The identification of a fluid collection within the central canal of the spinal cord in a child often results in anxiety for parents and uncertainty for clinicians. Without a readily identifiable proximate cause, the significance of these fluid collections is unclear, with a paucity of data in the literature to guide the discussion about the natural history of these lesions. For a better understanding of the implications of the presence of an idiopathic syrinx in the pediatric population, we sought to study these lesions through a collaborative effort. We present the combined data of 2 major pediatric neurosurgical centers to describe our experience with this condition.

THE IDIOPATHIC SYRINX IN THE PEDIATRIC POPULATION: A COMBINED CENTER EXPERIENCE

Clinical article

SURESH N. MAGGE, M.D.,1 MATTHEW D. SMYTH, M.D.,2 LANCE S. GOVERNALE, M.D.,1 LILIANA GOMNEROVA, M.D.,1 JOSEPH MADSEN, M.D.,1 BECCA MUNRO,2 STEPHEN V. NALBACH, M.D.,1 MARK R. PROCTOR, M.D.,1 R. MICHAEL SCOTT, M.D.,1 AND EDWARD R. SMITH, M.D.1

1Department of Neurosurgery, Children’s Hospital Boston, Harvard Medical School, Boston, Massachusetts; and 2Department of Neurosurgery, St. Louis Children’s Hospital, Washington University in St. Louis, Missouri

OBJECT

Discovery of a syrinx in a child, without a readily identifiable proximate cause such as a Chiari malformation, tumor, or site of tethering, is often a cause of concern for families and a source of consternation for clinicians. There is a paucity of data describing the natural history of an idiopathic syrinx in the pediatric population. The authors present the combined data of 2 major pediatric neurosurgical centers to describe their experience with this condition.

METHODS

Data were collected at Children’s Hospital Boston and St. Louis Children’s Hospital according to institutional review board–approved protocols and captured visits over a 2.5-year interval (October 2006–March 2009), with records reviewed if the patient had a preexisting diagnosis of syrinx. Patients were identified by ICD-9 codes derived from departmental databases. All pediatric patients (age < 19 years) in whom idiopathic syrinx had been diagnosed, as defined by MR imaging findings (dilated central canal in the spinal cord of ≥ 1 mm in axial dimension and extending over at least 2 vertebral levels), were included.

RESULTS

Forty-eight patients met the criteria for idiopathic syrinx during this period, and in 32 of them detailed follow-up imaging was available. Discovery of a syrinx was incidental in 6 patients, whereas the others were referred for imaging because of the presence of pain, neurological symptoms, scoliosis, or skin markings. The average age at the first MR imaging session was 9.7 years, with a mean syrinx size of 4 mm (range 1.2–9.4 mm). The majority (52%) of patients had a thoracic syrinx, with the average lesion spanning 7.1 vertebral levels. The average follow-up was 23.8 months (range 2–64 months), and subgroups of patients with < 3 years and ≥ 3 years of follow-up were independently reviewed. Overall, symptoms improved in 34% and worsened in 9%; 57% of the patients remained asymptomatic or stable. Radiographically (in the subgroup of 32 patients with detailed follow-up imaging), syrinx size decreased in 25% of patients, increased in 12.5%, and remained unchanged in 62.5%, with no apparent correlation between change in syrinx size and clinical symptoms.

CONCLUSIONS

Clinically, children with an idiopathic syrinx remained asymptomatic, stable, or improved in 91% of cases. The majority of syringes (87.5%) remained stable or shrank over time, with no apparent correlation between changes in size and changes in symptoms. Although longer follow-up is needed, these data suggest that the natural history of an idiopathic syrinx in children is benign, and that repeated imaging may not be necessary.

