Chiari malformation Type I is a common pediatric condition that often prompts a neurosurgical referral following diagnosis. The relationship between CM-I and a spinal cord syrinx is well established.4–8,47,50 Chiari malformation Type I can lead to spinal cord syrinx formation as a result of abnormal flow of the CSF at the foramen magnum.4,8,10,18,24,27,35,36,38,47 The true prevalence of syrinx in CM-I is not well defined, as most estimates have been derived from case series of patients undergoing surgery.4,14,26,27,38,42 Studies describing groups of patients that have been surgically treated for CM-I tend to have a bias for including symptomatic cases and excluding those that are more minimally affected. Syringomyelia is found in as many as 75% of patients undergoing surgical treatment for CM-I.9,26,27,32,43 Since surgeons are more likely to recommend surgery if a spinal syrinx is present, the true prevalence of a syrinx in patients with CM-I is lower.17,41

The radiographic characteristics of CM-I and syrinx have not been described in a large series, with the exception of descriptions of surgical outcomes following decompression.4,27,38,42 The relationship between patient age and other clinical and imaging characteristics, and the prevalence of syrinx in CM-I has never been properly studied. An improved understanding of these relationships may lead to better treatment decisions for children with CM-I.25,41

Chiari malformation Type I and syrinx in children undergoing magnetic resonance imaging

Clinical article

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Object. Chiari malformation Type I (CM-I) with an associated spinal syrinx is a common pediatric diagnosis. A better understanding of the relative age-related prevalence and MR imaging characteristics of these associated conditions may lead to improved treatment decisions.

Methods. The authors performed a retrospective review of 14,116 consecutive individuals 18 years of age or younger who had undergone brain or cervical spine MR imaging at the University of Michigan between November 1997 and August 2008. In the patients with CM-I, demographic, clinical, and radiographic information was recorded.

Results. Five hundred nine children (3.6%) with CM-I were identified. Among these patients, 23% also had a spinal cord syrinx, and 86% of the syringes were found in the cervical spine. The MR imaging prevalence of CM-I with a syrinx was 1.2% in girls and 0.5% in boys (p < 0.0001). The severity of impaired CSF flow at the foramen magnum was associated with the amount of tonsillar herniation (p < 0.0001) and conformation of the tonsils (p < 0.0001). Patients with CM-I were treated surgically in 35% of cases; these patients exhibited more severe tonsillar herniation (p < 0.0001) and impaired CSF flow (p < 0.0001) as compared with those who did not undergo surgery. On imaging, 32% of all the patients with CM-I were considered symptomatic by the treating physician. Patients were more likely to be considered symptomatic if they were female, had a syrinx, displayed abnormal tonsillar pulsations, or had a greater amount of tonsillar herniation.

Conclusions. In this study the authors describe the age-related prevalence and MR imaging characteristics of CM-I and its association with a syrinx and other abnormalities in a large group of children who underwent MR imaging for any indication. Syringes are more common in older children, in girls, and in patients with a greater degree of tonsillar descent and CSF flow impairment. (DOI: 10.3171/2011.5.PEDS1121)

Key Words • Chiari malformation Type I • syrinx • prevalence • magnetic resonance imaging
Methods

Following approval by the University of Michigan Institutional Review Board, we performed a retrospective review of the electronic medical records of all patients 18 years of age or younger who had undergone brain or cervical spine MR imaging at the University of Michigan between November 1997 and August 2008. Brain or cervical spine MR imaging was performed in 14,116 children during this period. All MR images were obtained on either a 1.5- or 3-T MR imaging device. All electronic records were reviewed using the Electronic Medical Record Search Engine (EMERSE), a search engine that queries all free-text documents within the electronic medical records of a specified patient population. We used EMERSE to identify a population in which the terms “tonsillar ectopia,” “tonsillar herniation,” “tonsillar descent,” “syrinx,” “syringomyelia,” “hydromyelia,” or “Chiari” were used in the text of the medical record. We then manually reviewed the medical and imaging records of all patients identified by EMERSE to select those who met the inclusion criteria. Cerebellar tonsillar descent ≥ 5 mm below the foramen magnum was considered diagnostic of CM-I. Patients were excluded from the study if tonsillar ectopia was believed to be caused by tumor mass effect, hydrocephalus, cysts, or cerebral edema or was associated with craniostenosis. Any patient who met the criteria for CM Type II, III, or IV, including a history of myelomeningocele repair, was excluded from the study. Any patient who had received surgical treatment for CM-I prior to their first MR imaging review at our institution was also excluded.

