Key Points

1. Scoliosis occurs in at least 25% of CM/SM patients
2. The exact link between syringomyelia and scoliosis is not known
3. Research has found no connection between the size of herniation and scoliosis
4. Research has also found no link between the size and location of syrinxes and scoliosis
5. In this study, researchers hypothesized that SM related scoliosis may be due to denervation of pa.htmlinal muscles
6. Study looked at two groups of children, one with CM/SM related scoliosis and one with idiopathic scoliosis
7. Biopsies revealed abnormal distribution of AChRs in 56% of children with SM related scoliosis
8. None of the idiopathic scoliosis children had abnormal AChR distribution
9. AChR is important in the nerve supply of muscles

Definitions

AChRs - acetylcholine receptors; a set of proteins at the nerve muscle junction which are important in the nerve supply of muscles

biopsy - removal of a small piece of tissue for examination

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

denervation - loss of a nerve supply to a muscle

idiopathic - due to an unknown cause

immunostaining - laboratory process which uses dyes to identify the presence of specific proteins in a sample

SM Related Scoliosis May Be Due To Spinal Muscle Denervation

January 31, 2008 -- Scoliosis, an abnormal curvature of the spine, is common in patients with Chiari and syringomyelia. In fact, studies have shown that at least 25% of such patients, and possibly more than 75%, have scoliosis accompanying the CM/SM. Despite the high prevalence, research has so far failed to identify the mechanism by which syringomyelia leads to scoliosis. Study after study has failed to identify any relationship between either the size of tonsillar herniation, or the size and location of a syrinx, and the presence or severity of scoliosis.

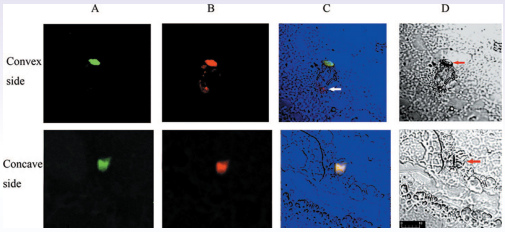
One theory on how a syrinx can lead to scoliosis involves the denervation of the muscles which are located next to the spine. All muscles are supplied by nerves which activate their use and essentially give them strength. It could be that a syrinx damages the nerves which feed the muscles along the spine. As these muscles become weak and atrophied, the natural support for the spine is removed, allowing it to bend into a scoliosis curve. A recent study from a group of Chinese researchers (Zhu et al.) published in the October, 2007 issue of the journal, Spine, provided some evidence to support this theory.

Zhu and his team hypothesized that if syrinxes were causing denervation of the pa.htmlinal muscles, that they could find indications of this within the muscles themselves. Specifically, the scientists decided to look at the distribution of acetylcholine receptors (AChRs) in children with syringomyelia related scoliosis as compared to children with idiopathic scoliosis. AChRs are a group of proteins at the nerve-muscle juncture which are critical to the nerve activation of muscles. When there is nerve damage to the point that it affects a muscle, the distribution of these proteins becomes abnormal, and can be identified through microscopic analysis.

For the study, 41 children with scoliosis were recruited. Twenty-five of the children had Chiari and syringomyelia related scoliosis and 16 suffered from idiopathic scoliosis. For the Chiari scoliosis group, the researchers used MRIs to identify the amount of tonsillar herniation, and the location and size of the syrinx (measured by what percent of the spinal cord width the syrinx comprised). Within the Chiari group, about half the children had herniations above the level of the first cervical vertebra, and about half had tonsillar herniations at or below the C-1 level. In terms of syrinx width, in 16 of the children, the syrinx was more than half the width of the spinal cord, and in 9 it was less than half.

For all the children, both syringomyelia related and idiopathic, a spinal muscle biopsy was taken during spinal surgery. The researchers then used a process called immunostaining (see Figure 1) to identify the distribution of AChRs.

Figure 1: Immunostaining To Identify AChR Distribution



In support of their theory, the scientists found that while none of the children in the idiopathic group showed an abnormal distribution of the receptors, more than half (56%) of the children in the CM/SM group did, indicating the denervation of the spinal muscles. The researchers used further testing to identify the abnormal presence of one specific subunit of AChR, which was present in 65% of the CM/SM group, but in none of the idiopathic children.

Interestingly, the team found no relationship between the amount of herniation or syrinx width and the presence of the abnormal AChRs (see Table 1). Further confusing the situation, they also failed to find a relationship between the abnormal AChRs and the severity of the scoliosis. So while the abnormal AChRs were present more than half of the syringomyelia related scoliosis children, they did not appear to be related to the progression of scoliosis.

Table 1
Percent of SM Patients With Abnormal AChR Distribution By Characteristic

Tonsillar Descent	Grade 1	62%
	Grade 2&3	50%
	>50%	50%

pa.htmlinal - muscles next to the spine

scoliosis - abnormal curvature of the spine

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

Zhu Z, Qiu Y, Wang B, Yu Y, Qian B, Zhu F. *Abnormal spreading and subunit expression of junctional acetylcholine receptors of pa.htmlinal muscles in scoliosis associated with syringomyelia.* Spine. 2007 Oct 15;32(22):2449-54.

Syrinx Cord Ratio	<50%	75%
Cobb Angle	<45 Degrees	50%
	>45 Degrees	60%

Notes: *None of the differences were found to be statistically significant.*

While this study offers some support of the hypothesis that syringomyelia related scoliosis is due to damage to the pa.htmlinal muscles, the results are far from definitive and one would have expected that if scoliosis in these cases is due to weakened spinal muscles then the abnormal distribution of the receptors should have been related to scoliosis severity. One possibility to explain this is that the study simply had too few cases to generate definitive results.

However, it is also important to keep in mind that it is far from certain that scoliosis is actually caused by syringomyelia. In fact, if anything, the consistent findings that syrinx size and location are not related to the presence or severity of scoliosis may indicate that something else is going on. It could be that both scoliosis and syringomyelia are a by-product of the same underlying problem in ways that are not yet understood.

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