Lumboperitoneal shunt combined with myelotomy for treatment of syringohydromyelia

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This study was undertaken to investigate whether a direct communication between the fourth ventricle and the syrinx exists in the majority of patients with syringohydromyelia and whether intraspinal pressure plays a role in the pathogenesis of this disorder. A series of 13 patients with syringohydromyelia delineated by magnetic resonance (MR) imaging was reviewed, and the intracranial pressure (ICP) and lumbar spinal pressure (LSP) were recorded concomitantly in three patients. Lumboperitoneal shunting was performed in seven patients, six of whom also underwent myelotomy of the caudal spinal cord. Magnetic resonance imaging visualized a cerebrospinal fluid (CSF) channel connecting the fourth ventricle with the syrinx in only one of the 13 patients, and in the remaining 12 patients it revealed a long segment of the spinal cord free of syrinx below the fourth ventricle. This finding is in accordance with recent MR studies of syringohydromyelia. The concomitant recording of ICP and LSP in our patients who had myelomeningocele and Chiari II malformation showed findings contradicting the reports of Williams. Lumbar spinal pressure was equal to ICP under steady-state conditions, and was elevated by straining or crying more markedly than ICP. Craniospinal pressure dissociation with ICP greater than with LSP, as described by Williams, was not observed. Lumboperitoneal shunting concomitant with myelotomy and syringopleural shunting 1 month after myelotomy resulted in marked and sustained shrinkage of the syrinx and neurological improvement in four patients. It was concluded that the majority of patients with syringohydromyelia and Chiari malformation lack a direct communication connecting the fourth ventricle with the syrinx. The results suggest that CSF moves under pressure into the spinal cord, contributing to the formation and maintenance of the syrinx, and that LP shunting combined with myelotomy can effect shrinkage of the syrinx.

KEY WORDS • hydromyelia • syringomyelia • lumboperitoneal shunt • myelotomy • magnetic resonance imaging

The precise pathogenesis of syringohydromyelia is unknown, and effective surgical treatment of this disorder remains to be established. To explain the formation and maintenance of syringohydromyelia, mechanisms involving hydrodynamics and spinal cerebrospinal fluid (CSF) have been suggested. The premise of the hydrodynamic mechanism is based on an assumption that, in patients with syringohydromyelia, the central canal of the spinal cord is always patent. Proponents postulate that CSF derived from the lateral ventricle, fourth ventricle, or the subarachnoid space of the posterior fossa is forced through the patent central canal and causes syringohydromyelia. In contrast, the spinal CSF mechanism is based on an argument that CSF moves under pressure into the spinal cord along the enlarged Virchow-Robin spaces or the dorsal roots to form syringohydromyelia.

The hydrodynamic mechanism would be valid only if the syrinx communicates with the fourth ventricle through a CSF channel located between the two chambers. In the past, the communication has occasionally been demonstrated by ventriculography, myelography, or syringography. However, no pathological or radiological investigation has established the existence of the communication in the majority of patients with syringohydromyelia. Proponents of this theory believed that the earlier radiological techniques were too limited to delineate the communication and, by the time of postmortem examination, the communication had collapsed, escaping detection on pathological examinations. Despite the lack of evidence supporting the existence of the communication, the hydrodynamic mechanism has led to treatment of syringohydromyelia with ventricular shunting, plugging of the obex.
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FIG. 1. Extent of the syrinx delineated by magnetic resonance imaging in 13 patients. Note that the rostral end of the syrinx is located below the C-1 level in 11 patients. Case 4 was found to have no recognizable opening into the syrinx on microsurgical examination of the fourth ventricle. Case 12 had a possible communication between the fourth ventricle and the syrinx.

decompression of the foramen magnum, drainage of the fourth ventricle, and terminal ventriculostomy. Magnetic resonance (MR) imaging is an excellent technique to delineate a syrinx. While reviewing our surgical results of obex plugging procedures in previously reported cases, we unexpectedly found that MR imaging often shows a long segment of normal spinal cord between the fourth ventricle and the syrinx. Since this observation may constitute important evidence against the hydrodynamic mechanism, we utilized MR studies to examine whether, in the majority of patients with syringohydromyelia, a direct communication exists between the fourth ventricle and the syrinx. In addition, the role of intraspinal pressure in the pathogenesis of syringohydromyelia was investigated by making simultaneous measurements of intracranial pressure (ICP) and lumbar spinal pressure (LSP), and performing lumbar peritoneal (LP) shunting in patients.