For patients who met all inclusion criteria, we recorded age, sex, presenting symptoms, associated radiological diagnoses, syrinx characteristics, indication given for the initial MR imaging, and any surgical treatment. We reviewed the MR images from each of the patients with CM-I and recorded the imaging characteristics of the malformation, including the degree of tonsillar descent in millimeters as well as tonsillar morphology. If a single patient had more than 1 MR image during the study period, the initial MR image on which the malformation was diagnosed was used for analysis.

Tonsillar descent was assessed by identifying the line from the basion to opisthion and then by measuring from this line to the inferior margin of the cerebellar tonsils on sagittal MR imaging. Tonsillar morphology was classified as either rounded or pegged. Cerebrospinal fluid flow data were recorded for all patients who had dedicated CSF flow sequences on MR imaging. At our institution, changes in signal intensity on sagittal phase-contrast CSF flow studies are observed in the CSF spaces anteriorly and posteriorly at the level of the cervicomедullary junction. The alternating bright and dark signals seen in the CSF spaces in the cine mode are diminished or absent when there is abnormal flow. On the axial and sagittal phase contrast images, any change in signal intensity of the cerebellar tonsils in the cine mode suggests tonsillar pulsations. Cerebrospinal fluid flow was categorized as normal, decreased anteriorly or posteriorly at the foramen magnum, or decreased at the foramen magnum with abnormal tonsillar pulsations, based on the initial report of the radiologist. The presence of scoliosis, defined as a ≥ 10° lateral Cobb angle on upright radiography, was recorded. Patients without adequate spine radiographs in our system were also designated as having scoliosis if a prior diagnosis of scoliosis was in the medical record. We noted any additional abnormal findings on the radiology report following MR imaging of the brain or spine, including retroverted dens or basilar invagination. Determining basilar invagination or retroflexed dens was based on the radiologist’s original interpretation of the images.

For the present analysis, a syrinx was defined as a spinal cord cyst (T1 hypointensity and T2 hyperintensity) ≥ 3 mm in width in the anteroposterior dimension on sagittal or axial MR imaging. Patients who had a presyrinx state (T2 hyperintensity within the cord parenchyma without cavitation) were considered separately. Either a neurologist or a neurosurgeon at our hospital had performed the clinical evaluation in 443 patients (87%). Patients were considered symptomatic if the treating physician thought the patient had symptoms caused by a CM or spinal syrinx.

Statistical significance calculations were obtained using ANOVA, chi-square, and Tukey multiple comparisons as well as univariate logistic regression. Data were analyzed using SPSS version 16.0 software (SPSS, Inc.).

Results

At our institution, 14,116 children underwent MR imaging of the brain or cervical spine over an 11-year interval. Of these, 509 patients (3.6%) met our imaging criteria for CM-I. Brain MR imaging had been performed in all of these patients, whereas cervical spine MR imaging had been performed in 397 (78%). At least one MR image of the entire spine had been obtained in 256 patients (50%). On imaging, 117 patients (0.83%) had both CM-I and spinal syrinx. The prevalence of CM-I in those who underwent MR imaging did not vary significantly by age (p = 0.54; Fig. 1). However, a spinal syrinx was more commonly found in older children with CM-I (p =
Chiari malformation Type I and syrinx on MR imaging

The prevalence of syrinx increased with age, with the greatest increase occurring in the first 5 years of life (Fig. 2). There was no significant increase in the prevalence of syrinx after 5 years of age. The prevalence of CM-I did not differ significantly by sex.

