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Chiari I Malformation Associated with Syringomyelia and Scoliosis

A Twenty-Year Review of Surgical and Nonsurgical Treatment in a Pediatric Population

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Study Design. Retrospective review of patients with Chiari I malformation with or without associated scoliosis.

Objectives. Determine the effect of decompression of Chiari I malformation with syringomyelia on stabilization or improvement of associated scoliosis.

Summary of Background Data. Chiari malformations are often associated with spinal deformities, including scoliosis. Studies have suggested a causal relation between syringomyelia and scoliosis.

Methods. Patients with Chiari I malformation and syringomyelia with or without scoliosis treated over the last 20 years were reviewed. Patients with any other anomalies were excluded. Scoliotic curves were classified by magnitude and curve type. All patients were treated with surgical decompression of the Chiari malformation with or without drainage of the syringomyelia.

Results. Twenty-five patients were identified, ranging in age from 19 months to 16.5 years. Nineteen patients (76%) had associated scoliosis. The majority of the patients with scoliosis (13 of 19) sought treatment for spinal deformity, and only 6 had for pain or neurologic symptoms. Eleven of 19 patients with scoliosis (58%) underwent fusion. Eight of 19 (42%) patients have not undergone fusion: 3 have experienced progress, 1 remains in a stable condition, and 4 have experienced improvement of curvature since undergoing decompression. The mean age of patients who experienced progress after decompression was 14.5 years, compared to 6 years for patients who experienced improvement.

Conclusion. Early decompression of Chiari I malformation with syringomyelia and scoliosis resulted in improvement or stabilization of the spinal deformity in 5 cases. Each of these patients underwent decompression before 8 years of age and before the curve was severe. However, this series represents a few patients demonstrating this trend, and further follow-up and investigation are warranted. [Key words: Chiari I malformation, scoliosis, syringomyelia, spinal fusion, decompression] **Spine 2002; 27:1451–1455**

Scoliosis has been defined as a 10° lateral curve as measured by Cobb angles with vertebral rotation on a standing upright radiograph of the spine.[18] Idiopathic scoliosis is a common disorder affecting 2–4% of children aged 10–16 years.[5,6,19,39] In curves less than 20°, the incidence is about equal in boys and girls. However, in curves greater than 20°, the ratio is 4–5 girls to 1 boy.[25] Despite the overwhelming prevalence of idiopathic scoliosis diagnosed through screening, studies of the natural history indicate that fewer than 10% of persons with positive screening results require active treatment.[5,21,24,33,40]

Scoliosis patients are often referred to orthopedic surgeons for evaluation and treatment. Organic causes of the scoliosis such as congenital vertebral anomaly, spinal cord tumor, Chiari malformation, or syringomyelia must be ruled out in each case. The incidence of spinal cord or brainstem anomalies in the patient with "idiopathic" scoliosis ranges from 4 to 58%.[1,3,4,20,27,36] These lesions need to be addressed before treatment of the scoliosis is undertaken, to prevent potential neurologic complications.

The Chiari malformation was first described in 1891 by Chiari. It is defined as cerebellar tonsil extension below the foramen magnum. Since Chiari's first description, variations have been described by Arnold and others to include the associated lumbar myelomeningocele and occipital-cervical encephalocele. The mechanism of syringomyelia formation with Chiari malformations is not clearly understood. Two theories of origin exist: the water-hammer theory and the one-way-valve theory. In the water-hammer theory, arterial pulses from the choroid plexus are thought to be transmitted down through an abnormal forth ventricle to the cord, "hammering" out a dilatation in the cord. In the one-way-valve theory, the abnormal communication between the fourth ventricle and the cord causes an unequal pressure gradient with Valsalva maneuver, resulting in high pressures in the spinal column that cause the resulting dilatation.[8–10,16,17,23,28]