Clinical Material and Methods

This review includes 13 patients in whom MR imaging revealed syringohydromyelia. All patients were studied using a spine surface coil on a 1.0-tesla Siemens system. Sagittal and transverse T₁-weighted spin-echo images were obtained with a repetition time (TR) of 500 msec and an echo time (TE) of 17 msec. Twelve of the 13 patients were younger than 16 years and one was 31 years old. Associated pathological abnormalities were Chiari II malformation and myelomenigocele (nine patients), Chiari I malformation (two patients), lipomyelomeningocele (one patient), and posterior fossa arachnoidal adhesions (one patient). Clinical manifestations were spasticity or weakness of the extremities, paresthesias in the hands, scoliosis, and urinary incontinence.

Simultaneous measurements of ICP and LSP were made in three patients. In one patient, an ICP bolt and an LP shunt tube were placed, and ICP and LSP were continuously monitored for 24 hours.* In the other two, ICP and LSP were measured while the patients were anesthetized in the operating room. In these patients, ICP was measured via a No. 23 needle placed in the reservoir of ventriculoperitoneal shunts and LSP was measured via LP shunt tubes placed in the lumbar theca. The transducers were referenced to the L-4 vertebral level for recording LSP and to the foramen magnum for recording ICP.

Seven patients underwent LP shunting (five patients with a Chiari II malformation, one patient with a Chiari I malformation, and one patient with a lipomyelomeningocele). Five of these patients underwent surgery after the syringomyelia failed to respond to myelotomy, syringosubarachnoid shunting, syringopleural shunting, or obex plugging procedures. The follow-up period ranged from 10 to 24 months. The remaining six patients were left untreated because they were asymptomatic and the syrinx was small.

In the seven patients with LP shunting, T-tube shunts or newly designed LP shunts were placed in the lumbar region where a capacious CSF space was noted on MR imaging.† In the myelodysplastic patients, the shunts

* Pressure transducers manufactured by Gould Inc., Oxnard, California; pressure recorder, Model 7754B, manufactured by Hewlett-Packard, Avondale, Pennsylvania.
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were inserted caudal to the neural placode. In six of these patients, a myelotomy was carried out at the caudal end of the spinal cord and the shunts were then placed in the spinal theca under direct vision. One patient whose syrinx was located at the cervicothoracic level had previously undergone syringopleural shunting. In this patient, only a shunting procedure was performed.

Results

MR Findings of Syringomyelia

Figure 1 illustrates the extent of the syrinx noted in our patients. The most significant finding is that in all except two patients, the rostral end of the syrinx was separated from the fourth ventricle by a varying length of syrinx-free spinal cord. In only one patient was there a possible CSF channel connecting the fourth ventricle and the syrinx (Fig. 2); however, the channel was markedly narrower than the syrinx. In one patient who had holocord syringohydromyelia associated with posterior fossa arachnoidal adhesions, MR imaging revealed that the rostral end of the syrinx terminated only a few millimeters from the caudal portion of the fourth ventricle. The fourth ventricle was explored in an attempt to plug the obex, but the floor of the fourth ventricle was completely sealed with no recognizable direct communication between the ventricle and the syrinx. It is of note that the syrinx often expanded in the caudal spinal cord and consisted of multiple compartments separated by thin septa.

Measurement of ICP and Lumbar Spinal Pressure

Williams has demonstrated in infants with myelomeningocele and Chiari II malformation that LSP is lower than ICP. Contrary to that report, the LSP measured in three of our patients was always higher than ICP. Figure 3 illustrates the continuous pressure recording in a boy with a Chiari II malformation. While he remained calm in the supine position, LSP was equal to ICP. When he cried in the supine position, both LSP and ICP rose, but the LSP rose much more than the ICP. When he was in the upright position, steady-state LSP exceeded ICP by approximately 30 mm Hg, a difference which was only partially accounted for by the difference in height of the two transducers. When he cried, the difference in pressures further increased. Figure 4 shows changes in LSP and ICP in response to abdominal compression in another boy with a Chiari

Fig. 3. An example of a continuous 24-hour recording of intracranial pressure (ICP) and lumbar spinal pressure (LSP) in a child with syringohydromyelia and myelomeningocele. When he was asleep in the supine position, ICP and LSP were equal; when he cried, both rose, the LSP more than the ICP (upper panel). When he was in the sitting position, steady-state LSP was higher than ICP; crying in the sitting position elevated LSP more than ICP (lower panel). The LSP was never lower than the ICP throughout the 24-hour recording period.
II malformation. Abdominal compression rapidly increased LSP in both the supine and the upright position, while it did not affect ICP. In a third child, in whom these measurements were made, changes in pressure followed a similar pattern.

