td>16</td>
<td>23</td>
</tr>
<tr>
<td>tumor related</td>
<td>11</td>
<td>12</td>
</tr>
<tr>
<td>arachnoid cyst</td>
<td>4</td>
<td>5</td>
</tr>
<tr>
<td>congenital tethering/diastema</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>residual central canal</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>unknown</td>
<td>4</td>
<td>4</td>
</tr>
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</table>

*Includes two patients who underwent reoperation elsewhere.*

**TABLE 4**

**Summary of 105 procedures performed in 64 patients**

<table>
<thead>
<tr>
<th>Procedure</th>
<th>No. of Cases</th>
</tr>
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<tbody>
<tr>
<td>decompressive laminectomy/duraplasty</td>
<td>32</td>
</tr>
<tr>
<td>syrinx shunt</td>
<td>29</td>
</tr>
<tr>
<td>subarachnoid space shunt</td>
<td>11</td>
</tr>
<tr>
<td>dural bypass</td>
<td>3</td>
</tr>
<tr>
<td>cyst wall resection</td>
<td>4</td>
</tr>
<tr>
<td>tumor resection</td>
<td>2</td>
</tr>
<tr>
<td>shunt revision</td>
<td>14</td>
</tr>
<tr>
<td>cyst aspiration</td>
<td>6</td>
</tr>
<tr>
<td>anterior decompression</td>
<td>4</td>
</tr>
</tbody>
</table>

**Treatment Options**

Recognizing that partial obstruction of the subarachnoid space is integral to syrinx formation in primary spinal syringomyelia, the first-line treatment should be directed at eliminating this partial obstruction, whenever feasible, and this can be undertaken in association with a reasonable risk-to-benefit ratio.8

The clinical presentation of syringomyelia in this group of patients may result from a combination of effects of the tumor and of the syringomyelic cavity. Such true syringomyelia cavities should be distinguished from tumor cysts. Tumor cysts are commonly associated with glial intradural tumors (Fig. 4) and are also occasionally seen with hemangioblastoma. The fluid contained within glial tumor cysts is often xanthochronic and, in general, has a high protein content, whereas fluid of true syringomyelic cavities is generally indistinguishable from CSF in composition.

Table 2 provides a summary of my experience in treating primary spinal syringomyelias of various origins.

**TABLE 3**

**Primary spinal syringomyelia**

<table>
<thead>
<tr>
<th>No. of Procedures in 64 Patients</th>
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<tbody>
<tr>
<td>No. of Ops</td>
</tr>
<tr>
<td>1</td>
</tr>
<tr>
<td>2</td>
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<tr>
<td>3</td>
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<tr>
<td>4</td>
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<tr>
<td>5</td>
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<tr>
<td>6</td>
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</tbody>
</table>

*Includes two patients who underwent reoperation elsewhere.*


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cally for primary spinal syringomyelia underwent more than one surgical procedure (Table 3). The procedures performed are listed in Table 4.

Of the 23 patients who underwent reoperation, status stabilized in 10, improved in four, and worsened in three. Four patients died, two of late-onset complications related to their disorder (pneumonia and respiratory failure) and two of unrelated causes. Two patients were lost to follow up.

Medical therapy for spasticity and neuropathic pain forms part of the treatment plan. Commonly used medications for neuropathic pain include opiate agents, which block neurotransmitter release, and gabapentin, which works by reducing neurotransmitter release. I have had no significant experience with newly released drugs reported to be helpful in neuropathic pain such as pregabalin and duloxetine. Pregabalin, although it acts like gabapentin, is reported to be absorbed more dependably. Baclofen is useful primarily for the control of spasticity related to syringomyelia. The dosage of baclofen has to be individually adjusted.

The results of my treatment of patients with primary spinal syringomyelia are represented graphically in Fig. 5 and Table 5. It is clear from these results that the treatment of primary spinal syringomyelia does not yield as good an outcome as we would expect in patients with cerebellar

FIG. 5. Bar graphs showing outcome related to pathological entity (upper) and surgical procedure (lower). Ant = anterior; arach = arachnoid; congen/teth = congenital tethering; decomp = decompression; inflamm = inflammatory; resid cc = residual central canal; subarachn = subarachnoid.

### TABLE 5

<table>
<thead>
<tr>
<th>Outcome</th>
<th>No. of Cases</th>
</tr>
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<tbody>
<tr>
<td>improved</td>
<td>15</td>
</tr>
<tr>
<td>stabilized</td>
<td>18</td>
</tr>
<tr>
<td>worsened</td>
<td>8</td>
</tr>
<tr>
<td>required reop</td>
<td>22</td>
</tr>
<tr>
<td>no information</td>
<td>1</td>
</tr>
</tbody>
</table>
ectopia–related syringomyelia. Only focal lesions, such as those seen with arachnoid cysts, focal scars, or tumors are associated with a high incidence of good outcome without the need for reoperation. One of my patients underwent cervical laminectomy and duraplasty before the true significance and high incidence of a residual central canal focus was recognized.

Conclusions

The following conclusions may be drawn. 1) Syringomyelia may complicate SCLs, meningitis, SAH, and surgery, as well as being present in conjunction with developmental webs and spinal cord tumors. 2) Establishing the extent and level of a subarachnoid block associated with syringomyelia is important in planning treatment. 3) In patients with primary spinal syringomyelia multiple surgical procedures may be required; the outcome is better in patients with focal, rather than diffuse, obstruction of the subarachnoid space. 4) Resection of the focal obstructing pathological entity should be the first choice in the treatment of these cases; placing a shunt in the syrinx cavity should be performed only when there are no other choices. Placement of a syringosubarachnoid shunt after a hemilaminectomy appears to be the best strategy available today. 5) Medical treatment of spasticity and neuropathic pain is an important adjunct in the treatment of these patients.

References


Manuscript received August 1, 2005.
Accepted in final form September 20, 2005.
An earlier version of the present review was presented at the 21st Annual Meeting of the American Association of Neurological Surgeons/Central Nervous System Section on Disorders of the Spine and Peripheral Nerves, March 12, 2005, Phoenix, Arizona.
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