The association between pediatric Chiari malformations and the development of scoliosis in children with myelodysplasia (spina bifida) has been well documented.[13–15,30,32,34,35,37] Several studies have suggested a causal relation between syringomyelia and scoliosis not associated with myelodysplasia.[2,9,10,16,28,29] The cause of the scoliosis is thought to be a result of abnormal intramedullary pressure in the spinal cord causing interference with the postural tonic reflexes. The incidence of scoliosis in patients with Chiari malformation and syringomyelia is higher in the pediatric population. Eight-two percent of patients younger than 20 years with syringomyelia have scoliosis, but only 16% of patients older

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than 20 years.[16] Treatment of the Chiari malformation with syringomyelia is by surgical decompression of the cerebellar tonsils and/or by shunting of the syringomyelia. These treatments have been shown to stabilize or improve the neurologic symptoms associated with Chiari malformation and syringomyelia.[9,11,12,29] With this in mind, early decompression of the Chiari I and syringomyelia should halt the damage to the spinal cord and ultimately the progression of scoliosis.

The purpose of this study was to determine whether early decompression of Chiari malformation with syringomyelia results in stabilization or improvement of the associated scoliosis, ultimately saving the patient from the need for spinal fusion. Attempts at determining the long-term outcome have been made, but small patient numbers and varying outcomes have left the results inconclusive.[10,12,26,38] Does the age of the patient at decompression of the Chiari malformation affect the outcome of the scoliosis? Does the severity of the curve at decompression also affect the outcome of the scoliosis?

■ Materials and Methods

This was a retrospective review of patients with Chiari I malformation and syringomyelia with or without scoliosis over the past 20 years at The Children's Hospital in Denver, Colorado. International Classification of Diseases-9 codes and surgeons' logs were reviewed. A total of 25 patients with Chiari I malformation and syringomyelia who underwent surgical decompression by a neurosurgeon at our institution were identified; 19 of them had associated scoliosis. Patients with myelomeningocele, tethered cord, lipoma, and other spinal cord or brain anomalies were excluded. Charts and radiographs were reviewed. Scoliotic curves were measured using the Cobb method, and the curve pattern and severity was noted. Curve severity was defined as follows: mild, 10–25°; moderate, 26–40°; and severe, greater than 41°. Gender, age at diagnosis, presenting symptoms, and Risser sign were also noted.

■ Results

Twenty-five patients from 1979 to 1999 were identified. There were 12 male and 13 female patients. Their ages at presentation ranged from 19 months to 16.5 years (mean 8.67 years). The follow-up times ranged from 12 to 240 months (mean 62 months). Nineteen of the patients had scoliosis associated with their Chiari I malformation and syringomyelia. In 13 of 19 patients with scoliosis and Chiari I malformation with syringomyelia, the initial sign/symptom was the spinal deformity. Three of the 19 patients had back pain as a presenting symptom. One patient presented with leg pain, one with foot pain, and one with a neurologic abnormality consisting of asymmetric sweating in the upper extremity.

Complete radiographs for 17 of the 19 patients with scoliosis were found. In those 17 patients, the curve patterns consisted of 9 right thoracic curves, 3 left thoracic curves, 2 right thoracolumbar curves, and 3 left thoracolumbar curves. The curves at presentation, as measured by Cobb angles, were categorized into groups according to severity: 3 mild, 8 moderate, and 6 severe.

Table 1. Patients Without Fusions

Presenting Curve	Follow-up Curve	Age of Decompression	Most Recent Follow-up (y postdecompression)
80	112*	11.25	5
38	45	10.5	4.5
28	32	8.5	6.0
23	24	7	5.0
49	42	7	4.0
37	24	6.33	6.5
33	18	6.5	3.5
37	13	4	4.0

* This patient refused spinal fusion.
These are the patients who have not undergone fusion at the conclusion of the study listed in decreasing age. Note that the patients under age 8 have stabilized or improved their curves after decompression of their Chiari malformation.

Ten patients underwent spine fusion before or immediately after the diagnosis of syrinx or Chiari malformation. In 1 patient, a severe postlaminectomy kyphosis developed subsequent to syrinx drainage that required spinal fusion. As a result, in these 11 patients we were unable to determine the effect of the decompression on their scoliosis.

