Treatment of Syringomyelia with a Syringosubarachnoid Shunt

Charles H. Tator and Carlos Briceno

ABSTRACT: The surgical results in 40 patients with syringomyelia, treated with a syringosubarachnoid shunt or other procedures are reviewed. The principal indication for surgery was that of significant neurological deterioration. There were 12 patients with idiopathic syringomyelia without tonsillar ectopia, 12 with an associated Chiari malformation, 11 with post-traumatic syringomyelia and five patients with spinal arachnoiditis. There were 38 syringosubarachnoid shunts performed in 35 patients, and an excellent or good result was achieved in 26 patients (74.3%). In terms of the type of syringomyelia, the best results were obtained in the idiopathic group without tonsillar ectopia and in the post-traumatic group. A short duration of pre-operative symptoms favoured a better outcome, and in our opinion, early surgical treatment is indicated for all patients with neurological deterioration. All eight patients in whom a posterior fossa decompression was performed as the initial surgical procedure required a second operation, either a syringosubarachnoid or syringoperitoneal shunt to achieve neurological improvement or stabilization. Thus, the syringosubarachnoid shunt is an effective therapeutic modality for patients with syringomyelia, particularly for the idiopathic and post-traumatic groups. More than one surgical procedure may be required to achieve cessation of deterioration. Overall, excellent or good results were achieved in 29 (72.5%) of the 40 patients.

RÉSUMÉ: Le traitement de syringomyélie employant un shunt syringo-sous-arachnoïdien Nous revoyons les résultats chirurgiques du traitement de la syringomyélie en employant un shunt syringo-sous-arachnoïdien ou des autres méthodes chez 40 patients. Douze patients souffraient de la syringomyélie idiopathique sans éctopies tonsillaires, 12 patients étaient atteints d'une malformation d'Arnold-Chiari associée, 11 autres avaient la syringomyélie suivante un traumatisme, et 5 patients montraient l'arachnoïdite spinale. Nous avons éffectué l'implantation de 38 shunts syringo-sous-arachnoïdiens chez 35 patients; un résultat excellent ou satisfaisant se réalisa chez 26 d'entre eux (74,3%). À propos du type de syringomélie, les meilleurs résultats ont été produits dans le groupe idiopathique sans éctopies tonsillaires et dans le group post-traumatique. Un cours court de symptômes pré-opératifs était en faveur d'un bon résultat et, à notre avis, le traitement chirurgique sans délai est indiqué pour tous les patients montrant la détérioration neurologique. Chez tous les 8 patients où nous avons effectué une décompression de la fosse postérieur comme la première mode opératoire, ils avaient besoin d'une seconde opération afin de réaliser une amélioration ou une stabilisation neurologique: un shunt syringo-sous-arachnoïde ou syringo-péritonéal. Le shunt syringo-sous-arachoïde est donc une thérapie éfficace pour les patients atteints de la syringomyélie, plus particulièrement les groupes idiopathiques et post-traumatiques. Plus d'une seule opération peut être exigée pour produire un arrêt de la détérioration. En somme un bon ou satisfaisant résultat a été réalisé chez 29 (72,5%) des 40 patients de cette étude.

Can. J. Neurol. Sci. 1988; 15:48-57

The neurological syndrome associated with longitudinal cavitation and gliosis of the spinal cord, medulla or both, was regarded until recently as a chronic progressive degenerative disorder. Although most investigators now consider syringomyelia to be treatable, the indications for surgery and the methods of surgical treatment are controversial.[1,2] In general, the surgical procedures are directed toward either drainage of the syrinx itself, or decompression of the posterior fossa and the foramen magnum, with the variability of treatment being at least partly due to the multifactorial pathogenesis of this disorder. We believe that identification of the pathophysiology of each case is a prerequisite for the selection of the optimal treatment, and that surgery should be recommended to patients with deteriorating neurological function. This paper describes the indications, methodology, and results of treatment of patients with syringomyelia and shows that the syringosubarachnoid shunt provides favorable results in most. The present series of 42 operative cases includes the 20 patients with syringomyelia reported previously,[3] and provides a longer follow-up on these cases.

From the Division of Neurosurgery and Playfair Neuroscience Unit, Toronto Western Hospital, University of Toronto
Reprint requests to: C.H. Tator, Toronto Western Hospital, 399 Bathurst Street, Toronto, Ontario, Canada M5T 2S8

https://doi.org/10.1017/S0317167100027190 Published online by Cambridge University Press

Clinical Material and Methods

Population and Etiology

This paper reports the personal experience of one of the authors (C.H.T.) from 1969 to 1986 in the management of 51 consecutive patients with syringomyelia managed at the Toronto Western Hospital or Sunnybrook Medical Centre, University of Toronto. Eight of the 51 patients have remained stable neurologically and have not required surgical treatment, and one has refused surgical treatment in spite of documented deterioration. Several patients had an initial surgical procedure performed elsewhere prior to referral.

Five other patients not included in this series had extensive syringomyelia secondary to spinal ependymomas. The tumours were removed completely and the syrinxes did not require shunting.

Table 1 shows the etiology of syringomyelia in these 51 patients. This paper will concentrate on the 42 patients treated surgically, 20 from the first series previously reported,[3] and 22 in the second series. There were 28 males and 14 females, a 2:1 ratio. Their ages ranged from 21 to 73 years with a mean of 44 years and a median of 40 years. The etiology of the syringomyelia in these 42 cases is summarized in Table 2. Twenty-four cases were classified as idiopathic syringomyelia and half the cases in this group were associated with tonsillar ectopia. Thirteen patients had post-traumatic syringomyelia (PTS) and in this group, seven had a complete spinal cord injury, and six, an incomplete lesion. The five cases secondary to spinal arachnoiditis included one case of each of the following: head injury and presumably traumatic subarachnoid hemorrhage; bacterial meningitis from a compound depressed skull fracture; spinal osteomyelitis and focal meningitis; local spinal trauma after laminectomy for an unrelated problem; and after spinal anesthesia.

