Syringomyelia associated with foramen magnum arachnoiditis

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Object. Syringomyelia is often linked to pathological lesions of the foramen magnum. The most common cause is hindbrain herniation, usually referred to as Chiari I or II malformation. Foramen magnum arachnoiditis without either Chiari I or II malformation is a rare cause of syringomyelia. The authors undertook a retrospective analysis of 21 patients with foramen magnum arachnoiditis (FMA) and syringomyelia treated between 1978 and 2000 to determine clinical course and optimum management.

Methods. In the review of records, 21 patients with FMA and syringomyelia were documented. A stable clinical course was demonstrated in three patients in whom surgery was not performed, and one patient refused surgical intervention. Seventeen patients underwent 23 operations to treat progressive neurological disease. Of these 23 operations, 18 involved opening of the foramen magnum, arachnoid dissection, and placement of a large dural graft. One patient underwent insertion of a ventriculoperitoneal shunt for treatment of accompanying hydrocephalus, one patient received a cystoperitoneal shunt for an accompanying arachnoid cyst; two syringoperitoneal and one syringosubarachnoid shunts were also inserted. Hospital and outpatient files, neuroimaging studies, and intraoperative photographic and video material were analyzed. Additional follow-up information was obtained by telephone interview and questionnaires.

Standard and cardiac-gated magnetic resonance imaging studies are the diagnostic procedures of choice in these patients. Sensory disturbances, dysesthesias, and pain were the only symptoms likely to improve after foramen magnum surgery. Motor weakness and gait disturbances, which were severe in a considerable number of patients, and swallowing disorders tended to remain unchanged. As a consequence of the rather severe arachnoid lesions in most patients, clinical recurrences were observed in 57% over a 5-year period.

Conclusions. Surgery for FMA and syringomyelia has to provide clear cerebrospinal fluid pathways between the cerebellopontine cisterns, spinal canal, and fourth ventricle. If this can be achieved successfully, the syrinx decreases in size and the clinical course of the patient may even improve. In patients with severe and widespread areas of arachnoiditis, however, multiple operations may be required at least to stabilize the clinical course.

KEY WORDS • arachnoiditis • foramen magnum • syringomyelia • chiari malformation
outpatient files, and neuroimaging studies were evaluated. Additional information was obtained from questionnaires and telephone interviews. All 17 surgically treated patients presented with progressive neurological symptoms.

**Neuroimaging Studies**

Preoperative imaging consisted of plain x-ray films of the cervical spine, including lateral flexion–extension radiographs to rule out additional instabilities, which may have to be accounted for during operative positioning. Magnetic resonance imaging was performed in all patients pre- and postoperatively. The spinal cord was examined with and without administration of Gd to demonstrate the lower extent of the syrinx and to rule out an associated spinal tumor. Since 1990, we have performed cardiae-gated MR imaging in nine patients to assess CSF flow pre- and postoperatively. Postoperative MR images were obtained before discharge and after 6 or 12 months. Additional studies were performed only if the syrinx did not regress and/or neurological symptoms recurred or progressed.

**Surgical Technique**

Eighteen operations were performed to explore the foramen magnum. For the majority of patients, the prone position was used. In selected patients in whom severe forms of arachnoiditis were suspected (that is, those with a history of foramen magnum surgery or meningitis), we preferred to use the semisitting position, which allows irradiation of the surgical wound and may provide significant advantages in the operative field during microsurgical dissection. After a midline incision and detachment of the neck muscles from the occipital bone, a small occipital craniectomy including the foramen magnum is performed. In the majority of cases, a C-1 laminectomy is undertaken as well. The dura mater is incised in a Y-shaped configuration and the arachnoid is inspected to determine the extent and severity of scarring. The arachnoid is then opened sharply and resected in the midline to ensure free passage of CSF toward the spinal canal. Once this is achieved, the tonsils are spread apart in the midline by using two microdissectors. This usually requires additional arachnoid dissection in the midline toward the foramen of Magendie. Bilaterally the PICAs are identified and the foramen of Magendie is inspected. If obstructed, the latter is opened. To obtain a good overview of the foramen of Magendie and to ensure a good outflow from the fourth ventricle, the size of the tonsils is reduced using bipolar coagulation or subpial suction if medially if necessary. Concluding the intradural portion of the surgery, the pathway toward the cerebellopontine cisterns is inspected. Dense adhesions lateral to the brainstem, however, are not dissected because this would risk injury to the perforating vessels or lower cranial nerves, which may be very difficult to identify and to preserve. After ensuring that the CSF pathway is unobstructed from the fourth ventricle and the cerebellopontine cisterns toward the spinal canal, a large cisterna magna is created using a large dural graft. To minimize the risk of postoperative adhesion of the graft, we avoided autologous tissue, placing instead artificial material such as Neuropatch (Braun, Melsungen, Germany) or Gore-Tex (Gore & Associates, GmbH, Putzbrunn, Germany). Finally, great care is taken to obtain a good soft-tissue closure, which is mandatory to avoid postoperative CSF leakages. Figure 1 provides an example of pre- and postoperative imaging and operative management.

