Management and outcome of posttraumatic syringomyelia

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Traumatic paraplegia is the most common cause of nonhindbrain–related syringomyelia. Fifty-seven patients with a mean age of 34.3 years at presentation were treated at the Midland Centre for Neurosurgery and Neurology between 1973 and 1993. A variety of treatment strategies have been used over the years, including syringosubarachnoid and syringopleural shunts, spinal cord transection, and pedicled omental graft transposition. More recently decompressive laminectomy, subarachnoid space reconstruction and formation of surgical meningocele have been used. A total of 81 operations were performed in these patients, 69 of them at the Syringomyelia Clinic. Combinations of strategies were often chosen; the use of one strategy such as drainage did not preclude another such as transection or augmentation of the cerebrospinal fluid pathways.

The overall postoperative complication rate was 12%. Problems specific to the operation type included dislodged, blocked, and infected drains (10 patients). Acute gastric dilation was seen following pedicled omental graft (one patient). At 6 years only 49% of the drains inserted still functioned. A higher than expected rate of cervical spondylotic myelopathy has been noted. Two patients developed Charcot’s joints. Thirty-six patients were asked to score themselves with regard to limb function and performance of daily living activities and 30% reported improvement, particularly in arm function.

Since the use of magnetic resonance imaging has become widespread, it has become apparent that decompressive laminectomy with subarachnoid space reconstruction is effective in controlling the syrinx cavity. In complete paraplegia, spinal cord transection is an effective alternative. Pedicled omental grafting was associated with poor outcome and an increased complication rate and has been abandoned.

KEY WORDS • spinal cord injury • meningeal fibrosis • posttraumatic syringomyelia • syringopleural shunt • syringosubarachnoid shunt

Clinical Material and Methods

Patient Population

Fifty-seven patients with posttraumatic syringomyelia treated at the Syringomyelia Clinic in Birmingham, England between 1973 and 1993 are reviewed in this series. They form part of a database of 620 patients with syringomyelia due to all possible causes who have been treated in this center over the same time. Patients with a follow-up period of at least 1 year have been included. Many of these cases have been included in other publications.32,36–39,41

There is a predominance of males among all paraplegic patients; 48 (84%) of those in our study were male. The mean age at injury was 23.6 years (range 14–45 years) and the mean age at diagnosis of syringomyelia was 34.3 years (range 17–58 years). The average interval between the spinal cord injury and the onset of syringomyelia symptoms was 7.6 years (range 6 months–26 years). The average interval between spinal cord injury and the diagnosis of syringomyelia was 10.7 years (range 1–33 years), which implies an average delay of 3.1 years between the onset of symptoms and the diagnosis. The onset of new symptoms in paraplegic patients is not always appreciated. An average follow-up duration of 7.4 years has been recorded postoperatively (range 1–17 years).
Forty-one (72%) of the patients had complete paraplegia (Frankel Grade A) following rehabilitation from their spinal injury. This does not necessarily imply a relationship between the density of the spinal injury and the likelihood of developing syringomyelia later, but may reflect the higher incidence of complete paraplegia following spinal injury.8 Regarding the level of spinal fracture, 13 (22.8%) of these patients had cervical fracture, 25 (43.9%) had an upper thoracic, 13 (22.8%) had a lower thoracic, and six (10.5%) a lumbar fracture. It is of interest that almost half of the patients had an upper thoracic spinal fracture. This should be contrasted with the fact that the majority of spinal injuries are found at the lower cervical spine, the lower thoracic spine, and the thoracolumbar junction5 and may be because the spinal canal is narrow in the upper thoracic region.

Five patients were referred to our institution following treatment failures elsewhere. In all cases the indications for surgery were progressive symptoms of syringomyelia and several related disorders such as hindbrain herniation. Nevertheless the cases have not been selected rigidly or operated on by predefined methods. The operative techniques have evolved progressively, with modifications being adopted on the basis of failures rather than successes.

It should be noted that the treatment of the majority of the patients falls into one of the two main groups: drainage procedures or subarachnoid space reconstruction. However, there was an overlap in methods in that a significant number of patients were treated by both methods at the same time. Following the evident success of reestablishment and augmentation of the subarachnoid spaces in hindbrain-related syringomyelia, augmentation of the subarachnoid spaces has been practiced in other types of syringomyelia for many years at our clinic.

