Scoliosis and Chiari malformation Type I in children

Clinical article

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Object. The identification of Chiari malformations Type I (CM-Is) has increased in recent years, commonly during MR imaging for evaluation of a possible cause of scoliosis. The treatment of this abnormality remains controversial, and the expected success of treatment is unclear. The goal of the present study is to evaluate the effects of a craniotomy for CM-I decompression on scoliosis in children and adolescents.

Methods. The authors conducted a 10-year retrospective review of pediatric patients who were found to have a CM-I during evaluation for scoliosis. Seventy-nine patients were identified, ranging in age from 6 months to 18 years (median 12 years). There were 42 girls (54%) and 37 boys (46%). All were noted on MR imaging to have hydrosyringomyelia of the spinal cord. Forty-nine patients had curvatures less than 20° prior to treatment. The other 30 patients had curves ranging from 25° to 80° and underwent orthopedic follow-up and treatment. None of these patients were referred for specific neurological complaints, but 12 (16%) had neurological signs on physical examination. All were treated with a craniocervical decompression in a standard fashion. Follow-up ranged from 6 to 93 months with a median of 35 months. Magnetic resonance images obtained at 6 months postoperatively and serial standing antero-posterior spine radiographs were used to evaluate outcomes.

Results. On the MR images obtained 6 months postoperatively, 70 patients (89%) had a significant reduction in the syrinx with an associated ascent of the cerebellar tonsils. Persistent large syringes were treated with reoperation in 6 patients, and shunts were inserted for hydrocephalus in 2 patients. None of the 49 patients with curves less than 20° had progression of their curvature postoperatively. Of the 30 patients with curves greater than 25°, 9 had no change in the scoliosis or had a reduction in curve magnitude after Chiari decompression. This group required no further therapy and was effectively treated by Chiari decompression alone. Twenty-one patients required further scoliosis treatment after Chiari decompression; 12 required orthotic treatment, 11 received spinal instrumentation and fusion surgery, and 2 received orthoses followed by fusion and instrumentation. The severity of the curvature beyond 20° did not predict the need for spinal surgery.

Conclusions. This large series reports on the efficacy of treatment for scoliosis associated with a CM-I and syrinx in children. A CM-I decompression alone was adequate treatment for mild scoliosis of less than 20°. Patients with scoliosis greater than 20° required bracing and/or spinal fusion surgery 70% of the time in addition to the CM-I decompression. (DOI: 10.3171/2010.10.PEDS10154)

Key Words • Chiari malformation • scoliosis • syringomyelia • Cobb angle • outcome study • pediatric neurosurgery

Abbreviation used in this paper: CM-I = Chiari malformation Type I.
Methods

Under an institutional review board–approved protocol, a 10-year retrospective review (between January 1992 and December 2001) was performed at Children's Hospital Los Angeles. One hundred four children who had surgical treatment for a CM-I were identified. Eighty-nine patients had a CM-I and a syrinx, and 79 patients had a CM-I, syrinx, and scoliosis. No patient had a CM-I and scoliosis without a syrinx. It is this group of 79 children and adolescents who comprise the study group. Over this period, 926 spinal instrumentation and fusion procedures were done for scoliosis treatment.

The charts of these 79 patients were reviewed in their entirety. Charts included operative, radiological, and pathological reports and records of neurosurgical and orthopedic outpatient visits. All patients underwent preoperative MR imaging of the entire neuraxis, including a Gd-enhanced sequence to evaluate for other pathological entities (such as spinal cord tumors). The ventricular size was evaluated, and if hydrocephalus was seen, surgical management was carried out. Patients with hydrocephalus or primary spinal column or cord pathology were not included in the current series. The indications for obtaining an MR image in each of these patients included abnormal findings during neurological examination, an atypical presentation of scoliosis either related to young age at onset or curve pattern, or atypical imaging characteristics of the scoliosis, including the lack of vertebral rotation in association with the lateral curvature.