**Clinical Evaluation**

Neurological examination was performed preoperatively and before discharge. Follow-up examinations were performed after 3 months. Additional follow-up information was obtained at outpatient visits on a yearly basis and from questionnaires and telephone interviews. Neurological symptoms were analyzed according to a grading system for swallowing function, gait, motor weakness, sensory changes, pain, and bladder function (Table 1). Scores between 0 and 2 indicated disabled or incapacitated function. Additionally, the KPS was used. Success of treatment was defined as sustained improvement of preoperative symptoms or stabilization of previously progressive symptoms. Failure of treatment was defined clinically as postoperative neurological deterioration, such as clinical recurrence, independent of imaging results.

**Statistical Analysis**

For comparison of clinical data the chi-square test and Student t-test were used, provided that results of the Komolgorov–Smirnov test indicated normal data distribution. Differences were considered significant at p = 0.05. Long-term results were analyzed using the Kaplan–Meier method. Data are presented as the mean ± standard deviation unless otherwise indicated.

**Results**

**Preoperative Neurological Symptoms**

Patients presented at a mean age of 38 ± 12 years; their mean duration of symptoms was 64 ± 80 months. In 10 patients, the cause of arachnoiditis could be identified or was suspected to be related to: meningitis in five, birth-related trauma (Fig. 1) or other traumatic incidents in four, subarachnoid hemorrhage in one, and a combination of subarachnoid hemorrhage and multiple operations in one patient. The mean interval between the presumed causative event and the development of FMA and syringomyelia–related symptoms was almost 10 years (mean 115 ± 117 months, range 8 months–36 years). The most common neurological symptom, which patients with FMA noticed first, was progressive motor weakness of one upper limb in 48%. Sensory disturbances or headaches were each documented in 20% of patients as the first symptom. Problems of gait, swallowing, or hydrocephalus were noted in 4% of patients as the first clinical problem. In contrast, patients with Chiari I malformation and syringomyelia are more likely to report headaches and sensory disturbances as the first complaint, with gait or motor-power problems manifesting later in the clinical course. At presentation in our hospitals, patients with FMA complained mainly of progressive motor weakness (40%), gait problems (24%), or occipital pain (24%). Patients with Chiari I malformation on the other hand, predominately
nantly complain of occipital headaches; motor weakness is less often the predominant problem.10

Neuroimaging Studies

In five patients in this group neuroimaging demonstrated evidence of hydrocephalus. In two of these, the hydrocephalus was related to meningitis, in one each to trauma or previous surgery, and no obvious cause was detected in the remaining patient. An arachnoid cyst in the posterior fossa was demonstrated in three patients, again related to meningitis or trauma in one patient each, and with no apparent cause in the third patient. Hydrocephalus and posterior fossa arachnoid cysts associated with a syrinx indicate a rather widespread problem of CSF circulation that may go beyond the area of the foramen magnum.

In the absence of hydrocephalus or an arachnoid cyst the arachnoid lesions at the foramen magnum level may be difficult to diagnose. Standard MR imaging may demonstrate indirect signs of arachnoiditis such as bulging of the upper cervical cord (Fig. 1 upper left) and medulla or displacement of the medulla on axial sequences due to arachnoid scarring. In patients in whom the diagnosis is uncertain, cardiac-gated MR imaging may demonstrate obstruction of CSF flow at the foramen magnum as indirect evidence of arachnoid disease in the absence of a Chiari malformation. This imaging modality was diagnostic in each of the nine patients in whom it could be used.

