Idiopathic syringomyelia: retrospective case series, comprehensive review, and update on management

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Object. A syrinx is a fluid-filled cavity within the spinal cord that can be an incidental finding or it can be accompanied by symptoms of pain and temperature insensitivity. Although it is most commonly associated with Chiari malformation Type I, the advancement of imaging techniques has resulted in more incidental idiopathic syringes that are not associated with Chiari, tumor, trauma, or postinfectious causes. The authors present a comprehensive review and management strategies for the idiopathic variant of syringomyelia.

Methods. The authors retrospectively identified 8 idiopathic cases of syringomyelia at their institution during the last 6 years. A PubMed/Medline literature review yielded an additional 38 articles.

Results. Two of the authors' patients underwent surgical treatment that included a combination of laminectomy, lysis of adhesions, duraplasty, and syrinx fenestration. The remaining 6 patients were treated conservatively and had neurologically stable outcomes. Review of the literature suggests that an etiology-driven approach is essential in the diagnosis and management of syringomyelia, although conservative management suffices for most cases. In particular, it is important to look at disturbances in CSF flow, as well as structural abnormalities including arachnoid webs, cysts, scars, and a diminutive posterior fossa.

Conclusions. The precise etiology for idiopathic syringomyelia (IS) is still unclear, although conceptual advances have been made toward the overall understanding of the pathophysiology of IS. Various theories include the cerebellar piston theory, intramedullary pulse pressure theory, and increased spinal subarachnoid pressure. For most patients with IS, conservative management works well. Continued progression of symptoms, however, could be approached using decompressive strategies such as laminectomy, lysis of adhesions, and craniocervical decompression, depending on the level of pathology. Management for patients with progressive neurological dysfunction and the lack of flow disturbance is unclear, although syringosubarachnoid shunting can be considered.

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Key Words • syringomyelia • idiopathic syringomyelia • syringomyelia pathophysiology

A syrinx is defined as a fluid-filled cavity that anatomically lies within the spinal cord parenchyma or the central canal. Although syringomyelia is clinically associated with a centromedullary syndrome with predominantly sensory symptoms such as pain and temperature insensitivity, in many cases it is an incidental finding. This entity is most frequently associated with a CM-I, although other known causes include spinal cord tumor, trauma, and posttraumatic or infectious adhesive arachnoiditis. With the increasing use of MR imaging, discovery of IS is more common. Various terms have been used to refer to IS including hydromyelia, idiopathic localized hydromyelia, and syringohydromyelia. Although not clearly defined, the general consensus seems to be that IS is not associated with any of the conditions mentioned previously. The management and natural history of such idiopathic syringes remain unclear. Here, we present our experience with IS in a retrospective case series of 8 patients, and we provide a comprehensive review of the current evidence. We add to our previous study examining nontraumatic cervicothoracic syringes and include several cases that were observed conservatively. We also formulate guidelines to assist neurosurgeons who encounter this pathological entity.

Methods

The study was approved by the institutional review board for patients seen in our neurosurgical clinic with

Abbreviations used in this paper: ACDF = anterior cervical discectomy and fusion; CM-I = Chiari malformation Type I; IS = idiopathic syringomyelia.
a diagnosis of syringomyelia. From this group, we retrospectively identified all patients who had been diagnosed with IS between 2005 and 2011. The patients had no evident etiology such as CM-I, tumor, trauma, or postlaminectomy adhesions. We did not have a minimal syrinx diameter for inclusion, given the overall paucity of syringes that were idiopathic. We also did not exclude patients with arachnoid webs, adhesions, or cysts, consistent with other studies on IS.

PubMed/Medline searches using the key words “idiopathic syringomyelia,” “pathophysiology of syringomyelia,” “classification of syringomyelia,” and “management of syringomyelia” resulted in 130 articles, which were independently analyzed by 2 authors (A.K.R. and N.P.S.). This group was further screened to include articles with a focus on IS; this process yielded 38 articles.

Results

A total of 8 patients were identified. The mean age was 39.3 years (25–61 years), and there were 7 men and 1 woman. Patient characteristics, type of surgery (if any), follow-up status, and duration are detailed in Table 1. Table 2 summarizes the evidence from key articles.

