Comparison of syringopleural and syringosubarachnoid shunting in the treatment of syringomyelia in children

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Cystic dilation of the spinal cord was described as early as the sixteenth century; the term syringomyelia was coined in 1827 by Ollivier and the condition was first surgically managed in 1892. Nevertheless, today, there is still considerable controversy regarding the pathogenesis of syringomyelia. Each theory that has been proposed is subject to criticism regarding clinical observations that it cannot by itself explain. One wonders if these different pathogenic hypotheses do not simply reflect the heterogeneous means by which syringomyelia develops.

The prevailing concepts of the pathogenesis of syringomyelia have given rise to variety of operations and their heterogeneity may explain why the surgical treatment of syringomyelia is still not standardized. Indeed, different surgical approaches to syringomyelia have been advocated, including: ventriculoatrial or ventriculoperitoneal shunting; posterior fossa decompression with or without plugging of the obex; terminal ventriculostomy; percutaneous drainage; lumboperitoneal shunting associated with myelotomy; and syringosubarachnoid (SSA), syringoperitoneal, syringocisternal, or syringopleural (SP) shunting. In the past, the efficacy of these different techniques was difficult to evaluate, given the poor reliability and the invasiveness of diagnostic procedures. The advent of magnetic resonance (MR) imaging has had a major impact on the early diagnosis and follow-up study of syringomyelia because this technique allows direct noninvasive visualization of the syrinx and its relation to other structures such as the cisterna magna, the cerebellar tonsils, and the fourth ventricle.

The purposes of this paper are to summarize our experience in treating childhood syringomyelia with spinal shunting, as evaluated with MR imaging, and to compare two treatment modalities: SSA shunting and SP shunting. We will also define circumstances in which shunting procedures can be valuable in the treatment of this condition.

KEY WORDS • syringomyelia • syrinx • hydromyelia • Chiari malformation • syringopleural shunt • syringosubarachnoid shunt
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Clinical Material and Methods

Patient Population

Case records from the Montreal Children’s Hospital containing the diagnosis of “shunted syringomyelia” were retrospectively reviewed. Thirty-one patients were identified. The patients were divided into two groups according to the type of surgical procedure performed to treat the syringomyelia. Group A included 19 patients who underwent SP shunting; Group B, 13 patients treated with an SSA shunt. One patient was included in both groups because her syrinx was treated initially with an SSA shunt, which had to be removed and replaced by an SP shunt 5 months later due to the patient’s clinical and radiological deterioration.

There were 13 female and six male patients in Group A. Their ages at operation ranged from 1 to 18 years (10 ± 5 years, mean ± standard deviation (SD)). There were nine females and four males in Group B with ages ranging from 2 to 16 years (9 ± 5 years, mean ± SD).

Associated Pathological Conditions

Associated diagnoses in both groups included Chiari I and Chiari II malformations, spina bifida aperta, spina bifida occulta, and hydrocephalus (Table 1).

Table 1

<table>
<thead>
<tr>
<th>Associated Diagnosis</th>
<th>No. of Patients†</th>
</tr>
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<tbody>
<tr>
<td>Chiari I malformation</td>
<td>Group A</td>
</tr>
<tr>
<td>Spina bifida aperta</td>
<td></td>
</tr>
<tr>
<td>Spina bifida occulta</td>
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<tr>
<td>Hydrocephalus</td>
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</table>

<table>
<thead>
<tr>
<th>Previous Surgery</th>
<th>No. of Patients†</th>
</tr>
</thead>
<tbody>
<tr>
<td>Chiari decompression</td>
<td>Group A</td>
</tr>
<tr>
<td>with obex plugging</td>
<td></td>
</tr>
<tr>
<td>without obex plugging</td>
<td></td>
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<tr>
<td>total</td>
<td></td>
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<tr>
<td>VP shunting</td>
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<td>VA shunting</td>
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<tr>
<td>LP shunting</td>
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</tbody>
</table>

TABLE 1

Associated diagnoses in 31 patients with syringomyelia*

* One patient underwent both syringopleural and syringosubarachnoid shunting.
† Group A = 19 patients undergoing syringopleural shunting; Group B = 13 patients undergoing syringosubarachnoid shunting.