Foramen Magnum Findings

Obstruction of CSF pathways was demonstrated in every

<table>
<thead>
<tr>
<th>Score</th>
<th>Impairment (sensory, dysphagia, pain, dysesthesia)</th>
<th>Motor Weakness</th>
<th>Gait Ataxia</th>
<th>Bladder Function</th>
<th>Bowel Function</th>
</tr>
</thead>
<tbody>
<tr>
<td>5</td>
<td>no symptom</td>
<td>full power</td>
<td>normal</td>
<td>normal</td>
<td>normal</td>
</tr>
<tr>
<td>4</td>
<td>present, not significant</td>
<td>movement against resistance</td>
<td>unsteady, no aid</td>
<td>slight, no catheter</td>
<td>slight, control</td>
</tr>
<tr>
<td>3</td>
<td>significant, function not restricted</td>
<td>movement against gravity</td>
<td>mobile w/ aid</td>
<td>residual, no catheter</td>
<td>laxatives, control</td>
</tr>
<tr>
<td>2</td>
<td>some restriction of function</td>
<td>movement w/o gravity</td>
<td>few steps w/ aid</td>
<td>rarely incontinent</td>
<td>rarely incontinent</td>
</tr>
<tr>
<td>1</td>
<td>severe restriction of function</td>
<td>contraction w/o movement</td>
<td>standing w/ aid</td>
<td>often, catheter</td>
<td>often incontinent</td>
</tr>
<tr>
<td>0</td>
<td>incapacitated function</td>
<td>plegia</td>
<td>plegia</td>
<td>permanent catheter</td>
<td>permanent incontinence</td>
</tr>
</tbody>
</table>

Foramen magnum arachnoiditis

TABLE 1

Neurological scoring system

FIG. 1. Upper Left: Sagittal T1-weighted MR image obtained in a 44-year-old woman, demonstrating a large syrinx extending from the foramen magnum to T-11. No Chiari malformation is present. There was a history of birth-related trauma, and she complained of progressive gait ataxia and motor weakness in the hand muscles for the past 2 years. There was no history of meningitis. Surgery was recommended after reaching a presumptive diagnosis of FMA. Upper Right: After dural opening, dense arachnoid adhesions, causing a complete block of CSF flow at the foramen magnum, became visible. Lower Left: Final stage of the dissection. A small layer of scar is left on the spinal cord because it did not interfere with CSF flow. The fourth ventricle is open and both PICA's are visible. Lower Center: A large artificial dural graft has been inserted. Lower Right: Sagittal MR image obtained 1 week after surgery, revealing a significant decrease of the syrinx size. Neurological symptoms remained unchanged.
patient who underwent foramen magnum surgery. The arachnoid scarring was so severe that no CSF flow was observed after dural opening in all but one patient (Fig. 1 upper right). Only after opening and sharp dissection of the arachnoid toward the foramen of Magendie, could the cerebellopontine cisterns and the spinal canal, CSF flow be observed. The foramen of Magendie was obstructed in each of these patients and opened (Fig. 1 lower left). If the PICA was embedded in thick arachnoid adhesions, no attempt was made to dissect it free. Likewise, no dissection was performed laterally toward the perforating arteries or caudal cranial nerves. The only objective of surgery was to provide free-flowing CSF pathways. To maintain this CSF passage, a large dural graft was inserted (Fig. 1 lower center).

Treatment-Related Outcome

The complication rate was 19% and CSF leakage was the predominant problem (14%). Postoperative dysphagia was observed in 5% of these patients. During the first year after opening of the foramen magnum, arachnoid dissection, and insertion of a dural graft, improvement was observed in sensory disturbances, dysesthesias, and pain, whereas motor weakness, gait, sphincter functions, and dysphagia tended to remain unaltered. Considering the severe preoperative motor deficits in these patients, this lack of postoperative improvement is reflected in the unchanged mean KPS score during the 1st postoperative year. Compared with patients with Chiari I malformations, these postoperative results are less satisfactory.10

Four patients required multiple operations: stabilization after foramen magnum surgery and placement of a SP shunt (one case); two foramen magnum procedures and placement of a SP shunt (one case); two foramen magnum procedures and placement of a VP shunt (one case); a foramen magnum procedure after placement of a syringosubarachnoid shunt. The patient in Case 14 (Table 2) underwent placement of a cystoperitoneal shunt, but he continued to suffer progressive neurological symptoms. He underwent a foramen magnum procedure at another hospital, and his symptoms stabilized.