Table 1: Etiology of Syringomyelia in 51 Cases

Etiology		Number of Cases
Idiopathic		29
with tonsillar ectopia	15	
without tonsillar ectopia	14	
Post-traumatic		15
Spinal arachnoiditis		5
Spinal cord infarction		1
Undetermined		1
Total		51

Table 2: Etiology of Syringomyelia in 42 Operated Cases

	Number of Cases				
	First Series		Second Series		Combined
Etiology					
Idiopathic	15		9		24
with tonsillar ectopia	4	8		12	
without tonsillar ectopia	11	1		12	
Post-traumatic	4		9		13
Spinal arachnoiditis	1		4		5
Totals	20		22		42

Symptoms and Signs

The symptoms and signs in these 42 patients are summarized in Table 3. Pain was a prominent symptom and was present in almost 60% of the patients. Dissociated sensory loss was found in almost three-quarters, but posterior column involvement was found in only one-quarter of the patients. Weakness was present in all but one patient, and in three-quarters the weakness was severe. Ninety-five percent showed muscle wasting, and 69% were spastic. Cranial nerve involvement and Horner's syndrome were detected in approximately 25% of the patients. Only six patients had symptoms for less than six months prior to surgery.

Radiographic Findings

Table 4 shows the pertinent radiographic findings in the 42 surgical cases. The myelographic findings using air or iophendylate (pantopaque) have been reported previously.[3] The last 22 patients were all studied with metrizamide or iohexol myelography and computed tomographic scanning (CT scan) and most also had magnetic resonance imaging (MRI) of the spinal cord. Overall, about 60% of the patients showed a widened cord irrespective of the type of radiographic study performed. In one patient an unintentional syringography was performed when the needle entered the syrinx in a patient with adhesive arachnoiditis.

In 16 patients metrizamide or iohexol CT scans were performed shortly after subarachnoid injection of the contrast medium, and then repeated after a delay of approximately eight hours. If the study was inconclusive, it was repeated 24 hours after injection. All patients investigated in this fashion showed evidence of intramedullary pooling of the contrast material or the so-called "bull's eye" appearance. MRI studies showed a low intensity intramedullary signal in all patients investigated with this technique.

Eight of the 10 patients with cranial nerve involvement showed evidence of a Chiari malformation and low tonsils. The ninth had syringobulbia and syringomyelia secondary to spinal arachnoiditis, which developed several years after he sustained a head injury, including a posterior fossa subdural hematoma. The final case had idiopathic syringomyelia without tonsillar ectopia.

Comparisons of the first and second series of patients showed an inversion in the frequency of idiopathic cases associated

Table 3: Symptoms and Signs in 42 Patients with Syringomyelia

	Cases	
Symptoms and signs	Number	Per Cent
Pain	25	59.5
Burn	13	31.0
Dissociated sensory loss	31	74.0
Posterior column sensory loss	11	26.0
Motor weakness	41	98.0
mild	11	26.0
severe	30	72.0
Spasticity	29	69.0
Cranial nerve abnormalities	10	24.0
Horner's syndrome	6/22*	27.0
Wasting 21/22*	95.5	
Non-dissociated sensory loss	2/22*	9.0

*These signs were reviewed in only 22 patients

https://doi.org/10.1017/S0317167100027190 Published online by Cambridge University Press

with tonsillar ectopia, with almost all the recent cases showing this feature, probably due to improved radiographic diagnosis afforded by CT metrizamide studies and MRI.

Overall Operative Management and Surgical Procedure

The indication for surgery was progressive neurological deterioration. Overall, 26 patients were treated with only one surgical procedure, and in this group, the SS shunt was performed as the primary procedure in 23 patients: one patient with spinal arachnoiditis had a syringoperitoneal (SP) shunt as the initial procedure; one patient had a syringostomy; and one patient had a needle aspiration as the primary procedure. The remaining 16 patients were treated with more than one surgical method as described in Table 8.

Our surgical technique for the SS shunt has been described in detail elsewhere,[3] and will be noted here only briefly. Selection of the level of the spinal cord for insertion of the shunt was based on two factors: the site of maximum enlargement of the syrinx in the radiographic studies; and the site of the most severely involved level of the cord on clinical grounds. In addition, intraoperative real time spinal ultrasonography was used for the identification of the largest part of the syrinx. At surgery, the myelotomy site was selected under direct vision with the operating microscope, in a relatively avascular zone at either the site of maximum extent of the syrinx or where the syrinx came closest to the dorsal surface of the cord. In most instances, the myelotomy was performed in the midline, but in some cases an area beside the dorsal root entry zone was selected because the syrinx approached the surface at that point.

Results

The results are based on an analysis of 40 of the 42 surgically treated patients, with two patients excluded because of recent operation and a follow-up of less than six months (both had SS shunts). A recent follow-up was obtained in every other patient. Overall, the postoperative follow-up period ranged from six months to 15 years, with an average of 4.6 years. For 19 of the 20 cases in the first series, the mean follow-up is now 7.9 years. Indeed, in 13, the mean follow-up is 10.1 years.