To date, 8 of the 19 (42%) patients have not undergone fusion. One of these patients presented with a severe curve of 80° at 11 years and 3 months of age. Her Chiari malformation has been decompressed but she has continued to experience progress over the past 2 years to 112°. The patient and her family have not been interested in spinal fusion thus far. Two other patients have experienced progress after undergoing decompression. One patient's curve remains in the moderate range (28–32°), and 1 has progressed to severe (38–45°). One patient has remained the same for 6 years after decompression, with a curve of 24°. The remaining 4 patients have all experienced improvement since undergoing decompression. All but 1 had curves that have gone from moderate to mild. One patient's curve started severe (49°) and remained severe (42°). Postdecompression follow-up times for these patients ranged from 3 to 6.5 years with an average of 4.8 years. Table 1 outlines the data on this group of patients.

The average age of the patients in the study was 8.67 years. In patients who experienced progress after decompression of the Chiari malformations, the average age was 14.5 years, whereas the average age for the patients who experienced improvement after decompression was 6 years.

Illustrative Cases

Case 1. A 4-year-old girl was brought to the orthopedic surgeon because of a noticeable deformity of her back. She was not noted to have any neurologic abnormalities. Because of her young age and her moderate right thoracic curve of 37° without any apparent cause, magnetic resonance imaging was performed and demonstrated a