In terms of symptom recurrence, the overall rate in patients with FMA and syringomyelia after a foramen magnum procedure was 57% within 5 years (Fig. 2). This rate was again considerably worse than that in patients with Chiari I malformation.10

Discussion

Neuroimaging in patients with syringomyelia has to...
Foramen magnum arachnoiditis

![Graph showing clinical recurrence rate](image)

Fig. 2. Graph demonstrating that the clinical recurrence rate in patients with FMA and syringomyelia after a foramen magnum procedure was 57% within 5 years.

demonstrate the entire syrinx with and without gadolinium to rule out an intramedullary tumor. Once this is accomplished and examination of the entire spinal axis has ruled out a malformation, the cause of the syrinx may remain far from obvious. Syringomyelia is generally believed to be related to disturbances of CSF flow. The presence of arachnoid cysts in the posterior fossa or hydrocephalus may indicate arachnoid disease in the posterior fossa and foramen magnum area. In some cases, a history of meningitis, trauma, hemorrhage, or surgery already suggests arachnoid scarring. In other instances, however, clinical and neuroimaging studies have to be evaluated carefully to determine the cause of a syrinx and to identify patients with FMA. In several earlier cases of this series, FMA was diagnosed preoperatively by exclusion of other diseases. Since cardiac-gated MR imaging became available, we have been able to demonstrate CSF flow obstruction at the foramen magnum in patients with FMA. Once a Chiari malformation or other brainstem- or spinal cord-compressing lesions are excluded in patients with clinical or radiological features suggesting a foramen magnum process, CSF flow obstruction at this level, as demonstrated on cardiac-gated MR images, was considered diagnostic for FMA and was proven in all nine patients with this constellation.

A syrinx originates at the level of the partial or complete CSF flow obstruction and extends in a predominantly caudal or cranial direction, depending on the level of the spinal obstruction. Therefore, it is reasonable to assume that the spinal level associated with the neurological symptoms that a patient reports as the first manifestations of his disease are in anatomical proximity to the level at which CSF flow is obstructed. If a patient harboring a holocord syrinx reports sphincter problems as his first clinical symptom, the flow obstruction is likely to be found in the lower part of the spinal canal. On the other hand, occipital headaches or upper-extremity motor weakness implicates the upper cervical or foramen magnum area. Therefore, a carefully taken clinical history is extremely important to guide further neuroimaging studies toward the anatomical region where the syrinx originated.

When examining standard MR images in such a patient, the caliber of the syrinx usually is largest close to the level of the CSF flow obstruction because the syrinx has evolved from there. Furthermore, arachnoid disease may compress, displace, or otherwise distort the MR image of the cord, whereas the syrinx tends gradually to decrease in size toward the contralateral end without displacement of the cord. Interestingly, the interval between the presumed causative event and the manifestation of syringomyelia-related symptoms is almost identical for patients with FMA, spinal trauma, or other causes of spinal arachnoid scarring.

The severity of the arachnoid disease, however, cannot be determined by this method. A history of foramen magnum surgery or meningitis foretells that thick arachnoid adhesions may be encountered.

### Surgical Management

In patients with associated hydrocephalus, this disorder should be treated first. If the patient continues to deteriorate clinically, the function of the ventricular shunt should be checked prior to considering an operation at the foramen magnum. Once hydrocephalus has been ruled out and CSF flow obstruction at the foramen magnum has been demonstrated, the treatment strategy should be to explore the foramen magnum, the objective being to establish a free-flowing CSF passage. It is very difficult to provide a reasonably accurate prediction of the postoperative outcome even when the operation succeeds and normal CSF flow is obtained. Therefore, we recommend surgery only in patients in whom the clinical situation clearly deteriorates.

Arachnoid dissection is associated with the potential risk of damaging small perforating arteries or even the PICA. To minimize such risks, we recommend using sharp dissection only and to limit surgery to the midline by using microsurgical techniques. Arachnoid disease lateral to the brainstem should be left untreated because it carries considerable risk to lower cranial nerves and small perforating arteries. Once a passage between spinal canal, fourth ventricle and cerebellopontine cisterns is achieved, no further intradural maneuvers are necessary.

Because any operation undertaken to improve a problem related to arachnoid scarring may create new arachnoid scars, the lesser the extent of dissection and the lesser the contamination of the surgical field with blood, the better the chance of achieving good long-term results. Presumably for this reason, the results of foramen magnum operations reported by Appleby, et al. were not very successful, and thus they recommended limiting surgical therapy to placement of a ventricular shunt. To reduce the

### Table 3

Preoperative and postoperative clinical symptoms in patients with FMA and Chiari I malformation