In those with CM-I, the mean tonsillar descent measurement was 10.2 mm below the foramen magnum. Cerebrospinal fluid flow studies were obtained at or near the time of CM-I diagnosis in 308 patients (61%). An increasing amount of tonsillar herniation was associated with more severe alterations in CSF flow at the foramen magnum. Patients with abnormal tonsillar pulsations had a mean tonsillar descent of 13.5 mm as compared with a mean of 11.5 mm in patients with decreased flow anterior or posterior to the foramen magnum (p < 0.0001). Abnormal tonsillar pulsations were also associated with a pegged tonsillar morphology. Pegged tonsils were found in 77 (93%) of 83 patients with abnormal tonsillar pulsations as compared with 46 (51%) of 91 patients with normal CSF flow at the foramen magnum (p < 0.0001).

### Table 1: Syrinx and scoliosis in 509 pediatric patients with CM-I

<table>
<thead>
<tr>
<th>Morbidity</th>
<th>Male No.</th>
<th>% MRI†</th>
<th>% CM‡</th>
<th>Female No.</th>
<th>% MRI†</th>
<th>% CM‡</th>
<th>Total No.</th>
<th>% MRI†</th>
<th>% CM‡</th>
</tr>
</thead>
<tbody>
<tr>
<td>CM-I</td>
<td>249</td>
<td>3.4</td>
<td>NA</td>
<td>260</td>
<td>3.8</td>
<td>NA</td>
<td>509</td>
<td>3.6</td>
<td>NA</td>
</tr>
<tr>
<td>CM-I &amp; syrinx</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>w/ scoliosis</td>
<td>22</td>
<td>0.3</td>
<td>8.8</td>
<td>51</td>
<td>0.8</td>
<td>19.6</td>
<td>73</td>
<td>0.5</td>
<td>14.3</td>
</tr>
<tr>
<td>w/o scoliosis</td>
<td>15</td>
<td>0.2</td>
<td>6.0</td>
<td>29</td>
<td>0.4</td>
<td>11.2</td>
<td>44</td>
<td>0.3</td>
<td>8.6</td>
</tr>
<tr>
<td>total</td>
<td>37</td>
<td>0.5</td>
<td>14.8</td>
<td>80</td>
<td>1.2</td>
<td>30.8</td>
<td>117</td>
<td>0.8</td>
<td>22.9</td>
</tr>
<tr>
<td>CM-I &amp; scoliosis</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>w/ syrinx</td>
<td>22</td>
<td>0.3</td>
<td>8.8</td>
<td>51</td>
<td>0.8</td>
<td>19.6</td>
<td>73</td>
<td>0.5</td>
<td>14.3</td>
</tr>
<tr>
<td>w/o syrinx</td>
<td>13</td>
<td>0.2</td>
<td>5.2</td>
<td>28</td>
<td>0.4</td>
<td>10.8</td>
<td>41</td>
<td>0.3</td>
<td>8.1</td>
</tr>
<tr>
<td>total</td>
<td>35</td>
<td>0.5</td>
<td>14.0</td>
<td>79</td>
<td>1.2</td>
<td>30.4</td>
<td>114</td>
<td>0.8</td>
<td>22.4</td>
</tr>
</tbody>
</table>

* NA = not applicable.
† Percentage of patients 0–18 years of age undergoing MR imaging.
‡ Percentage of patients 0–18 years of age with CM-I on MR imaging.