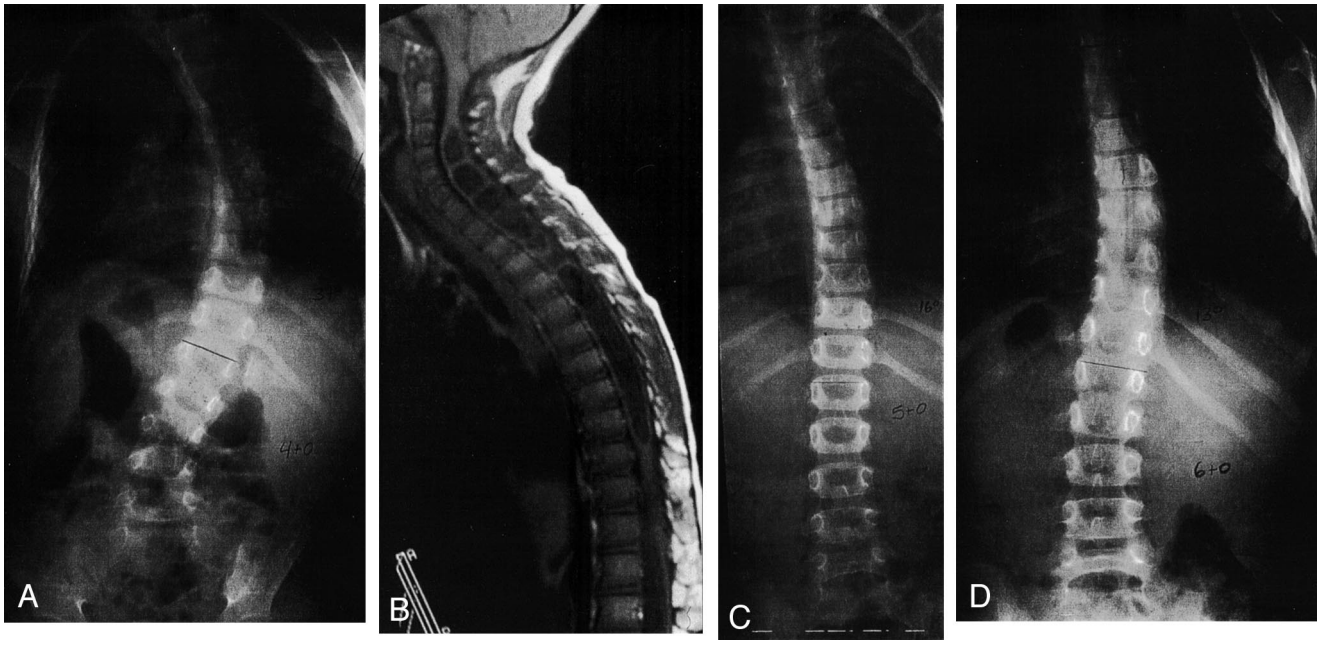

Figure 1. **A,** Posteroanterior radiograph of a 37° right thoracolumbar curve and no neurologic abnormalities observed in a 4-year-old girl. **B,** Coronal magnetic resonance imaging scan of the spine showing the Chiari I malformation and the extensive syringomyelia in the cord. **C,** Posteroanterior radiograph 1 year after decompression of the Chiari malformation, showing a curve of 16°. **D,** Posteroanterior radiograph 2 years after decompression, showing a curve of 13°.

Chiari malformation with associated syringomyelia of nearly the entire cord. She underwent hindbrain decompression by a neurosurgeon shortly after presentation. Her curve has improved over 4 years to 13° (Figure 1).

Case 2. A 6-year-old girl was brought to the orthopedic surgeon with a noted scoliosis. No apparent abnormalities were observed on physical examination except for her right thoracolumbar scoliosis at a young age. Magnetic resonance imaging demonstrated a Chiari malformation with associated syringomyelia. She underwent hindbrain decompression by a neurosurgeon and was treated in a brace. Her curve improved dramatically over 1 year from 33° to 18°. Her curve continues to remain stable at 18°, 3.5 years later (Figure 2).