(DOI: 10.3171/2010.10.PEDS1057)

KEY WORDS • syringomyelia • hydromyelia • syringohydromyelia • idiopathic syrinx • pediatric neurosurgery • scoliosis
Idiopathic syrinx in the pediatric population

Methods

This was an Institutional Review Board–approved retrospective study of all pediatric patients (age < 19 years) in whom idiopathic syrinx had been diagnosed during the period available for review (October 2006–March 2009), as documented on MR imaging. This study was done at Children’s Hospital Boston and St. Louis Children’s Hospital. Departmental databases were searched for patients with diagnosis codes including “syringomyelia,” “cyst spinal meninges,” and “anomaly brain/spine,” as per ICD-9 billing codes. Chart records were then reviewed to identify appropriate patients.

Inclusion criteria were that patients were < 19 years of age, with the documented presence of a syrinx (which was identified on MR imaging, reported by a radiologist, and had a diameter of 1 mm or greater in axial measurement, and which extended over at least 2 vertebral levels in a rostrocaudal axis). Charts and MR imaging studies of these patients were reviewed to exclude any who also had a Chiari Type I malformation, spinal cord tumor, vascular malformation, tethered cord, history of CNS infection, history of spinal cord trauma (vertebral fracture, penetrating injury), hydrocephalus, or previous spinal surgery. A total of 48 patients were ultimately identified who met the criteria for the diagnosis of idiopathic syrinx.

Although the searchable databases covered the interval from October 2006 to March 2009, the children identified were a mix of newly diagnosed patients and individuals who already were known to have a syrinx and who were undergoing routine follow-up during this period. Therefore, the follow-up of some patients covers a period of several years.

Once identified, charts and imaging studies were reviewed for a number of criteria. Routine demographic data, including age and sex, were recorded. Presenting symptoms that led to a scan were identified and categorized into 5 groups: 1) scoliosis; 2) presence of a cutaneous marker or developmental anomaly (midline hair patch, hemangioma, and so on) considered to be a cause for concern as a possible indication of a spinal pathological entity; 3) pain in the lower extremities or back; 4) neurological symptoms (toe–walking, constipation, incontinence, abnormal reflexes, lower-extremity weakness); and 5) discovery incidentally or after screening for other unrelated conditions (clinician noting a syrinx after chest MR imaging, screening of the neuraxis for possible neurofibromatosis, and so on).

All patients had initial scans, and all patients received clinical follow-up. Changes in clinical symptoms were reviewed and documented at the longest follow-up time point. Thirty-two of the 48 patients underwent subsequent follow-up imaging to complement the clinical examination. These images were reviewed and compared with those obtained at presentation to assess for changes in the size of the syrinx and, if present, changes in the degree of scoliosis.

The MR imaging studies of the patients with an idiopathic syrinx were reviewed. The following data were obtained from the MR imaging studies: location of syrinx; size of the syrinx in millimeters in the axial plane; the number of vertebral levels involved in the sagittal plane (with each level defined as the vertebral body between disc spaces); and, in follow-up studies, documentation of any apparent progression in the size of the syrinx over time with serial images. If present, the degree of scoliosis was measured and any changes over time were documented.

Results

Forty-eight pediatric patients were found to have an idiopathic syrinx between October 2006 and March 2009. The average age of the patients was 9.7 years (range 0.2–19.3 years). Thirty patients were female and 18 were male.

Presenting Symptoms

Presenting symptoms included scoliosis in 12 patients (of note, 2 of these had associated structural causes, with the presence of hemivertebrae), a cutaneous marker or developmental anomaly causing concern in 6, lower-extremity or back pain in 13, and neurological symptoms in 15; a syrinx was found incidentally or as part of a screening study in 6 patients (4 of the 48 patients had multiple presenting signs or symptoms).

Radiographic Evaluation

The MR imaging studies were reviewed, and the average syrinx size in axial diameter was 4 mm (range 1.2–9.4 mm) (Fig. 1). The average syrinx spanned 7.1 vertebral levels in the rostrocaudal axis (range 2–17 levels), with most involving the thoracic spine. Distribution included 2 lesions contained only within the cervical spine, 25 lesions within the thoracic spine only, 14 that involved the cervicothoracic spine, 5 that were thoracolumbar, and 2 that involved cervical, thoracic, and lumbar levels.

