

Key Points

1. Research on Chiari related scoliosis has identified what types of scoliosis are likely to be related to Chiari and that decompression surgery alone is sometimes enough to halt curve progression
2. However outcome studies have produced wide ranging results and it is not clear why some scoliosis cases improve while others don't
3. Study from Johns Hopkins reviewed 21 pediatric, Chiari related scoliosis cases to look for predictors of success and failure post-operatively
4. Overall 38% of the children experienced improvement in their scoliosis following decompression surgery while 48% continued to get worse
5. Found that a thoracolumbar curve and failure of syrinx to resolve were both associated with an increased likelihood of curve progression
6. Also found that every degree of Cobb angle was associated with an increased chance of curve progression
7. Did not find any connection between any presenting symptoms, patient demographics, or herniation size and curve progression

Definitions

cervical - upper region of the spine; neck area

Cobb angle - measurement used to determine severity of scoliosis, in degrees

comorbidity - an additional disease or health problem

duraplasty - surgical technique where a patch is sewn into the dura (the covering of the brain) to create more room

lumbar - lower part of the spine

scoliosis - abnormal curvature of the spine

How Decompression Surgery Affects Scoliosis Progression

May 31, 2008 -- Scoliosis, an abnormal curvature of the spine, is found in as many as 30% of Chiari patients and 60% of syringomyelia patients. The curves can develop in different regions of the spine and be of varying severity (which is measured in degrees and known as the Cobb angle). One of the most troubling aspects of scoliosis is its tendency to progress, or get worse, which can lead to the need for corrective surgery involving braces, rods, and screws. In adults with Chiari, the presence of scoliosis has been shown to result in poorer long term outcomes.

Although the exact nature of the link between Chiari, syringomyelia, and scoliosis remains elusive, in recent years, a growing body of research has produced some significant advances in understanding and treatment. Specifically, models have been proposed as to what types of scoliosis warrant an MRI to check for Chiari and the standard of care has evolved such that Chiari decompression surgery is now performed before any type of scoliosis surgery in an attempt to halt curve progression. To date, such studies have reported a fairly wide range of outcomes using this approach, and it is not clear why scoliosis improves with decompression surgery in some cases, but not in others.

It is this precise question which a team of researchers from Johns Hopkins tried to address with a recent publication in the Journal of Neurosurgery: Pediatrics (Attenello et al.). Specifically, the researchers reviewed the records of children they had treated with Chiari related scoliosis between 1995-2005. Out of 258 total Chiari cases, they identified 21 children with significant scoliosis who had undergone decompression surgery and for whom follow-up information was available. Children who had had a planned fusion as part of their surgery were excluded from the review.

Seventeen of the children were female and only four were male. The average age of the group was 9 years old and every child had Chiari, scoliosis, and a syrinx. The most common location of the scoliosis curve was the thoracic region (48%, See Figure 1), followed by curves which started in the thoracic region but extended to the lumbar region as well (thoracolumbar, 33%). The average curve severity was 28 degrees. Each child underwent decompression surgery, although not all the surgeries were the same. There were variations involving whether a duraplasty was part of the procedure and whether the cerebellar tonsils themselves were burned back. The children were followed for an average of 39 months after surgery.

Figure 1: Scoliosis Curve Location (21 Patients)

Location	Percent
Cervical	5%
Thoracic	48%
Lumbar	14%
Thoracolumbar	33%

For each case, the researchers reviewed presenting symptoms, neurological deficits, demographic information, co-morbidities, radiographic images, operative reports, and clinical records. They were looking for predictors of which scoliosis cases improved after surgery and which ones continued to get worse, or progress. For the purposes of this research, they defined both improvement and progression as a change of at least 10 degrees in the Cobb angle. Similarly, syrinx improvement was defined as a reduction in size of at least 20%.

Overall, the scoliosis improved in 8 of the children (38%, see Figure 2), continued to progress in 10 (48%), and stabilized in 3 (14%). Three of the 10 children whose scoliosis continued to worsen eventually required spinal fusion surgery. When the researchers looked for predictors of the scoliosis outcome, they found that children whose syrinxes did not improve (shrink by at least 20%) were 4 times more likely to have scoliosis progression than children whose syrinxes did improve. While it has long been speculated that syrinxes are somehow involved in the development of scoliosis, the research on this is mixed. Several studies have failed to find a link between syrinx size or location and the presence or severity of scoliosis. And while at first glance the syrinx related result from this study suggests a causal link between the two, this is not necessarily the case. It could be that both syrinxes and scoliosis are due to a more fundamental underlying problem which in these cases was not resolved by the decompression surgery.

Figure 2: Scoliosis Curve Outcome After Decompression Surgery (21 Patients)

Outcome	Percent
Improved	38%

thoracolumbar - refers to a scoliosis curve which starts in the thoracic region but extends to the lumbar region

thoracic - middle part of the spine, chest area

cerebellar tonsils - portion of the cerebellum located at the bottom, so named because of their shape

cerebellum - part of the brain located at the bottom of the skull, near the opening to the spinal area; important for muscle control, movement, and balance

cerebrospinal fluid (CSF) - clear liquid in the brain and spinal cord, acts as a shock absorber

Chiari malformation I - condition where the cerebellar tonsils are displaced out of the skull area into the spinal area, causing compression of brain tissue and disruption of CSF flow

decompression surgery - general term used for any of several surgical techniques employed to create more space around a Chiari malformation and to relieve compression

syringomyelia - condition where a fluid filled cyst forms in the spinal cord

Source

Attenello FJ, McGirt MJ, Atiba A, Gathinji M, Dato G, Weingart J, Carson B, Jallo G. [Suboccipital decompression for Chiari malformation associated scoliosis: risk factors and time course of deformity progression.](#) J Neurosurg: Ped. 2008 June; 1(6).

No Change	14%
Progressed (Worse)	48%

Note: Improvement or progression, was defined as a change of at least 10 degrees in Cobb angle.

The researchers also found that children with thoracolumbar scoliosis curves were five times more likely to have their curves continue to progress after surgery than the other children. The authors did not speculate on possible reasons for this finding. Finally, the scientists also found, perhaps not surprisingly, that the more severe the scoliosis prior to surgery, the more likely it was to progress. This is in line with other research studies which found that severe curves were less likely to improve with decompression surgery. However, contrary to other research this study found no correlation between the age of the patients and the scoliosis outcome. Previous research has found that age can play a role in outcomes. Interestingly, presenting symptoms and the size of the tonsillar herniation were not at all related to the scoliosis outcome.

Figure 3: Factors Predicting Curve Progression After Surgery

Factor	Increased Likelihood of Progression
Thoracolumbar	5X
Failure of syrinx to resolve	4X
Cobb angle	1 degree = 11% increase

Note: symptoms, size of herniation, and age were not related to curve progression

While there have been a number of studies published on Chiari related scoliosis, their results do not always agree. This is likely a by-product of the type of research, namely retrospective studies involving just a few patients, which has inherent limitations. What is needed in this area is a structured theory on the link between Chiari, syringomyelia, and scoliosis from which predictions can be derived and tested through rigorous studies.

-- Rick Labuda

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