The clinical results were graded as "excellent", "good", or "poor": excellent was defined as an improvement of neurological deficit; good as a cessation of the progression of the neurological deficit; and poor as further deterioration. These criteria are similar to those used by Love and Olafson.[4] Overall, the clinical results from surgery were excellent or good in 29 patients (72.5%), and poor in 11 patients (27.5%) because of continuing deterioration despite one or more surgical interventions (Table 5).

The duration of pre-operative symptoms ranged from five weeks to 35 years (mean 6.9 years). In only five patients was the duration of pre-operative symptoms less than six months, and this was correlated with good prognosis since four of them had an excellent result (Table 6).

Table 7 shows the clinical results achieved with the SS procedure when performed as a primary surgical treatment or after a previous surgical method had failed. The Table lists 38 SS procedures performed on 35 patients, and all have had adequate follow-up. Of the 30 patients in whom the shunt was the initial treatment, 22 (73.3%) had excellent or good results. When the shunt was performed as a subsequent procedure, only four (50%) of eight had excellent or good results. However, three of the four with good or excellent results were patients having a second SS shunt. The five remaining patients treated with an SS shunt, having received this treatment after other surgical procedures had failed, and only one of them had a good or excellent result. Thus, of the 35 patients who had an SS shunt as the principle or only form of surgical treatment, an excellent or good result was achieved in 26 (74.3%). Three required a second SS shunt.

Eight patients had a posterior fossa decompression (PFD) or Gardner's procedure:[5] seven had herniation of the cerebellar tonsils through the foramen magnum; and one was an idiopathic case with platybasia but without tonsillar ectopia. There were no patients in whom this procedure alone was sufficient to stop the progression of the neurological deficits. Indeed, in six of these cases, an SS shunt was required following the PFD because of continuing deterioration; four achieved an excellent or good result, and two a poor result. The seventh patient with a PFD had a syringostomy performed simultaneously and was graded excellent for eight months, but then began to lose some of the recovered neurological function. The MRI showed a persistent distended syrinx although it was smaller than preoperatively (Figure 1). An SS shunt was then inserted with a dramatic improvement, but the follow-up to date is only two months (Figure 1). The eighth patient continued to do poorly after a PFD with shunting of the fourth ventricle and subsequent treat-

Table 4: Radiographic Findings in the 42 Operated Cases

	Myelogram			CT Scan	
Findings	**Air**	**Pantopaque**	**Metrizamide**	**Metrizamide**	**MRI**
No. of cases	9	20	17	16	17
Collapsing cord sign	7	—	—	—	—
Cord Size					
— widened	—	12	10	10	10
— normal	—	6	3	1	4
— atrophic	—	1	2	4	2
— not visualized	—	1	2	1	1
Tonsillar/cerebellar ectopia	—	3	4	5	7
Arachnoiditis or tethered cord	—	2	3	1	1
Arachnoid cyst	—	1	—	—	—
Bull's eye appearance	—	—	—	16	—
Intramedullary low signal intensity	—	—	—	—	17

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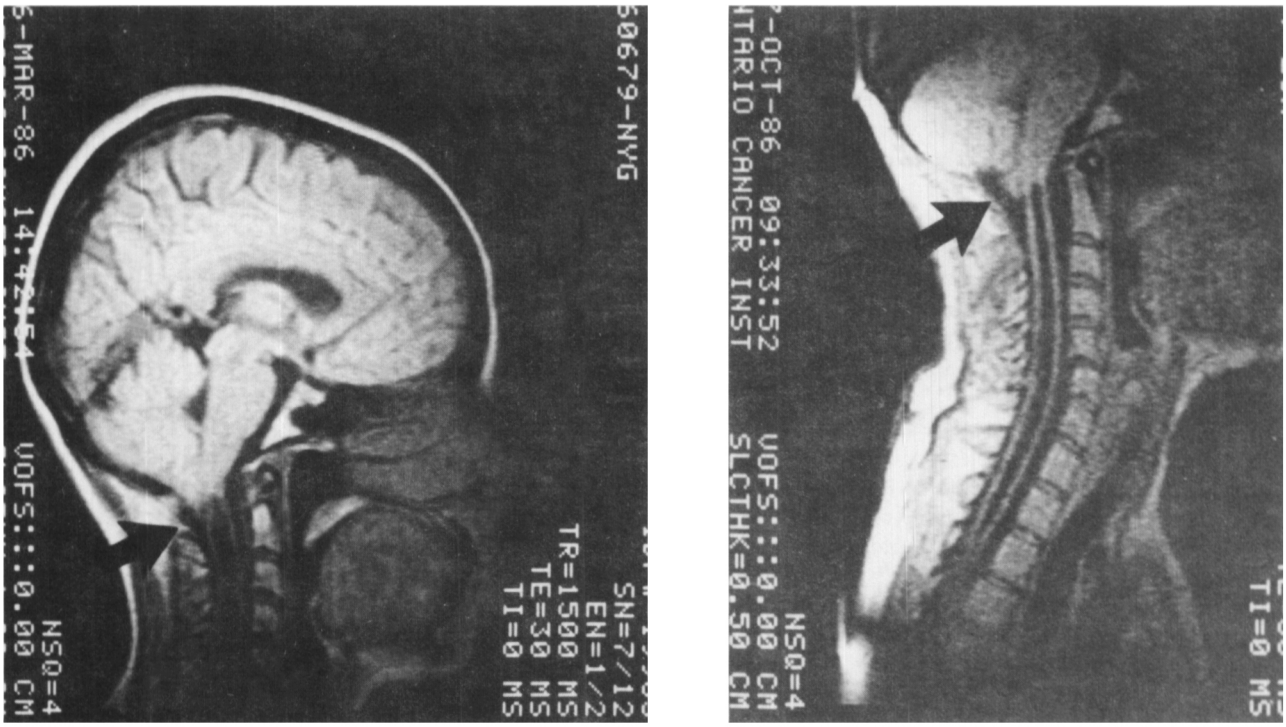

Figure 1 — Left. Pre-operative MR shows Chiari I malformation and syringomyelia. There is a narrow subarachnoid space around the tonsils (arrow). Right. MR 8 months after PFD with duraplasty and myelotomy (syringostomy). The subarachnoid space around the tonsils is wider (arrow), but the syrinx is only moderately smaller.

ment by a ventriculoperitoneal shunt; this patient refused to have further surgical treatment.

