Syringoperitoneal shunt for treatment of cord cavitation

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SUMMARY A series of twenty-nine patients with cord cavitation were treated by syringoperitoneal shunt. Twenty-two of them showed improvement after operation, five were unchanged and two worsened. Operation was performed in patients with post-traumatic cystic myelopathy, idiopathic syringomyelia, intramedullary tumours associated with cysts, and patients who had developed cystic myelopathy in association with spinal arachnoiditis. Pain improved in twenty patients, sensory symptoms and signs in eleven and weakness in ten.

Cord cavitation was first described in 1564¹ although the aetiology and natural history of the disease is variable. The surgical treatments that have been proposed are also widely varied and include direct syrinx decompression, aspiration of the fluid from the syrinx, and syringostomy. Gardner² performed decompression of the hindbrain with closure of the obex and this procedure has been the surgical treatment of choice in many centres for the foramen magnum dysplasia associated with idiopathic syringomyelia. All of these procedures carry a recognisable morbidity and mortality.³ There have been several reports of shunt procedures of various types,^{4,5} but these have frequently been unsuccessful because of the development of arachnoiditis and shunt malfunction.⁶ The development of microsurgical techniques together with new and improved shunt devices now allows direct drainage to be effectively performed.^{6,7} One of us (LS) started performing syringoperitoneal shunts in 1980, prior to Phillip and Kindt's description of this procedure.⁸

Patients and methods

From 1980 to 1983 twenty-nine patients with cord cavitation were treated with a syringoperitoneal shunt at the National Hospital, Queen Square. There were twenty-four male and five female patients ranging from 18 to 63 years with a mean age of 39 years. As shown in table 1, 17

patients had post-traumatic cord cavitation, seven had idiopathic syringomyelia, three were secondary to meningitis and two had had spinal tumours removed with subsequent irradiation before admission to this hospital. There is a close link between this Neurosurgical Unit and the National Spinal Injuries Unit at Stoke Mandeville Hospital, England, thus reflecting the higher proportion of post-traumatic cases. Table 5 shows the various surgical procedures performed on nine patients prior to our shunt surgery for cord cavitation.

Symptoms and signs

Table 2 summarises the main symptoms and signs of cord cavitation in the twenty-nine patients. The chief complaints were pain, in the face in two patients, in the neck and the upper limb in 19 and in the trunk in four patients. On examination advancing ascending or descending sensory loss was detected in all but one patient. Spinothalamic sensory disturbance was present in 28 patients and dissociated sensory loss was demonstrable in 19 patients. Disturbance of the posterior columns was noted in 11 patients. New areflexia was observed in 17 patients unilaterally or bilaterally. Motor weakness, most frequently in the upper and/or lower limbs, was present in 79% of cases although

Table 1 *Aetiology of cord cavitation in 29 patients*

Aetiology	No of cases
Post-traumatic cord cavitation	17
Idiopathic cord cavitation	7
with cerebellar ectopia	5
without cerebellar ectopia	2
Post meningitic cord cavitation	3
tuberculous meningitis	2
pyogenic meningitis	1
Spinal tumour	2
low grade astrocytoma	1
haemangioblastoma	1

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Table 2 Preoperative symptoms and signs attributed to cord cavitation in 29 patients

Symptoms and signs	Cases	
	No of cases	Per cent
Pain	23	79
Spinothalamic disturbance	28	97
Dissociated sensory disturbance	19	66
Posterior column	15	52
Motor weakness	23	79
Areflexia	19	59

in the majority this had been associated with the original trauma. Three patients were admitted complaining of increasing bilateral leg weakness, two of them had been operated on for spinal tumours elsewhere and another was a post-traumatic case. Duration of symptoms varied, being less than one year in seven cases while the remaining 22 had symptoms for over a year, the average being six years. There were five patients in the series with cerebellar ectopia, four of whom had previously undergone decompression of the foramen magnum at the National Hospital. The fifth case had not had a foramen magnum decompression (although minimal ectopia had been demonstrated) because there was no evidence of significant foramen magnum block to contrast medium.