There was little variation in syrinx size when analyzed by presenting symptom. Patients presenting with scoliosis had an average syrinx size of 3.8 mm, in those with cutaneous lesion/developmental anomalies it was 4 mm, in those with pain it was 3.9 mm, in those with neurological symptoms it was 4.1 mm, and in those in whom a syrinx was found incidentally or by screening it was 3.8 mm.

Scoliosis Evaluation

The presence of significant scoliosis was defined as a convex curve > 10°. Twelve patients (25%) of 48 met this criterion, and 2 of them were found to have associated hemivertebrae as an underlying structural cause of their deformity. Of the 10 patients without hemivertebrae, the average syrinx size was 4.3 mm (vs an average syrinx size of 3.8 mm in patients without scoliosis), and the average angulation was 20.7° (range 10°–70°).

Follow-Up and Outcomes

Follow-up data for patients in this series were analyzed both in the aggregate and also at different time points, including patients who had < 3 years of follow-up (40 patients) and those with ≥ 3 years of follow-up (8). Of these patients, 16 had only clinical follow-up, with no additional imaging after the initial study. These pa-
patients without follow-up imaging were included because they could provide data on progression or regression of symptoms over time. In the patients with only clinical follow-up, the average follow-up time was 15.5 months (range 3–56 months).

Obviously, the patients who had both clinical and radiographic follow-up were more valuable; they provided a correlation between serial radiographic images and clinical status. There were 32 patients in this group, with an average imaging follow-up of 23.8 months (range 2–64 months) and an average clinical follow-up of 24.1 months (range 1–84 months). Within this group, 24 patients had < 3 years of follow-up and 8 had ≥ 3 years of follow-up.

Outcomes were assessed by grouping patients in 2 different ways: categorizing them by presenting symptom (scoliosis, cutaneous marker/developmental anomaly, pain, neurological findings, or screening/incidental) or by length of follow-up (< 3 years or ≥ 3 years).

### Outcome by Presenting Symptom

**Scoliosis.** Of the 12 patients who presented with scoliosis, 10 had no apparent structural cause for their curve and 2 had hemivertebrae as an underlying structural cause of their deformity. Both of the latter patients underwent surgical correction of their deformity with instrumented spinal fusion, and both have remained stable with regard to their curvature postoperatively. There were 10 patients with idiopathic syringomyelia who had scoliosis without underlying structural anomalies (Table 1). Of these patients, 4 were treated with bracing and 6 had no specific treatment for their scoliosis. The average follow-up was 33 months. Evaluation of these patients revealed that 1 of 10 was marginally worse radiographically (with a minor progression of 4°; from 25° to 29° over 2 years), 4 of 10 remained stable (average curvature of 10°, range 10°–11°), and 5 of 10 improved (average change 10.1°, range 2°–40°). There was no consistent pattern of the location of the syrinx relative to the curve.

**Cutaneous Marker/Developmental Anomaly.** In a total of 6 patients, a syrinx was found secondary to a cutaneous marker or developmental anomaly. The average follow-up in these children was 41 months, and all but 1 remained stable. The child who had a change developed mild, intermittent leg weakness following an attempt to decompress the syrinx surgically (the operation was performed at another institution).

**Pain.** Thirteen patients presented with pain. A variety of interventions were used, including medications, counseling, and observation. With an average follow-up of 19 months, the pain was worse in 1 of 13 patients, unchanged in 4, and improved in 8.

**Neurological Symptoms.** Fifteen patients presented with neurological symptoms. The average follow-up for this group was 20.6 months, and within this population 2 patients were worse (increased constipation and nighttime enuresis), 9 were unchanged (unsteady gait, intermittent incontinence, tight heel cords), and 4 were improved (improved continence, better gait, and increased strength).

