In this report current surgical procedures that are performed to treat patients with primary spinal syringomyelia are summarized.

**PRIMARY SPINAL SYRINGOMYELIA**

The treatment of syringomyelia due to abnormalities originating within the spinal canal remains a subject of ongoing discussion and investigation. The underlying causes of these forms of syringomyelia are as follows: 1) posttraumatic, either without significant spine deformity or with significant spine deformity; 2) postsurgical; 3) post-inflammatory, either postinfectious or postchemical inflammation; 4) intradural or extradural tumor; and 5) extradural mass effect caused by disc protrusion.

Myelocystoceles have deliberately not been included in the previous list as they are believed to represent a different clinical entity. It is also noted that the tumor-associated syrinx cavities are distinct from tumor-associated neoplastic cysts, the latter being contiguous with the tumor and usually containing proteinaceous fluid. Determining the underlying cause of syrinx formation is important because the therapeutic approach varies. For example, treatment of syrinx cavities associated with tumor or disc protrusion should be directed at the primary problem only because, in many cases, the syrinx cavity will collapse after removal of the tumor or disc.10,22

Compression, or at least partial obstruction, of the subarachnoid space is postulated to be a factor common to all of these conditions. Arachnoid scars or bands appear to be present in patients with a history of trauma and inflammation, as well as in those in whom a syrinx has developed postsurgically.4 The extent of subarachnoid obstruction that will result in syrinx formation has never been quantitated but must vary considerably. It must be assumed that in most patients with a history of spinal trauma, meningoitis, and intradural tumor surgery some scar tissue is formed as part of the natural healing process, although they do not go on to develop syringomyelia. The estimates for incidence of syringomyelia among spinal cord–injured patients vary widely, from 1 to 2%5 to as high as 64%.8 Obviously the advent of MR imaging has had a major impact on the frequency with which the diagnosis of syringomyelia is established.

In patients with intra- or extradural tumors, the subarachnoid obstruction is caused not by scarring but by compression of the subarachnoid space by the tumor. Removal of the tumor usually results in spontaneous resolution of the syrinx cavity.10 Cases of posttraumatic spinal deformity, usually kyphosis, may represent a combination of extradural bone compression and intradural scar formation.

The spinal level at which the arachnoid lesion lies (cervical, high thoracic, or low thoracic) may also have some bearing on the development of a syringomyelic cavity, as much as the CSF dynamics vary in each region.
TREATMENT OF POSTTRAUMATIC AND POSTINFLAMMATORY SYRINGOMYELIA

There are three modes of treating posttraumatic and postinflammatory-related syrinx cavities: drainage, expansion of the subarachnoid space, and cyst obliteration. Results by any currently used technique are generally more favorable for posttraumatic than for postinflammatory syringomyelia.

Drainage Procedures

Drainage of the Syrinx Cavity. Historically, drainage procedures were the first to be used, originally as an extrapolation of techniques used to treat Chiari malformation-related syringomyelia. Whereas small openings in the spinal cord were made as the only treatment, this technique is now generally thought to be ineffective because the holes presumably close over.12 Thus, cyst drainage was not widely performed until the development of shunts that drain into the subarachnoid space and into other body cavities.1,5,25 The advantages of shunt placement are the immediacy of cyst drainage and of reduction in cyst size, as well as the fact that the procedure is generally technically less complicated. The disadvantages of draining a cyst via a shunt include: development of some degree of myelomeningeal adhesions, although often not functionally significant, such as a truncal sensory deficit; drainage of trauma-induced cysts that communicate with the subarachnoid space at the level of injury14 requiring a valve-regulated shunt system that may be ineffective; and drainage of septated or multichannel cysts that is, at best, difficult. Drainage of a syrinx cavity located below an adherent arachnoid scar may also place some traction forces on the spinal cord. Complications related to shunt placement in syringomyelic cavities have been studied by Sgouros and Williams18 and our group3 and can be categorized as follows: 1) shunt obstruction, proximal and distal; 2) tethering of the spinal cord by the shunt; 3) low CSF pressure state; 4) shunt-related infection; and 5) shunt dislocation. In our own studies we have found an overall shunt failure or complication rate of 50%.3