Drainage procedures performed at our clinic involved laminectomy at the injury site, opening of the dura, dissection of the subarachnoid adhesions to produce some form of subarachnoid space reconstruction, and insertion of a syringosubarachnoid or syringopleural shunt. The dura was left open at the end of the procedure. With this technique 24 patients underwent insertion of syringopleural and six of syringosubarachnoid shunts.

Two syringopleural shunts were inserted in sites away from the level of spinal cord injury. One of these procedures was performed in Birmingham. The syringopleural drains failed in 7 months and 4 years, respectively. In addition, in contrast to our technique for inserting syringopleural drains, such procedures performed elsewhere did not involve reconstruction of the subarachnoid space and the dura was closed in the five patients referred to our center following treatment failure.

Subarachnoid space reconstruction involves laminectomy at the injury site, opening of the dura, dissection of the subarachnoid adhesions to reestablish communication cranially and caudally to the injury site, and augmentation of the subarachnoid space with formation of surgical meningocele by leaving the dura open at the end of the procedure (Fig. 1). The technique has been described in detail elsewhere.16 Dissecting adhesions is an unrewarding surgical task. The major effort was to open the surgical

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**TABLE 1**

Operative procedures performed in 57 patients with post-traumatic syringomyelia treated at the Syringomyelia Clinic (SC) and at other institutions

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**Functional Assessment**

All of the patients were asked to assess their own functional status and judge the effect of surgery on their performance by answering a questionnaire about activities of daily living as well as limb motor and sensory function and bladder function. For the assessment of limb and bladder function, a modification of the scale for cervical myelopathy of the Japanese Orthopedic Association was used.16 Responses were received from 36 patients.

**Surgical Techniques**

Table 1 shows a list of the various procedures performed in these patients. A total of 81 operations were performed in 57 patients, 69 at our clinic. This study is prospective in the sense that care has been taken in preoperative evaluation and close postoperative scrutiny of patients with all varieties of syringomyelia and several
meningocele into the subarachnoid spaces above and below the fracture site. Some dissection opposite the fracture site was necessary to retract the dura; however it was usually not possible to open the subarachnoid space extensively and permanently at the level of the paraplegia. Releasing the dura from the pia at this level may allow primary cysts to relax but they usually fill up again. Stents have been placed in the subarachnoid space to discourage early adhesion formation postoperatively. These stents were removed by the 5th postoperative day. We use the term “stent” as originally envisaged by C. Stent, who observed that the soft-tissue healing could be aided by a hard support that was later removed. Using this technique, 18 patients underwent reconstruction and augmentation of the subarachnoid space at the injury site. Five of these operations had been preceded by drainage procedures that failed to control the syrinx.

Two patients underwent myelotomy at the injury site as an extended syringosubarachnoid shunt and one other underwent terminal ventriculostomy. In five patients pedicled omental grafting was performed to improve the vascularity of the spinal cord. It was believed that their clinical deterioration was due to progressive spinal cord failure, rather than to syringomyelia, which was stable on serial imaging. A limited myelotomy was performed and the omental graft was laid inside the syrinx cavity, with the dura remaining open at the end. Omental grafting procedures were followed by significant morbidity, including chest wound infection and ileus secondary to devascularization of the greater curve of the stomach. In view of the unfavorable outcome and the high complication rate, this procedure was abandoned.

The bar graph in Fig. 2 upper shows the evolution of treatment strategies at our clinic. This graph is based on 69 operations performed in our institution, excluding the 12 that were performed in referring hospitals. In the 1970s and 1980s the majority of the patients underwent drainage procedures including syringosubarachnoid and syringopleural shunts. From the late 1980s there has been a swing away from drainage procedures in favor of subarachnoid space reconstruction. Drainage procedures have not been performed as part of the initial operation since 1991. Initially a temporary drain, which was removed in the first 3 to 5 days postoperatively, was inserted in the syrinx cavity following subarachnoid space reconstruction. In all cases the output from that drain was minimal and therefore temporary drainage was abandoned.

**Results**

**Surgical Outcome**

Table 2 shows a list of postoperative complications, with an overall rate of 12.3%, including operations performed in the referring hospitals. Although this percentage may seem high, the numbers are small in each group. It is of interest that only one patient developed a cere-
brospinal fluid (CSF) leak, although in the majority of the patients the dura was left open at the end of the procedure. Similarly, only one patient had wound breakdown requiring resuturing. In two patients who underwent cervical laminectomy while in the sitting position, postoperative pneumocephalus caused temporary confusion. There were no postoperative deaths.