Symptoms and Signs

One patient (Case 1) presented with a history of worsening right arm pain and had myelopathic signs on examination that were evidenced by gait abnormalities, urinary urgency, and hyperreflexia in the lower extremities. Another patient (Case 2) presented with progressive difficulties in walking and also had myelopathic signs evidenced by hyperreflexia in the lower extremities.

Five patients (Cases 3–7) presented with milder symptoms that mostly included pain in different distributions. None of these patients had any abnormalities on examination. The patient in Case 8 had an initial diagnosis of cervical stenosis, which was treated by a C3–4 ACDF to which he responded well. Five years later, the patient presented with some recurrent radicular symptoms, which led to the diagnosis of a lower thoracic syrinx.

Table: Summary of patients with IS

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (yrs), Sex</th>
<th>Sx &amp; Signs</th>
<th>Imaging Finding; Diameter (mm), Length (mm)</th>
<th>Treatment</th>
<th>Time/Outcomes</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>43, M</td>
<td>rt UE pain, urinary urgency, patellar hyperreflexia</td>
<td>C7–T2 syrinx; 7, 38.73</td>
<td>T1–3 laminectomy w/ lysis of adhesions</td>
<td>1 yr, collapse of syrinx &amp; resolution of Sx</td>
</tr>
<tr>
<td>2</td>
<td>45, M</td>
<td>Lt leg weakness</td>
<td>C5–T5 syrinx; 5.6, 119.25</td>
<td>T4–6 laminectomy w/ lysis of adhesions &amp; fenestration of syrinx</td>
<td>5 yrs, initial resolution w/ LE pain at 5 yrs</td>
</tr>
<tr>
<td>3</td>
<td>27, M</td>
<td>mid-thoracic pain</td>
<td>T5–12 syrinx; 2, unclear</td>
<td>conservative</td>
<td>6 mos, stable syrinx &amp; resolution of Sx</td>
</tr>
<tr>
<td>4</td>
<td>43, M</td>
<td>thoracic dyesthesia</td>
<td>T6–9 syrinx; 0.78, 76.66</td>
<td>conservative</td>
<td>no FU available</td>
</tr>
<tr>
<td>5</td>
<td>40, M</td>
<td>neck &amp; thoracic pain</td>
<td>C5–T2 syrinx; 8.39, 128.13</td>
<td>conservative</td>
<td>10 yrs, stable syrinx &amp; resolution of Sx</td>
</tr>
<tr>
<td>6</td>
<td>25, M</td>
<td>low-back &amp; LE pain</td>
<td>T5–10 syrinx; 4.92, 178.81</td>
<td>conservative</td>
<td>2 yrs, stable syrinx &amp; resolution of Sx</td>
</tr>
<tr>
<td>7</td>
<td>31, F</td>
<td>neck &amp; shoulder pain</td>
<td>C4–T2 syrinx; 1.24, 61.84</td>
<td>conservative</td>
<td>no FU available</td>
</tr>
<tr>
<td>8</td>
<td>61, M</td>
<td>tingling in neck, low-back &amp; LE pain</td>
<td>T8–11 syrinx; 1.16, 75.77</td>
<td>C3–4 ACDF at time of initial presentation</td>
<td>5 yrs, episodic radicular Sx</td>
</tr>
</tbody>
</table>

* Two patients underwent surgical intervention after signs of myelopathy were evident, which was progressive in nature. All other patients had a stable follow-up for different periods of time. Abbreviations: FU = follow-up; LE = lower extremity; UE = upper extremity.

Imaging Studies

The patients in Cases 1 and 2 underwent extensive imaging, which revealed CSF flow abnormalities. In the patient in Case 1, initial MR imaging revealed a syrinx at the cervicothoracic junction. Further studies, including cine MR imaging and CT myelography, revealed abnormal flow at the T1–2 level and possible scarring/adhesions at the same level. Initial MR images obtained in the patient in Case 2 revealed a C5–T5 syrinx, and with the addition of cine MR imaging and CT myelography revealing abnormal flow at the C5–T1 levels and subarachnoid compartmentalization from T-4 to T-6.