Surgical Results of LP Shunt

Of the seven patients who underwent insertion of LP shunts, postoperative MR imaging in four patients showed unequivocal shrinkage of the syrinx. The cases of two patients are described below. Two other patients on whom myelotomy and LP shunting were performed showed noticeable postoperative expansion of the syrinx. An isotope shuntogram of these two patients disclosed complete shunt malfunction and their shunts were subsequently revised. In one of these patients, repeat MR imaging 3 months after the shunt revision showed no change in the syrinx. Another patient developed low-pressure headaches after the shunt revision, which led to incorporation of a low-pressure valve into the LP shunt. This surgical procedure relieved the headaches but resulted in infection of the LP shunt, requiring removal of the shunt. This patient is to undergo LP shunt replacement; myelotomy will be also performed if the previous myelotomy has closed. In one patient, LP shunting has not changed the syrinx over a period of 6 months.

Case Reports

Case 1

This 14-year-old boy first developed progressive left hemiparesis, the upper extremity being weaker than the lower extremity, and then weakness of the right lower extremity. In addition, he noted pain in the left shoulder. On examination, he showed normal sensation but mild muscle atrophy and weakness of the upper extremities. Reflexes were brisk throughout, and sustained ankle clonus was elicited bilaterally. Metrizamide computerized tomography (CT) myelography demonstrated caudal displacement of the cerebellar tonsils to C-1 and intramedullary contrast enhancement from C-1 to T-4, suggesting a Chiari I malformation and syringomyelia. With the aid of intraoperative ultrasonography, a midline myelotomy and insertion of a syringosubarachnoid shunt at the C-6 level were carried out in May, 1986. The fluid aspirated from the cavity was clear and contained protein (15 mg/dl). Motor strength was significantly improved after the operation.

Two months later, the patient developed recurrent motor weakness and became ataxic. An MR study showed caudal displacement of the cerebellar tonsils 2.5 cm below the foramen magnum and an intramedullary cystic cavity with low signal intensity extending from C-2 to C-7. From C-3 to C-6 the syrinx enlarged the anteroposterior diameter of the spinal cord to completely fill the spinal canal. In July, 1986, the posterior rim of the foramen magnum was removed and the cervical cord was reexplored. The syringosubarachnoid shunt was completely blocked and a new syringopleural shunt was inserted.

Despite these efforts, the patient’s motor weakness progressed to the point where he became bedridden. Repeat MR imaging showed a persistent syrinx at the same level (Fig. 5A). In August, 1986, an LP shunt was placed. On the 1st postoperative day, motor strength of the upper extremities began to improve noticeably. By 3 weeks after the operation, motor strength returned to 4+/5+ in the right upper extremity, 3+/5+ in the left upper extremity, and 4+ in the lower extremities. Isotope shuntography in September, 1986, confirmed normal shunt function. By November, 1986, the patient became ambulatory and since August, 1988, he has been able to walk independently. In keeping with his neurological improvement, postoperative MR imaging 3 weeks and 7 months after LP shunting disclosed marked collapse of the syrinx (Fig. 5B and C).

Case 2

This myelodysplastic infant had a shunt placed for treatment of hydrocephalus. At the age of 6 weeks he developed symptoms of a Chiari II malformation, for which he underwent a decompressive cervical laminectomy from C-1 to C-3 in October, 1983. At 2 1/2 years of age, he showed slowly increasing spasticity of the right lower extremity. Metrizamide CT myelography revealed a syrinx involving the upper thoracic and the caudal spinal cord. The conus medullaris was tethered at the L-4 level and was markedly thinned by the syrinx. In September, 1985, a myelotomy was performed on the conus medullaris to drain the syrinx and the tethering was released. Repeat CT myelography 8 months after the operation showed no interval changes in the
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**FIG. 5.** Case 1. **A:** Preoperative magnetic resonance (MR) image disclosing a persistent syrinx in the cervical cord despite syringosubarachnoid and syringopleural shunting at the C-6 level. **B:** Marked shrinkage of the syrinx was demonstrated on this MR image 3 weeks after lumboperitoneal shunting. **C:** The syrinx remained shrunken on repeat MR imaging 7 months after the shunting.