Of the 34 syringopleural and syringosubarachnoid drains inserted, 10 (29.4%) were associated with some form of mechanical or infective complication (Table 2). On reoperation, three drains were found dislodged from the syrinx cavity and another four drains were occluded by gliotic tissue. On one occasion the insertion of a syringopleural shunt caused formation of a bronchopleural fistula. In this case the pleural cavity was obliterated by adhesions secondary to chest injury at the time of the spinal trauma. Dissection of the pleural cavity damaged the underlying lung, causing a bronchopleural fistula through which the patient was coughing CSF immediately postoperatively. In addition to the two drains that were found infected at reoperation, another patient was successfully treated with antibiotic medications for presumed low-grade infection of one of his drains. He had previously undergone four operations in another unit, leaving him with three implanted drains, two different syringopleural shunts and one thecoperitoneal shunt. Later he underwent a successful subarachnoid space reconstruction procedure in our unit. He was admitted 2 years after this procedure with clinical signs of meningitis and leukocytosis not associated with bacterial growth in the CSF.

The long-term effectiveness of the various procedures was calculated from the outlined data using the Kaplan–Meier product limit method, producing cumulative proportional “survival” curves. The graph in Fig. 2 lower compares 34 drainage procedures with 18 subarachnoid space reconstruction procedures. A procedure was considered a failure when it resulted in direct clinical deterioration or when further recurrent symptoms associated with radiological deterioration of the syrinx occurred following initial postoperative improvement. It is clear from the graph in Fig. 2 lower that after 4 years only 53% of the drains remained effective. In comparison, 83% of the subarachnoid space reconstruction procedures continue to effect clinical stability. The difference is statistically significant (Wilcoxon signed-rank test, p = 0.0912). It is notable that the effectiveness of the subarachnoid space reconstruction procedures remains stable in time (the curve remains flat after the 1st year), in contrast to the continuing decline of the curve for drainage procedures. At 6 years only 49% of drains remain effective. This comparison should be interpreted with caution in view of the difference in follow-up times between the two groups. Nevertheless, the observed difference is clinically significant. The reasons behind the failure of drainage operations have been analyzed elsewhere.32

Radiological Outcome

Postoperative magnetic resonance (MR) images were available in 42 patients. In 37 (88%) of these patients the appearance of the syrinx had improved postoperatively. In
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one case (2%) the appearance was unchanged and in four patients (10%) the radiological appearances of the syrinx postoperatively was worse and this was associated with clinical deterioration. Control was eventually established in every case. In the 37 patients with radiological improvement in their syringes postoperatively, discrepancies were noted between clinical and radiological outcome. Following successful surgery, 26 patients were rendered asymptomatic or improved substantially. In seven of these a persistent primary cyst was seen at the injury site (Fig. 3).

Of the 37 patients with improved postoperative radiology, 11 had persistent symptoms. Of these, four patients had a persistent syrinx cavity, although it was substantially smaller than preoperatively, one had a persistent primary cyst, and six had a completely healed syrinx.

Functional Outcome

Cervical spondylosis is commonly observed in paraplegics. This may reflect the increased workload of the arms and neck in wheelchair-bound paraplegic patients, who have to use their arms to compensate for the loss of leg function. Of the 56 patients, five developed significant cervical spondylosis. Similarly, Charcot’s neuropathic joints have been observed in paraplegics. A total of three joints including one shoulder and two elbow joints were seen on the same patient. These outcomes were seen in patients with syringes extending up to the cervical cord.

Figure 4 shows the results of self-assessment of functional status and the effect of surgery on function in the 36 patients who responded to the questionnaire. Of these, 17 patients (47% of the responders or 30% of the whole group) reported postoperative deterioration of their overall functional status. A small number reported improvement in activities of daily living. It is clear that arm motor and sensory functions are the ones that primarily improved. Not surprisingly, there was little change in leg function. Two patients reported deterioration of bladder function.