The patients in Cases 3–8 were only investigated using MR imaging, given their mild symptoms and intact neurological examinations. All these patients had some evidence of degenerative changes apart from the syringes.

The combined mean syrinx diameter was 3.88 mm (range 0.78–8.39 mm), and the mean length was 97 mm (range 38.7–178.8 mm). A cursory analysis does reveal a larger mean diameter in the surgically versus conservatively treated cases (6.3 vs 3.08 mm); however, given the low case numbers, we have not run further statistical tests on these results. Most syringes spanned the cervicothoracic cord. The mean length was 78.9 mm for the surgical cases and 104.2 mm for the conservative cases. Only the imaging report was available for the initial scan in 1 patient (Case 3), which did not include the actual length of the syrinx.

Outcomes

Given the presence of myelopathic signs on examination with a demonstrable CSF flow abnormality, 1 patient (Case 1) underwent T1–3 laminectomy with lysis of adhesions. Figure 1 shows pre- and postoperative images of this case with collapse of the syrinx. At the 1-year postoperative visit, the patient had an intact neurological
**TABLE 2: Summary of papers on IS patients including presentation, imaging findings, and outcomes**

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Type of Study</th>
<th>No. of Pts &amp; Sex</th>
<th>Age (yrs)</th>
<th>Location</th>
<th>Sx</th>
<th>Treatment†</th>
<th>Outcome†</th>
<th>FU Period</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ataizi et al., 2007</td>
<td>case report</td>
<td>1 F</td>
<td>28</td>
<td>C5–T1</td>
<td>neck &amp; back pain</td>
<td>conservative (pt refused op)</td>
<td>resolution of pain; spontaneous collapse of syrinx</td>
<td>16 mos</td>
</tr>
<tr>
<td>Bogdanov et al., 2004‡</td>
<td>cross-sectional: 17 idiopathic, 17 CM-I, 32 control</td>
<td>17 idiopathic: 2 F &amp; 15 M</td>
<td>mean 49</td>
<td>cervical</td>
<td>segmental sensory loss, pyramidal signs, muscle atrophy</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
</tr>
<tr>
<td>Chen et al., 2004</td>
<td>case report</td>
<td>1 F</td>
<td>19</td>
<td>C2–6</td>
<td>proximal upper limb weakness, diminished pain &amp; temperature</td>
<td>suboccipital craniectomy w/ C-1 and C3–5 laminectomy</td>
<td>improved motor strength but unchanged sensory deficit; syrinx reduced</td>
<td>12 mos</td>
</tr>
<tr>
<td>Chen et al., 2011</td>
<td>retrospective case series</td>
<td>15: 6 F &amp; 9 M</td>
<td>mean 10.5</td>
<td>multiple</td>
<td>scoliosis, HA, neck pain</td>
<td>suboccipital craniectomy w/ C-1 laminectomy</td>
<td>resolved (4), improved (6), stable (3), worse (1), persistent HA (1)</td>
<td>12–75 mos</td>
</tr>
<tr>
<td>Holly &amp; Batzdorf, 2002§</td>
<td>prospective study</td>
<td>32: 14 F &amp; 18 M</td>
<td>mean 40</td>
<td>cervical (16), thoracic (12), cervico-thoracic (4)</td>
