Scoliosis in pediatric Chiari malformations without myelodysplasia

MICHAEL G. MUHONEN, M.D., ARNOLD H. MENEZES, M.D., PAUL D. SAWIN, M.D., and STUART L. WEINSTEIN, M.D.

Division of Neurosurgery and Department of Orthopedic Surgery, The University of Iowa College of Medicine, Iowa City, Iowa

A prospective study was undertaken in 1985 to better understand how the surgical manipulation of hindbrain herniation affected abnormal spinal curvature. Eleven patients under 16 years of age with Chiari malformation (not associated with myelodysplasia) and scoliosis of at least 15° were studied. The mean curve angle at the time of original treatment was 29°, with the convexity to the right in seven patients. The curvature was rapidly progressing in four patients. The most common presenting signs were myelopathy and weakness. Investigative procedures included spine radiographs with the patient standing and magnetic resonance (MR) imaging of the brain, spinal cord, and craniovertebral junction. Eight children had associated hydroxyringomyelia.

Surgical intervention consisted of a dorsal posterior fossa decompression in all patients and a transoral ventral decompression of the cervicomedullary junction in five. All patients were followed at 3, 6, and 12 months, and at yearly intervals thereafter with clinical evaluations, spine radiographs in the standing position, and postoperative MR imaging. The mean follow-up period was 35 months. The scoliosis improved in eight patients, stabilized in one, and progressed in two. Only one child required postoperative spinal fusion and instrumentation for progression of scoliosis. Hematomyelia or hematomyelia was associated with persistent scoliosis in two patients. The presence of hydroxyringomyelia and bone erosion did not preclude curve improvement. All patients under 10 years of age had resolution of their scoliosis, despite preoperative curves of more than 40°. These findings emphasize the importance of early surgical intervention, with the restoration of normal cerebrospinal fluid dynamics at the craniovertebral junction in children with symptomatic Chiari malformations.

Key Words • Chiari malformation • hematomyelia • hydroxyringomyelia • scoliosis • children

The association between pediatric Chiari malformations and the development of scoliosis has been well documented in the spina bifida population. Scoliosis occurs in 50% to 70% of these patients, and is therefore a common cause of functional decline. The scoliosis is thought to be secondary to a generalized paresis of the trunk musculature, to congenital structural changes in the vertebrae, and to the effects of abnormal intramedullary pressure with interference of the postural tonic reflexes. The association between scoliosis and hindbrain herniation without myelodysplasia has not previously been analyzed and followed after surgical therapy of the Chiari malformation.

A prospective study was undertaken in the pediatric population beginning in 1985 in an attempt to better understand the relationship between the development of scoliosis and Chiari malformations not associated with myelodysplasia. Patients with myelomeningocele, tethered spinal cord, lipoma, or diastematomyelia were excluded. Eleven children were assessed: they were all aged 2 to 15 years, had scoliosis of at least 15°, and had undergone surgical intervention for symptomatic Chiari malformations without myelodysplasia between the years 1985 and 1989. The purpose of this study was to analyze the progression of the scoliosis before and after surgical manipulation of the craniovertebral junction compression. The patients were grouped to allow comparison of those who experienced postoperative resolution of their scoliosis with those who had scoliosis that worsened or stabilized but did not improve despite surgical intervention. This group analysis allowed us to study the relationship among age, duration of symptoms, associated signs, and progression of the scoliosis.
The ultimate goal was to use these data in the preoperative analysis of patients to predict which factors may contribute to progression of the spinal curvature.

Clinical Material and Methods

Case Material

Between 1985 and 1989, 16 children without myelodysplasia underwent surgical treatment for symptomatic hindbrain herniation at our institution. The surgical treatment was individualized, but a standard decision-making protocol was followed in each case. All operations were performed by one surgeon (A.H.M.). The diagnosis was established by preoperative magnetic resonance (MR) imaging of the brain, spinal cord, and craniovertebral junction and by spine radiographs with the patient standing. Of the 16 patients, 11 had a spinal curvature of at least 15° and these are the subject of this report.