### Table 2: Incidental or associated findings on MR imaging in patients with CM-I

<table>
<thead>
<tr>
<th>Finding</th>
<th>No. (%)</th>
<th>Tonsillar Herniation</th>
<th>Syrinx</th>
<th>CSF Flow</th>
</tr>
</thead>
<tbody>
<tr>
<td>retroverted dens</td>
<td>33 (6.5)</td>
<td>&lt;0.00001</td>
<td>0.30</td>
<td>0.006</td>
</tr>
<tr>
<td>basilar invagination</td>
<td>24 (4.7)</td>
<td>&lt;0.00001</td>
<td>0.006</td>
<td>0.001</td>
</tr>
<tr>
<td>arachnoid cyst</td>
<td>18 (3.5)</td>
<td>0.21</td>
<td>0.58</td>
<td>0.81</td>
</tr>
<tr>
<td>callosal dysgenesis</td>
<td>15 (2.9)</td>
<td>0.85</td>
<td>1.00</td>
<td>0.31</td>
</tr>
<tr>
<td>venous angioma</td>
<td>15 (2.9)</td>
<td>0.60</td>
<td>1.00</td>
<td>0.81</td>
</tr>
<tr>
<td>Klippel-Fell syndrome</td>
<td>14 (2.7)</td>
<td>0.64</td>
<td>1.00</td>
<td>0.56</td>
</tr>
<tr>
<td>absent septum pellucidum</td>
<td>12 (2.4)</td>
<td>0.86</td>
<td>0.31</td>
<td>0.081</td>
</tr>
<tr>
<td>hemivertebra</td>
<td>12 (2.4)</td>
<td>0.086</td>
<td>1.000</td>
<td>0.22</td>
</tr>
<tr>
<td>platybasia</td>
<td>9 (1.8)</td>
<td>0.002</td>
<td>0.44</td>
<td>0.13</td>
</tr>
<tr>
<td>butterfly vertebrae</td>
<td>8 (1.6)</td>
<td>0.14</td>
<td>0.086</td>
<td>0.18</td>
</tr>
<tr>
<td>spina bifida occulta</td>
<td>8 (1.6)</td>
<td>0.85</td>
<td>0.086</td>
<td>0.34</td>
</tr>
</tbody>
</table>

* In each category, patients in whom these additional diagnoses had been made were tested against all other patients with CM-I for the amount of tonsillar herniation (in mm), the presence of a syrinx, or abnormal CSF flow at the foramen magnum. Significant p values suggest that a greater amount of tonsillar herniation, the presence of a syrinx, or more severe CSF flow alterations at the foramen magnum were associated with the listed clinical condition as compared with patients with CM-I but without the listed condition.
spine imaging. Girls were more likely than boys to have undergone complete spine imaging. Of the patients with CM-I who had undergone at least cervical and thoracic spine imaging, 155 were girls (155 [60%] of 260 girls) and 108 were boys (108 [43%] of 249 boys). A syrinx was found in 117 patients (23%) with CM-I (Table 1). In contrast to CM-I alone, CM-I together with syrinx was more common in girls (1.2%) as compared with boys (0.5%; p < 0.0001). More girls had both CM-I and scoliosis (1.2%) as compared with boys (0.5%; p < 0.0001). There was overlap between patients with syrinx and those with scoliosis. Seventy-three patients had CM-I, spinal syrinx, and scoliosis. Chiari malformation Type I with scoliosis but no syrinx was found in 41 patients. Chiari malformation Type I with spinal syrinx but no scoliosis was found in 44 patients (Table 1).

