

SIGNIFICANT SCOLIOSIS REGRESSION FOLLOWING SYRINGOMYELIA DECOMPRESSION: CASE REPORT

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ABSTRACT

We present the case of a 5-year-old boy presenting with a 54-degree scoliosis secondary to a Chiari I malformation with a holocord syringomyelia extending from C1 to T10. Neurosurgical treatment involved posterior fossa craniectomy with decompression, and partial C1 laminectomy. At follow-up 7 years later, at age 12, radiographs revealed only a 4-degree scoliosis, and follow-up MRI revealed a deflated syrinx. We report this case to reveal the most significant scoliosis regression seen in our experience that may occur in younger patients after neurosurgical syringomyelia decompression for Chiari I hindbrain herniation.

CASE REPORT

A five-and-a-half year-old Caucasian boy presented to his pediatrician with rounding of his shoulders and scoliosis noticed over the course of one month. A pediatric orthopaedic surgeon (S.L.W.) evaluation revealed abnormal neurologic exam findings, including a diminished gag reflex, and 3+ knee hyperreflexia. Radiographs demonstrated a 54-degree right thoracic scoliosis (Figure 1), and a 42-degree thoracic kyphosis. Given the rapid onset scoliosis and kyphosis, and the abnormal neurologic exam, an MRI was indicated to evaluate for primary central nervous system pathology. MRI identified a Chiari I malformation with a holocord syringomyelia extending from C1 to T10 (Figure 2).

Successful neurosurgical treatment (A.H.M.) involved posterior fossa decompression via craniectomy and partial C1 laminectomy (Figure 3A). The superior two-thirds of the C1 lamina were removed. Intraoperatively, the cerebellar tonsils were found to be shrunken

and medially approximated, and had descended below the C1 lamina (Figure 3A). A pathologic "veil" was occluding the egress of cerebrospinal fluid (CSF) from the fourth ventricle to the subarachnoid space (Figure 3B). The "veil" was surgically opened to restore CSF outflow from the fourth ventricle (Figure 3C). A fascia duraplasty was then performed.

The patient gradually returned to all activities. The scoliosis regressed. Six month post-operative radiographs showed a scoliosis decrease to 33 degrees; and at 7 years after surgery, a further decrease to 4 degrees (Figure 4). After 9 years, the curve clinically increased slightly, likely related to pubertal growth, but it remained stable thereafter, with no orthopaedic intervention indicated. Follow-up MRI showed a deflated stable syrinx. At age 17, he had no deformity or back pain, and was the starting running back for his high school varsity football team.

DISCUSSION

A series of very young children with Chiari I malformation with hindbrain herniation without myelodysplasia has been reported from our institution.¹ In this report, 31 patients were seen at the University of Iowa from 1985 to 2000. Most patients presented similar to our patient with oropharyngeal impairment and scoliosis. In the above reported series, patients presented most commonly with a chief complaint of impaired oropharyngeal function (35%), scoliosis (23%), headache or neck pain (23%), sensory disturbance (6%), and weakness (3%). Patients were treated surgically with posterior fossa decompression, duraplasty, and cerebellar tonsillar shrinkage, as described in our patient. Syringomyelia improved in all patients, as in our case. Scoliosis resolved in 2 of 8 patients, improved in 5, and stabilized in 1. However, our patient in this case report represents a substantial scoliosis resolution, from 55 degrees to 4 degrees. This is in our experience the largest secondary scoliosis regression that we have seen after neurosurgical decompression.

Age was recently identified by Brockmeyer et al.² to correlate with scoliosis improvement in children with Chiari I malformations and scoliosis treated with posterior fossa decompression. Thirteen patients of the 21 study patients (62%) had curve improvement or stabilization following neurosurgical treatment. Eight of 21

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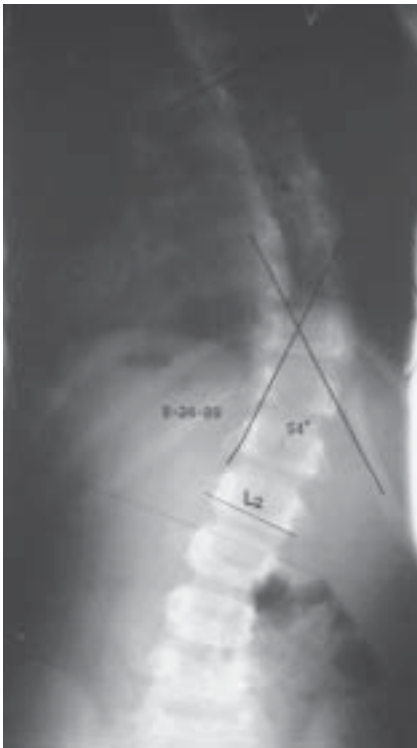


Figure 1. Scoliosis at presentation with a 54-degree Cobb angle.