Preoperative Studies

All patients were investigated preoperatively with MR imaging (Signa 1.5-tesla; General Electric, Milwaukee, WI) or myelography/spinal metrizamide–computerized tomography (CT) (model CT 9800; quick HiLight detectors, General Electric). They also underwent CT scanning of the head. All cases except one were followed postoperatively with serial MR imaging studies. Patients in whom a Chiari malformation was diagnosed on MR imaging had an ear, nose, and throat examination to assess vocal cord movement and a sleep study aimed at detecting apnea episodes. Urological examinations (with urodynamic investigations when necessary) were performed on a regular basis both pre- and postoperatively.

Operative Techniques

The syrinx was approached via a three-level laminotomy and was entered using a cerebrospinal fluid (CSF) cardiac catheter with a reduced diameter tip (model 125-B; Pudenz-Schulte Medical, Goleta, CA). Using ultrasound guidance (echocamera SSD-500; Aloka, Tokyo, Japan) the catheter was advanced through the dorsal root entry zone or the median sulcus and tunneled either to the pleural or subarachnoid space. The valveless tube was anchored to the pia mater for both shunt types and to the dura mater as well as the fascia for SP shunts.

Results

Only four patients in each group exhibited the classic dissociated sensory deficit characterized by thermalgesic loss with preservation of the postural sense. A new onset weakness or a progressive deterioration of a preexisting motor handicap was the most relevant clinical feature in this series. It was encountered in 13 Group A cases and in eight Group B cases. Urological difficulties were the presenting clinical manifestation in one Group A and in five months, mean ± SD) prior to receiving an SSA shunt. The obex was plugged in four cases (Table 2).
The syringomyelia did not recur afterward despite no reinter-
tervention at the spinal cord level. After the occurrence of
this last complication, a right-angle anchoring piece was
systematically intercalated in subsequent cases and no
such problem was encountered thereafter.

In Group A 11 patients improved neurologically and
eight stabilized. No patients, including the two patients
who were reoperated on for pleural effusion and shunt
migration, deteriorated. In Group B, one patient improved
clinically, eight stabilized, three worsened, and one was
lost to follow-up review. The three Group B patients with
poor postoperative outcome exhibited a rapid clinical and
radiological deterioration characterized by increased hand
weakness, progressive sensory deficit with ataxia, and an
increase in the size of their syringomyelic cavity on MR
imaging. In these three cases, deterioration occurred a few
weeks after SSA shunting. One of these patients required
reoperation and underwent an SP shunt placement. She
remained clinically stable for the next 4 years. A second
patient underwent repeat SSA shunting 4 months later. He
remained clinically and radiologically stable for the next 3
years. The last patient who exhibited deterioration refused
reoperation.

Postoperative Radiological Findings

With two exceptions, all patients underwent early post-
operative MR imaging studies 1 to 6 months after surgery
and most of them had late postoperative MR examinations
1 to 4 years after surgery. Two Group B patients under-
went early postoperative metrizamide–CT evaluations. In
one of these cases, the spinal cord appeared to be less
dilated; however, this patient was subsequently lost to fol-
low-up review. In the other case, two spinal metrizamide–
CT scans, performed 1 and 2 years after surgery, revealed
expansion of the spinal cord. This finding correlated well
with the clinical deterioration exhibited in that case. The
patient was offered reoperation but refused. In 12 Group
A patients, disappearance of the syringomyelic cavity
was revealed on MR imaging; the only signal remain-
ing was produced by the catheter itself. In addition, the
syringomyelic cavity was reduced by more than 50% in
five cases (Fig. 1). One patient exhibited an asymptomat-
ic recurrent thoracic syrinx 2 years after receiving an
SP shunt; her early postoperative MR image had shown
a reduced syringomyelic cavity. In the patient with the
pleural effusion imaging was obtained only after her SP
shunt had been replaced by a syringoperitoneal shunt.
Nevertheless, her cavity was revealed to have collapsed.
In Group B, the syringomyelic cavity was collapsed in one
case, reduced in seven, unchanged in two, and enlarged in
three cases.