**Incidentally Found.** Of the 6 patients with lesions found incidentally, follow-up was 23 months, with 5 pa-

### Table 1: Outcome by symptom in 48 pediatric patients with idiopathic syrinx

<table>
<thead>
<tr>
<th>Presenting Symptoms</th>
<th>Mean FU Time</th>
<th>No. of Patients (%) total</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Worse</td>
<td>Unchanged</td>
</tr>
<tr>
<td>nonstructural scoliosis</td>
<td>33</td>
<td>1 (10)</td>
</tr>
<tr>
<td>cutaneous marker &amp;/or development anomaly</td>
<td>41</td>
<td>1 (17)</td>
</tr>
<tr>
<td>pain</td>
<td>19</td>
<td>1 (8)</td>
</tr>
<tr>
<td>neurological symptoms</td>
<td>21</td>
<td>2 (13)</td>
</tr>
<tr>
<td>incidental finding</td>
<td>23</td>
<td>1 (17)†</td>
</tr>
</tbody>
</table>

* FU = follow-up.
† Single episode of priapism.
Idiopathic syrinx in the pediatric population

patients unchanged and 1 patient with a single episode of priapism in the setting of neurofibromatosis Type I.

Outcome by Length of Follow-Up

Thirty-two patients had both clinical and radiographic follow-up. These patients were grouped according to the length of follow-up available (< 3 years and ≥ 3 years), and they were evaluated both clinically and radiographically (Table 2).

Follow-Up Less Than 3 Years. Twenty-four patients had < 3 years of follow-up. Clinically, 1 of 24 underwent an operation for spinal fusion (in the setting of a hemivertebra), 1 of 24 was worse (an increase of 4° of scoliosis in the setting of no new symptoms and a decrease in syrinx size by 2 mm), 12 of 24 were unchanged, and 10 of 24 were improved (pain was better in 6, scoliosis lessened in 2, and neurological symptoms were better in 2).

Radiographically, the size of the syrinx decreased in 3 patients (1 had a decrease of 2 mm in the setting of scoliosis worsening by 4°, 1 had a decrease of 1.8 mm in the setting of a fusion for hemivertebra, and the third patient had a decrease of 2 mm with no change in pain). The syrinx size remained unchanged in 18 and increased in 3 patients (1 had an increase of 1 mm with no change in neurological symptoms; 1 had an increase of 0.5 mm while remaining asymptomatic; and the other had an increase of 3.3 mm, with a corresponding decrease in symptomatic pain).

Follow-Up 3 Years or More. A total of 8 patients had ≥ 3 years of follow-up. Clinically, 2 of these 8 patients were worse (1 had increasing incontinence although MR imaging showed a slight decrease of the syrinx, by 0.5 mm; the other had increased leg weakness after an operation to shunt the syrinx contents, with a 3-mm decrease in syrinx size). Five of 8 patients were unchanged, and 1 of 8 was improved clinically (with improved urinary continence in the setting of a decrease in syrinx size by 2.4 mm). Radiographically, there was an increase in the size of 1 syrinx, with 2 of 8 unchanged and 5 of 8 decreased in size. Of those whose syrinx decreased in size, 2 patients were clinically worse, 1 was better, and 2 were unchanged. Increased and stable lesions were identified in association with unchanged clinical presentations.

Surgical Treatment

Two patients were treated surgically. In 1 patient, who was treated initially at another institution (for whom operative indications are unfortunately not available), the syrinx was shunted to the subarachnoid space, with subsequent new weakness of the lower extremities and a worsened, unsteady gait at 4 years postoperatively. These clinical findings were in the context of a documented decrease in syrinx size from 7 to 4 mm. The other patient was treated with fenestration of the syrinx and sectioning of the filum at one of our institutions, with reported indications for surgery centering on continued concerns about unrelenting leg pain related to the obvious large anomaly of the syrinx seen on imaging. The child had no change in the clinical symptoms of intermittent leg pain and sensory changes, and no change in the size of the syrinx, which remained stable at 8.5 mm.