The mean syrinx width was 7.8 mm, and the average length was 9 vertebral levels. Individuals with a syrinx had a greater amount of tonsillar descent at the time of CM-I diagnosis. Those with a syrinx had a mean tonsillar descent of 12.6 mm; those without a syrinx had a mean tonsillar descent of 9.5 mm (p < 0.0001; Fig. 3). More severe CSF flow alterations at the foramen magnum occurred in patients with a syrinx: 52% of patients with abnormal tonsillar pulsations had a syrinx compared with 13% of patients with normal CSF flow at the foramen magnum (p < 0.0001). Furthermore, only 6% (7 of 126) of patients with rounded tonsils had a syrinx compared with 31% (109 of 357) of patients with pegged tonsils (p < 0.0001). Patients with basilar invagination were more likely to have an associated syrinx (p = 0.006). A syrinx was found in 46% of patients with basilar invagination compared with 22% (106 of 485) of patients without. Although the degree of tonsillar herniation did correlate with the presence of a syrinx, the degree of tonsillar herniation was not associated with syrinx width (p = 0.6). Similarly, alterations in CSF flow were not associated with syrinx dimensions such as length or width (p = 0.15 and 0.28, respectively). Syrinx length was not associated with the degree of tonsillar descent. A test of slope was conducted using linear regression to determine whether an increase in tonsillar herniation was associated with a linear increase or decrease in syrinx length. The slope was not significantly different from 0 (p = 0.9). Tonsillar descent was also evaluated for a relationship with the cranial extent of the syrinx by comparing tonsillar descent in groups of syringes with a cranial extent at C-3 or above, between C-4 and C-7, and in the thoracic spine. The tonsillar descent was not significantly different between these groups (p = 0.6). Syringes were found to have their cranial extent within the cervical spine in 86% of cases (Fig. 4). In the 16 patients with a syrinx that was entirely within the thoracolumbar spine, 5 had scoliosis and 11 did not.

Of those patients with CM-I on MR imaging, 32% were considered to be symptomatic at the time of CM-I diagnosis by the treating physician. A higher proportion of girls (41%) were considered to be symptomatic as compared with boys (22%; p < 0.0001). Patients who were symptomatic were more likely to have a syrinx (p < 0.0001). Older children were more likely to be classified as symptomatic (p = 0.0002). The mean age of asymptomatic patients at diagnosis was 8.0 years, and the mean age of symptomatic patients at diagnosis was 9.9 years. Symptomatic patients had more severe alterations in CSF flow at the foramen magnum (p < 0.0001). Finally, symptomatic patients had a greater amount of tonsillar descent (p < 0.0001). Patients with symptoms had a mean tonsillar descent of 13 mm, whereas those without had a mean descent of 9 mm. Associated clinical conditions discovered at the time of CM-I diagnosis are presented in Table 3.
Treatment by decompression occurred in 180 patients (35%). The mean tonsillar descent in these patients was 12.9 mm, and the mean descent in those treated without surgery was 8.7 mm (p < 0.0001). The severity of CSF flow alterations at the foramen magnum was also associated with the surgical treatment of CM-I. Sixty-five (78%) of 83 patients with abnormal tonsillar pulsations had decompression surgery compared with 25 (27%) of 91 patients with normal CSF flow. Patients treated using decompression were older at the time of CM-I diagnosis compared with those whose disease was managed without surgery, but this difference was not statistically significant (p = 0.1). Patients who underwent surgery for CM-I had a mean age of 9.3 years at diagnosis, and those without surgery had a mean age of 8.4 years at diagnosis (Fig. 5).

The indications for obtaining the initial MR image as provided by the ordering physician are listed in Table 4. In many cases, the indication for imaging was considered a potential diagnosis that was not confirmed on MR imaging. Chiari malformation Type I was diagnosed at the time of the first MR image in 482 patients (95%). Another 27 patients had an initial MR image that was not diagnostic for CM-I but later had at least 1 other MR image that met our criteria for CM-I diagnosis. In these patients, the mean interval between the first and the subsequent scan demonstrating a CM was 1087 days.

