Percutaneous thecoperitoneal shunt for syringomyelia

Report of three cases

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Between January and April, 1990, three consecutive cases of syringomyelia were treated by percutaneous placement of thecoperitoneal shunts. Two of these patients had undergone craniocervical decompression earlier at other centers and the third was treated primarily by a thecoperitoneal shunt. In each case, the syrinx was associated with Chiari I malformation, although the clinical presentation was due to a myelopathy. All three patients obtained unequivocal benefit from this simple procedure. Postoperative magnetic resonance images showed considerable shrinkage of the cysts corresponding with clinical improvement.

Key Words: syringomyelia • thecoperitoneal shunt • Chiari I malformation • shunt

Thecoperitoneal shunting is now a well-established technique for treating communicating hydrocephalus. It has also proved its usefulness in cases of cerebrospinal fluid (CSF) rhinorrhea, to arrest an intractable CSF leak, and in benign intracranial hypertension to prevent impending loss of vision. We have found this technique rewarding in three consecutive cases of syringomyelia treated between January and April, 1990, and would like to place our experience on record for future evaluation of the technique.

All three patients presented clinically with a progressive myelopathy. In all three, preoperative magnetic resonance (MR) studies showed the cysts to be located in the cervical cord and revealed an associated Chiari I malformation. Two patients had been treated earlier at other centers by craniocervical decompression, but the symptoms had persisted. In the third case, a thecoperitoneal shunt was used as the primary procedure.

Case Reports

Case 1

This 45-year-old woman was admitted to Prince Alykhan Hospital, Bombay, on December 16, 1989, with a 6-month history of progressive difficulty in walking. She also had some weakness of the hands, with minimal clawing.

Examination. The patient had spasticity of all four limbs, hyperreflexia, and upgoing plantar reflexes on both sides. There was no objective sensory loss and cerebellar signs were absent. She had undergone surgery on the craniocervical region 15 years earlier, but could furnish no further details.

Plain x-ray films of the cervical spine showed evidence of craniocervical decompression. On December 25, 1989, MR imaging showed a fusiform syrinx within the cervical cord extending from C-2 to C-7 (Fig. 1 left). Associated with the syrinx was a Chiari I malformation of the cerebellar tonsils. The syrinx had no continuity with the fourth ventricle, which was normal in size and shape.

We recommended re-exploration of the craniocervical region for duroplasty and syringotomy; however, the patient refused major surgery at the same site as before. It then occurred to one of the authors (U.S.V.) that a thecoperitoneal shunt might prove beneficial in this case by providing controlled drainage to the subarachnoid space. This presumption was based on observations made by radiologists during investigations for a syrinx and on the hypotheses put forward by some workers to explain the development and expansion of a syrinx by abnormal hydrodynamic forces within the subarachnoid space. These can be summarized as fol-
lows. 1) When air myelography was the investigation of choice for suspected cervical cord compression, it was observed that, in syringomyelia, the expanded cord shadow seen on the initial films had collapsed on the films taken after further exchange of CSF (the "collapsing cord" sign). This was obviously due to emptying of the cyst following CSF drainage. 2) In metrizamide-computerized tomography scans performed to delineate a syrinx, it was observed that the cyst invariably filled with dye on the follow-up study. This is also thought to be due to the passage of dye into the cyst through the cord parenchyma. 3) While explaining the development and expansion of the syrinx, most recent workers have stressed the role of abnormal hydrodynamic forces created in the subarachnoid space as a result of a Chiari I malformation situated critically at the craniovertebral junction. The forces ultimately act on the cord parenchyma either via the fourth ventricle above or (more likely) through the perivascular spaces of the cord itself. Drainage of subarachnoid CSF should, logically, minimize the effect of these forces. We therefore decided to perform this simple procedure, to which the patient consented.

Operation. On January 2, 1990, a thecoperitoneal shunt was successfully inserted under general anesthesia (Fig. 2). A fine multiperforated thecal catheter was introduced percutaneously into the lumbar subarachnoid space through a Touhy needle. The peritoneal catheter was introduced into the pelvic peritoneum through a small Macburney incision. The two catheters were connected in the flank by a tapering plastic connector. The flushing capsule was not used in order to minimize resistance and to ensure a smooth continuous flow of CSF. The fine caliber of the thecal catheter and the slit valves of the peritoneal catheter ensured a pressure-regulated flow.

Postoperative Course. During the immediate postoperative period, the patient noted improved function of her hands. At the time of discharge on January 11, 1990, her gait had improved and she was able to walk freely without support. Repeat MR imaging on January 14 showed complete collapse of the cyst (Fig. 1, right).

Case 2

This 24-year-old man was admitted to B.Y.L. Nair Ch. Hospital, Bombay, on March 20, 1990, with a 1-year history of inability to stand, walk, and sit without support. He had undergone craniovertebral decompression in September, 1988, following which he had regained control over his bladder but the motor disability continued.

Examination. He was found to have a spastic quadriparesis of 4/5 with hyperreflexia, bilateral ankle clonus, and bilateral extensor plantar response. When supported, he attempted to walk with a broad-based gait. His sensations were normal. An MR study on April 3, 1990, showed the presence of basilar invagination, Chiari I malformation, and a syrinx within the cervical cord (Fig. 3, left).

Operation and Postoperative Course. A thecoperitoneal shunt was performed on April 6, as described in Case 1. Postoperatively, the patient had remarkable improvement of his motor function. His spasticity had decreased considerably and he was able to walk without support. The deep-tendon reflexes continued to remain brisk and ankle clonus was also present. Repeat MR imaging on April 10 showed collapse of the syrinx (Fig. 3, right).