Outcomes Summary

During the period of time studied, no patients with follow-up imaging (32 individuals) were subsequently found to have an identifiable proximate cause for their syrinx (that is, there were no delayed discoveries of tumor, Chiari malformation, and so on). Overall, clinically 24 (75%) of 32 patients improved, 3 (9.4%) of 32 worsened (1 developed weakness after a surgical intervention, 1 had increased incontinence frequency, and 1 had a 4° increase in scoliosis), and 18 (56.2%) of 32 were unchanged. (To be transparent about the need for intervention, in Table 2 we moved from the “unchanged” to the “worse” category a patient whose symptoms did not change clinically, but who ultimately underwent surgery for a congenital hemivertebra.)

Radiographically, the average syrinx size at presentation was 4 mm (range 1.2–9.4 mm), and 20 children (62.5%) had no change in the size of their syrinx. Of the 12 of 32 lesions that were noted to change over the period of time studied (average 23.8 months), 4 (12.5%) increased (by an average of 1.6 mm), and 8 (25%) decreased (by an average of 1.8 mm). There was no correlation between symptom change (for better or worse) and change in syrinx size.

Discussion

Syrinx is a term used to describe the presence of a fluid-filled cavity oriented in a rostrocaudal axis within the parenchyma of the spinal cord. The syrinx can have an ependymal or glial lining, and is presumed to be filled with a derivative of CSF. Nomenclature related to this subject can be confusing, with the terms “dilated central canal,” “hydromyelia,” “syringomyelia,” and “syringo-hydromyelia” often used interchangeably in describing this condition. Hydromyelia refers to an ependymal-lined cavity that may be a dilated central canal. This is contrasted with syringomyelia, which refers to a cavity within the

<p>| TABLE 2: Outcome by length of follow-up in 32 patients with both clinical and radiographic findings |
|-----------------------------------------------|----------------|----------------|</p>
<table>
<thead>
<tr>
<th>Parameter</th>
<th>&lt;3 Yrs</th>
<th>≥3 Yrs</th>
</tr>
</thead>
<tbody>
<tr>
<td>total no. of patients</td>
<td>24</td>
<td>8</td>
</tr>
<tr>
<td>symptoms</td>
<td></td>
<td></td>
</tr>
<tr>
<td>improved</td>
<td>10 (42)</td>
<td>1 (12.5)</td>
</tr>
<tr>
<td>unchanged</td>
<td>12 (50)</td>
<td>5 (62.5)</td>
</tr>
<tr>
<td>worse</td>
<td>2 (8)*</td>
<td>2 (25)</td>
</tr>
<tr>
<td>syrinx size</td>
<td></td>
<td></td>
</tr>
<tr>
<td>decreased</td>
<td>3 (12.5)</td>
<td>5 (62.5)</td>
</tr>
<tr>
<td>unchanged</td>
<td>18 (75)</td>
<td>2 (25)</td>
</tr>
<tr>
<td>increased</td>
<td>3 (12.5)</td>
<td>1 (12.5)</td>
</tr>
</tbody>
</table>

* One patient in the category had surgery for a hemivertebra, not for worsening of syrinx symptoms.
spinal cord, which has a gliotic (not ependymal) wall. Syringohydromyelia is a broad term used to encompass either of these cavities, because they can be difficult, if not impossible, to distinguish on imaging. In practice, it is not clear that there is any clinical relevance to discriminating between the 2 types of cavities. For the sake of simplicity, and following precedent, the term “syrinx” will be used to describe the radiographic findings reviewed here.2

Identification of a syrinx in a patient is important to clinicians because it may herald the presence of one of several treatable conditions. These include Chiari malformations (Types I and II, and possibly Type 0 as well); spinal cord tethering (including fatty fila, especially those > 2 mm in diameter, lipomas, and sinus tract); spinal cord tumors; and vascular malformations. In situations in which the syrinx is secondary to a proximate cause, treatment of the underlying disorder (if possible) is of paramount importance. Successful treatment of the primary problem will often result in subsequent reduction or resolution of the syrinx. A spinal cord syrinx may also arise following trauma or infection, due to presumed loss of neural tissue and scarring, leading to a cavity that fills with CSF.

