Key Points

1. The relationship between Chiari, SM and scoliosis has been studied in children, but has not been examined in adults
2. Japanese group looked at 27 CM/SM patients and compared those with and those without scoliosis across a number of parameters
3. Found that length of syrinx and duration of symptoms were longer among those with scoliosis
4. Also found that most patients with scoliosis suffered from upper body muscle atrophy, while those with no scoliosis did not
5. Also found that those with scoliosis had lower pre-op and post-op scores on a functional assessment
6. Interestingly, the width of the syrinx was not related to the presence of scoliosis; neither was the length of tonsillar herniation
7. In general, the presence of scoliosis resulted in poorer outcomes

Definitions

cervical - the upper part of the spine; the neck area

Cobb angle - measurement taken from an X-ray which quantifies the amount, or degree, of scoliosis

craniectomy - surgical technique where a piece of the skull is removed

dura - thick, outer layer covering the brain and spinal cord

duraplasty - surgical technique where a patch is sewn into the dura, thus making it bigger

laminectomy - surgical technique where part of one or more bony vertebra are removed

lumbar - the lower back area

Scoliosis Affects Surgical Outcomes In Adults

March 31, 2007 -- While the link between scoliosis, an abnormal curvature of the spine, and CM/SM has been studied in children, little work has been done to examine the role it plays in adult CM/SM patients. Pediatric scoliosis is actually one of the most active topics of Chiari research, and work in that area has resulted in the recognitions of what types of scoliosis warrant an MRI, and the understanding that decompression surgery should be tried before corrective, orthopedic surgery.

Despite the high level of attention, the fundamental link between scoliosis and CM/SM remains a mystery. Specifically, it is not known whether a syrinx leads directly to scoliosis by weakening the spine or back muscles, whether scoliosis somehow influences the development of a syrinx, or if both are secondary to the compression and disrupted CSF flow associated with Chiari. While it seems logical that a syrinx could lead to scoliosis, several studies have failed to find a connection between the size and location of a syrinx and the presence or severity of scoliosis.

Now, a recent study out of Japan, while not answering these questions conclusively, has shed some light on the role that scoliosis plays in the outcomes of adult CM/SM patients. The Japanese team, led by Dr. Atushi Ono, looked at 27 consecutive, adult CM/SM patients treated between 1995-2002. Although the criteria for inclusion in the study was those older than 20, the average age of the group was a much higher 55.

To study the role of scoliosis, the patients were divided into two groups based on whether they had 10 degrees or more of abnormal spine curvature. Using this method, 15 patients were placed in the scoliosis group and 12 in the no scoliosis group. In the scoliosis group, the average curve was a sizeable 23 degrees, with 10 of the patients exhibiting a single curve, four a double curve, and one a triple curve.

All the patients underwent a similar surgical procedure which involved a C1 laminectomy and removal of the outer layers of the dura (it is interesting to note that for some surgeons, the presence of a syrinx serves as a trigger to always open the dura and insert a patch, but these surgeons chose not open the dura completely).

The researchers then compared the two groups across a number of parameters, including:

- length, width and shape of the syrinx
- degree of tonsillar herniation
- duration of symptoms
- muscle atrophy in the upper extremities
- abnormal leg reflexes
- cranial nerve symptoms
- pre-op and post-op clinical status as measured by the Japanes Orthopedic Association (JOA) scale
- calculated recovery rate

Their analysis revealed a number of significant differences between the groups (see Table 1) which were published in the March, 2007 issue of the Journal of Neurosurgery: Spine. Specifically, the average length of the syrinx in the scoliosis group was almost 13 vertebral segments long compared to 7 in the no-scoliosis group. Perhaps most strikingly, nearly three-fourths of the scoliosis group suffered from upper extremity muscle atrophy , but only 8% of the no-scoliosis group did. Similarly, a whopping 93% of the patients with scoliosis also exhibited abnormal leg reflexes, while less than half of the patients without scoliosis did. Both pre and post-op clinical scores, along with the calculated recovery rate, were significantly worse for the scoliosis group as well.

The researchers also found that the length of the syrinx and the duration of symptoms were correlated to the degree of scoliosis. In other words, patients with longer syrinxes, or who had suffered from symptoms for longer, tended to have worse cases of scoliosis. Interestingly, neither the width of the syrinx, nor the amount of tonsillar herniation were found to be related to the amount of scoliosis or the clinical scores.

Finally, the authors built a statistical model to determine which factors influenced the final JOA score (at the last follow-up). While the primary factor was the pre-op JOA score - meaning how bad symptoms were prior to surgery - they also found that the degree of scoliosis and the duration of symptoms significantly influenced outcomes.

Based on their findings, the doctors conclude that adults with CM/SM related scoliosis tend to have poorer

scoliosis - abnormal curvature of the spine

thoracic - the middle part of the spine; the chest area

vertebra - the individual bony segments of the spine; often referred to by region and number, such as C3 for the third cervical vertebra

vertigo - dizziness, spinning sensation

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

Source

Ono A, Suetsuna F, Ueyama K, Yokoyama T, Aburakawa S, Numasawa T, Wada K, Toh S. Surgical outcomes in adult patients with syringomyelia associated with Chiari malformation type I: the relationship between scoliosis and neurological findings. J Neurosurg Spine. 2007 Mar;6(3):216-21.

outcomes. This is further supported by the fact that even with successful decompression surgery, the scoliosis did not improve more than five degrees in any of the patients. While the small number of patients makes it difficult to say definitively, it would appear that pediatric scoliosis related to CM/SM responds better to decompression surgery than adult scoliosis.

Despite the fact that the syrinx width did not correlate with the degree of scoliosis, the researchers believe that scoliosis is essentially a symptom of syringomyelia. They point out that there is evidence that the shape and size of a syrinx changes naturally over time while the damage it causes can be permanent. This can make correlating what a syrinx looks like at any given point in time with symptoms difficult, because the damage could have been caused earlier when the syrinx had a different size and shape. While it seems reasonable that scoliosis may be secondary to syringomyelia, and may even represent a late, advanced stage of damage, it is not clear if every patient with a syrinx would eventually develop scoliosis.

While the exact connection between scoliosis and CM/SM remains somewhat fuzzy, it is clear, at least from this study, that doctors should pay attention to the presence of scoliosis in adult CM/SM patients.

Table 1
Significant Differences Between Adult CM/SM Patients With and Without Scoliosis

	W/Scol	W/out Scol
Avg Length of Syrinx	12.8	7.2
Duration of Symptoms	14 yrs	7 yrs
UE Muscle Atrophy	73%	8%
Abnormal Leg Reflex	93%	42%
Avg. Preop JOA Score	10.1	14.4
Avg Postop JOA Score	11.9	15.8

Notes: syrinx length is measured in vertebral segments; JOA stands for Japanese Orthopedic Association scale; UE=upper extremity; significant refers to the difference being statistically significant and likely to be due to chance

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