Discussion

Although the clinical presentation of syringomyelia after spinal cord injury can be variable, it should be emphasized that the occurrence of new symptoms following a period of stability indicates occurrence of syringomyelia until proven otherwise.37,38,41,42 This particularly applies to pain, although sensory, motor, and autonomic symptoms such as disturbance of sweating can be involved35,36,37, and may be unilateral, even though the syrinx may occupy the center of the cord.37,38 Symptoms below the established level of paraplegia may be more difficult to appreciate. In patients with complete paraplegia, alteration of sweating patterns and improvement of leg spasms can indicate causal extension of the syrinx. In patients with incomplete paraplegia, deterioration of sensory and motor function can be observed, as well as progressive autonomic dysfunction, bladder and bowel deterioration, and lessening of spasms. New contralateral symptoms in a patient who underwent drainage treatment for a deteriorating syrinx and had been stable for a while could indicate the excavation of a new cavity beside the previous one.

The pathology of posttraumatic syringomyelia is interesting. As a result of the injury, spinal cord contusion leads to edema, blood effusion, and subsequent liquefaction and myelomalacia. In a proportion of cases there is cavity formation.17,25 On MR imaging almost 50% of injured spinal cords have a small cyst opposite the fracture site that can be called a primary cyst.2,31 There may be more than one of these. This should be differentiated from the syringomyelic cavity, which extends more than two vertebral bodies cranially and/or caudally from the injury site. This distinction is of clinical importance because following successful surgical treatment the syrinx can become flat but the primary cyst may remain unchanged (Figs. 3 and 5). The primary cyst probably represents established spinal cord damage that took place at the time of the injury, which was followed by gliosis, and surgical treatment directed toward it is unlikely to result in any benefit. The widening of the cord at the site of the primary cyst may contribute to the subarachnoid obstruction at the injury site, which seems to be the main factor involved.
in the filling mechanism of the syrinx. Direct puncture of the primary cyst at operation in a number of the patients in this series did not prevent refilling postoperatively, even in the presence of a collapsed syrinx. The walls of the cyst may be lined with collagen, which is stiff and noncompliant and keeps the cyst dilated. Persistence of the primary cyst does not seem to influence the clinical state in these patients.

As the acute injury settles, the inflammation is followed by adhesions in the subarachnoid space and gliosis in the cord. Obstruction of the CSF pathways is caused primarily by meningeal fibrosis, which causes adhesions in the subarachnoid space, but may also be compounded by narrowing of the bony spinal canal from the spinal fracture. Recent animal studies reinforce these observations.

All these factors contribute to obstruction in the subarachnoid space opposite the fracture site. This may lead to dissociation of pressures on either side of the block.

The fluid pathway into the syrinx has not yet been clearly established. It may enter at the fracture site or close to it. As we have discussed previously, the spinal cord is not waterproof and potential spaces along the vessels of the cord have been described by Virchow and Robin. The cord is usually thin-walled at operation and fluid can sometimes be seen oozing through its wall. It has been postulated that CSF tracks along the perivascular spaces to gain access to the center of the cord. Every time a Valsalva maneuver takes place (for example, coughing or straining), the intrathoracic and intraabdominal pressure is transmitted inside the spinal canal via the valveless venous plexus around the vertebral bodies. Normally pressures are readily equalized between the top and bottom of the spine, with a half-life of less than 0.1 second. In the presence of a partial subarachnoid block, fluid can be forced upward past the block more efficiently than it can run down again. This may lead to a collapsed theca below the block, thus creating a suction effect that promotes entry of CSF into the syrinx cavity. This is the "suck" mechanism. Syrinx fluid may move more readily than the CSF in the subarachnoid space, because the latter fluid finds some resistance from the arachnoid strands, the dentate ligaments, the vessels, and the nerve roots that connect the cord to the dura. The movement of the syrinx fluid can be violent enough to extend the cavity both cranially and caudally. This is the "slosh" mechanism.

These two mechanisms explain the generation and propagation of the syrinx, but are not universally accepted as yet.

There are still some observations awaiting explanation, including the formation of septations inside the syrinx cavity and the evidence that different cavities are under different pressures. The excavation of new cavities beside old ones that have apparently healed appears to represent failure of correction of the filling mechanism and is often associated with drainage problems.

Until recently, most surgical strategies had unreliable success, and this has caused some neurosurgeons to avoid performing early surgery, even in the presence of developing neurological deterioration. With the advent of MR imaging it has been possible to image the spinal cord with accuracy, detect the presence of cavitation within it, and correctly diagnose the cause of neurological deterioration.
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in the paraplegic patient following a period of stability after recovery from the spinal injury, as well as monitor the results of surgery.1,2,6,11,13,27,28,31

Regarding the timing of surgical intervention, it has been our policy to favor early surgery.37,38 Sudden onset of symptoms, as well as the presence of syrinx occupying the cervical cord, potentially threatening the arms of a paraplegic patient, have both been considered indications for emergency surgery. All the patients in this series had advancing symptoms and neurological deficits. Some of them had been undergoing observation in other units. As has been observed previously, delay in surgical treatment can lead to unfavorable clinical results, despite satisfactory radiological improvement of the syrinx.19 This is demonstrated in our series of six patients with completely healed syringes but persistent symptoms following successful surgery. Gliosis has been demonstrated in relation to the syrinx walls,14,32 and this may be responsible for this irreversible change.