The syrinx and the spasticity had worsened. In June, 1986, the posterior fossa was reexplored and a piece of muscle was placed to plug the obex. It was evident at that time that the fourth ventricular outlets were completely occluded by arachnoidal adhesions. An initial MR image 4 months after the obex plugging revealed that the syrinx involved the lower cervical cord and the rest of the caudal spinal cord (Fig. 6A). The spinal cord terminated in the cystic collection at the upper lumbar region. In February, 1987, an LP shunt was inserted, and MR imaging 2 months later showed marked collapse of the syrinx in the thoracic cord (Fig. 6B). The cervical syrinx remained unchanged. Repeat MR imaging in August, 1988, showed sustained collapse of the syrinx.

**Discussion**

Three important observations were made in this small series of patients. First, in the majority of patients with syringohydromyelia, no CSF channel was found between the fourth ventricle and the syrinx on MR imaging. Second, in syringohydromyelia patients who had associated myelomeningocele and Chiari II malformation, the LSP was equal to or higher than ICP. Third, drainage of CSF via LP shunts effected shrinkage of the syrinx. These observations support the theory presented by Aboulker,1 and Ball and Dayan2 and cast doubts on the validity of the hydrodynamic mechanism proposed by Garder, et al.5-10 with respect to the causes of syringohydromyelia.

The MR findings concerning the communication between the fourth ventricle and the syrinx were contrary to what we had initially anticipated but coincide with recent reports on MR studies of syringohydromyelia.17,23,26,28 These reports, including the present one, describe a total of 127 patients with syringohydromyelia documented by MR imaging. The most pertinent finding in this total series is that none of the patients had an evident communication between the fourth ventricle and the syrinx. Only three previously reported patients17,26 and one of our patients showed narrow CSF channels connecting the fourth ventricle and the syringeal cavity; thus, the communication was probably present in only 3% of the 127 patients. Since MR imaging can delineate a small CSF channel, such as the aqueduct of Sylvius, and its resolution in outlining the brain stem and upper cervical cord is excellent, it should demonstrate a communication greater than 1 mm in diameter between the fourth ventricle and the syrinx and leave only microscopic communications unvisualized. However, there are no pathological data to show that microscopic communications exist in the majority of syringohydromyelia patients. The absence of such pathological data stands out in the literature.2 It seems inconceivable that microscopic communications could always escape detection at pathological examinations.

Moreover, the mere presence of a microscopic channel does not indicate that CSF actually flows through it. As suggested by Ball and Dayan,2 one can calculate pressures required to move fluid through a tubular
channel, such as the dilated central canal, using the Poiseuille-Hagen formula. The formula states: 

\[ P = \left( \frac{8 \eta L}{\pi r^4} \right) \times F \]

where \( P \) is the pressure difference between the fourth ventricle and the syrinx (in mm Hg), \( F \) is flow (in ml/sec), \( \eta \) is viscosity, and \( L \) is the distance between the fourth ventricle and the syrinx (in mm). Assuming that flow is 0.05 ml/sec, radius of the channel is 0.5 mm, and length of the channel from the fourth ventricle to the syrinx is 5 mm, ICP must exceed intraspinal pressure by a minimum of 10.2 mm Hg in order to move CSF from the fourth ventricle into the syrinx. However, the ICP in our patients was equal to or lower than the LSP. We therefore argue that, even if microscopic communications exist, they would not function as a pathway for CSF flow from the fourth ventricle to the syrinx.