Radiological findings

Myelography was performed in all twenty-nine cases (table 3) using metrizamide or iohexol via cervical or lumbar puncture and showed a widening of the spinal cord in 23, a normal cord in one case, an apparently collapsed cystic cord in three and atrophy of the cord in two cases. Diagnosis was made in the latter cases by CT scan with contrast medium. In two cases after tumour resection, myelography failed to show the shape of the spinal cord.

Endomyelography was performed on six patients. Confirmation of the collapse of the cord cavity after surgery was obtained in two cases, by contrast medium injection into the shunt tube.

CT scanning was performed in twenty-six patients 6 and 24 hours after myelography using metrizamide or iohexol.^{9,10} (table 4) Twenty-five cases showed expansion of the cord and in two patients contrast medium was seen in the syrinx 6 hours post injection. In 23 cases, delayed

Table 3 Myelographic findings

Findings	Cases	
	No of cases	Per cent
Myelography:	29	
Size of spinal cord		
widening	23	79
atrophy	2	7
normal	1	3
not visualized	3	10
Block	8	28
Sign of collapse	2	7
Cerebellar ectopia	5	17
Endomyelography:	6	
Cavitation	6	100

Table 4 Findings of computed tomography

Findings	No of cases
CT: with contrast medium	26
expanded cord	25
contrast within cord	24
immediately after injection	1
24 hours after injection	23

scanning performed 24 hours later showed density within the cord. In one patient dense cavity contrast was visible 6 hours after injection of subarachnoid contrast medium but was not demonstrable 24 hours later: there was no change in this patient's symptoms after operation. In two cases density within the cavity could not be seen at six or 24 hours after injection. One patient showed subsequent symptomatic improvement, the other remained unchanged.

Cerebellar ectopia was identified in all the "idiopathic" cases by supine myelography, and, though sought, in none of the post-traumatic cases.

Surgical procedure and findings

In most cases a two or three level laminectomy was performed, the patient lying in the prone position under general anaesthesia with the right upper limb abducted, allowing sufficient exposure for insertion of the syringoperitoneal shunt. After exposure of the dura in the mid-line, the operating microscope was used to facilitate dissection. Attention was directed to preserving the arachnoid membrane which was opened and both edges clipped to the sides of the opened dura. A small, longitudinal mid-line incision in an avascular area of the spinal cord was made without the use of electrical coagulation between the posterior columns and a Pudenz ventricular catheter was inserted several centimeters into the cavity rostrally. Microsurgical dissection is useful to minimise the damage to the spinal cord and to minimise bleeding into the subarachnoid space which could lead to meningeal irritation and arachnoiditis. The dura was closed, fixing the Pudenz catheter *in situ* using an encircling "Tiller" stitch. Fluid coming from the cavity could be observed within the catheter, the level changing with respiration. An appropriate length of connecting catheter was passed subcutaneously from a right flank incision to the laminectomy site. This was attached to the effluent catheter from the syrinx by a simple metal connector which was then fixed to the side of an intact spinous process at the lower end of the laminectomy incision. After closure of the laminectomy incision, the patient was turned to a supine position with the right

Table 5 Previous surgery for syrinx

Operative procedure	No of cases
Laminectomy	3
Syringostomy	3
Foramen magnum decompression and myelotomy	1
Foramen magnum decompression, myelotomy and terminal ventriculostomy	1
Gardner's operation	1
Total cases	9

Fig 1 (a, b) Myelography with CT scanning showing large post-traumatic ascending syrinx from the C6 level.

upper limb again abducted, and a low pressure Raimondi peritoneal catheter placed in the peritoneal cavity through a right paramedian incision or subcostal incision. A track was then burrowed from the abdomen to the flank incision and the two catheters connected with an appropriate connecting device. In two instances a double flushing reservoir was connected in series to allow flushing rostrally and caudally and for dye investigation. We did not find this helpful. We have not used the posterior approach to the peritoneum as this may require rib resection, an unnecessary complication particularly in the presence of a kyphosis. Neither do we advocate the sitting position as used by Barbaro *et al*,¹¹ as many of these patients have very poor spinal cord "reserve" and may experience hypotension during anaesthesia.