It must, however, be stated that at several institutions the surgeons still perform syringomyelic shunt procedures that are associated with a low immediate complication rate and seemingly good follow-up results. Notable is the experience reported by Iwasaki, et al.,9 who have continued to refine their technique of subarachnoid shunt placement and now report good results when placing the cyst shunt from the dorsal root entry zone to the anterior subarachnoid space, in front of the dentate ligaments.

They have treated nine patients with primary spinal syringomyelia, reporting significant improvement in pain as well as sensory and motor impairment in patients with posttraumatic or postmeningitic syringomyelia at a mean follow up of 33 months. The Toronto group has obtained similar good results by using subarachnoid shunts in 16 patients with posttraumatic and postinflammatory syringomyelia, with good to excellent results in 63% of the patients.19 This procedure may be less likely to cause spinal cord tethering than that in which shunts drain into the peritoneal or pleural cavities; however, it assumes that the distal subarachnoid space is open and able to absorb CSF, which may not be the case in patients in whom diffuse scarring of the subarachnoid space has formed. However, concerns regarding septated or multichannel cysts apply to these procedures, just as they do to other types of syrinx cavity shunts.

Drainage of the Subarachnoid Space. Diversion of CSF from the spinal subarachnoid space as a means of reducing the size of the syringomyelic cyst was described in 1991 by Vengsarkar, et al.,21 and termed "thecoperitoneal shunting." In their study, however, the authors treated patients with Chiari-related syringomyelia, and the risks of lumbothecal shunt placement in these cases have been addressed by Williams24 and Pillay.16 Of greater interest is the subsequent report of Vassilouthis, et al.,20 who performed this approach in three patients (two with primary spinal syringomyelia) and obtained successful resolution of the syrinx. Myelographic studies were not conducted in any of these patients, but the high CSF pressures recorded suggested that arachnoid scarring resulted in a total or subtotal block of the spinal subarachnoid space. Our experience with this procedure, in which we used a pressure valve shunt system, has yielded mixed results; however, in at least two patients, one who developed postoperative adhesions following tumor removal, the other with postinflammatory adhesions, dramatic clinical improvement was demonstrated, which correlated with significant reduction in the size of the syrinx cavity.

Subarachnoid Space Expansion

Scar Resection and Expansive Duraplasty. The concept of resecting scar tissue and expanding the subarachnoid space in patients with primary spinal syringomyelia was introduced by our group (unpublished data) and by Williams23 and is analogous to the suboccipital decompression and duraplasty performed in patients with syringomyelia related to Chiari malformation. This technique is most applicable in patients with very focal areas of scarring or an arachnoid band. It is probably not appropriate to apply this technique in patients with arachnoid scarring that extends over several spinal segments, as is seen, for example, in patients with tuberculous meningitis and in relatively few of the patients who sustained trauma-related spinal cord injury. It is often difficult to define the exact point of scar adhesion on MR images. For this reason, we recommend performing myelography in any patient for whom there is doubt about the location and extent of the arachnoid scar (Fig. 1). In the event of complete blockage of the flow of contrast material that has been introduced in the lumbar region, we recommend an additional injection of contrast material at C1–2 to define the upper level of the block. Through careful observation of flow dynamics by the radiologist, a partial obstruction to the flow of contrast material may be identified.

There are two main advantages of scar tissue resection and expansive duraplasty in the treatment of syringomyelia: 1) avoidance of making an incision into the spinal cord itself and 2) avoidance of introducing a foreign body that may act as a nidus for infection or as a source of spinal cord tethering and is subject to mechanical blockage by glial overgrowth. The existence of syrinx septations or multiple channels is of no concern when using this procedure, which is performed on the outer surface of the spinal cord.
Primary spinal syringomyelia

cord. Furthermore, restoration to a more normal pattern of subarachnoid spinal fluid flow is a more physiological approach, and it appears to counteract the filling mechanism of the syrinx cavity, the precise nature of which is not yet completely clear. The disadvantages are the possibility that the scar may reform, the greater technical challenge involved, and the fact that collapse of the cavity may not occur immediately.