A syrinx may be present without evidence of a proximate cause, either radiographically or by history. Identification of an idiopathic syrinx can present a challenge to clinicians who may be unsure of the implications of this finding. Many children with an idiopathic syrinx are referred to neurosurgeons for evaluation.

Following the requisite detailed neurological examination, history, and documentation of complete imaging of the entire spine to exclude treatable causes, there is a paucity of literature to guide the neurosurgeon in his or her discussion with the family about the natural history of this condition.

A review of the literature reveals 8 reports comprising 68 total cases of patients found to have an idiopathic syrinx (Table 3). Of the patients in the literature review, we were only able to find 2 children under the age of 18 years. Given these extremely limited data on the natural history of pediatric idiopathic syrinx, we undertook this study to describe the combined experience with this condition at 2 major pediatric neurological centers.

With the increasing availability and more powerful resolution of MR imaging, the ability to identify the presence of a syrinx continues to expand. There are approximately 8.4 new cases of a syrinx reported per year per 100,000 people.5 This radiographically observed entity is often found in association with clinical symptoms such as sensory changes, pain, spasticity, weakness, and urinary difficulty. In many cases, the syrinx is presumed to be a consequence of another, primary disorder (as noted previously). If the syrinx is a result of a primary process, then treatment of this process (if possible) is usually recommended—often with subsequent marked resolution of the syrinx.

Contrasted against the syrinxes found in association with a known cause are those found in the absence of other pathological conditions—the idiopathic syrinx. Using a broad definition of syrinx, which included “dilated central canals,” one group reported the prevalence of these cavities as 1.5% in the general population.11 Although debate continues on whether there are criteria for distinguishing between a dilated central canal and a syrinx, it is apparent that there are a substantial number of central fluid-filled cavities being identified on MR imaging that do not have an apparent proximate cause. Once these are found, clinicians—often neurosurgeons and neurologists—are being asked for advice on the natural history and management of these entities. Unfortunately, the appropriate course of action for such cases is often unclear, given the limited data on the subject (Table 3).

In response to the need for better data on this entity in children, we have described the experience with the diagnosis and management of idiopathic syrinx in the pediatric population at our respective institutions. For the sake of clarity, we have looked to the literature for guidance and have decided to define a syrinx as a centrally located fluid-filled cavity within the spinal cord, with a diameter of at least 1 mm in the axial plane and extending over at least 2 vertebral levels in a rostrocaudal axis.9,10

It was our objective to provide information for the treating physician by framing our analysis in the context of how patients may ask their questions—if a child presents with scoliosis in the setting of an idiopathic syrinx, for example, how might the curve change over time? Alternatively, one might ask if changes—either clinical or radiographic—occur more commonly shortly after diagnosis, or if they become apparent only after the passage of time.

### TABLE 3: Prior reports of idiopathic syringomyelia*

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>No. of Cases</th>
<th>Avg Pt Age</th>
<th>Spine Segment</th>
<th>Avg Time of FU</th>
<th>Change in Syrinx</th>
</tr>
</thead>
<tbody>
<tr>
<td>Yeager &amp; Lusser, 1992</td>
<td>1</td>
<td>19 mos cervical</td>
<td>cervical</td>
<td>2 yrs</td>
<td>resolved on MRI</td>
</tr>
<tr>
<td>Ataizi et al., 2007</td>
<td>1</td>
<td>28 yrs cervical</td>
<td>cervical</td>
<td>17 mos</td>
<td>resolved on MRI</td>
</tr>
<tr>
<td>Jinikins &amp; Sener, 1999</td>
<td>3</td>
<td>27 yrs cervical/thoracic/lumbar</td>
<td>cervical</td>
<td>2.5 yrs</td>
<td>clinically stable (no repeat MRI)</td>
</tr>
<tr>
<td>Chen et al., 2004</td>
<td>1</td>
<td>19 yrs cervical</td>
<td>cervical</td>
<td>1 yr</td>
<td>decreased after cervical decompression</td>
</tr>
<tr>
<td>Kastrup et al., 2001</td>
<td>1</td>
<td>61 yrs cervical/thoracic/lumbar</td>
<td>cervical</td>
<td>8 yrs</td>
<td>resolved on MRI</td>
</tr>
<tr>
<td>Petit-Lacour et al., 2000</td>
<td>12</td>
<td>34 yrs cervical</td>
<td>cervical</td>
<td>3 mos–9 yrs stable on FU</td>
<td></td>
</tr>
<tr>
<td>Bogdanov et al., 2004</td>
<td>17</td>
<td>49 yrs cervical</td>
<td>cervical</td>
<td>none</td>
<td>advocated PF decompression</td>
</tr>
<tr>
<td>Holly &amp; Batzdorf, 2002</td>
<td>32</td>
<td>16–63 yrs cervical 16, thoracic 12, cervicothoracic 4</td>
<td>cervical</td>
<td>32 mos (mean)</td>
<td>stable on MRI (all cases)</td>
</tr>
</tbody>
</table>