Details of previous surgery are shown in table 5.

Results

The average postoperative follow-up period was one

year. There was no surgical mortality in this series. Table 6 shows the overall clinical results.

For the purposes of this study patients were categorized in three main groups: *Improved*: Cases in which there was full or partial improvement of symptoms or at least one of the main symptoms that is pain, sensory disturbance or motor weakness caused by cord cavitation. *Unchanged*: Cases still manifesting the main symptoms without significant alteration. *Worse*: Cases where there was deterioration of at least one of the above main symptoms.

Twenty-two of the 29 patients showed improvement of some symptoms. Five patients remained "unchanged" and two experienced a deterioration of at least one symptom. As shown in table 6 14 of 17 post-traumatic patients improved and none deteriorated. Four of seven patients with idiopathic

Fig 2 (a, b) Post-operative appearance in the same patient shows cord cavity to be collapsed and the shunt tubing is visible in the CT scan image.

cavitation improved, two were unchanged and one worsened. The latter case had a secondary meningo-myelocoele on the site of previous laminectomy and following syringoperitoneal shunt leakage of cerebrospinal fluid occurred necessitating repositioning of the shunt, at a lower level of the cavity. Four months after operation this patient showed some relief of pain, but deterioration of lower limb function and worsened sensory loss.

In the patients with postmeningitic cavity formation, two of three showed improvement and one worsened. Surgery was successful in both spinal tumour patients who had initially complained of deterioration in the power of both legs.

Postoperative changes are summarised in table 7. Pain had a better prognosis than the other two modalities showing improvement in 20 of 23 patients. Four of 20 patients had complete remission of their

Table 6 Summary of results in 29 cases

Result	Types of syringomyelia									
	Traumatic		Idiopathic		Meningitic		Spinal tumour		Total	
	No	%	No	%	No	%	No	%	No	%
Improved	14	82	4	57	2	67	2	100	22	76
Unchanged	3	18	2	29	0	0	0	0	5	17
Worse	0	0	1	14	1	33	0	0	2	7
Total	17		7		3		2		29	

pain, three of them had pain in the upper extremities and the other had pain in the thorax. All four were patients with post-traumatic cord cavitation. Symptoms of pain in the neck and/or upper limbs were most frequently improved followed by those in the face and trunk as shown in table 8. Sensory disturbance and motor weakness were the least likely deficits to improve after operation. Improvement of motor weakness was seen in three patients, two of whom had cord cavitation extending both rostrally and caudally from the level of the tumour.

Of 22 improved patients six had undergone previous operations for their symptoms prior to admission to the National Hospital and the remainder underwent a syringoperitoneal shunt procedure as the first manoeuvre. All seven patients with a one year history or less showed improvement after operation. Three patients have had a shunt revision, having experienced improvement in symptomatology followed by relapse. In these cases confirmation of an expanded cyst was made with CT contrast myelography. In one case the upper end was found to be blocked and in two the lower catheter. Both were cleared without difficulty.

Discussion

The term syringomyelia has been used to refer to a chronic, progressive syndrome caused by cavitation of the spinal cord adjacent to the central canal or to any cyst of the spinal cord or dilatation of the central canal and/or its extension into the cord substance selectively.¹² Such dilatation was thought to be primarily developmental in origin but some post-natal conditions were found to induce the same condition.¹² It would be preferable to use the term "cord cavitation" to define pathological cord cavitation without reference to specific factors.