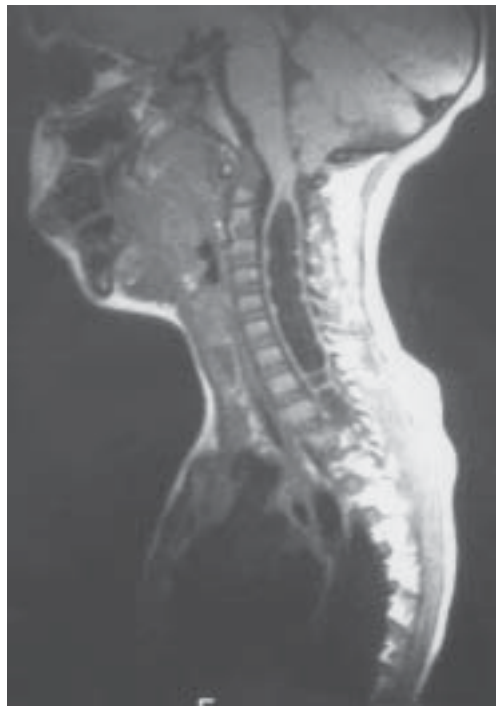


Figure 2. Sagittal MRI revealing holocord Chiari I syringomyelia.

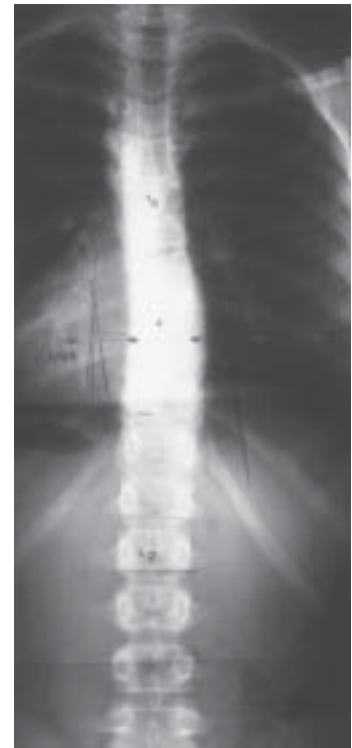


Figure 4. Scoliosis 7 years post-operatively with a 4-degree Cobb angle.

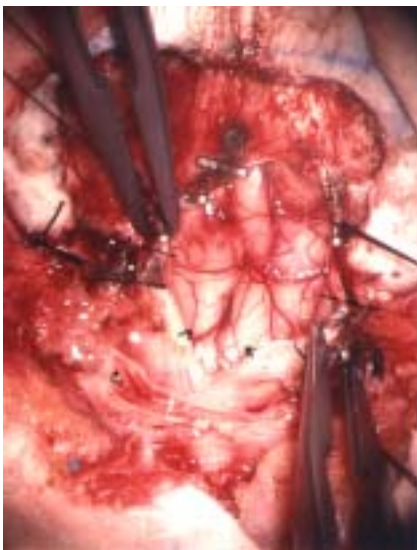


Figure 3. Posterior fossa craniectomy with removal of superior two-thirds of C1 lamina. Interrupted dotted line (3A) represents foramen magnum plane. Arrows (3A) point to herniated medially approximated cerebellar tonsils. Arrows (3B) point to a pathologic "veil" covering the fourth ventricle obstructing CSF outflow. Label V4 refers to fourth ventricle (3C) after the "pathologic" veil has been opened to allow egress and to restore normal CSF dynamics.

patients (38%) had curve progression. Specifically, 10 of 11 patients (91%) who were less than 10 years of age at the time of neurosurgical decompression have had their curves improve or stay the same during follow-

up. In contrast, 5 of 7 female patients (72%) older than 10 years old with a curve greater than 40 degrees before neurosurgical decompression have either been fused or are awaiting fusion.

In a prior series by Mulhonen et al. from Iowa City,³ all Chiari I patients with scoliosis who were under 10 years of age had resolution of their scoliosis, after neurosurgical hindbrain decompression, despite preoperative curves of more than 40 degrees.

The literature, and our case report emphasizes the importance of early diagnosis of Chiari I malformation with syringomyelia in the very young child with scoliosis. The physical exam finding of an abnormal gag reflex is critical. Our case reveals that such young children who undergo surgical manipulation for hindbrain herniation result in improvement of syringomyelia, and also result in improvement of the secondary scoliosis, which in our case was a significant correction of 41 degrees.

REFERENCES

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