<td>mechanical spinal pain, radicular pain, numbness</td>
<td>conservative (31), ACDF at C6–7 (1)</td>
<td>improved (6), worsened (7), unchanged (19)</td>
<td>6–110 mos (mean 38 mos)</td>
</tr>
<tr>
<td>Jinkins &amp; Sener, 1999</td>
<td>case series</td>
<td>3: 2 F &amp; 1 M</td>
<td>mean 27.3</td>
<td>lumbar, cervical, thoracic</td>
<td>low-back pain, HA, nonfocal back pain</td>
<td>conservative</td>
<td>stable w/ resolution of pain (2), migraines (1)</td>
<td>2–4 yrs (mean 3 yrs)</td>
</tr>
<tr>
<td>Kastrup et al., 2001</td>
<td>case report</td>
<td>1 F</td>
<td>61</td>
<td>C1–conus medullaris</td>
<td>burning pain</td>
<td>carbamazepine</td>
<td>subsequently collapsed syrinx but Sx did not change</td>
<td>8 yrs</td>
</tr>
<tr>
<td>Kyoshima et al., 2002¶</td>
<td>retrospective case series</td>
<td>4: 3 F &amp; 1 M</td>
<td>mean 37.8</td>
<td>holocord (2), medulla–T12 (1), C1–T9 (1)</td>
<td>touch &amp; pain impairment, weakness, hypalgesia</td>
<td>cranio cervical decompression</td>
<td>all improved Sx &amp; decreased syrinx except for 1</td>
<td>2.5–11 yrs (mean 8 yrs)</td>
</tr>
<tr>
<td>Lin et al., 2006</td>
<td>case report</td>
<td>1 M</td>
<td>35</td>
<td>T2–9</td>
<td>leg weakness over 5 yrs, decreased pinprick &amp; touch below Lt T-7</td>
<td>T6–8 laminectomy, SS shunt</td>
<td>JOA score improved from 10 preop to 14 on POD 30</td>
<td>5 yrs (30 days postop)</td>
</tr>
<tr>
<td>Magge et al., 2011</td>
<td>retrospective case series</td>
<td>48: 30 F &amp; 18 M</td>
<td>mean 9.7</td>
<td>2–17 levels, mostly thoracic</td>
<td>scoliosis, cutaneous marker, LE or back pain, neurological Sx incidental</td>
<td>SS shunt (1), fenestration of syrinx (1), rest conservative</td>
<td>op (2), 1 w/ worsened gait &amp; weakness, 1 w/o no change in clinical Sx</td>
<td>clinical: 3–56 mos (mean 15.5 mos); clinical + radiographic: 2–64 mos (mean 23.8 mos)</td>
</tr>
<tr>
<td>Mallucci et al., 1997</td>
<td>retrospective case series</td>
<td>10: 2 F &amp; 8 M</td>
<td>mean 48</td>
<td>not described</td>
<td>sensory disturbance, less commonly weakness</td>
<td>laminectomy &amp; excision of web, cyst, SP shunt (2)</td>
<td>all improved (Sx better &amp; syrinx decreased) except for 2 who underwent shunting</td>
<td>not described</td>
</tr>
<tr>
<td>Mauer et al., 2008‡</td>
<td>prospective case series</td>
<td>125: 76 F &amp; 49 M</td>
<td>mean 36</td>
<td>1–18 levels not clearly defined</td>
<td>w/o op: pain, sensory impairment; w/ op: bowel or bladder dysfunction, gait abnormalities, paralysis</td>
<td>arachnoid scar or web resection (10); conservative (115)</td>
<td>op: improvement (4), remaining pts showed no further progression; conservative: precise outcomes not described</td>
<td>pts selected over 3-yr period; FU not described</td>
</tr>
</tbody>
</table>

(continued)
TABLE 2: Summary of papers on IS patients including presentation, imaging findings, and outcomes* (continued)