* Avg = average; NA = not available; PF = posterior fossa; PT = patient.
Idiopathic syrinx in the pediatric population

Our data revealed that the presentation of children with an idiopathic syrinx could be categorized into 5 major groups based on the rationale for obtaining spinal cord imaging: scoliosis; cutaneous marker/developmental anomaly; pain; neurological findings; or screening/incidentally found. Of those presenting with scoliosis, the only patients who required surgery were those with documented bony structural anomalies (hemivertebrae). For those without hemivertebrae, 1 patient worsened minimally (4°), and the remaining 90% were stable or improved. The size of the syrinx did not correlate with the degree of the curve or outcome. These findings suggest that patients with idiopathic syringomyelia and scoliosis should be carefully evaluated for bone abnormalities, but—if none are found—can be reassured that the clinical course will probably be benign, with the curve amenable to treatment with bracing or observation.

Patients who presented with cutaneous marker/developmental anomalies were stable throughout their follow-up, with the only exception being a child who worsened after surgery to decompress the syrinx. Similarly, patients in whom a syrinx was found incidentally did well, with the only reported issue being a single episode of priapism in 1 patient. Our data support the premise that children in whom a syrinx is identified in the absence of clinical symptoms will remain asymptomatic over time.

Children who present with some type of pain in the setting of a syrinx generally do well, with 61% improving, 31% remaining stable, and only 8% worsening over the period of time studied. Outcomes appear to be similar whether children are followed for <3 years or ≥3 years. It does not appear that there is any correlation between syrinx size or location and the symptom of pain. The patients who presented with an identifiable neurological finding had less variability in their outcomes; in 13% symptoms were worse, 27% had improvement, and the remaining majority were unchanged. It is tempting to speculate that the radiographic finding of the syrinx is a physical manifestation of an error in a developmental process or a perinatal injury that may herald an absence of properly functioning spinal cord parenchyma, which in turn results in a relatively fixed deficit.

Radiographically, the size of the syrinx did not appear to play a role in the presentation or outcome of the patients in this study. There was a great deal of variation in individual syrinx sizes, but negligible differences between groups of patients when categorized by symptoms. Moreover, there was no clear predictive utility to observed changes in syrinx size; 12.5% increased over time, 25% decreased, and the rest remained unchanged, with no apparent correlation between symptom change and change in syrinx size. Specifically, an increase in syrinx size did not necessarily mean worsened symptoms, a decrease in size did not mean that symptoms improved, and patients with no change in size could still manifest changes in clinical presentation over time. All in all, there does not appear to be any clear connection between radiographic findings and symptoms.