Patient age ranged from 2 to 15 years (mean 10.1 years) at the time of surgery. The duration of symptoms ranged from 1 day to 5 years and tended to correlate with age. All of the three patients under 10 years of age had symptoms for 1 year or less (mean 8 months), while the mean duration of symptoms in patients 10 years of age or over was 2.5 years (Fig. 1, p = 0.04, Fisher test). Nine patients had Chiari I malformations, and two had Chiari II malformations. Five had basilar invagination in addition to hindbrain herniation. The presenting signs and symptoms were varied and included motor, sensory, and autonomic dysfunction. The most frequent presentations were progressive myelopathy (72%), weakness (54%), headache (45%), sensory loss (45%), and ataxia (27%).

The scoliosis ranged from 15° to 54°, with a mean of 29° at the time of presentation. The curvature was to the left in four patients and to the right in seven. Rapidly progressing scoliosis was present in four patients (defined as a progression of > 1°/month). The scoliosis was painful in three patients, and only one patient presented with an asymptomatic curvature. All of the curves involved a portion of the thoracic spine. Eight children had associated hydrosyringomyelia, diagnosed by MR imaging. A portion of the hydrosyringomyelic cavity was in the cervical region in these eight patients and the entire cord was involved in three. Hematobulbia and hematomyelia were each present in one patient.

Five patients had basilar invagination, resulting in ventral brain-stem compression. This high incidence of basilar invagination (five of 16 patients with Chiari malformation) does not reflect the true association be-

![Fig. 1. Graph showing patient age at the time of surgery and duration of preoperative symptoms in 11 children with Chiari malformation and scoliosis. The straight line is a simple linear regression, the slope being significantly greater than a line of slope = 0 (p = 0.04, Fisher test). The curvilinear lines represent the 95% confidence intervals.](image)

![Fig. 2. Posteroanterior thoracolumbar radiographs in Case 2. Left: Preoperative film showing levo-scoliosis of 40° with a compensatory lumbar curve to the right. Center: Three months after a posterior fossa decompression, the scoliosis has improved to 20°. Right: One year after surgery, the scoliosis has nearly resolved.](image)
Scoliosis with Chiari malformations

tween these two pathological states, but is more likely related to the referral pattern to The University of Iowa Hospitals and Clinics.

**Radiological Studies**

Neuroradiological investigations consisted of plain radiographs and MR images of the posterior fossa and the spinal cord. Due to the sensitivity and noninvasive nature of MR imaging, this technique was employed as the imaging study of choice to establish the diagnosis of Chiari malformation as well as to determine the presence and extent of associated hydrosyringomyelia. In addition to plain radiography and MR imaging, five patients with basilar invagination or other craniovertebral junction anomalies underwent pluridirectional polytomography of the craniocervical region.

The 11 patients were followed postoperatively with upright posteroanterior and lateral scoliosis survey radiographs at 3, 6, and 12 months, and annually thereafter. All patients underwent postoperative MR imaging. Scoliosis was measured using the Cobb technique as recommended by the Scoliosis Research Society. To derive this measurement, lines are drawn tangential to the superior endplate of the most rostral end vertebra and to the inferior endplate of the most caudal end vertebra. The end vertebrae are those that possess the greatest tilt from the horizontal plane. Perpendicular lines to these tangential lines construct the Cobb angle. There is a standard error of 2.2° to 3.0° when employing the Cobb method.

All radiographs were examined for evidence of bone abnormalities. Parameters evaluated included the presence or absence of spinal curvature, direction of curve (defined as the direction to which the convex portion of the curve projects on posteroanterior radiographs), segment of the spine involved by the curve, presence of compensatory curve, evidence of bone erosion, rotation of the curve, and presence of congenital bone abnormality. Spinal canal diameter, vertebral body diameter, and interpedicular distance were measured at the C-5 vertebral body level. This region was chosen because of the propensity of hydrosyringomyelia in this population to affect the cervical spine. In accordance with the guidelines of the Scoliosis Research Society, curves measuring less than 10° were not considered to be pathological and, thus, patients with this degree of curvature were excluded from the study.

**Operative Procedures**

An anatomical and physiological approach was used to determine the appropriate decompressive procedure. All of the children had lower brain-stem compression, resulting from dorsal neural impaction at the foramen magnum or from ventral compression of the craniovertebral junction. If ventral compression was present, the patient was placed in traction in an attempt to reduce the impingement on the brain stem. If the compression persisted, a transoral cervicomedullary decompression was performed before the dorsal decompression. All 11 patients underwent a dorsal decompression, which consisted of a posterior fossa craniectomy and dissection of arachnoid adhesions.