Twelve patients required more than one procedure in an attempt to stabilize the neurological deterioration after failure of the initial operation, and this was achieved in 50% (Table 8). Five had associated tonsillar ectopia, three of whom improved or became stable. Two were idiopathic cases without tonsillar ectopia, of whom one became stable, and the other continued to deteriorate. There were three cases of PTS requiring multiple procedures: two improved, and one was a poor result. Finally, two patients had spinal arachnoiditis and both had poor results in spite of an additional lumboperitoneal shunt and an SP shunt in one, and a terminal ventriculostomy in the other after an initial SS shunt had failed.

The relationship between the etiology of syringomyelia, the method of surgical management and the clinical result is presented in Table 8. Six of the 12 patients with idiopathic syringomyelia and tonsillar ectopia had a PFD procedure and drainage of the syrinx with a shunting procedure or a syringostomy, performed simultaneously or at a subsequent operation, and there was an excellent or good result in five patients and only one continued to deteriorate. The result was poor in one patient following a PFD and shunting of the fourth ventricle, but without drainage of the syrinx, and in another patient following simple needle aspiration of the cavity. In the remaining four patients in this group, an SS shunt was performed as the only procedure and three had excellent or good results.

In 10 of the 12 patients in the idiopathic group without evidence of tonsillar ectopia, drainage of the syrinx with an SS shunt arrested the neurological deterioration. One patient with a thoracic arachnoid cyst in whom an initial SS shunt did not stop the progression of the neurological deficit, required an SP shunt because of arachnoidal adhesions at the thoracic level, and a good result was obtained. In the other patient, platybasia

Table 5: Overall Clinical Results of Surgical Treatment in 40 Patients with Adequate Follow-up

	Cases	
Result	**Number**	**Per Cent**
Excellent	19	47.5
Good	10	25.0
Poor	11	27.5

Table 6: Effect of Duration of Preoperative Symptoms on Clinical Results of Surgical Treatment in 40 Patients with Adequate Follow-up

Duration of symptoms	**Excellent**	**Good**	**Poor**	**Total**
less than 6 months	4	0	1	5
more than 6 months	15	10	10	35
Total	19	10	11	40

Table 7: Clinical Results in 35 Patients Treated with a Syringosubarachnoid Shunt with Follow-up More Than Six Months

Procedure	**Excellent**	**Good**	**Poor**	**Total**
SS shunt as Initial Treatment	14*	8	8	30
SS shunt as Subsequent Treatment**	3	1	4	8
Total Number of Procedures	17	9	12	38***

* One case underwent PFD simultaneously.

** Three of the eight cases had had a poor result from the initial SS shunt and the other five had had a poor result from another operative procedure.

*** Thirty-eight SS shunts were performed in these 35 patients who achieved excellent results in 17, good in 9, and poor in 9. Thus, the SS shunt produced an excellent or good result in 26 (74.3%) of 35 patients.

https://doi.org/10.1017/S0317167100027190 Published online by Cambridge University Press

was the only abnormality associated with the syrinx, and the outcome was poor despite a PFD followed by syrinx aspiration and then, an SS shunt.

In the 11 patients in the PTS group, seven patients were treated with the SS shunt as a primary surgical procedure (Table 8) and the result was excellent or good in six and poor in the seventh. Three other patients required more than one surgical procedure because of further deterioration: two had an excellent result, but in the third, the final result remained poor despite three shunting procedures, including an SP shunt. The remaining patient in this group had, at operation, multiple small cavities suggestive of myelomalacia rather than a definite unilocular cyst: a syringostomy was performed, and the result was poor.

The worst results were in the five patients with syringomyelia associated with spinal arachnoiditis, although two of the patients had a good result from SS shunts. In the third patient, an SP shunt gave a poor result, and in the remaining two, the results remained poor despite multiple drainage procedures (Table 8).

A summary of the relationship between the etiology and the clinical results is given in Table 9. In idiopathic syringomyelia, excellent or good results were obtained in eight of the 12 patients with a Chiari malformation, and in 11 of the 12 patients without evidence of tonsillar herniation. In the group with PTS 8 of 11 patients had an excellent or good result. In the five patients with syringomyelia secondary to spinal arachnoiditis, none improved, two had good results and three continued to deteriorate.

The long-term results of surgical treatment were evaluated in the 20 cases in the first series. The initial post-operative follow-up was extended from five years to 10.1 years in 13 patients, but the remaining 7 patients could not be located for further follow-up. We found that the initial result was unchanged in 11 of these patients but in the other two, the initial neurological deterioration which placed them in the poor category had ceased. One of these patients had idiopathic syringomyelia associated with a Chiari malformation and underwent an SS shunt followed by a simultaneous PFD, and then an SS shunt revision a year later. He was graded as a poor result after three years of follow-up. However, seven years after the second operation, he became stable neurologically and has remained so for the subsequent five years. The second case had idiopathic syringomyelia without tonsillar ectopia, and the continuing neurological deterioration was arrested by a second procedure (SP shunt) performed five years after the initial one.

