Key Points

1. Small studies have shown that decompression surgery may be effective in treating scoliosis related to CM/SM
2. Studied 21 pediatric patients with scoliosis, Chiari I, and syringomyelia (19 out of 21) and no spinal fusion
3. All patients underwent similar decompression surgery
4. The curve of 13 out of 21 patients (62%) was improved or unchanged over time
5. Males less than 10 years old and with curves less than 40 degrees at surgery were more likely to have their curves improve with decompression surgery

Definitions

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

Chiari malformation - 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

Cobb Angle - technique used to measure the severity of a spinal curve - in degrees - from spinal images

decompression surgery - common term for any of several variations of a surgical procedure to alleviate a Chiari malformation

duraplasty - surgical technique where a patch is sewn into the dura, the tough covering of the brain and spinal cord

fusion - surgical procedure where vertebra are joined together using bone grafts and often instruments such as rods, screws, etc.

laminectomy - surgical removal of

Chiari, Syringomyelia, Scoliosis & Surgery

It is well known that scoliosis - especially in children - is associated with Chiari and syringomyelia, and this publication has reported on several small studies which appeared to indicate that decompression surgery improves the abnormal spinal curve in many cases. Now, researchers from the University of Utah have provided additional evidence that decompression surgery is often an effective treatment for Chiari related scoliosis and have even begun to identify what type of cases may benefit most from the procedure.

Dr. Douglas Brockmeyer, a neurosurgeon at the University of Utah, and his orthopedic colleagues, reviewed the neurological, orthopedic, image, surgical, and medical records of 21 pediatric patients who initially sought treatment for scoliosis and were subsequently found to have a Chiari malformation. They published their results in the November 2003 issue of the journal Spine.

The researchers found their subjects out of a group of 85 pediatric patients who had undergone Chiari decompression surgery at some point over the past decade. To be included in the study, the children had to have first been evaluated for scoliosis, then identified to have a Chiari malformation, not be treated previously with any type of spinal fusion, and then undergone the decompression surgery (all the children underwent a similar procedure which included a laminectomy and duraplasty).

The children in the study ranged in age from 3 to 19 years and there were 14 girls and 7 boys. All had demonstrable Chiari malformations on MRI and 19 out of the 21 had a syrinx as well. After surgery, the children were followed medically for an average of 2.1 years using MRI to evaluate the decompression and the syrinx and plain X-rays - or radiographs - to evaluate the progression of the scoliosis. A method known as the Cobb angle was used to quantify the degree of scoliosis and a change in more than 5 degrees was defined as improvement (or worsening). A change less than this amount was considered insignificant and the status of the scoliosis defined as unchanged.

The doctors found that 13 of the 21 curves (62%) either improved or stabilized after the Chiari surgery and 8 curves (38%) got worse over time (see Table 1). Four out of the 21 required spinal fusion surgery in addition to the Chiari decompression and 3 more will probably undergo spinal fusion in the future. Interestingly, the MRI's revealed that all the syrinxes improved over time and there appeared to be no correlation between syrinx improvement and scoliosis improvement.

Table 1
Surgery Results (21 Patients)

Age	Sex	Initial Cobb Angle	Final Cobb Angle	Out come
4	M	42	9	Better
3	M	28	0	Better
13	F	56	39	Better
3	M	30	16	Better
5	F	42	28	Better
4	F	28	15	Better
4	M	28	20	Better
6	F	30	23	Better
10	M	29	24	Better
19	F	44	40	Same
15	F	12	8	Same
14	F	25	22	Same
4	F	42	46	Same
13	F	38	43	Worse
3	F	23	28	Worse
5	M	25	33	Worse
11	F	44	52	Worse Fusion
12	F	30	40	Worse Fusion
9	F	42	57	Worse Fusion

part (the bony arch) of one or more vertebrae

MRI - Magnetic Resonance Imaging; diagnostic device which uses a strong magnetic field to create images of the body's internal parts

radiograph - another name for an X-ray; diagnostic machine which uses radiation to create an internal image of the body

scoliosis - abnormal curve of the spine

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

syrinx - fluid filled cyst in the spinal cord

vertebra - segment of the spinal column

Source

Brockmeyer D et al. *Scoliosis Associated With Chiari I Malformations: The Effect of Suboccipital Decompression on Scoliosis Curve Progression.* Spine Nov 15 2003 28(22): 2505-09.

12	F	25	50	Worse
13	M	52	90	Worse Fusion

Note: *Cobb angles in degrees; Outcome refers to scoliosis progression*

In an attempt to identify characteristics associated with successful surgery, the researchers also looked at the age of the children and the amount of curve present before the decompression surgery. They found that an amazing 91% of the children under 10 (10 out of 11) either improved or stabilized after surgery. This stands in contrast with 5 of 7 girls (71%), older than 10 and with curves greater than 40 degrees, who worsened after surgery and have either had, or are waiting to have, spinal fusion to attempt to correct their scoliosis.

While the follow-up period is a little short to say definitively, this study supports previous research which has shown that the younger children are at the time of surgery, and the less severe their scoliosis is, the more likely decompression surgery is to help. It is also interesting that these researchers, like others, failed to correlate the syrinx size or progression directly with scoliosis.

Given that a young age seems critical in arresting what can be a devastating progression of spinal curvature associated with Chiari, it is important for doctors - and parents - to know when an MRI should be performed when a child develops scoliosis. Luckily for children and parents alike, there are doctors performing research to do just that; namely, establish guidelines and criteria for when an MRI is necessary. As research along both these lines - when an MRI is necessary and how to best treat Chiari related scoliosis - progresses, it will certainly improve the outcome for children with CM/SM and should provide some measure of hope and relief for their parents as well.