Technically, the procedure is performed via a laminectomy centered over the area of the focal scar and extending one spinal level above and one spinal level below the scar (unpublished data). The laminectomy should be as wide as possible without violating the facet joints. A midline dural incision is made under the microscope; every attempt is made to leave the arachnoid intact initially, and dural retention sutures are placed.

The injection of a very small amount of indigo carmine (0.2–0.3 ml, via a 27-gauge needle attached to a tuberculin syringe) into the exposed subarachnoid space rostrally may help define the arachnoid band or scar intraoperatively (Fig. 2). Microdissection and resection of the scar is then performed, extending as far to each side of the spinal cord as possible without actual manipulation of the cord. In a few cases fluid has been aspirated from the syrinx cavity via a 27-gauge needle to "jump start" the process of syrinx cavity collapse. The dura is closed using a dural graft. Autologous fascia lata may be used except in paraplegic patients in whom this tissue layer becomes quite atrophic; bovine pericardium may be used, as well as other biological membranes. Use of an inner layer of Gore-tex dura graft substitute may reduce the risk of scar tissue adhesion to the inside of the biological dural graft membrane. Some surgeons also like to place dural tenting sutures before suturing the graft, in an attempt to keep the dural graft away from the underlying spinal cord. This can be accomplished by tacking the dura to the soft tissue (muscle) wall of the wound cavity; occasionally, we have placed a tenting suture into the graft, but this is not generally recommended as, in one case, such a suture tore and resulted in a spinal fluid leak.

In 1997 our group described their experience with scar resection and expansive duraplasty.11 We have now performed this procedure in approximately 35 patients. The major problems we encountered were formation of pseudomeningocele or recurrence (the latter occurred in a patient in whom multiple previous procedures had been performed through the same incision), and failure of the procedure to stabilize or reduce the size of the syrinx cavity. This was attributed to a presumed communication of the syrinx cavity with the subarachnoid space at the level of injury (one patient) and failure to correct the kyphotic spinal deformity contributing significantly to construction of the subarachnoid space. Stabilization of the syrinx or reduction in size occurred in 83% of patients in whom there was focal arachnoid scarring, but in only 17% of patients in whom extensive scarring had been demonstrated.11 Other complications include those common to surgical procedures in general.

Bone Decompression and Correction of Kyphosis. In five patients ventral decompression of the subarachnoid space has been achieved by creating a kyphotic spinal deformity; resolution of symptoms or stabilization has been demonstrated in all patients.7 Two of these patients required an additional procedure in which resection of intradural scarring was performed, as described earlier.
Cyst Obliteration

Martin first suggested obliteration of the syringomyelic cyst as a means of eliminating the dynamic forces within the cyst cavity. Omental transposition was reported by Min-Shu, et al., and by Sett and Crockard; there has only been a small number of cases, and no long-term follow-up study has been reported. Recently, Falci, et al. and Fessler (personal communication, 1999) have used embryonic tissue grafts, and it will be interesting to learn the results of this experimental approach.

CONCLUSIONS

Based on our experience to date, we recommend surgical expansion of the subarachnoid space as the preferred first approach in patients with primary spinal syringomyelia. The area of subarachnoid space narrowing should be defined using myelography, in addition to MR imaging, whenever necessary. Anterior bony decompression should be performed as the first step whenever a significant kyphotic spinal deformity is present. Resection of arachnoid scars and expansive duraplasty have resulted in superior long-term benefit in patients with syringomyelia. Placement of a shunt may be appropriate in certain situations, but we prefer to use this option as a last resort.

References


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