Each patient was offered a CM-I decompression surgery, and all underwent a stereotypical surgical procedure. An occipital craniectomy limited to 2.5 × 2.5 cm was performed. A C-1 laminectomy was performed in all cases, with a C-2 laminectomy performed in 12 cases in which the tonsils descended to C-2. The dura mater was opened in all cases, followed by a lysis of arachnoid adhesions, ensuring a free egress of CSF from the fourth ventricle in the midline as well as the lateral apertures. The dura was loosely approximated, but no duraplasty was performed.

The median follow-up was 35 months and ranged from 6 to 93 months. All patients underwent at least 1 MR imaging session 6 months postoperatively. Subsequent MR images were obtained initially yearly and then as indicated. Standing anteroposterior spine radiographs were also obtained during the pre- and postoperative periods as indicated. These were measured according to the Cobb method, with a change in the Cobb angle of the primary scoliosis curve of greater than 5° being considered significant. All patients underwent follow-up until they reached skeletal maturity and were finished growing.

Results

Seventy-nine children were identified who presented with scoliosis and were found to have a CM-I and hydroxyringomyelia of greater than 6 mm in diameter. This group included 42 girls and 37 boys. The patients’ ages ranged from 6 months to 18 years, with a median age of 12 years.

All of these children presented for medical evaluation because they were noted to have scoliosis. None had a neurological complaint initially. However, on further evaluation, 12 patients (16%) were discovered to have a neurological finding. Nine had gait abnormalities, 4 had upper-extremity weakness, 8 had sensory changes, and 1 had ophthalmoplegia; 4 patients had more than 1 finding. On questioning, 26 patients had a history of headaches.

Scoliosis was measured using the Cobb method. On initial evaluation, 49 of the patients had curves less than 20°, whereas 30 patients had curves ranging from 25° to 80°. A disproportionate number of patients with curves less than 20° were young, a finding in line with prior reports of spinal cord abnormalities on MR imaging in about 30% of children with the onset of a significant scoliosis before the age of 10 years (Table 1).

Postoperatively, 10 of the 12 patients with neurological signs had resolution of these on physical examination 6 months following decompression surgery. Only 8 of the 26 patients with headaches reported persistent headaches at the time of follow-up.

There were no deaths in this series. One patient had a cerebellar hematoma that required surgical evacuation. Thirteen patients (16%) had headache, nausea, and/or vomiting in the postoperative period consistent with aseptic meningitis. All cases were self-limited. Seven patients (9%) had CSF leakage, which required bedside suturing. One of these patients developed an infection, which required a course of antibiotics.

All patients underwent MR imaging 6 months postoperatively to evaluate the hydrosyringomyelia. Sixty (76%) of the 79 patients had total or near-total (< 2 mm of residual syrinx) resolution of the syrinx. An additional 9 patients had greater than 50% reduction of the syrinx. Thus, 69 (87%) of 79 patients had a significant size reduction of the syrinx, along with the ascent and decompression of the cerebellar tonsils.

Ten of the patients did not have significant reduction of the syrinx on MR imaging studies at the time of follow-up. Two patients had progressive hydrocephalus and underwent placement of ventriculoperitoneal shunts with subsequent resolution of the syrinx. Two patients had resolution of the CM-I and no abnormal enhancement and placement of syringopleural shunts, with syrinx resolution. Six patients underwent additional surgical exploration of the foramen magnum region with lysis of arachnoid adhesions, and 1 patient required 2 such procedures.