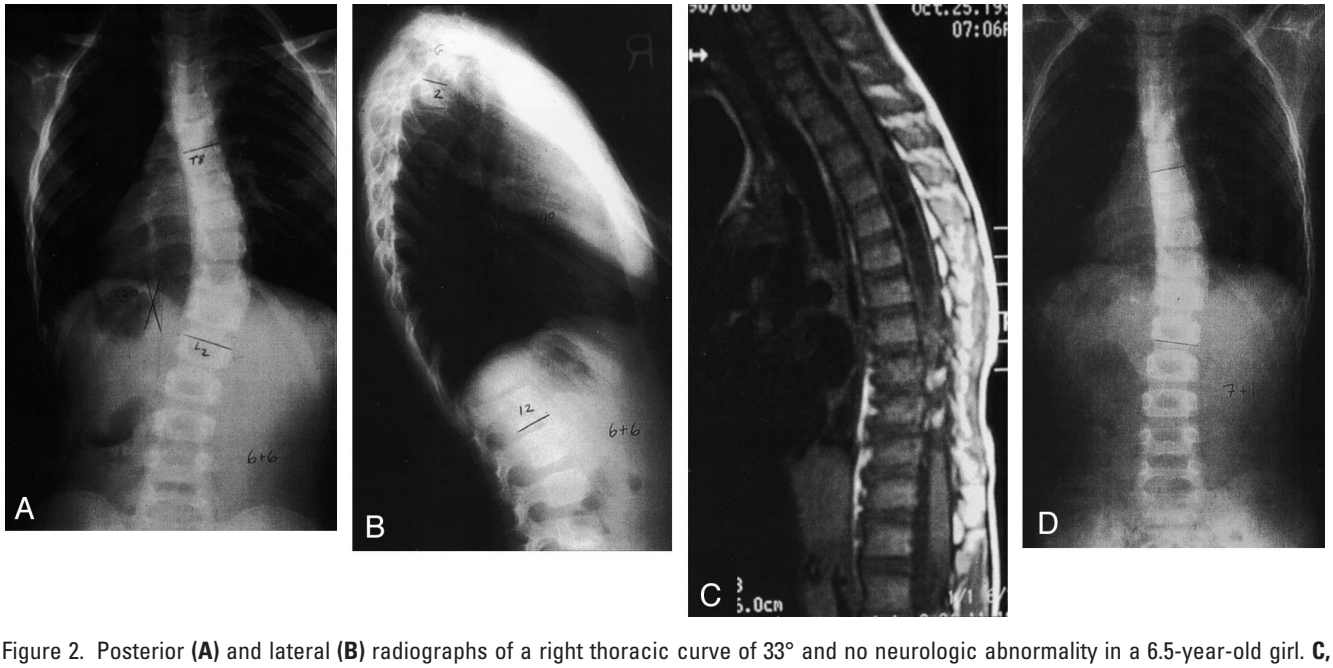

Figure 2. Posterior **(A)** and lateral **(B)** radiographs of a right thoracic curve of 33° and no neurologic abnormality in a 6.5-year-old girl. **C,** Coronal magnetic resonance imaging scan of the spine showing the syringomyelia in the cord. **D,** Posteroanterior radiograph 7 months after decompression of Chiari malformation, showing a curve of 18°.

■ Discussion

The association between Chiari malformation (exclusive of myelodysplasia) and scoliosis has been recognized but not completely understood.[8–10,23,42] Multiple theories have been proposed to explain this association. Huebert and MacKinnon theorized that the syringomyelia compromises cells in the cord that are responsible for muscle balance of the trunk, leading to the scoliosis. Decompression of the Chiari malformation with syringomyelia has been shown to halt or improve neurologic abnormalities associated with this condition.[9,11,17,29] One can then theorize that with improvement of the neurologic condition, the scoliosis could improve, provided decompression is performed early enough before permanent structural changes occur in the spine. The incidence of scoliosis in patients with Chiari malformation and syringomyelia is higher in the pediatric population: 82% of patients younger than 20 years with syringomyelia have scoliosis, and only 16% of patients older than 20 years.[16] In our study, we found a similar incidence of scoliosis (76%) in pediatric patients with Chiari malformation and syringomyelia.

Scoliosis in patients with Chiari malformation and syringomyelia can occur at very young ages and can rapidly progress, necessitating early diagnosis and treatment to prevent the need for spinal fusion. In this study, only 1 patient under 8 years of age with a curve less than 40° experienced progression of the curve enough to require fusion after being decompressed. All other patients who were treated before their curves were severe and before the age of 8 years have experienced improvement or stabilization. Moreover, 1 patient with a severe curve has experienced improvement of her scoliosis after decompression of the Chiari malformation.

The left thoracic curve has been implicated to be associated with underlying causes in scoliosis.[7,41] The incidence of left thoracic curves in this study was only 18% (3 of 17) of the patients. The more common right thoracic curve was present in 53% (9 of 17) of the patients. This contradicts the reports in the literature of 36–50% brain stem/cord abnormalities.[1,17,26,38] The average age of the patients in this study was 8.67 years, putting these children in the juvenile "idiopathic scoliosis" age group. With this in mind, patients presumed to have "normal" idiopathic scoliosis should be heavily scrutinized, and the threshold for further evaluation should be lowered, to catch these abnormalities early in the hope of preventing progression of the curve and ultimate fusion of the spine.

In summary, early decompression of Chiari malformation with syringomyelia showed improvement of the associated scoliosis in 4 cases. In each of these cases, the decompression was performed before the patient was 8 years old and before the curve was severe in all but 1 of the patients. Our current postdecompression follow-up times range from 3 to 6.5 years (average 4.8 years). Obviously, these patients will need to be monitored until they reach skeletal maturity, at least, to enable concrete conclusions about their scoliosis to be drawn. However, we believe that decompression at a young age in patients with smaller curves will give the best opportunity to delay or diminish the chances of worsening scoliosis. Therefore, the authors strongly suggest magnetic resonance imaging in all patients with juvenile scoliosis when treatment such as bracing or fusion is being considered, especially associated with a rapidly progressive curve or neurologic abnormalities.[22,31]

■ Key Points

- Chiari I malformation is associated with scoliosis.
- Scoliosis can improve after decompression of Chiari I malformation.
- Early decompression appears to have more favorable results on scoliosis.

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