Patients with unremarkable symptoms whose syrinx is small and not progressing are difficult to treat. This situation is sometimes encountered postsurgery when the patient improves symptomatically but the syrinx remains moderately full. It may be that these patients are likely to deteriorate at some stage if left untreated and thus merit close monitoring with MR imaging. It might be thought best for such patients to avoid strenuous exercise. The use of extracorporeal “shortwave” lithotripsy is contraindicated because it has been reported to aggravate syringomyelia.21

The type of surgery used to treat syringomyelia has evolved over the years. Syringosubarachnoid and syringopleural shunts were widely used in the 1970s and 1980s with encouraging early results,4,7,11,15,18,20,22,24,27,29,30,33,41 although long-term effectiveness has been questioned recently.32 As clinical observations gradually improved our understanding of the filling mechanism and with the improved radiological imaging of MR studies, the philoso-

FIG. 7. Magnetic resonance (MR) images obtained in a patient with a T11–12 fracture showing the effects of a previous drainage operation performed at the widest part of the syrinx. Upper Left: The drain is seen on the left-hand image with a recurrent syrinx below and a shape that looks like a primary cyst, perhaps caused by the effect of upward sloshing abutting against the zone of gliosis provoked by the myelotomy and the drain. The right-hand image depicts the adjacent sagittal slice with the tip of the drain at the level of the inferior aspect of C-2. It should be noted that the new syrinx is located away from the drain. The symptoms had changed sides from right to left. Upper Right: Low cervical axial MR image showing the tense circular syrinx in the same patient. Compare with the typical postoperative appearance of Fig. 6d. Lower Left: Postoperative axial MR image showing collapsed syrinx and the tube of the syringopleural shunt that was inserted previously. Compare with upper right. Lower Right: An MR image showing that the entire syrinx has collapsed. The T2-weighted image shows persistent excess fluid in the cord in relation to the drain, which would have been avoided by prompt and drainless surgery. Compare with upper left.
The current treatment of posttraumatic syringomyelia in our clinic is decompressive laminectomy and subarachnoid space reconstruction by opening of the subarachnoid pathways past the fracture site and formation of surgical meningocele (Figs. 3, 5, and 8). The filling mechanism of the syrinx seems to be disabled as the partial subarachnoid block is bypassed via the meningocele, which may be producing a dumping effect by absorbing the energy of CSF pulsations.

In cases in which subarachnoid space reconstruction failed to control the syrinx, it is believed that the dissection of the subarachnoid adhesions may have been inadequate. In that situation reoperation to complete the subarachnoid space reconstruction should be considered, as well as spinal cord transection, which is an effective alternative in patients with complete paraplegia.\textsuperscript{3,32,33,34} It would be overly dogmatic to state that drains have no place in the treatment of syringomyelia but, in the same way that syrinx drains have been almost abandoned in the treatment of hindbrain-related syringomyelia, it has proved possible to achieve success without using drains for posttraumatic syringomyelia.

The surgical technique of subarachnoid space reconstruction and augmentation has been shown to result in radiological improvement or healing of the syrinx in 88% of the patients so treated. Despite this, a significant number of patients continue to deteriorate clinically. It is believed that progressive gliosis around the syrinx walls may be responsible for the continued deterioration. Clinical and pathological observations support this view although concrete proof has yet to become available. Medical problems of paraplegic patients markedly exceed those of the normal population at all ages and for quadriplegic patients the outlook is worse by a factor of approximately 1.4.\textsuperscript{35} A similar multiplier may be applied for patients with posttraumatic syringomyelia. If timely treatment is not instituted, neurological deficits may advance and become irreversible. An aggressive surgical policy is necessary to preserve function in these patients.

Acknowledgment

The Editor notes that this article is Bernard Williams' final submission to the Journal of Neurosurgery before his untimely death in September of 1995.

It is our privilege to publish this manuscript on his behalf.

References


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