Discussion

Syringomyelia is a disease traditionally believed to af-
fect principally young adults and this was abundantly
reported in the pre–MR imaging era.3,5,7,9,31–33,45 The advent
of MR imaging has not only allowed more accurate24,26,34
but also earlier diagnosis of this disorder. Consequently
syringomyelia is now increasingly recognized throughout
the pediatric population at earlier ages when the neuro-
logical deficits caused by this condition may still be
minor57,58 and potentially reversible.3,7,23 Magnetic reso-

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nance imaging, by also providing a method of reliable noninvasive follow-up study, is likely to have a favorable impact on the management of syringomyelia. 3,4,6

The literature would lead one to suggest that the clinical presentation of syringomyelia may be quite heterogeneous, depending on the extent, shape, and location of the syrinx and on the duration of the disease. 2,3,5,7,8,10,13–16,20–22, 30,32–34,45–47,50,58

In this series, the complete syndrome of syringomyelia, with muscular atrophy of the upper extremities accompanied by dissociated sensory loss and long-tract signs in the lower extremities, was virtually never encountered. This may be related to the young age of the population studied; the frequent association of other conditions, such as spina bifida, 40 which because of their accompanying neurologic deficits may mask the signs of syringomyelia; and, finally, the relatively precocious investigation with MR imaging. Indeed, in pre-MR imaging studies, an estimated one-fourth of syringomyelic cavities were missed. 29

In total, at an average follow-up period of 2 years, clinical and radiological improvement or stabilization was observed in 18 of 19 Group A cases, the exception showing clinical improvement not correlated radiologically. Even if the disparity between our two treatment groups and the retrospective nature of our study preclude any valid comparison, results obtained with SSA shunting appear less impressive. On the whole, the SSA group of patients was afflicted with less severe associated conditions (Table 1) and yet, at a mean follow-up review of 4 years, only one patient had improved clinically whereas three patients had deteriorated both clinically and radiologically. Of note, all treatment failures in this group occurred within weeks of the operation. The reasons for the higher failure rate with SSA shunting are numerous and may include: the absence of a significant pressure gradient between the syringomyelic cavity and the subarachnoid space; the fact that the frequently encountered arachnoiditis precludes normal perimedullary CSF circulation; and, finally, the fact that with significant arachnoiditis it may be difficult to establish with certainty whether the distal tube is, indeed, in the subarachnoid space. We, therefore, agree with others 40 that when a syringostomy is indicated, the greatest likelihood of emptying the syrinx and keeping it empty is to drain it into an area of low pressure, such as the pleural or peritoneal cavity. We prefer SP over syringoperitoneal shunting because the laminotomy and pleural entry sites are closer, the operation can be entirely conducted with the patient in the prone position, and positioning the distal catheter in the pleural space, one avoids the potential complication of distal obstruction by viscera or omentum. 50

In our series, a total of 14 patients who previously underwent posterior fossa decompression developed clinical manifestations of syringomyelia. We believe that the uniform operative finding of arachnoiditis (encountered either at the time of craniovertebral decompression or that of syringostomy) in conjunction with altered craniovertebral CSF circulation may be an important pathogenetic factor in the production of syringomyelia in these patients. The less than uniform success rate of craniovertebral decompression in the face of syringomyelia and the associated morbidity of the treatment 3,5,6,8,13,15,19–21,29,30,32,40,44–46, 52,57,58 have led to the development of alternative therapeutic modalities. These include syrinx aspiration, 2,28,45 termino-

nal ventriculostomy, 18,55 and SSA, 21,30–32,39,46,47,50 syringoperitoneal, 5,13,21,27,32,35,39,41,45 and SP shunting. 21,28,42,56–58 The failure rates of these techniques in the control of syringomyelia are said to vary from none in small series with a short follow-up period to 42% 5,27,41,46,47,58 or more when terminal ventriculostomy is included. 55 With increased experience in the use of these alternative methods, some authors 46,56 have advocated the use of shunting procedures for the primary treatment of syringomyelia not associated with posterior fossa symptomatology. Others reserve this type of surgery for cases without hindbrain anomaly 47 or for patients previously treated by craniovertebral decompression, in whom a sizeable syringomyelic cavity persists. 56

Our results indicate that SP shunting is a valuable technique in the management of syringomyelia that is refractory to craniovertebral decompression: eight patients who underwent previous Chiari decompression, yet still suffered from persistent syringomyelia, responded well to subsequent SP shunting. Furthermore, eight of the 14 Group A patients who had an associated Chiari II malformation without exhibiting hindbrain signs were first treated with an SP shunt. Their Chiari II malformation never required craniovertebral decompression because the syrinx responded well to the SP shunting. In Group B, all six patients with Chiari malformation received decompression first (Table 2).

The low morbidity and high long-term success rate of SP shunting renders it appealing as a primary treatment in either idiopathic or posttraumatic syringomyelia, as well as in syringomyelia that is refractory to Chiari decompression. It may also represent a viable option in the primary treatment of syringomyelia associated with myelodysplasia and Chiari II malformation, if the latter does not cause hindbrain signs.

References


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