Gardener² believed that a disorder in the outflow tract of the fourth ventricle is central to the development of syringomyelia and has advanced his hydrodynamic theory to explain the progressive nature of syringomyelia. He postulated that an arterial pulsatile force from the choroid plexus was transmitted via the fourth ventricle into the central canal. On the other hand, according to Williams¹³ the distending force in communicating syringomyelia is related to movement of cerebrospinal fluid and this movement is dependent on intraspinal

Table 7 Results by symptom and signs

Result	Symptoms and signs					
	Pain		Sensory		Motor	
	No of cases	%	No of cases	%	No of cases	%
Improved	20	87	11	39	10	43
completely	9	39	3	11	5	22
partially	11	48	8	28	5	22
Unchanged	3	13	16	57	11	48
Worse	0	0	1	4	2	9
Total cases	23		28		23	

Table 8 Relationship between improved symptom and sign and the site

Symptom and signs	Improved site			
	Face	Neck, shoulder, upper limb	Trunk	Lower limb
Pain	1 (2)	14 (19)	2 (4)	0 (0)
Sensory disturbance	2 (7)	8 (19)	2 (11)	0 (2)
Motor weakness	0 (0)	7 (17)	0 (1)	3 (7)

() Number of impaired patients

and intracranial venous distension which varies with constriction of the neck, stooping, muscular exertion, coughing, sneezing and vomiting. These pressure changes are responsible for displacement of cerebrospinal fluid through the foramen magnum. Taking into account these pathological factors, surgical therapy can only aim at correction of the cerebrospinal pathway.

Barnett *et al*¹² divided syringomyelia into a communicating and non-communicating type and recommended different types of treatment for each. The non-communicating type developed as a late sequela of spinal cord trauma, arachnoiditis or intramedullary tumour. However, it may be difficult to apply this classification satisfactorily since there is often radiological evidence of communication between the cavity and the subarachnoid space in the post-traumatic patient.

Ascertaining whether the cavity is communicating or non-communicating might be less important than assessing the degree of cerebellar ectopia. In our series 14 of 17 patients with post-traumatic cord cavitation improved in contrast to four of seven patients with idiopathic cord cavitation.

On the basis of his hypothesis Gardner¹⁴ proposed exploration of the posterior fossa to remove arachnoidal adhesions and decompression of structures at the foramen magnum in order to promote cerebrospinal fluid flow from the fourth ventricle; and occlusion of the central canal at its origin from the fourth ventricle in an attempt to prevent flow into the central canal and spinal cavity. This procedure has been recognised as an acceptable operation with favourable results; indeed 175 such procedures have been undertaken during the last twenty years at the National Hospitals for Nervous Diseases, but there is, as with most surgery for syringomyelia, some mortality and morbidity;^{2 15} also complications and deterioration of symptoms have been reported.

Ventriculo-atrial shunting was recommended to relieve the pressure in the cavity by Kräyenbühl¹⁶ who reported improvement in 13 of 22 patients and seven patients who further deteriorated. This treatment he felt should be recommended as a first or additional operative step when hydrocephalus is demonstrated, a view which we have not adopted.

Operative procedures for cord cavitation are designed to drain the fluid from the cavity, divert the flow and prevent pulse pressure in the cavity. Percutaneous syrinx aspiration has been advocated as a possible mode of therapy.^{17 18} However, aspiration of the fluid is followed by prompt refilling of the cavity from the ventricular system or subarachnoid space where there is communication.

Gardner¹⁵ advocated a terminal ventriculostomy, which involves opening the central canal at the tip of

the conus medullaris. This procedure will not apply in all cases, only to those in which the cavity extends into the tip of the conus medullaris.