**Table 3:** Additional clinical diagnoses in patients at the time of CM-I diagnosis

<table>
<thead>
<tr>
<th>Condition</th>
<th>No. (%)</th>
<th>Tonsillar Herniation</th>
<th>Syrinx</th>
<th>CSF Flow</th>
</tr>
</thead>
<tbody>
<tr>
<td>scoliosis</td>
<td>114 (22.4)</td>
<td>0.001</td>
<td>&lt;0.0001</td>
<td>0.001</td>
</tr>
<tr>
<td>sleep apnea†</td>
<td>65 (12.9)</td>
<td>0.002</td>
<td>0.009</td>
<td>‡</td>
</tr>
<tr>
<td>seizure disorder</td>
<td>64 (12.6)</td>
<td>0.020§</td>
<td>0.0007§</td>
<td>0.140</td>
</tr>
<tr>
<td>shunted hydrocephalus</td>
<td>42 (8.3)</td>
<td>0.310</td>
<td>0.010</td>
<td>0.007</td>
</tr>
<tr>
<td>autism</td>
<td>26 (5.1)</td>
<td>0.21</td>
<td>0.34</td>
<td>0.29</td>
</tr>
</tbody>
</table>

* In each category, patients with these additional conditions were tested against all other patients with CM-I for the amount of tonsillar herniation (in mm), the presence of a syrinx, or abnormal CSF flow at the foramen magnum. In most cases, the p values indicate that a greater amount of tonsillar herniation, the presence of a syrinx, or more severe CSF flow alterations at the foramen magnum were associated with the listed clinical condition as compared with patients with CM-I but without the listed condition.
† Fifty-seven patients were diagnosed with obstructive sleep apnea; 4 patients each had central sleep apnea or combined obstructive and central sleep apnea.
‡ Insufficient data available for statistical analysis.
§ Patients had less tonsillar herniation, no syrinx, and less severe CSF flow at the foramen magnum.

**Discussion**

Cerebellar tonsillar descent of at least 5 mm below the foramen magnum is usually considered consistent with an imaging diagnosis of CM-I. Several prior studies estimated the prevalence of CM-I to be between 0.6% and 1% in those undergoing imaging. Meadows et al. found CM-I in 0.77% of patients undergoing MR imaging at a single referral center. All age groups were included in their analysis, but children composed a relatively small proportion of their study participants. Age-specific or even age group-specific prevalence is not reported by Meadows et al., and it is not clear whether the prevalence is different in children as compared with adults in their series. Their report is remarkable for the relatively few asymptomatic cases of CM-I discovered on imaging; only 25 (14%) of 175 patients with CM-I were thought to be asymptomatic. In contrast, we found a much higher percentage of asymptomatic cases (68%) at the time of CM-I diagnosis. It is certainly possible that this percentage reflects differences between institutions in the relative sensitivity of CM-I diagnosis as well as in referral biases for imaging. Our determination of symptom-
atic or asymptomatic CM-I was made according to the judgment of the treating physician. The complex clinical presentation of CM-I as well as the retrospective nature of this study does not allow for a more precise definition. In general, at our institution, patients with headaches are considered symptomatic if the headaches have at least some of the features considered compatible with CM-I headaches, including a tussive component, a short duration, and a lack of migrainous features. Other symptoms assigned to CM-I include sleep apnea, swallowing difficulty, scoliosis, and motor or sensory disturbances in the extremities of patients with spinal syrinx.

In our series as well as in several prior reports,2,11,30,34 patients who were symptomatic on presentation were older at the time of CM-I diagnosis. To some extent, this finding may be a result of the difficulty of eliciting a history of consistent CM-I symptoms in very young patients. Infants with CM-I often present with atypical symptoms, and diagnosis may be delayed. It is possible that the greater prevalence of asymptomatic cases in our series reflects our focus on children rather than on a group consisting mostly of adults.

Patient sex appears to be an important factor in CM-I presentation. Some groups have reported a female predominance for CM-I, but this finding is not universal.9,14,25,27,34,50 Although our results do not support any sex predilection for CM-I diagnosis on imaging, we did find that girls are more likely to be considered symptomatic, to have an associated spinal syrinx, and to have associated scoliosis. As a result, girls appear to be more likely to present for neurosurgical treatment, probably explaining the female predominance in some series.9,23,27,40 In our own series, girls were more likely to undergo surgical treatment of CM-I.