There was no operative mortality in the 42 operated patients, but mild to moderate transient neurological deterioration occurred postoperatively in seven patients. The majority improved within a few days, although only after two months in one case. All seven patients ultimately improved to at least their pre-operative state. One patient with a poor result after an SS shunt and a PFD, developed subluxation at C5-6 four years later and required

Table 8: Relationship Between Etiology, Method of Surgical Treatment and Clinical Results in 40 Cases with Adequate Follow-up

Etiology and Procedure	Results			
	Excellent	Good	Poor	Total
I. Idiopathic:				
A) with tonsillar ectopia	6	2	4	12
SS(a) shunt only	2	1	1	4
Terminal ventriculostomy, then PFD(b), then SS shunt	1	—	—	1
PFD & IV ventricle shunt, then VP(c) shunt	—	—	1	1
PFD, then SS shunt	1	—	—	1
PFD and syringostomy	1	—	—	1
SS shunt, then PFD & SS shunt revision	—	1	—	1
PFD and SS shunt	1	—	—	1
PFD, then SS shunt, then terminal ventriculostomy	—	—	1	1
Needle aspiration only	—	—	1	1
B) without tonsillar ectopia	7	4	1	12
SS shunt only	7	3	—	10
Removal of spinal arachnoid cyst, then SS shunt, then SP(d) shunt	—	1	—	1
PFD, then VA(e) shunt, then syrinx aspiration, then SS shunt	—	—	1	1
II. Post-traumatic:	6	2	3	11
SS shunt only	4	2	1	7
Syringostomy	—	—	1	1
SS shunt, then SP shunt, then higher SS shunt	—	—	1	1
SS shunt and lysis of arachnoid adhesions, then lower SS shunt, then LP(f) shunt, then SP shunt	1	—	—	1
Cordectomy and shunting, then SS shunt	1	—	—	1
III. Spinal Arachnoiditis:	—	2	3	5
SS shunt only	—	2	—	2
SP shunt only				
SS shunt, then LP shunt, then SP shunt	—	—	1	1
SS shunt, then terminal ventriculostomy	—	—	1	1
Total	19	10	11	40

(a) SS: syringosubarachnoid
(b) PFD: posterior fossa decompression
(c) VP: ventriculoperitoneal
(d) SP: syringoperitonal
(e) VA: ventriculoatrial
(f) LP: lumboperitoneal

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Table 9: Relationship Between Etiology and Clinical Result in 40 Operative Cases with Adequate Follow-up

Type of syringomyelia	Excellent	Good	Poor	Total
Idiopathic	13	6	5	24
with tonsillar ectopia	6	2	4	12
without tonsillar ectopia	7	4	1	12
Post-traumatic	6	2	3	11
Secondary to spinal arachnoiditis	0	2	3	5
Total	19	10	11	40

an anterior cervical fusion at that level. Two years later, subluxation occurred at C4-5 and C6-7 as well, and fusions were also required. Another patient with PTS who benefited from an SS shunt, required an anterior cervical decompression and fusion one year after the shunting procedure due to further deterioration produced by an anterior bony compression. The patient subsequently improved to his pre-operative state.

Table 10 shows the relationship between the size of the cord as assessed radiographically, and the clinical result. Eighteen of the 26 patients (69%) with widened cords, and eight of nine (89%) with a normal cord diameter had a good or excellent result.

Discussion

The clinical findings were similar to those reported in previous series,[6-10] and were quite variable. Pain, especially axial or radicular pain, was the most common symptom, and was present in almost 60%. Dissociated sensory loss, muscle wasting and weakness, and leg spasticity were the most common signs. Muscle wasting in the upper extremities was found in 95% of cases. One-quarter of the patients had cranial nerve abnormalities, of which involvement of the trigeminal spinal tract was the most common. As reported previously[3] no single symptom, sign or group of symptoms and signs were of value for predicting the results of surgery.

The radiological diagnosis of syringomyelia has been remarkably facilitated by new techniques which have improved both the imaging of the syrinx and tonsillar ectopia. The usefulness of water-soluble contract media and the combination of myelography and CT for the diagnosis of syringomyelia have been well documented,[11-14] and their value for postoperative evaluation has been stressed.[15] Magnetic resonance imaging (MRI) has proven to be as accurate as the metrizamide CT scan for the diagnosis of intramedullary cavities.[16-18] Because it is non-invasive and provides excellent visualization of the spinal cord and the cranio-vertebral junction, it is the optimal technique for the diagnosis and follow-up assessment of patients with syringomyelia (Figure 2). However, there have been false positive diagnoses of syringomyelia. For example, Pojunas et al[19] reported a biopsy proven case of viral myelomalacia where the MRI gave a signal intensity indistinguishable from that of an intramedullary cyst. We have had two patients with traumatic spinal cord injury in whom both the metrizamide CT scan and MRI showed typical signs of syringomyelia. However, the intraoperative spinal ultrasonography showed no evidence of a large syringomyelic cavity, and the myelotomy revealed a combination of myelomalacia and multiple microcystic degeneration. These examples of false positive radiographic studies are the

Table 10: Relationship Between the Radiographic Size of the Cord and Clinical Results in 40 Operative Patients with Adequate Follow-up

Cord size	Excellent	Good	Poor	Total
widened	16	2	8	26
normal	2	6	1	9
atrophic	0	2	2	4
not visualized	1	0	0	1
Total	19	10	11	40

subject of a separate publication.[20] Lee et al[17] have reported that MRI was less sensitive than metrizamide CT in detecting cavities within normal-sized or atrophic spinal cords. In the present series all patients studied with MRI showed a low intensity intramedullary signal, typical of a syrinx.