<table>
<thead>
<tr>
<th>TABLE 1: Outcome of scoliosis by age</th>
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<tbody>
<tr>
<td><strong>Age (yrs)</strong></td>
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<tr>
<td>&lt;6</td>
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<tr>
<td>6–8</td>
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<tr>
<td>8–10</td>
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<tr>
<td>10–12</td>
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<tr>
<td>12–14</td>
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<tr>
<td>14–16</td>
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<tr>
<td>&gt;16</td>
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</table>
Scoliosis and Chiari malformation Type I

All of these patients had MR imaging studies that indicated control of the syrinx after these surgical procedures. Scoliosis was also evaluated preoperatively and postoperatively in an on-going manner (Tables 2 and 3). Forty-nine patients initially presented with curves of 20° or less, and the average age of this group was 11.8 years. All of these patients had either no further curve progression or resolution of their scoliosis following the CM-I decompression, and none of these patients required any additional orthopedic intervention.

Thirty patients presented with curves of 25°–80°; the average age of this subgroup was 12.4 years. At the time of latest follow-up, 9 patients had no further curve progression or had improvement in scoliosis and did not require further orthopedic treatment, except for periodic radiographic follow-up. Scoliosis bracing had been begun in 2 of these patients before the CM-I decompression. The other 21 patients in this subgroup had postoperative progression of the scoliosis despite the CM-I decompression surgery. Twelve patients required a new thoracolumbosacral orthosis, and 11 (including 2 of the 12 patients who had originally received orthoses) patients eventually required spinal instrumentation and fusion surgery. Curve patterns in those requiring brace treatment included 5 patients with a left thoracic scoliosis, 6 patients with a right thoracic scoliosis, and 1 patient with a double thoracic-lumbar curve pattern. Children who required spinal instrumentation and fusion surgery were slightly older on average at the time the diagnosis of a CM-I was made, in comparison with the group as a whole (12.9 vs 11.9 years) and tended to have larger curves. In the patient group requiring spinal fusion surgery, 4 patients had a left thoracic curve, 5 had a right thoracic curve, and 2 had a single thoracolumbar curve (2 patients had a double thoracolumbar curve pattern). In this group with significant progressive scoliosis of greater than 25°, there was no direct correlation between the magnitude of the curvature preoperatively and the need for subsequent spinal instrumentation and fusion surgery. There was also no correlation seen between the size of the syrinx preoperatively on the MR imaging studies and the eventual need for corrective spinal surgery. An abnormal neurological examination also did not predict outcome.

Discussion

Scoliosis is a well-recognized common condition in childhood. It is estimated that 2%–4% of all children between the ages of 10 and 16 years have some degree of abnormal curvature of the spine. However, in most cases, the curvature is relatively mild and nonprogressive, with only approximately 10% of those with spinal symmetry requiring treatment other than periodic observation.

One of the challenges for the orthopedist evaluating a child or teenager with scoliosis is to differentiate between patients with idiopathic scoliosis and those with a clear etiology. Idiopathic scoliosis typically is seen in adolescent girls during the pubertal growth spurt. A right thoracic scoliosis is the most common curve pattern, with the convex side to the right and with a right-sided rib prominence on physical examination. In these adolescent girls with at least a year of growth remaining and with a scoliosis that progresses to greater than 25°, brace treatment is recommended, using a thoracolumbosacral orthosis worn about 20 hours daily until skeletal maturity. If the scoliosis is greater than 50° in the thoracic area and greater than 40° in the thoracolumbar or lumbar area, spinal instrumentation and fusion surgery is recommended. Patients with an atypical presentation or with neurological signs or symptoms warrant a workup for a causative etiology. Examples of atypical presentation include a left thoracic curve pattern, onset of significant scoliosis before the age of 10 years, and little or no vertebral rotation on the anteroposterior spine radiograph showing the lateral spinal curvature. It is estimated that up to 10% of cases of significant scoliosis have a demonstrable underlying etiology, with syringomyelia being among the most commonly seen in younger patients with scoliosis. In a study by Schwend et al., 14 of 95 cases of scoliosis were found to have a causative spinal cord anomaly, most commonly a syrinx. In that report, all of the cases were predictable by abnormalities discovered in the neurological history or by examination.