Chiari malformation Type I is known to cause spinal cord syrinx in some patients.4,6–8,10,18,24,32,35,38 Most surgical studies report that between 60% and 85% of patients with CM-I have an associated syrinx.26,27,43 Because the presence of a syrinx is an indication for surgery at many centers, published surgical studies have overestimated the frequency with which a syrinx occurs in patients with CM-I.15,37,41 Therefore, the true age-related prevalence of spinal syrinx in children with CM-I is not well established. Meadows et al.25 found only 1 patient with a spinal syrinx in their series of 25 asymptomatic patients with CM-I. It is possible that syringes were uncommon in their study because of screening methods that may have been less sensitive in detecting spinal syringes. Most of the patients in our series underwent cervical spine MR imaging to screen for syrinx, and half of our patients underwent complete spine imag-

**TABLE 4: Indication for initial MR imaging in patients with diagnosed CM-I***

<table>
<thead>
<tr>
<th>Indication</th>
<th>Frequency (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>headache</td>
<td>111 (22.8)</td>
</tr>
<tr>
<td>scoliosis</td>
<td>77 (15.8)</td>
</tr>
<tr>
<td>neurological change</td>
<td>60 (12.3)</td>
</tr>
<tr>
<td>seizure</td>
<td>58 (11.9)</td>
</tr>
<tr>
<td>hydrocephalus/macrocephaly*</td>
<td>43 (8.8)</td>
</tr>
<tr>
<td>other</td>
<td>29 (6.0)</td>
</tr>
<tr>
<td>cranial nerve palsy</td>
<td>27 (5.6)</td>
</tr>
<tr>
<td>developmental delay</td>
<td>22 (4.5)</td>
</tr>
<tr>
<td>trauma</td>
<td>22 (4.5)</td>
</tr>
<tr>
<td>pituitary/endocrine</td>
<td>19 (3.9)</td>
</tr>
<tr>
<td>tumor/cyst/mass*</td>
<td>18 (3.7)</td>
</tr>
<tr>
<td>total</td>
<td>486† (100)</td>
</tr>
</tbody>
</table>

* Indication for imaging was provided on the MR imaging requisition. In many cases, the indication for imaging was considered a possible diagnosis that was not confirmed on MR imaging.
† Indication for initial imaging was unknown in 23 patients.
Chiari malformation Type I and syrinx on MR imaging

ing. We found on initial imaging that a syrinx was present in 23% of all patients with CM-I. It is possible that even this rate represents an overestimate of the true population prevalence of syrinx in children with CM-I, especially given that a population of children undergoing spine imaging may have an increased likelihood of harboring an associated syrinx as compared with children with CM-I who do not come to medical attention. In patients with CM-I, age appears to be a relevant factor in spinal syrinx prevalence. In contrast to CM-I, which was found throughout the pediatric age range with a similar age-related prevalence, syringes were much less common during the first few years of life but do appear to reach a stable age-related prevalence by 5 years of age. This finding supports our current understanding of the causal relationship between the malformation and the spinal syrinx and suggests some potential utility for spine imaging follow-up in the children with CM-I diagnosed at a very young age.

The relevance of the degree of tonsillar herniation and the likelihood of syringomyelia formation is controversial. Although some studies have shown that syringes are associated with a greater amount of tonsillar herniation, others have suggested that an intermediate degree of herniation between 9 and 14 mm is more likely to be associated with a syrinx than either lesser or greater degrees of tonsillar descent. In contrast, we found that an increased amount of tonsillar herniation was associated with a greater likelihood of an associated spinal syrinx (Fig. 3). It seems possible that the prior report assigning pathophysiological significance to an intermediate degree of tonsillar herniation may have suffered from a small sample size.