Our experience with "endomyelography" is minimal. This procedure can be performed percutaneously, and gives an accurate delineation of the morphology and the rostral and caudal extent of the syrinx, provides cyst fluid for biochemical and cytological analysis and, through decompression, plays a therapeutic role.[21-23] More recently it has been suggested that endomyelography with a small dose of metrizamide (1 ml, 170 mg of iodine/100 ml) in combination with CT scan increases the sensitivity and specificity of the diagnosis of syringomyelia, especially when other radiographic studies have been inconclusive.[24]

Parallel to these dramatic improvements in the neuroradiological diagnosis of syringomyelia, there has been an evolution in the understanding of its pathophysiology and surgical treatment. The multiple reviews of the pathophysiology of syringomyelia[1,5,8,11,25-39] indicate that the etiology of spinal cord cavitation is multifactorial. Although there are multiple "initiating causes" of intramedullary cavitation, there may be only one mechanism for the extension of the cavity within the spinal cord parenchyma. The "slosh" mechanism of Williams'[37] may be responsible for "the hydrodynamic dissection of the spinal cord" by causing rapid movement of fluid in the syrinx in response to compression of the cerebrospinal fluid (CSF) within the subarachnoid space. This compression of CSF may be initiated by engorgement of the epidural venous plexus due to changes in thoraco-abdominal pressure associated with coughing, sneezing, straining and the Valsalva manoeuver. This concept is supported by Martins,[30,31] who showed in an ovoid cavity, such as a spinal syrinx, that the sites of maximal tension are located where the curvature is the greatest. In a syrinx these sites are at the ends, and may explain the extension of the syrinxes at the poles.

Several factors may govern the maintenance of fluid inside a syrinx, including the transport of CSF along perivascular spaces, cord ischemia and venous congestion. For example, in patients with idiopathic syringomyelia and tonsillar ectopia who continue to deteriorate after decompression of the hindbrain, the "suck" phenomenon, which results from a "craniospinal pressure dissociation"[36,37] may be responsible for the continuing entry of fluid into the syrinx. Transudation of proteinaceous fluid probably accounts for the maintenance of fluid in the syrinx in cases associated with spinal cord tumours, although the finding that these cavities opacify in delayed metrizamide CT scans[15] suggests direct passage of the fluid through the cord.

In our series of 40 operative cases with adequate follow-up, we achieved neurological improvement or cessation of deterioration in 29 (72.5%). The surgical treatment of syringomyelia

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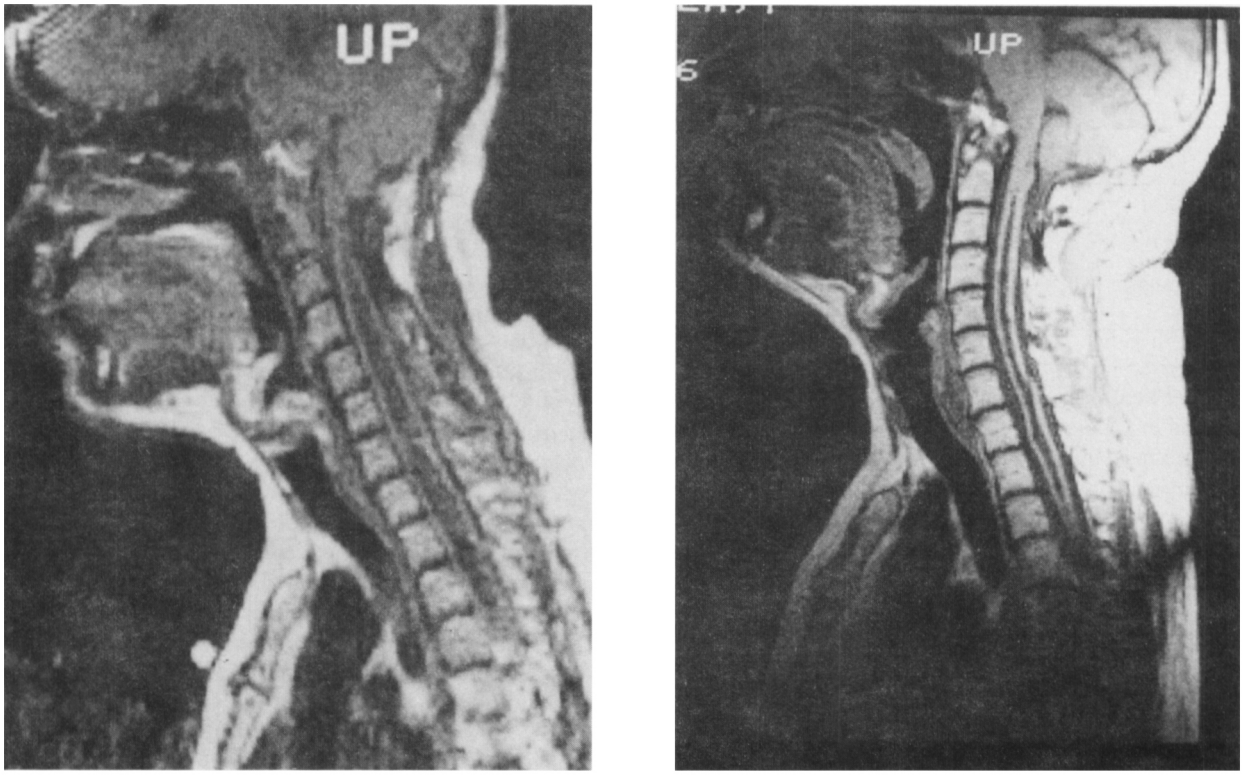