A right-angled blunt probe was used to assess the patency and width of the communication between the obex and central canal. If the connection was patent, as it was in six of the children, a plug of compressed muscle was fashioned and gently introduced into this opening to obliterate the communication. The fourth ventricle was shunted to the cervical subarachnoid space using a Silastic catheter with the distal end positioned ventral to the dentate ligament. The catheter was sutured at its midpoint to the overlying dura to prevent it from slipping out of position. A duraplasty was then performed, completing the decompressive procedure. Five patients with basilar invagination underwent transoral ventral decompressions. Of these, three underwent dorsal fusion subsequent to their transoral decompression for treatment of cervical instability, and all five had a posterior fossa decompression. The fusion was performed at the time of the posterior fossa decompression.

**Illustrative Cases**

**Case 2**

This 2-year-old girl presented with a 6-month history of nonspecific back pain and rapidly progressing thoracic levoscoliosis, as noted by her personal physician. At the time of referral to our institution, she had a long left thoracic curve, measuring 40° with minimal rotation, and a 26° compensatory lumbar curve to the right (Fig. 2 left), but she was otherwise neurologically intact. An MR image revealed a Chiari I malformation with cervicothoracic hydrosyringomyelia (Fig. 3). The pa-

---

**Fig. 3.** Case 2. Preoperative magnetic resonance T1-weighted image, midsagittal view, demonstrating a Chiari I malformation and cervicothoracic hydrosyringomyelia.
tient underwent a posterior fossa craniectomy, C-1 laminectomy, obex plugging, fourth ventricle-to-subarachnoid space shunt, and duraplasty. Postoperatively, her back pain resolved almost immediately. At her 3-month follow-up examination, her scoliosis had improved to 20° (Fig. 2 center), and by 12 months it was less than 10° (Fig. 2 right). She has remained without symptoms for 23 months after her decompression.

Case 3

This 5-year-old boy presented with a rapidly progressing thoracic dextroscoliosis, measuring 54° at the time of admission (Fig. 4 left). He had evidence of myelopathy on physical examination, with exaggerated reflexes and upgoing toes to plantar stimulation. An MR image revealed a Chiari I malformation and associated holocord-septated hydrosyringomyelia (Fig. 5 left). Cervical spine films revealed the anteroposterior diameter of the canal at C-5 to be 2.9 cm, whereas the corresponding vertebral body was only 1.5 cm (Fig. 5 right). After a posterior fossa decompression similar to that of Case 2, this patient had a dramatic resolution of his myelopathy by 3 months after the operation. His scoliosis had improved to 33° by this time (Fig. 4 center), and at 1 year had improved to 17° (Fig. 4 right). He has remained without symptoms for 2 years after his decompression.

Case 7

This 13-year-old boy presented with a 2-year history of headaches and several months of severe nausea and frequent vomiting. Clinical workup at this time revealed basilar invagination and a Chiari II malformation. After a posterior fossa decompression combined with a fourth ventricle-to-subarachnoid space shunt and duraplasty failed to relieve his symptoms, the patient was transferred to our institution. Spine radiographs revealed a Klippel-Feil deformity and a 15° right thoracic curve. An MR image showed persistent anterior indentation of the pons by an abnormal clivus-odontoid articulation and an adequate posterior fossa decompression (Fig. 6 right). Compared with the MR image taken before the

Fig. 3. Preoperative studies in Case 3. Left: Magnetic resonance T1-weighted image, midsagittal view, demonstrating a Chiari I malformation with a large, septated spinal cord cavity. Right: Lateral cervical spine x-ray film showing the increased canal-to-vertebral body diameter secondary to the pressure effects of the hydrosyringomyelia.

Fig. 4. Posteroanterior spinal radiographs in Case 3. Left: Preoperative film showing dextroscoliosis of 54°. Center: Three months after surgery, the scoliosis has improved to 33°. Right: One year after surgery, the spinal curvature is reduced to 17°.
dorsal decompression (Fig. 6 left), the subarachnoid space was increased behind the cervicomedullary junction. However, a persistent ventral pontomedullary compression existed secondary to the abnormal anterior craniovertebral bone complex. After a transoral resection of the anterior craniovertebral junction (odontoidectomy), the patient had an uneventful recovery with resolution of his preoperative symptoms. His scoliosis had resolved by the time of his 12-month follow-up visit.