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Type of Study</th>
<th>No. of Pts &amp; Sex</th>
<th>Age (yrs)</th>
<th>Location</th>
<th>Sx</th>
<th>Treatment†</th>
<th>Outcome†</th>
<th>FU Period</th>
</tr>
</thead>
<tbody>
<tr>
<td>Nakamura et al., 2009</td>
<td>retrospective case series</td>
<td>15: 4 F &amp; 11 M</td>
<td>mean 45.1</td>
<td>localized C3-T2</td>
<td>upper limb numbness, neck pain, extended C1-T8 (3)</td>
<td>conservative (12), SS shunt (3)</td>
<td>conservative: no changes; op: decreased syrinx, mean JOA decreased from 11.8 to 11.2</td>
<td>7–20 yrs (mean 10 yrs)</td>
</tr>
<tr>
<td>Porensky et al., 2007</td>
<td>case report</td>
<td>2 M</td>
<td>43 &amp; 44</td>
<td>T1–2, C5–5</td>
<td>ataxia, neck pain, progressive lt leg paresis, pinprick deficit rt leg</td>
<td>laminectomy, lysis of adhesions, duraplasty</td>
<td>asymptomatic, collapsed syrinx (Case 1); refilled syrinx, unchanged neurological exam (Case 2)</td>
<td>1 yr (Case 1), 7 mos (Case 2)</td>
</tr>
<tr>
<td>Roser et al., 2010</td>
<td>prospective case series</td>
<td>40: 25 F &amp; 15 M</td>
<td>mean 36.7</td>
<td>cervical (23%), thoracic (51%), cervicothoracic (25%)</td>
<td>pain or dysesthesia of limbs</td>
<td>conservative</td>
<td>no radiological changes; neurologically stable</td>
<td>6–93 mos (mean 36.9 mos)</td>
</tr>
<tr>
<td>Struck &amp; Haughton, 2009</td>
<td>retrospective case series</td>
<td>8: 4 F &amp; 4 M</td>
<td>mean 12.1</td>
<td>lower cervical or thoracic</td>
<td>variable (scoliosis, HA, back pain, extremity numbness, family history, nausea &amp; vomiting)</td>
<td>posterior fossa decompression (4); no op at time of study (4)</td>
<td>postop: decreased Sx (4); syrinx reduction (1)</td>
<td>pts selected over 7-yr period; FU not described</td>
</tr>
</tbody>
</table>

* HA = headache; JOA = Japanese Orthopaedic Association; POD = postoperative day; NA = not available; pt = patient; SP = syringopleural; SS = syringosubarachnoid.
† Numbers in parentheses indicate the number of patients.
‡ Findings from imaging studies.
§ Ten patients had a history of trauma.
¶ Technically these cases were not idiopathic since a tight cisterna magna is a definite lesion.
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examination with resolution of symptoms. Under similar operative indications, another patient (Case 2) underwent a T4–6 laminectomy with lysis of adhesions. Although this patient did well for almost 2.5 years after surgery, he subsequently developed bilateral pain and weakness in his lower extremities. Cine MR imaging at this time revealed diminished dorsal and ventral flow from C5–T4, although we did not identify a clear pathology for surgery. In a subsequent 5-year follow-up, this patient has had some resolution of the pain.

The patients in Cases 3–8 were treated conservatively. The follow-up periods ranged from 6 months to 10 years. In 3 of these 6 patients, radiographic follow-up was also available and revealed no changes in syrinx size. Figure 2 shows images that were obtained at the initial visit and the 10-year follow-up in 1 patient (Case 5). All conservatively treated patients had resolution of their symptoms during follow-up visits, with the exception of the patient in Case 8 who had occasional radicular symptoms in his legs. This responded well to epidural steroid injections.

Discussion

Pathophysiology

There are few studies that directly address the pathophysiology behind IS. Most work in the field of syringomyelia comes from work done on CM-I. Here, we include a review of this evidence as background for possible similarities in mechanisms behind IS. Koyanagi and Houkin divided the CM-I syringomyelia evidence into 3 categories depending on the source of origin: CSF entrance from the fourth ventricle, CSF entrance from the subarachnoid space, and extracellular fluid origin.

Previous evidence by Gardner and Angel regarding CSF entrance from the fourth ventricle was supported by success of their procedure that involved closure of the obex along with a medial suboccipital craniectomy. The validity of this theory is now questioned by cine-mode MR imaging studies that did not demonstrate any CSF entrance from the fourth ventricle into the syrinx. The procedure performed by Gardner and Angel has also been reported to work as well without closure of the obex. In their review of pathological specimens, Milhorat et al. reported that syringes that communicated with the fourth ventricle were mostly found in children with hydrocephalus. This included an association with postmeningitic and posthemorrhagic hydrocephalus along with CMs Type II and Dandy-Walker cysts.

