

Key Points

1. Research has shown a link between Chiari, syringomyelia and scoliosis, but the nature of the link is unknown
2. Adolescent idiopathic scoliosis affects up to 4% of 10-16 year olds
3. Previous work has shown that children with AIS tend to have low-lying cerebellar tonsils
4. This study compared skull measurements and CSF flow of AIS patients with healthy controls
5. Found that children with AIS had low cerebellar tonsils and large foramen magnums; also found that level of tonsils correlated with degree of scoliosis
6. However, did not find any difference between the groups in peak CSF velocity at the level of the foramen magnum
7. AIS children clearly have some abnormal anatomical characteristics which are similar to Chiari, but it is not clear why some have Chiari symptoms and some don't

Definitions

AIS - adolescent idiopathic scoliosis; refers to scoliosis affecting a teenager for which no underlying cause can be found

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

craniocervical junction - anatomical region where the skull and spine meet

foramen magnum - opening at the base of the skull through which the spine and brain connect

idiopathic - due to an unknown cause

morphological - referring to the physical shape, size and structure of something; in this article the skull

Looking For Signs Of Chiari In Idiopathic Scoliosis

July 31, 2007 -- Adolescent idiopathic scoliosis (AIS) is a fairly common problem affecting teenagers. Defined as an abnormal curvature of the spine of at least 10 degrees, AIS may affect up to 4% of 10-16 year olds. Although the curvature will not get worse for the majority of children, in some it can progress relentlessly and in others it can be associated with serious neurological problems, such as Chiari.

While clinical guidelines have been developed to help doctors determine when an MRI should be used to look for neurological issues, in the case of Chiari, the true link between the two conditions is not understood. For example, although it has been shown that decompression surgery can stop the progression of Chiari related scoliosis, research has also shown that the location and severity of scoliosis is not related to the amount of tonsillar herniation or the location or size of syrinxes.

In other words, researchers have yet to find features of Chiari which are strongly related to features of scoliosis which would help reveal the link between the two. Because of this, it is not known if Chiari somehow causes scoliosis, if scoliosis can lead to Chiari, or if both are in essence a symptom of a more fundamental anatomical problem.

One group of researchers who has been exploring a link between AIS and Chiari is based out of Hong Kong (Chu et al.). Previously, this group found that AIS patients tend to have low-lying cerebellar tonsils compared to healthy children, and that the level of the tonsils was related to abnormal neurological testing (somatosensory evoked potentials).

Based on this work, the research team decided to study whether there were other Chiari-like similarities in AIS children. Specifically, they decided to look again at the position of the cerebellar tonsils, but this time they also took dimensional measurements of the skull base and used phase-contrast MRI to look at the peak CSF velocity at the craniocervical junction. Recall that Chiari research has shown (Iskandar, Haughton) that Chiari patients tend to have elevated CSF velocity at the level of the foramen magnum.

To accomplish this, the team recruited 105 girls, aged 11-18. Sixty-nine of the girls had scoliosis, with an average curve of 35 degrees, and 36 of the girls were healthy and acted as a control group. All of the girls were found to have no abnormal neurological signs and anyone with a head injury, back injury, or history of headaches was excluded. Each girl underwent a standard MRI and a phase-contrast MRI to measure CSF velocity. In addition, the girls were given SSEPs to see if there were any problems with the nerves along their spine. They published their results in the July 1st, 2007 issue of the journal, Spine.

As they had with their previous study, the researchers found a significant difference between the position of the cerebellar tonsils in the girls with AIS as compared to the healthy girls (See Table 1). Specifically, the average position of the tonsils for those with AIS was 1.2mm below the foramen magnum (note, this would still be considered a mild herniation by many doctors) compared to 3.5 mm above foramen magnum for the healthy girls. The team also found that the position of the tonsils was correlated with the degree of scoliosis, meaning that those with more severe curvatures of the spine, tended to have lower lying tonsils.

Table 1
Significant Differences Between Normal Controls and AIS Patients

	AIS	Control
Cobb Angle (degrees)	36	0
Tonsillar Position (mm)	1.2	-3.5
FM Area (mm ²)	807	767
FM Diameter (mm)	34.1	33

Notes: Tonsillar position refers to the level of the cerebellar tonsils relative to the foramen magnum with a positive value below the FM and a negative value above; FM refers to foramen magnum;

In addition, the scientists found that the AIS girls tended to have larger foramen magnums (the opening at the base of the skull) than the healthy controls. While abnormal skull base anatomy has been demonstrated with Chiari, unfortunately the researchers in this study did not look beyond the foramen magnum at other measurements which have been found to be unusual in Chiari. It is also interesting to note that the size of the foramen magnum in the AIS group, although large, did not correlate with the degree of scoliosis.

What was surprising to the researchers is that they could find no difference in the peak CSF velocities between

scoliosis - abnormal curvature of the spine

somatosensory evoked

potentials (SSEPs) - test which evaluates nerve function by stimulating a nerve with an electrical signal and measuring how long the signal takes to travel along the nerve

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

the AIS group and the control group. Although the author speculate that the large opening of the foramen magnum may allow for normal CSF flow, this interpretation is not entirely clear. In fact, Chiari research has shown that measuring CSF velocity is tricky and the results can vary depending on the specific technique used.

In terms of the SSEP tests, the researchers found that 19% of the AIS group actually had abnormal results (even though their neurological exams were normal), indicating some impairment of the spinal nerves. When they tried to link the SSEP results with the other parameters they were studying, they found that there was a small association with increased CSF velocity, but the finding was not statistically significant.

The findings from this study are difficult to interpret. While it is very interesting that the girls with scoliosis tended to have low-lying cerebellar tonsils, Chiari research has shown that the actual position of the tonsils is not related to symptoms or clinical outcome.

One aspect that was not explored in this publication was the possibility of a genetic link between scoliosis and Chiari. It is believed that AIS may have a large genetic component, much as it is believed that Chiari may as well. Since some of the anatomical features of AIS are similar to Chiari, one has to wonder if the genes involved in the two conditions are closely related. Perhaps some people have a scoliosis gene (for lack of a better word), some people have a Chiari gene, and some people have both.

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Source

Chu WC, Man GC, Lam WW, Yeung BH, Chau WW, Ng BK, Lam TP, Lee KM, Cheng JC. *A detailed morphologic and functional magnetic resonance imaging study of the craniocervical junction in adolescent idiopathic scoliosis.* Spine. 2007 Jul 1;32(15):1667-74.

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