Although our understanding of syringomyelia remains imperfect, treatment is still best directed at preventing distension of the cavity. Additionally, the pressure differential between the cavity and surrounding tissues is an important concern.¹⁷ It seems clear that distension of the cavity causes dysfunction of neural tissue. To provide continuous drainage local syringostomy has been performed by many surgeons for some time.^{6 7 20-24} Numerous materials and methods have been used including gutta percha drain, rubber drains, polythene tubing, cotton wick, silk sutures, tantalum wire sutures⁴ and silastic wick.²⁵ These materials are mainly used to maintain the patency of the outlet rather than as a conduit for drainage and some are not recommended because possible reactions to them may cause arachnoiditis and shunt malfunction. Faulhauer²⁶ introduced a ventricular catheter into the cavity as a conduit for drainage. As reported by Pitts⁵ shunt operations showed favourable results, the rate of post operative improvement varying from 20% to 60%.

Simple comparison of the results is difficult because of the differences in surgical indication, surgical technique and the criteria for the evaluation of results. In addition there is the variable natural history of the diseases which often shows long periods of apparent non-progression. In our series 22 of 29 patients (76%) showed neurological improvement after an average one year follow-up period. Subsequent deterioration has been evident in four patients who have shown a later return of symptoms following improvement after shunt, despite radiological demonstration of a collapsed spinal cord and verified functioning shunts. One must, therefore, assume that the process of cystic myelopathy has progressed despite relief of intramedullary pressure. It has, nevertheless, continued to be our policy to confirm shunt function both radiologically with myelography and CT scan and by simple disconnection of the tube at the level of the flank incision or at the lower end of the laminectomy to check the patency of both ends.

Drainage into the subarachnoid space following syringo-subarachnoid shunt is not always free, absorption of fluid from the cavity may be imperfect, and reflux from the subarachnoid space to the syrinx can occur. Eight cases in our series showed subarachnoid block demonstrated by myelography and adhesions between the arachnoid and pia mater which were subsequently confirmed at surgery. In patients with high protein content in the fluid, absorption of cyst fluid is diminished and may result in shunt malfunctions. One of the four patients who

had undergone surgery for syngo-subarachnoid shunt previously showed a blocking of the shunt tube by brown inspissated material. The peritoneal cavity on the other hand is capacious and can readily absorb fluid from the syrinx. A low pressure Raimondi catheter prevents reflux and overdrainage which may cause complete collapse of the cord cavity, tending to encourage obstruction of the tube. However, the system is essentially low flow and one must expect to have to revise these shunts periodically, particularly if the cyst fluid protein content is raised.

In those cases with idiopathic cavitation, the results were worse than those of the traumatic cases. All three patients with idiopathic cavitation who remained unchanged or worsened had undergone previous surgery and the duration of symptoms before admission was 6, 16 and 21 years. Prolonged duration of symptoms may be a factor in the subsequent deterioration of these patients.

It is difficult to determine the cause of pain in patients with cord cavitation, signs ranging from pain from cerebellar ectopia, distortion of nerve roots to pain originating from within the cord itself. In our series 20 of the 29 patients showed improvement of pain and in particular, pain of the neck, shoulder and upper limbs in fourteen patients (74%). In these 14 patients cavitation was demonstrated by radiography from the cervical to the thoracic spine and all underwent syngo-peritoneal surgery. Six patients were shunted from the cervical cavity and the remainder from the thoracic cavity. Shunting from the thoracic cavity also relieved the pain in the neck, shoulder and upper limbs.²⁷

In conclusion, it would appear that syngo-peritoneal shunting is a logical and reasonably effective treatment for cord cavitation of whatever cause. Our results are not dissimilar to those reported by Barbaro *et al.*,¹¹ although the selection of patients is not absolutely comparable. The procedure is technically simple and as is general in neurological disease, more effective in patients where the disease is not too advanced. The preferred site of cyst drainage has been, in our hands, the upper dorsal spine. Syngo-peritoneal shunting may become the initial treatment of choice in idiopathic syngomyelia with ectopia cerebelli in which there is no radiological evidence of foramen magnum compression, although it is too early as yet to make this claim with confidence.

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