Greitz questioned the theory about increased subarachnoid pressure propelling CSF into the syrinx since such a pressure might actually compress the cavity. Moreover, most studies have not consistently identified a pressure gradient that would favor CSF flow into the syrinx from the subarachnoid space. It has been noted that delayed CT myelography after intrathecal injection of metrizamide has demonstrated enhancement of syringomyelic cavities. In their review, Koyanagi and Houkin reported that these results are not specific to syringomyelia and that delayed clearance of the contrast from syrinx cavities may explain the delayed visualization on CT myelography. In an animal model of posttraumatic syringomyelia, Brodbelt et al. showed increased perivascular flow at the level of the syrinx, although it is unclear if such flow actually causes the syrinx to develop or is simply a byproduct of localized inflammation and edema. Iwasaki et al. demonstrated that syringosubarachnoid shunting is effective at collapsing CM-I syringes; this does not appear to be in concordance with the theory postulating CSF entrance from the subarachnoid space. The piston theory by Oldfield et al. suggests that increased downward motion by cerebellar tonsils in CM-I causes increased CSF pressure, driving subarachnoid CSF into the central canal. This, however, has been questioned by the more recent studies mentioned above.

Papers that support an extracellular origin of CSF include those by Klekamp, Levine, Greitz, and Koyanagi and Hougin. The exact mechanism, however, is still debated. Greitz described the concept of intramedullary pulse pressure causing cord distension and subsequent cavitation. Koyanagi and Hougin argued that a reduced compliance of posterior spinal veins leads to reduced absorption of extracellular fluid and thus syringomyelia. The interested reader is referred to these reviews for an exhaustive coverage of these theories and historical evidence.

The intramedullary pulse pressure theory by Greitz...
deficits and mainly present with pain that could be radicular, burning, or musculoskeletal. This constellation of symptoms is similar to those described by Holly and Batzdorf.\(^{11}\) Struck and Haughton\(^ {37}\) also reported abnormal CSF flow velocities in patients with IS and raised the possibility of similarities in flow patterns at the foramen magnum between patients with CM-I and those with IS. The idea of similar morphometric abnormalities in IS and CM-I seems to follow the reasoning by Klekamp,\(^ {12}\) who questioned whether a true idiopathic syrinx actually exists. Poresky et al.\(^ {13}\) reported on 2 patients with IS; the report postulated that subtle microtrauma may have been contributory.

**Imaging and Classification**

On imaging, it is critical to differentiate between a true syrinx and residual enlargement of the central canal.\(^ {38}\) As we age, the central canal normally involutes, such that by adulthood it is often not easily seen on images.\(^ {38}\) Holly and Batzdorf\(^ {33}\) examined 32 patients with slitlike syrinx cavities, which they termed “asymptomatic persistent central canals.”\(^ {33}\) The authors found symmetrically enlarged central spinal cord cavities with a mean diameter of 2 mm (range 1–5 mm) with no enhancement after intravenous Gd injection. It was noted that 10 patients did have a history of trauma, although the study did not classify the syrinxes as posttraumatic. The study also found that 50% of the patients had alternate diagnoses for their symptoms. Holly and Batzdorf\(^ {33}\) argued that these slitlike syrinx cavities do not represent true syringomyelia and are possibly even different from a presyrinx-like state. In the literature, there is no uniformity regarding this opinion. For example, Roser et al.\(^ {14}\) differentiated hydromyelia as referring simply to a dilated central canal due to IS. These authors postulated that IS is accompanied by different clinical and radiological signs. According to Roser et al.,\(^ {14}\) patients with hydromyelia have no neurological deficits and mainly present with pain that could be radicular, burning, or musculoskeletal. This constellation of symptoms is similar to those described by Holly and Batzdorf.\(^ {14}\) Roser et al.\(^ {14}\) suggested that hydromyelia is a congenital condition; in the setting of trauma, syringomyelia could develop.\(^ {14}\) Based on these studies, it is difficult to determine if slitlike syringes, hydromyelia, and IS are truly different entities or simply a continuum on a spectrum.