Results

Clinical Outcome

There were no major surgical complications or deaths. The follow-up duration ranged from 20 to 68 months (mean 35 months). Seven patients had complete resolution of their preoperative symptoms, and four showed stabilization or improvement. All patients who did not achieve complete preoperative symptom resolution had presented with chronic symptoms lasting longer than 1 year. Three of those who stabilized exhibited muscle atrophy preoperatively and had persistent postoperative weakness. One patient with headaches and a sensory deficit suffered persistent headaches after decompression. Myelopathic signs resolved in five of the eight patients who had a preoperative myelopathy, including all who did not present with atrophy. Ataxia resolved in all three patients who presented with this complaint.

Improvement in Scoliosis

The scoliosis improved in eight patients, stabilized in one, and progressed in two. The mean spinal curvature at 1 year after the operation improved from 29° to 16°. The most dramatic improvement was noted in the two children under 7 years of age, where the mean curve improvement was 34° over a 1-year period. The improvement in scoliosis was seen as early as 3 months after surgery. The mean Cobb angles of all patients were calculated, grouped by age, and plotted in relation to time after surgery (Fig. 7). Despite the small sample size, several trends were noted. Children under 10 years of age at the time of the operation presented with a greater degree of scoliosis than those over 10 years of age. However, the younger children showed a more rapid and more complete resolution of their scoliosis after surgery. These findings did not take into account the presence or absence of postoperative external bracing. This graphic improvement seen in the younger age group is largely because the three children who failed to improve were in the older age group.

Failure of Scoliosis to Resolve

Three patients failed to exhibit improvement in their scoliosis. One patient stabilized at 33° postoperatively and continued to wear an external orthotic brace. Because of the persistent rapid progression of the curvature in one child, despite an external orthotic brace, Harrington rods were placed 1 year after the initial

![Fig. 7. Graphic representation of the resolution of scoliosis in eight children age 10 or older and three children under 10 years of age after surgical therapy for a symptomatic Chiari malformation. N = number of patients.](image-url)
operation with subsequent improvement of the scoliosis from 55° to 15°. This patient had hematobulbia as documented on preoperative MR imaging. A second patient also exhibited progression of his curvature, from 24° to 33° over 1 year. During his decompressive surgery, a resolving clot was removed from the hydrodysmolytic cavity. On postoperative MR imaging, hemosiderin changes persisted in the walls of the collapsed cavities in both cases.

All of the patients who exhibited no improvement in their scoliosis were 14 years of age or older (mean 14.6 years). Although these patients tended to be older than those who showed improvement in their scoliosis (mean 9.9 years), age was only slightly statistically significant (p = 0.06, Fisher two-tailed test) due to the small sample size.

Although both patients who had postoperative progression of their scoliosis had pedicle erosion, the presence of bone erosion did not preclude resolution of the scoliosis. Similar bone erosion was present in two of the patients who improved postoperatively. The mean anteroposterior diameter of the spinal canal at the level of the C-5 vertebral body was 23 mm in those with hydrodysmolytic myelomes 15 mm in those without. The vertebral body:canal diameter ratio (measured at C-5) was smaller in those patients with hydrodysmolytic myelomes (0.42:1) compared with those without (1.1:1). This is indicative of the pressure effects of the cavity on the spinal canal and eventually the vertebrae. Postoperative MR imaging studies revealed that the hydrodysmolytic myelomes had resolved in seven of the eight patients.

Discussion

Historical Perspective

The association between Chiari malformations (exclusive of myelodysplasia) and scoliosis has been recognized but not completely understood, and the long-term outcome of these curvatures after treatment of the hindbrain herniation has not previously been reported. The development of scoliosis associated with a Chiari malformation typically has been ascribed to the presence of hydromyelia. The incidence is higher in patients presenting before skeletal maturity and is between 20% and 80% overall. In our series, the overall incidence of Chiari malformation and scoliosis was 73% whereas, in the three patients under 10 years of age, the incidence was 100%. The presence of hydromyelia did not correlate with failure of the scoliosis to resolve postoperatively. This is in agreement with the findings of Carmel, who noted that these children have the capacity to reverse the deficits related to severe syringomyelia and to reduce the degree of scoliosis. There was, however, a correlation between the presence of a hydromyelic cavity and the spinal canal diameter. This confirms the progressive, destructive nature of hydromyelia. The bone erosion that resulted did not significantly impair resolution of the scoliosis.