Case 3

This 35-year-old man was admitted to B.Y.L. Nair Ch. Hospital on April 17, 1990. Four years previously he had suffered persistent pain in the right shoulder after lifting a heavy weight, and had noted numbness.
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Fig. 2. Operative sketches of the suggested simplified management of syringomyelia in which a thecoperitoneal shunt is performed without a myelotomy. The shunt is placed as a percutaneous procedure, avoiding the necessity for an open laminectomy. Arrows indicate flow of cerebrospinal fluid.

of the right side of the face and neck, the right upper extremity, and the left trunk and left lower extremity from below the level of the navel. He also had decreased sweating over the right side of the face and minimal weakness of the right upper extremity.

Examination. The patient had loss of touch sensation as well as pain and temperature in the above-mentioned areas, while his joint and position sense was preserved in all extremities. Power was 4/5 in the right upper extremity. An MR image obtained on April 20 showed a long syrinx within the spinal cord extending from the C-2 to the T-2 level, associated with a Chiari I malformation (Fig. 4 left).

Operation and Postoperative Course. A thecoperitoneal shunt was placed on April 25, as described in Case 1. Postoperatively, the patient showed 50% improvement of sensation on the right side of the face and right upper extremity, and 25% improvement on the left side of the trunk and left lower extremity. Power in the right arm improved to 5/5 but remained 4/5 in the fingers. Repeat MR imaging on April 28 showed complete collapse of the cyst (Fig. 4 right).

Discussion

Until recently, our policy in cases of syringomyelia and associated Chiari malformation has been to perform a craniovertebral decompression with duroplasty and a midline syringotomy. However, the clinical results of this treatment have not been altogether satisfactory. It appears that only 30% of patients show some degree of improvement following this operation. Encouraged by the success of thecoperitoneal shunt place-

Fig. 3. Magnetic resonance images, sagittal views, in Case 2. Left: Image obtained on April 3, 1990, showing a syrinx in the cervical cord, basilar invagination, and a Chiari I malformation. Right: Postshunting image obtained on April 10 showing collapse of the syrinx.
ment in Case 1, we performed this simple procedure in two further cases. Surprisingly, all three patients have shown excellent clinical benefits with radiographic evidence of shrinkage of the cyst.

We studied the recent literature on the relative merits of various operative options for syringomyelia with Chiari malformation. Matsumoto and Symon, in an extensive review, advocated craniovertebral decompression and duroplasty as the primary operation of choice for all syringes associated with Chiari I malformations. This procedure, they believed, corrects the abnormal hydrodynamic forces created by the malformation and ultimately results in collapse of the syrinx. In patients whose cysts and symptoms persist following the primary operation, they advocated syringoperitoneal shunt placement as a secondary procedure. They recommended implanting a syringoperitoneal shunt as a primary operation only in cases where the syrinx is not associated with hindbrain abnormality. However, they mentioned the increasing success of shunting procedures such as placement of syringosubarachnoid or syringoperitoneal systems because of modern microsurgical techniques and the improved quality of shunts.

Gardner's original operation of craniovertebral decompression and plugging of the obex is now rarely performed because, first, it is associated with high mortality and morbidity rates, and, second, in most cases the communication between the syrinx and the fourth ventricle is either thin or nonexistent. Plugging of the obex, therefore, serves no purpose. Gardner's simplified procedure of terminal ventriculostomy also has no rational basis because, in many cases, neither the syrinx nor the central canal reaches the filum terminale.

Park, et al., have stressed the value of lumboperitoneal shunting with myelotomy, performed by an open technique, to achieve collapse of the syrinx. Their valuable experimental work supports the hypothesis of Aboulker and Ball and Dayan that abnormal hydrodynamic forces are created in the subarachnoid spaces of the cervical spine because of the Chiari malformation and that these forces act through the perivascular space of cord parenchyma on the central canal to create the syrinx. Their experiments do not bear out the hypothesis of Williams that a differential rise in pressure in the cranial subarachnoid space acts through the fourth ventricle to open out the central canal and form the syrinx.

Against the background of these options, we believe that the thecoperitoneal shunt may, in the future, play a preferential role in the operative treatment of syringomyelia. The reasons are as follows. 1) It is a simple, safe, and effective technique for draining the syrinx. 2) Standardized thecoperitoneal shunts of good quality are now readily available. This ensures patency of the shunt over a long period of time. Patency can be easily tested at frequent intervals by subarachnoid injection of a radioisotope and performing a scintillation scan of the abdomen. Revision of the shunt, if and when necessary, is also easy. 3) By direct controlled drainage of the subarachnoid space, a thecoperitoneal shunt can empty all kinds of syringes: single, multiple, or multiseptate. In this sense it is superior to syringoperitoneal or syringosubarachnoid shunts which drain only a single cavity or a single compartment at a time. 4) Thecoperitoneal shunting effectively diffuses the abnormal hydrodynamic forces created by a Chiari malformation within the subarachnoid space. It is these forces which, during the patient's straining, act through the cord parenchyma and lead to the development and expansion of the cyst.

In conclusion, we believe that thecoperitoneal shunting can be tried as primary treatment for all cases of syringomyelia associated with Chiari malformation that present clinically with a progressive cord dysfunction.
The coperitoneal shunt for syringomyelia

Craniovertebral decompression and duroplasty, which is a relatively major procedure, should be reserved for cases where a patient presents with clinical features of occipital and neck pains, cerebellar ataxia with nystagmus, and lower cranial nerve palsies, where Chiari malformation is strongly suspected of being the causative pathology.

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