Figure 2 — Left. Pre-operative MR showing tonsillar ectopia and a large syrinx extending from C2 to the thoracic cord. Right. Seven months after an SS shunt inserted at C7 the MR shows a significant reduction in the size of the syrinx which correlated with the excellent clinical result. The defect in the signal intensity at C7 is most likely the shunt itself.

should be directed toward drainage of the syrinx and/or correction of the primary lesion allowing entrance of fluid into the cord, such as a hindbrain abnormality or a spinal cord tumour. In our opinion, surgery is indicated in all patients with syringomyelia who show neurological deterioration. In those with the Chiari malformation and significant tonsillar herniation, we have, until recently, favoured a PFD with plugging of the obex. In these cases, Williams' "craniospinal pressure dissociation" theory[36,37] may apply. Peerless and Durward[32] suggested that the additional placement of a shunt from the fourth ventricle to the subarachnoid space may improve the results as compared with PFD alone or with plugging of the obex, although this has not been confirmed in other series.[6,10,40-42] Seven of our 12 patients with syringomyelia and tonsillar ectopia underwent a PFD operation. However, six of the seven ultimately continued to deteriorate and required another surgical procedure; in the seventh case, a simultaneous SS shunt was performed, and the excellent result has persisted from 1972 to the present. Of these seven cases, only two had simultaneous plugging of the obex, and a third had simultaneous placement of a shunt between the fourth ventricle and the cervical subarachnoid space. Four of the 12 patients in this group underwent an SS shunt as the only surgical procedure and three of them had excellent or good results. A poor result was obtained in the only patient who underwent a needle aspiration alone. The failure of the PFD procedure in the long term in these cases has been documented by others,[6,10,36,37,42-45] and therefore, we suggest in patients with hindbrain abnormalities who present with symptoms related primarily to syringomyelia, that drainage of the syrinx should be the primary surgical procedure (Figure 2), with simultaneous PFD reserved for patients with brainstem compression due to a highly compromised foramen magnum, based on clinical and/or radiographical evidence. Thus, accurate preoperative assessment of the degree of brainstem compression and tonsillar herniation is crucial for the selection of the optimum surgical procedure. Patients with Chiari malformations with or without syringomyelia whose primary symptoms are due to brainstem compression will still require PFD.

The PFD procedure is not free of serious morbidity, and its mortality rate ranges from none[41,45,46] to as high as 15%.[1,36,42,47,48] In contrast, the SS shunt as performed at our institution, has been associated with no operative mortality and low morbidity.

To reduce operative neurological damage during the placement of an SS shunt, we have recently been using intraoperative monitoring of somatosensory and spinal evoked potentials. The monitoring attempts to identify any decrease in the amplitude or increase in the latency of the waveforms during the entire procedure, but especially during the myelotomy and insertion of the shunt into the syrinx. Our data are still preliminary, and no definite conclusions can be drawn about the usefulness of this technique.

Early reports of the SS shunt showed neurological improvement in only 20% to 29%.[10,44,49] However, Pitts and Groff[50] achieved a 60% rate of improvement or stabilization of neurological deficits using a gutta percha drain. In the present series of 35 patients treated with an SS shunt an excellent or good result was achieved in 26 (74.3%), although three patients required a second SS shunt (Table 7). In the 30 cases in whom the SS shunt was the primary surgical treatment of syringomyelia, 22 (73.3%) achieved an excellent or good result. Indeed, in patients with idiopathic syringomyelia but without tonsillar ectopia, shunting of the syrinx is the treatment of choice since neurological improvement or stabilization was achieved in 11 of 12 patients in this group (Tables 8 and 9). The high incidence of favourable results in our series is at least partly due to the use of

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microsurgical techniques and silastic shunt materials, as discussed previously.[3] The optimal site for the insertion of the proximal end of the shunt is the area of maximal dilatation of the syrinx as shown radiologically, or the region of the cord accounting for the maximum neurological deficit. Intraoperative real time ultrasonography is also helpful for locating the area of maximal dilatation of the syrinx before opening the dura. Typically, these studies have shown an anechoic, smooth and regular, intramedullary cavity with thinning of the surrounding cord tissue (Figure 3). Conversely, a complex collection of echoes suggests the possibility of an intramedullary mass.[51] Furthermore, in the two patients with the clinical diagnosis of PTS who had false positive metrizamide CT scans and MRI studies, the sonography accurately excluded the presence of a syrinx and correctly predicted that myelomalacia would be found at the exploratory myelotomy.[20] Placement of a shunt was not pursued in these cases, thus avoiding the possibility of inducing further neurological deficit.

Williams et al[38] suggested inserting the distal end of the shunt tube into the cephalad subarachnoid space to give flow preference to the fluid draining from the syrinx, and to ensure that the distal end is above the site of possible postoperative adhesions. We have not found any difference in the clinical results between patients in whom the distal end of the shunt was placed in a cephalad or caudad direction.

It has been suggested that establishing a communication between the syrinx and the subarachnoid space might allow fluid to flow into the syrinx,[36,37] and accordingly, several investigators have recommended shunting the fluid from the syrinx to a low pressure extraspinal site to create a favourable pressure-differential.[7,39,52,53] We believe that this reversal of flow would only occur in patients with tight impaction of the tonsils at the foramen magnum where the "cranio-spinal pressure dissociation" caused by the Valsalva manoeuvre might produce a "sucking" mechanism. In support of our belief, Ellertson and Greitz[54] reported that the intramedullary spinal pressure exceeded the subarachnoid pressure when measured by percutaneous puncture of the syrinx, which would favour continued drainage of the syrinx by the SS shunt, provided that the shunt remained patent.