Classically, the idiopathic form of syringomyelia does not have an anatomical abnormality readily seen on imaging. Some of the evidence presented in the pathophysiology section earlier, however, raises the concern that this may be too simplistic of a definition. Struck and Haughton\(^ {37}\) have reported increased peak CSF flow velocities in IS. The study by Bogdanov et al.\(^ {15}\) also found a small posterior fossa and narrow CSF spaces in IS. Kyoshima et al.\(^ {20}\) described a series of 4 patients with IS but also documented all as having a “tight cisterna magna,” referring to the cisterna magna being impacted by the tonsils. Mallucci et al.\(^ {27}\) evaluated 10 patients with apparent idiopathic syringomyelia, with subsequent CT myelography revealing CSF blockage in 9 of them. Patients who subsequently underwent laminectomy and adhesions of arachnoid webs performed much better than those who underwent shunt treatment. Clifton et al.\(^ {8}\) also reported on a patient with a T2–T5 syrinx along with a spinal intradural arachnoid cyst between T-6 and T-3 found by aqueous myelography. This patient improved after laminectomy and collapse of the cyst. A later study by Mauer et al.\(^ {28}\) evaluated 125 patients with IS using cardiac-gated phase-contrast CSF flow studies and found blockage of flow in 33 patients. The most common level of blockage was T-6. Fifteen of the 33 cases had a ventrally open circular shape syrinx cavity. In 8 of the 33 patients with unequivocal evidence of CSF flow blockage on MR imaging studies, Mauer et al.\(^ {28}\) also performed CT myelography, which revealed blockage in 2 patients. These authors concluded that conventional myelography is not a useful tool in diagnosis and that cardiac-gated CSF flow studies should suffice. According to this study, IS is not entirely idiopathic and a closer look at the anatomy may reveal structural problems.

Other descriptors of IS include a distinction of localized versus extended IS. In their retrospective case series on IS, Nakamura et al.\(^ {31}\) described localized IS as being fewer than 3 vertebrae and extended IS as 4 or more vertebrae. The localized variant had milder symptoms and was treated conservatively. Nakamura et al.\(^ {31}\) also referred to the localized syringomyelia variant as a possible congenital enlargement of the central canal. Kyoshima et al.\(^ {20}\) detailed the numerous classification systems to delineate terms such as hydromyelia, simple hydromyelia, syringomyelia, and syringohydromyelia. It is unclear if this aids in the diagnostic or management process.

**Presentation**

Symptoms that lead to the diagnosis of IS can fall into a variety of categories. In a population of 48 children with IS, Magge et al.\(^ {26}\) divided the presentation into the following 5 groups: scoliosis, cutaneous marker/developmental anomaly, pain, neurological findings, and screening/incidental finds. Chern et al.\(^ {7}\) also recently published their evidence on patients with CM Type 0 and syringomyelia, and the presenting symptoms included scoliosis, headaches, and neck, back, or leg pain.

Specifically, regarding the issue of pain, Magge et al.\(^ {26}\) noted that there does not appear to be any correlation between syrinx size or location and the symptom of pain. Although we did find an overall increased syrinx diameter in the 2 surgical cases, the relevance is unclear given the few cases we have. Nonspecific symptoms such as headaches, neck pain, and back pain may be a part of the presentation of IS.
Idiopathic syringomyelia

as pain may be a coincidental finding rather than a direct result of the syrinx. Even in our population of adults who were conservatively treated, it is difficult to localize the symptoms to the syrinx. Thus, it is critical to stress that IS is generally an incidental, asymptomatic finding. While the classically described centromedullary syndrome may accompany a syrinx, these are usually found in surgical cases.

In the few cases in which we can attempt to localize the symptoms to the syrinx, it is highly dependent on the location within the spinal cord. Patients may actually have comorbid symptoms from degenerative changes of the spine and musculoskeletal complaints, prompting medical evaluation. Although syringomyelia symptoms are classically described as pain and temperature insensitivity in a capelike distribution, few of our patients, and indeed few of the studies reviewed, actually demonstrated this. Mallucci et al.27 discussed that while the most common presenting symptom is pain followed by paresthesias, numbness, and unnoticed hand injuries, it is not uncommon for patients to present with long tract signs.