Etiology of Hydrosyringomyelia

All of the current etiological theories of syringomyelia place emphasis on pathological cerebrospinal fluid (CSF) hydrodynamics. Williams proposed that it develops from a craniospinal fluid pressure dissociation, with enlargement of the cavity as the result of a valve-like mechanism produced by anomalies around the foramen magnum. Transient venous pressure changes within the spine and skull could then result. Gardner thought that imperforation of the foramina of Luschka and Magendie allows CSF to flow into and dilate the central canal. The atresia of the foramina also contributes to hydrocephalus with subsequent cerebellar herniation. This theory does not explain the lack of patency between the fourth ventricle and the central canal (absent in 20% of the University of Iowa series reported in 1990), and it does not easily account for spinal cord cysts associated with tumors and trauma. Ball and Dayan proposed that an obstruction at the craniovertebral junction increases the CSF pressure in the subarachnoid space, forcing CSF to track along the perivascular Virchow-Robin spaces into the substance of the spinal cord. As these small pools of CSF coalesce, a syrinx forms, initially independent of the central canal but eventually connecting with it.

Neurogenic Scoliosis

Multiple theories have been proposed to explain the association of neurogenic scoliosis with hydromyelia and the Chiari malformation. In the presence of hydromyelia, the cord does not funnel in a normal fashion. Huebert and MacKinnon theorized that the cysts of the syrinx compromise the medial nuclear groups of cells innervating the muscles of the trunk. As the lower motor neurons in the gray matter of the anterior horns are progressively affected, scoliosis ensues. Liszka showed that division of the posterior nerve roots produced a greater scoliosis than if the anterior nerve roots alone were divided. Wyatt et al. found that patients with idiopathic scoliosis have pathological vibratory responses because of malfunction of the posterior column pathways. These patients have an equilibrium dysfunction traceable to the brain stem, suggesting that the curve may develop secondary to a disturbance of the postural reflex system. This would partially explain the scoliosis that was present in three patients in our series who had a Chiari malformation without hydromyelia.

Duration of Symptoms vs. Outcome

The duration of symptoms associated with pediatric Chiari malformations has been reported to correlate with the long-term outcome. The age-related findings in our study show that patients 10 years of age and over tended to have a longer chronic duration of symptoms. All patients under 10 years of age were asymptomatic after an average of 2.6 years of follow-up study; they were treated early in the course of their disease before permanent neurological damage had occurred.
Scoliosis with Chiari malformations

The two patients who exhibited progression of scoliosis and the one who stabilized but did not improve were all over 10 years of age. This would support the concept that there is a gradual progression of disease with this condition.2,7,23,35

Hematomyelia and Hematobulbia

Hematomyelia and hematobulbia were each present in one patient in this series and were associated with persistent scoliosis despite appropriate surgical management. In both cases, the hemorrhage appeared to have entered an already existing chronic hydromyelic cavity. Typically, spinal cord edema follows the intramedullary blood clot. The blood can dissect longitudinally for several segments rostral and caudal to the hemorrhage site, most significantly affecting the gray matter and adjacent white matter. Clinical improvement often accompanies the resolution of the hematoma, but the ultimate neurological outcome is difficult to predict and is probably directly related to the degree of initial insult as well as to the extent of reactive gliosis invoked. In this series, the presence of hematomyelia or hematobulbia was a poor prognostic sign for the resolution of scoliosis, but it was not otherwise associated with further progression of symptoms.