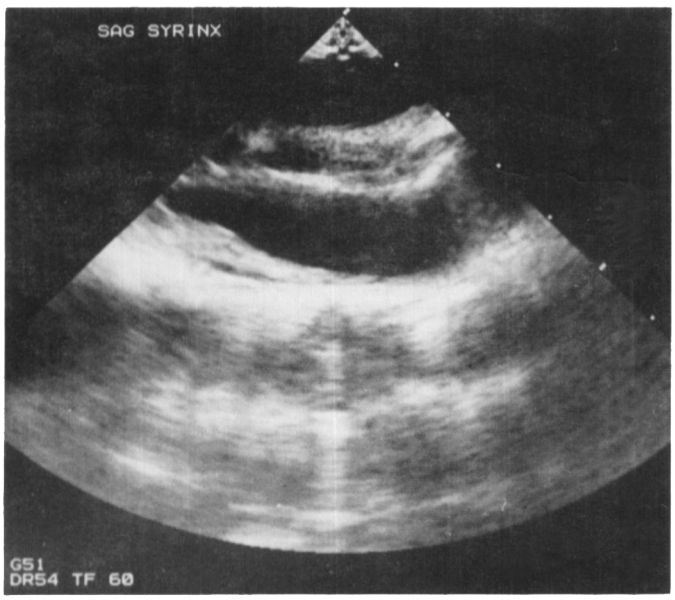

Figure 3 — Sigittal view of intraoperative sonography of the spinal cord at C7 and T1, demonstrating an anechoic cavity with smooth walls. The thin residual cord is seen around the widest part of the cystic cavity which tapers superiorly to C7 on the left of the photograph.

The peritoneal cavity has been the most frequently used extraspinal site for shunting in syringomyelia, and the reported clinical results with the SP shunt have been encouraging.[9,52,53] Furthermore, Barbaro et al[7] found that a higher percentage of patients experienced neurological improvement with the SP shunt than with the SS shunt or any other method, but that the former procedure had a higher complication rate. However, the reported experiences with the SP shunt include only short-term follow-up, and more time is needed before final conclusions can be drawn. Peerless and Durward[32] compared the two methods and found the SP shunt was only marginally better than the SS shunt. Finally, the results in the present series compare favourably with those reported for the SP procedure.

A terminal ventriculostomy was performed in three cases in the present series, and the result in all was poor. Although, theoretically, it might relieve the intrasyringeal pressure when the syrinx extends into the conus medullaris, we agree with Williams and Fahy[39] who concluded that this procedure should not be the operation of first choice for the treatment of syringomyelia. In two of our cases it was performed after an initial procedure had failed, and even then it was unsuccessful. In our opinion, the indications for this procedure are uncertain.

The incidence of post-traumatic cystic degeneration of the spinal cord has been reported to vary from 1.48%[55,56] to 3.2%[57] in patients with spinal cord injury. Favourable results from shunting have almost invariably been reported in symptomatic patients.[32,38,55,57-59] After complete spinal cord injury it has been shown that cordectomy or cord transection are acceptable surgical treatments.[8,55,60] In our single case of cordectomy or transection, a poor result was achieved, perhaps because of worsening of the adhesive arachnoiditis at the site of the previous injury. This patient improved neurologically after an SS shunt was done at a higher level. Our results show that the SS shunt is an effective treatment for symptomatic PTS: of the 10 patients treated with the SS shunt, five improved, and two stabilized (Tables 8 and 9). One patient improved after a subsequent SP shunt, and the remaining two patients had a poor result. An eleventh patient in this group did not undergo the shunting procedure when myelotomy and exploration showed microcystic degeneration (one of the false positive cases, to be reported separately).

Syringomyelia secondary to arachnoiditis continues to be a difficult therapeutic problem, perhaps because progression of the arachnoiditis overwhelms any benefits from treatment of the syringomyelia. The free flow of CSF in the subarachnoid space is severely impaired by the arachnoidal adhesions. The use of SP shunts has been reported to be favorable,[32] although our two patients treated with this modality had a poor result.

In our opinion, failure of the SS shunt is often due to technical problems, especially incorrect placement of the distal end of the catheter into the subdural space instead of the subarachnoid space, or occlusion of the catheter by postoperative arachnoidal adhesions. It is therefore essential to identify and preserve the arachnoid membrane to ensure correct insertion of the distal end of the catheter into the subarachnoid space, and to allow closure of the arachnoid membrane over the myelotomy and the shunt. These manoeuvres maintain the integrity of the

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subarachnoid space, and may help to prevent postoperative arachnoiditis. Similarly, hemostasis is crucial to prevent blood from reaching the subarachnoid space. Dislodgement of the catheter from the syringeal cavity is prevented by suturing the catheter to either the pia or the arachnoid at the site of the myelotomy. Unfortunately, septation and loculation of the syrinx may prevent adequate drainage and account for further clinical deterioration. Postoperative radiological investigations should be helpful for assessing the morphologic changes of the syrinx. Indeed, useful information about the morphology and continuity of the cavity has been obtained with delayed metrizamide CT scanning.[15] Syringography by percutaneous puncture might also help, although in our opinion, MR is the procedure of choice for postoperative studies.

Finally, the present series suggests that a short duration of preoperative symptoms favours a better outcome. Barbaro et al,[7] found that the severity of the preoperative neurological deficit was inversely related to the outcome, and we agree that early surgical treatment is warranted in patients with progressive, symptomatic syringomyelia.

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