<table>
<thead>
<tr>
<th>Symptom</th>
<th>Preop</th>
<th>Postop</th>
<th>3 Mos</th>
<th>6 Mos</th>
<th>1 Yr</th>
</tr>
</thead>
<tbody>
<tr>
<td>sensory deficits</td>
<td>2.9 ± 0.5</td>
<td>3.3 ± 0.6</td>
<td>3.4 ± 0.8</td>
<td>3.4 ± 0.8</td>
<td>3.3 ± 0.8</td>
</tr>
<tr>
<td>dysesthesias</td>
<td>4.2 ± 0.8</td>
<td>4.3 ± 0.8</td>
<td>4.3 ± 0.8</td>
<td>4.3 ± 0.8</td>
<td>4.4 ± 0.8</td>
</tr>
<tr>
<td>pain</td>
<td>3.5 ± 1.0</td>
<td>3.9 ± 0.8</td>
<td>4.0 ± 0.8</td>
<td>4.0 ± 0.8</td>
<td>3.8 ± 0.9</td>
</tr>
<tr>
<td>motor power</td>
<td>2.9 ± 1.3</td>
<td>2.9 ± 1.4</td>
<td>3.1 ± 1.4</td>
<td>3.0 ± 1.5</td>
<td>2.9 ± 1.6</td>
</tr>
<tr>
<td>gait ataxia</td>
<td>3.4 ± 1.6</td>
<td>3.5 ± 1.6</td>
<td>3.7 ± 1.3</td>
<td>3.5 ± 1.4</td>
<td>3.4 ± 1.6</td>
</tr>
<tr>
<td>bladder function</td>
<td>4.3 ± 1.0</td>
<td>4.2 ± 1.0</td>
<td>4.2 ± 1.0</td>
<td>4.2 ± 1.0</td>
<td>4.3 ± 1.0</td>
</tr>
<tr>
<td>bowel function</td>
<td>4.8 ± 0.6</td>
<td>4.8 ± 0.6</td>
<td>4.8 ± 0.6</td>
<td>4.8 ± 0.6</td>
<td>4.8 ± 0.6</td>
</tr>
<tr>
<td>dysphagia</td>
<td>4.8 ± 0.6</td>
<td>4.9 ± 0.4</td>
<td>4.9 ± 0.4</td>
<td>4.9 ± 0.4</td>
<td>4.9 ± 0.4</td>
</tr>
<tr>
<td>KPS score</td>
<td>65 ± 14</td>
<td>67 ± 14</td>
<td>70 ± 14</td>
<td>69 ± 16</td>
<td>66 ± 20</td>
</tr>
</tbody>
</table>

*Values are presented as the means ± standard deviations.*
risk of postoperative adhesions, which may cause recurrent CSF flow obstruction, we use a large dural graft to create as extensive a cisterna magna as possible. In our experience, artificial material yields better results compared with autologous graft, because the tendency for adhesions is considerably lower (Fig. 1 lower right).

The risk of CSF fistulas, however, is higher. Therefore, a meticulous soft-tissue closure is an important adjunct to contain any CSF leak and to avoid penetration toward the skin.

Shunting of the syrinx will not influence the mechanism that caused the syrinx. It will also not influence those symptoms that may be directly related to the arachnoiditis itself. Therefore, shunting of the syrinx should be performed only in those cases in which the CSF flow obstruction cannot be corrected—that is, patients with extensive arachnoid disease after meningitis.

Treatment-Related Outcome

Compared with patients with Chiari I malformation, results in those with syringomyelia and FMA are considerably worse. The optimum clinical result in patients with FMA is stabilization of a previously progressive clinical neurological course. Even this can only be achieved in approximately 50% of the patients in whom a single foramen magnum operation has been performed. Two major reasons may account for this outcome. First, the rather severe arachnoid disease in this group of patients may preclude a better outcome because the objective of surgery—free-flowing CSF pathways at the foramen magnum—cannot be fully achieved in many instances. Often the surgeon has to compromise to avoid undue risks. Second, unlike patients with Chiari I malformation in whom brainstem decompression generally yields postoperative improvement, such compression does not play a role in FMA.

Conclusions

Foramen magnum arachnoiditis is a rare cause of syringomyelia. Without progressive neurological symptoms, patients with the lesions should be treated conservatively. Patients with neurological deterioration are candidates for operative intervention. The goal of surgery should be to establish free-flowing CSF pathways between the fourth ventricle, cerebellopontine cisterns, and spinal canal, with careful arachnoid dissection and creation of a cisterna magna by placing a spacious dural graft. Surgery-related results are less satisfactory compared with those in patients with Chiari I malformation.

Stabilization of the clinical course is the realistic postoperative aim in these patients, which can sometimes only be achieved after multiple operations. Placement of a syrinx shunt is indicated in patients in whom the arachnoiditis is so severe that attempts at arachnoid dissection carry undue risks.

References


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