The present study reports on 79 patients who presented with scoliosis and were found to have a CM-I and a spinal cord syrinx. During this 10-year period, 926 patients underwent operative correction of their scoliosis. Most of these 926 patients did not undergo MR imaging. Patients with scoliosis underwent imaging if they presented at an anomalous age (significant scoliosis before age 10 years and before the pubertal growth spurt) or by unusual characteristics of the curve. However, 12 patients (16%) had positive neurological signs or symptoms on examination, which prompted further workup. Thus, at our institution, fewer than 10% of the patients with the need for surgical treatment of scoliosis were found to have a CM-I and a

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**TABLE 2: Outcome of scoliosis by initial Cobb angle**

<table>
<thead>
<tr>
<th>Cobb Angle (°)</th>
<th>Total</th>
<th>w/ Stable Scoliosis</th>
<th>w/ Progressive Scoliosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt;20</td>
<td>49</td>
<td>49</td>
<td>0</td>
</tr>
<tr>
<td>21–30</td>
<td>10</td>
<td>3</td>
<td>7</td>
</tr>
<tr>
<td>31–40</td>
<td>12</td>
<td>4</td>
<td>8</td>
</tr>
<tr>
<td>41–50</td>
<td>4</td>
<td>1</td>
<td>3</td>
</tr>
<tr>
<td>&gt;51</td>
<td>4</td>
<td>1</td>
<td>3</td>
</tr>
</tbody>
</table>

**TABLE 3: Cobb angle based on age and presentation**

<table>
<thead>
<tr>
<th>Age (yrs)</th>
<th>&lt;20°</th>
<th>21–30°</th>
<th>31–40°</th>
<th>41–50°</th>
<th>&gt;51°</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt;6</td>
<td>16</td>
<td>3</td>
<td>4</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>6–8</td>
<td>5</td>
<td>2</td>
<td>4</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>8–10</td>
<td>3</td>
<td>2</td>
<td>1</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>10–12</td>
<td>4</td>
<td>2</td>
<td>1</td>
<td>0</td>
<td>0</td>
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<tr>
<td>12–14</td>
<td>4</td>
<td>1</td>
<td>2</td>
<td>1</td>
<td>1</td>
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<tr>
<td>14–16</td>
<td>10</td>
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<td>0</td>
<td>1</td>
<td>1</td>
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<tr>
<td>&gt;16</td>
<td>7</td>
<td>0</td>
<td>0</td>
<td>2</td>
<td>1</td>
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</table>
This rate is similar to those reported by other authors, who have described MR imaging–evident intraspinal abnormalities in 2%–20% of their children with scoliosis.\textsuperscript{7,12,18-20} We emphasize our findings and the findings of previous authors\textsuperscript{2-26} and recommend that a patient with an abnormal neurological examination, an atypical age at presentation, or atypical physical examination or radiological characteristics of the scoliosis should undergo a complete neuraxis MR imaging study (cervical, thoracic, and lumbar spine) as part of the evaluation of the scoliosis. We emphasize that in our series, 84% of the patients had normal findings on a neurological examination, and imaging was undertaken because of the atypical age at presentation or atypical nature of the curvature.

The association between CM-I, syrinx, and scoliosis is well recognized\textsuperscript{7,12,13,18,19,21-23,24} It is assumed, but not definitively proven, that the syrinx is the cause of the scoliosis. Based on this assumption, most clinicians advocate addressing the CM-I and syrinx as an initial step in the management of the scoliosis.\textsuperscript{17} However, no large study has previously evaluated the outcome of CM-I decompression on the subsequent course of the scoliosis. Smaller series of fewer than 25 patients have yielded variable results. Muhonen et al.\textsuperscript{25} reported that 9 of 11 patients had improvement or stabilization of their scoliosis after CM-I decompression and did not require spinal surgery. Kontio et al.\textsuperscript{20} reported that only 1 of 5 patients required spinal instrumentation and fusion after the neurosurgical procedure. However, Sengupta et al.\textsuperscript{27} Eule et al.,\textsuperscript{14} Farley et al.,\textsuperscript{15} and Ghanem et al.\textsuperscript{16} all reported that more than half of their patients still required spinal fusion surgery after the CM-I decompression was performed. The largest prior series, presented by Brockmeyer et al.\textsuperscript{7} showed intermediate results between these other studies, with 8 (38%) of 21 patients having scoliosis progression after the CM-I decompression. These authors reported that age older than 12 years and scoliosis greater than 50° increased the likelihood that the patient would require additional scoliosis surgery after the CM-I decompression was performed. In another series, Anttenello et al.\textsuperscript{3} showed that 10 (48%) of their 21 patients had a progression of their curvature, and that risk factors included thoracolumbar junction scoliosis and failure of the syrinx to improve. They showed a linear relationship between the prospective Cobb angle and curve progression.