For noncommunicating syringes, symptoms can include spastic weakness of the lower extremities, paresthesias, or dysesthesias and segmental sensory loss.20 Bogdanov and Mendeleichiv reported that the pace of neurological deterioration in syringomyelia is initially rapid but slows down after neurological signs become well established.

Management

Table 2 provides a review of the key literature regarding idiopathic syringomyelia. The reader is cautioned that only some of the details for each study were included and that some of the patients were eventually found to have other structural anomalies. The studies discuss a large number of treatment options including posterior fossa and foramen magnum decompression, laminectomy, lysis of adhesions, syrinx fenestration, and syringosubarachnoid, syringoperitoneal, and syringopleural shunting. Most recent studies have reported on the importance of improving CSF flow dynamics regardless of the treatment strategy used.2,20,21,34

The management strategies outlined above should only be reserved for clearly symptomatic patients with progression on serial examinations. The majority of the patients in our series were conservatively treated, giving credence to the idea that serial imaging is an option for patients. The reader is especially cautioned in tying the symptoms to the syrinx and proceeding with surgical treatment, since the symptoms may be purely coincidental as mentioned earlier. In particular, a nonoperative approach is justified when a patient is either asymptomatic or experiences relatively mild symptoms. In their study on IS, Magge et al.26 also suggested that surgical management of the syrinx may not offer much utility since both their surgical cases either worsened or showed no changes clinically. We routinely may obtain follow-up MR images on an annual basis or more frequently as dictated by symptoms.

Historically, shunting strategies have been used and have led to clinical and radiological improvement in the treatment of IS. However, some studies have reported a variety of complications including shunt failure, syrinx relapse, catheter tip migration, and comorbidities from mechanical damage to cord tissue.2,25 Mallucci et al.27 also discussed that shunts are not an effective solution in preventing the progress of syringomyelia given the subsequent gliosis that can follow within the cord. We believe that shunting should be used as a measure of last resort when no etiology is evident after repeated imaging studies and surgical exploration do not reveal any pathology around the site of the syrinx.

In cases in which the etiology is clearly evident such as a tight cisterna magna or small posterior fossa, cranio-cervical decompression is the best option for restoring CSF flow dynamics. Holly and Batzdorf,12 however, have cautioned that too wide of a suboccipital craniectomy can lead to cerebellar ptosis. Kyoshima et al.20 also mentioned that wider opening of the foramen magnum and not the posterior fossa is the key. If CSF flow obstruction originates from spinal subarachnoid pathology such as a cyst, web, or scar, treatment should be targeted toward decompressing the spinal subarachnoid space and reconstituting flow. This is illustrated by the 2 patients in our series who underwent surgical treatment. There is no clear evidence on precisely what such decompression and reconstruction should include. Laminectomy followed by scar, web, or cyst resection has been commonly used in other studies2,21,27 and has been used with good results by the senior author (A.G.). In the event of no evidence of any anatomical abnormality in the setting of progressive neurological dysfunction attributable to a syrinx, surgical exploration is a reasonable option with a shunt as the last resort.

We have combined evidence on IS and our experience into an algorithm (Fig. 3) to assist in the decision-making process. In most cases of incidental, asymptomatic findings, annual imaging should be sufficient. For the more challenging symptomatic cases, the key focus is to resolve CSF flow problems since most of the evidence points to disrupted flow dynamics leading to the development of syringomyelia.

Conclusions

Idiopathic syringomyelia is a pathological entity in which no overt etiology is evident for a syrinx; numerous cases of IS are now attributed to CSF flow abnormalities. It is important to understand that most incidental cases of IS can be successfully managed using conservative approaches. Most patients in our series were treated conservatively. With regard to surgical options for continued progression of symptoms, syrinx shunting is generally a less favored approach as it does not resolve the underlying etiology and is associated with high failure rates. A particular challenge to the neurosurgeon is surgical treatment of syringes with no overt etiology and worsening symptoms even after a complete diagnostic workup including flow studies. While we recommend surgical exploration in these cases, future studies will hopefully reveal a more systematic approach for these patients. We have presented our institution's recent experience with IS management, and we have detailed the existing literature.

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Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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