Our observation that the LSP was higher than the ICP contradicts the report of Williams. He reported that, in patients with myelomeningocele and Chiari II malformation, steady-state LSP was lower than ICP and, after a period of straining, the difference in pressures of the two compartments increases. He also demonstrated that, in patients with Chiari I malformation and syringohydromyelia, LSP and ICP are equal at a steady state and during straining but, immediately after straining, LSP falls to a greater degree than does ICP and remains low until it rises to the level of ICP, producing a transitory pressure differential. Williams termed the difference in pressure “cranspinal pressure dissociation” and claimed that pressure dissociation causes “suck” of CSF from the fourth ventricle to the syrinx. Since the pressure dissociation in his patients sometimes approached the order of 10 mm Hg, this phenomenon could be an important pathogenetic factor in his patients. However, the pressure dissociation noted in our myelomeningocele patients differed from that reported by Williams in that the baseline LSP was equal to ICP and straining always raised LSP more markedly than ICP. The reasons for the disparate observations are unclear, but a major difference in the two investigations is the age of patients at the time of the pressure measurements. All of the 10 patients examined by Williams were neonates or infants and the pressures were measured in some of the patients before or shortly after repair of the myelomeningocele. In contrast, we measured pressures in patients older than 3 years of age and long after myelomeningocele repair. It is possible that leakage of CSF from the spinal canal in Williams’ patients contributed to the low LSP. Whether patients with Chiari I malformation and syringohydromyelia very often show pressure dissociation as suggested by Williams remains to be confirmed.

Ball and Dayan were the first to propose a possible role of intraspinal pressure in the pathogenesis of syringohydromyelia. Aboulker later endorsed this view. Although the proposal was made on the basis of pathological findings of the spinal cord with syringohydromyelia, evidence from radiological and physiological studies also provides support for it. First, metrizamide and fluorescein instilled into the spinal theca are recovered in the syringeal cavities confined to the caudal spinal cord. This indicates that certain diffusible substances such as CSF can reach the syringeal cavity from the spinal subarachnoid space across the spinal cord parenchyma. It is possible that these substances can diffuse through the spinal cord and that diffusion is facilitated by an intermittent rise of LSP above intrasyringeal pressure. The increased pressure is known to facilitate diffusion of substances across the permeable tissue. An osmotic gradient of metrizamide and fluorescein cannot account for diffusion of the substances into the syrinx because the osmotic pressure caused by intrathecal instillation must be highest at the spinal subarachnoid space. Moreover, metrizamide and fluorescein were discovered in the syringeal fluid with low protein content. Second, syringohydromyelia is frequently seen distal to the level of underlying lesions which may augment increases in the intraspinal pressures in response to straining. The prime examples of the underlying lesions are spinal cord injury, lipomyelomeningocele, extradural tumor, and arachnoiditis. In these cases, a weakened site in the spinal cord may provide a pathway through which CSF would enter the spinal cord more easily than otherwise. Third, as shown in the present investigation, in patients with Chiari malformation and syringohydromyelia, LSP markedly fluctuates but remains equal to or higher than ICP. This pressure gradient makes it impossible for CSF to flow from the ventricle into the syrinx. The fact that LP shunting reduced the syrinx in some of our patients is additional evidence supporting the role of intraspinal pressure in the pathogenesis of syringohydromyelia.

Whether LP shunting will prove clinically useful in the treatment of syringohydromyelia remains to be determined. Nevertheless, several potential problems with LP shunting for syringohydromyelia should be discussed. One of them is that, as demonstrated in previous studies of humans and dogs, syringeal pressure may be higher than spinal subarachnoid pressure. Although the reports do not address variations of the pressures in different body positions and straining, it is conceivable that a fall in LSP after LP shunting leaves the syringeal pressure higher than LSP, leading to expansion of the syrinx. We have no direct evidence to support this speculation, but to avoid the potential complication in our patients we carried out myelotomy immediately before LP shunting. Perhaps needle aspiration of the syringeal fluid followed by LP shunting may achieve the same results. Another potential complication is caudal displacement of the brain stem and cerebellum aggravated by LP shunting. In certain cases, the decompression of the foramen magnum before LP shunting may be required to prevent this complication. Inasmuch as a decompressive procedure can result in collapse of the syrinx, this procedure should be considered before LP shunting is carried out. Lastly, as noted in our patients, low-pressure headaches, shunt...
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blockage, and infection may occur. The risk of shunt infection and malfunction may be reduced by employing various surgical techniques. Prevention of low-pressure headaches is a challenging problem because the syrinx may not shrink if too little CSF is drained via the shunt.

In conclusion, our observations indicate that, at the time of clinical presentation, patients with syringohydromyelia only rarely harbor a direct communication between the fourth ventricle and the syrinx. The spinal CSF is an important source of syringal fluid, and LP shunting can result in collapse of the syrinx.

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References


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