The current report of 79 patients evaluated the efficacy of CM-I decompression in terms of syrinx resolution, neurological improvement, and scoliosis stabilization. Magnetic resonance imaging studies demonstrated resolution of the syrinx 6 months postoperatively in 87% of the patients, and syrinx control was achieved in the remaining patients with additional procedures. Resolution of the neurological findings was seen in 80% of the patients, and 69% had resolution of headache symptoms. No further orthopedic intervention was required in 74% of the patients who had stabilization or improvement in the scoliosis after CM-I decompression. Given the large size of this series, we believe that these numbers accurately represent the expected and attainable outcome of neurosurgical treatment.

However, 27% of our patients (21 of 79) had progression of their scoliosis and required additional orthopedic treatment. Although this percentage is lower than that in the other reported series, this represents a sizeable number of patients in whom the CM-I decompression was not the definitive treatment for the scoliosis. Although the data do not achieve statistical significance, patients requiring additional scoliosis treatment tended to be those with larger curvatures in the early part of the adolescent growth spurt, similar to findings described by others.\textsuperscript{3,14,21,27} Peak growth velocity occurs in females for the 2 years following the onset of breast development and before the onset of menarche. If a scoliotic curve is large at the time of diagnosis of the CM-I and syrinx, which usually occurs before the adolescent growth spurt in most, the years of growth remaining will tend to lead to uneven spinal growth and a progressive scoliosis requiring spinal surgical treatment, despite the prior CM-I decompression and syrinx resolution.

While most orthopedists treating children with scoliosis are aware of the association between atypical spinal deformity and CM-I and syrinx, many primary care physicians may not be. It is clear from our data and from that of others that early detection of scoliosis is important, and early recognition of atypical spinal deformity is key. In our series, in which no patient with a scoliotic curve less than 20° at the time of diagnosis and treatment of the CM-I and syringomyelia needed further scoliosis treatment, it appears safe to conclude that early intervention is the most important factor to improve neurological signs, to improve headache symptoms, and to prevent the need for later spinal fusion surgery.

Conclusions

In this large series of patients who presented with scoliosis and were found to have a CM-I with a syrinx, a CM-I decompression alone was adequate treatment for mild scoliosis of less than 20°. For patients with curves greater than 25°, the risk of curve progression is high even after a CM-I decompression and syrinx resolution, with 70% (21 of 30 patients) in this subgroup requiring further orthopedic treatment of bracing or surgery. Patients who present at an older age and with a larger degree of curvature are more likely to require spinal instrumentation and spinal fusion surgery. These patients require close periodic orthopedic follow-up at least until skeletal maturity and may eventually need orthotic or surgical treatment.

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: Krieger, McComb. Acquisition of data: Krieger, Bowen, Falkinstein, Tolo. Analysis and interpretation of data: Krieger, Bowen, Falkinstein, Tolo. Drafting the article: Krieger, Bowen. Critically revising the article: Krieger, Bowen, McComb. Reviewed final version of the manuscript and approved it for submission: all authors. Statistical analysis: Krieger, Bowen. Administrative/technical/material support: Krieger, Bowen. Study supervision: Krieger, Bowen.
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References


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