In our series, in addition to increased tonsillar descent, syringes were associated with pegged tonsillar morphology and decreased CSF flow at the foramen magnum. Not surprisingly, we also found that patients with basilar invagination and hydrocephalus were more likely to have a spinal syrinx (Tables 2 and 3). Furthermore, a retroverted dens and basilar invagination were both associated with increased tonsillar descent. These associations are consistent with current morphometric theories of CM-I pathogenesis. For the present analysis, we used the radiological interpretation as the basis for determining the presence of basilar invagination and retroflexed dens. It is possible that the presence of basilar invagination or a retroflexed dens is more likely to be noted by a radiologist when a syrinx is present, a potential source of bias in this analysis. The negative correlation that was found between spinal syrinx and seizures, a diagnosis that is unrelated to CSF flow at the foramen magnum, may be a result of a selection bias for children with this condition who were undergoing MR imaging.

Although CM-I–associated syringomyelia can occur at any level of the spine, even as low as the conus, the cervical spine is most frequently affected. This tendency follows a general principle for syrinx formation at or immediately caudal to any pathological narrowing of the subarachnoid space within the craniocervical junction or spinal canal. We found that most syringes had their cranial extent in the cervical spine, but a significant number of patients had a syrinx only in the lower segments (Fig. 4). This finding should be considered when ordering imaging studies. If the presence of a syrinx would change the treatment recommendation, total spine imaging should be considered in patients with CM-I. Of the 16 patients with a spinal syrinx entirely within the thoracic or lumbar spine, 11 did not have associated scoliosis.

Patients with CM-I were identified by examining medical and imaging records from a cohort of patients undergoing MR imaging at our institution. This method allowed an analysis of all patients with a diagnosis on MR imaging without respect to their ultimate treatment or clinical evaluation. The 509 patients with CM-I described here reflect the selection bias that is anticipated for a group that has been referred for neuroimaging at a medical center. This bias is evident in the indications provided for obtaining the initial MR image as well as the frequency of comorbidities in this patient group. Although there are many reasons to obtain an MR image in a child, the population of children undergoing MR imaging is different from the general population. Therefore, our reported prevalence estimate for CM-I should be considered the MR imaging prevalence rather than the true population prevalence. It seems likely that MR imaging prevalence is an overestimate of CM-I prevalence in the general population. Several groups have reported on intracranial findings in healthy adult volunteers. Although each of these studies was small, a meta-analysis by Morris et al. revealed CM-I on 71 of 15,559 MR imaging studies from combined data of multiple reports in adults. The sensitivity for detecting CM-I in each of these studies and in the subsequent meta-analysis is not clear, and in some reports contained in the meta-analysis, no cases of CM-I were found. Given these concerns about the sensitivity of CM-I detection, it is possible that the estimated prevalence of CM-I provided by those studies may be less than the true population prevalence. Vernooij et al. recently studied 2000 healthy adults over the age of 45 years and found 18 volunteers (0.9%) with CM-I. To obtain a true age-based population prevalence of CM-I in children, screening the general pediatric population with MR imaging would be preferable to reviewing a cohort of children for whom scanning was thought to be medically necessary. Nevertheless, an analysis of MR imaging prevalence in the group undergoing MR imaging for any reason can provide insight into associated conditions, age and sex differences, and imaging characteristics of CM-I and syrinx.

Conclusions

At our institution, 3.6% of children undergoing MR imaging of the brain or cervical spine had CM-I, with a near equal sex distribution. Syrinx and scoliosis were much more common in girls with CM-I than in boys with CM-I. A greater amount of tonsillar herniation and pegged tonsillar morphology were associated with spinal cord syrinx and more severe alterations in CSF flow at the foramen magnum. A syrinx was found in 23% of children with CM-I and was found much less frequently in the first 5 years of life.

Disclosure

Dr. Muraszko is a consultant for Stem Cells, Inc.
Author contributions to the study and manuscript preparation include the following. Conception and design: Maher, Muraszko, Garton. Acquisition of data: Maher, Muraszko, Strahle, Kapurch. Analysis and interpretation of data: all authors. Drafting the article: Maher, Strahle, Kapurch. Critically revising the article: all authors. Approved the final version of the manuscript on behalf of all authors: Maher. Study supervision: Maher, Muraszko, Garton.

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