Surgical Management

The first procedure in the management of scoliosis associated with a Chiari malformation is the application of a spinal orthotic brace, if the scoliosis is shown to be progressive and detrimental. If hydrocephalus is present, the initial surgical management should be the placement of a ventriculoperitoneal shunt.11,25,28 None of the patients in this series exhibited hydrocephalus, although it was present in two (12%) of the 16 children we reviewed before 1985.11 Subsequent intervention should be directed at the offending compressive abnormality. When there is ventral compression, a transoral clivus odontoid resection is indicated.13,25,26,39 Case 7 exemplifies the importance of treating the ventral pathology before undertaking the dorsal decompression (see Fig. 8). When ventral compression is not present, decompression of the posterior fossa is performed. If a hydrosyringomyelic syndrome persists and the cavity is still present on MR images, a syringosubarachnoid shunt is placed. If, after the restoration of normal CSF dynamics, the patient continues to exhibit progression of the scoliosis despite a spinal orthotic brace, surgical correction of the scoliosis should be attempted. Only one of our patients required a spinal fusion and instrumentation for the treatment of curvature progression (from 44° to 55° over 12 months).

Resolution of the scoliosis was not limited to those undergoing only a posterior fossa decompression. It was also seen in the five patients who had a ventrally directed procedure. Although three of these five later underwent posterior cervical fusion and all five received posterior fossa decompression, some of their symptoms were relieved immediately after a transoral decompression. Two explanations for this improvement are offered. The first is that the transoral procedure relieves the brain-stem compression, allowing return to normal brain-stem function.11,25 The second possibility is that CSF dynamics return to normal after a ventral decompression. Abnormal CSF dynamics due to cerebellar impaction and arachnoid adhesions have been suggested, based on clinical and experimental observation.42,44 Figure 8 left depicts the abnormal CSF circulation that results when a Chiari malformation and basilar invagination are present. After an adequate ventral and posterior fossa decompression, brain-stem compression is relieved and physiological CSF circulation is restored (Fig. 8 right); Williams48 was one of the first to emphasize the re-establishment of CSF pressure/flow relationships at the lower hindbrain as the primary goal of the operation.

Idiopathic vs. Neurogenic Scoliosis

It is critical to differentiate idiopathic scoliosis from neurogenic scoliosis secondary to a Chiari malformation, hydrosyringomyelia, or other causes of spinal cord dysfunction. Forceful surgical correction of scoliosis in association with syringomyelia, without treating the underlying pathology, is potentially disastrous. Huebent and MacKinnon31 reported two patients treated surgically for scoliosis, one of whom became paraplegic after the operation. Norrwall and Wikkelso27 documented the case of a 15-year-old boy who developed spastic paraparesis and a myelographic block 10 days after fusion and placement of Harrington distraction rods for the treatment of a rapidly progressing curvature of 55°. Several clinical findings different from those ex-
pected in idiopathic scoliosis were the clue to the diagnosis of neurogenic scoliosis in our series. Four (36%) of the 11 children exhibited a left thoracic curvature pattern. This is in sharp contrast to the expected findings with idiopathic scoliosis. Coonrad reviewed the records of 1622 consecutive scoliosis patients, and found only 27 (1.7%) with left thoracic curve patterns (RW Coonrad, et al., unpublished data). Other atypical findings in our patients included painful curvatures (27%), rapidly progressing curvatures (36%), and a curvature appearing in the first 2 years of life, not associated with other congenital abnormalities. Other cases in which scoliosis progresses in spite of adequate bracing or after skeletal maturity also warrant a high index of suspicion.

Conclusions

The observations from this study are several-fold: 1) patients older than 10 years of age tended to have a longer duration of preoperative symptoms; 2) hematomyelia and hematobilubia were associated with persistent postdecompression scoliosis; 3) patients under 10 years of age exhibited a rapid extensive resolution of their scoliosis; 4) the preoperative presence of hydro-syringomyelia did not impair the postoperative resolution of the scoliosis; and 5) the canal diameter and vertebral body changes correlated with the presence of hydro-syringomyelia. These findings emphasize the importance of early surgical intervention in a child with a symptomatic Chiari malformation.

The significance of the observations from this study is limited due to the small sample size, but these findings can contribute to a better understanding of the pathophysiology of the events occurring at the craniovertebral junction and within the spine. Once the pathology is appropriately identified, surgery can be tailored to correct the abnormality.

References


M. G. Muhonen, et al.
Scoliosis with Chiari malformations


Manuscript received July 22, 1991.
Accepted in final form December 11, 1991.
Address reprint requests to: Arnold H. Menezes, M.D., Division of Neurosurgery, 42GH, The University of Iowa Hospitals and Clinics, Iowa City, Iowa 52242.