Although there was a limited number of cases (2 of 48 patients, or 4%), our data suggest that surgical intervention aimed at decreasing the size of the syrinx may not offer much utility. One patient remained unchanged clinically, with no change in the size of the syrinx, whereas the other was treated with a syringosubarachnoid shunt. Following this intervention, the size of the syrinx decreased and the patient concomitantly developed new leg weakness. Because the weakness progressed over time and had some waxing and waning presentation, it is unclear whether the weakness was secondary to injury at surgery, to physiological changes to the cord resulting from syrinx decompression, or to some combination thereof. When the limited experience with surgical intervention is compared with the overall outcomes of the 46 other, nonsurgically treated patients, the data appear to support the route of nonsurgical management. Overall, only 1 nonsurgically treated patient had any worsening clinically (increased incontinence); the other reported adverse outcome was radiographic evidence of a mild scoliosis progression (4°). A little more than one-third of all patients demonstrated clinical improvement over the course of the study, whereas the rest remained stable.

This study is the largest on the subject to date, and provides both clinical and radiographic outcomes, which help to define the natural history of idiopathic syringomyelia in children. However, there are a number of limitations inherent to this work. Many of the clinical findings are subjective, such as pain and neurological findings, and the very definition of what constitutes syringomyelia is debatable. Most importantly, this is a retrospective study, with relatively small numbers of patients and limited follow-up. Ideally, one would have clinical and radiographic data spanning a longer period, such as a decade. Although we are continuing to collect this information in a prospective fashion and hope to be able to present greater longitudinal follow-up at a future date, we believe that the data described in this study offer new and useful information to practicing clinicians. The ability to cite a combined center experience with defined groups of patients—some of whom have more than 3 years of both clinical and radiographic follow-up—offers physicians some meaningful answers to questions that both they and patients’ families may have.

In summary, we recommend that all patients who present with a syrinx undergo detailed imaging of their neuraxis to look for proximate causes of the lesion. Part of this initial evaluation may include studies that enable identification of congenital and acquired lesions, sites of tethering, and compartmentalization of the subarachnoid space (as has been described in entities such as the so-called Chiari Type 0, or spinal arachnoid cysts). If a cause is found, treatment of the primary problem (if possible) will often result in diminution or obliteration of the syrinx. If no proximate cause is found initially, our data suggest that it is highly unlikely that subsequent imaging would reveal a new pathological entity at a later date. However, if a patient experiences notable worsening in his or her clinical examination, we will often consider reimaging the spine to assess for changes.

Clinically, children with an idiopathic syrinx remained asymptomatic, stable, or improved in 91% of cases. Radiographically, the majority of syringes (87.5%)
remained stable or shrank over time, with no apparent correlation between changes in size and changes in symptoms. Our data suggest that surgical intervention targeted at reducing the size of the syrinx in these cases may not be effective, and that symptomatic management by appropriate specialists (such as orthopedic surgeons, neurologists, or urologists) may be the appropriate course to follow. Parents can be reassured that most children will remain stable or improve with time.

Conclusions

This is the largest study to date focused on the entity of the idiopathic syrinx in the pediatric population. Initial evaluation should include adequate imaging of the CNS to exclude treatable causes. In the majority of our patients with an idiopathic syrinx, there was no radiographically noted change in syrinx size over the period of the study, and 91% of patients remained clinically asymptomatic or stable, or improved over time. Our data suggest that children with an idiopathic syrinx generally follow a benign clinical course in the first few years following diagnosis, and may require only limited follow-up.

Disclosure

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

Author contributions to the study and manuscript preparation include the following. Conception and design: Smith, Magge, Smyth. Acquisition of data: Smith, Magge, Smyth, Madsen, Munro, Nalbach. Analysis and interpretation of data: Smith, Magge, Smyth, Governale, Munro, Nalbach. Drafting the article: Smith, Magge, Smyth, Governale, Munro, Nalbach. Critically revising the article: Smith, Magge, Smyth, Goumnerova, Munro, Nalbach, Proctor, Scott. Reviewed final version of the manuscript and approved it for submission: Smith, Smyth.

References


Manuscript submitted February 4, 2010. Accepted October 15, 2010. Address correspondence to: Edward R. Smith, M.D., Department of Neurosurgery, Children’s Hospital of Boston/Harvard Medical School, 300 Longwood Avenue, Boston, Massachusetts 02115. email: edward.smith@childrens.harvard.edu.