

## Key Points

1. Despite significant research into Chiari related scoliosis, the nature of the link between the two conditions is not known
2. It is also not clear when an MRI should be ordered to check for problems like Chiari in an adolescent with scoliosis
3. Study looked at 87 children with Chiari related scoliosis to identify patterns that could be used
4. Found no relation between the amount of tonsillar herniation, or the size and shape of syrinx to severity of scoliosis
5. Group also looked at atypical curve patterns, atypical features of curves, and amount of kyphosis
6. Only kyphosis was found more frequently among children with more severe curves
7. Authors suggest an MRI is warranted if there is a left thoracic curve, if there are atypical features to a curve, or if there is thoracic kyphosis

## 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

**kyphosis** - abnormal front/back curve of the spine

**lumbar** - lower part of the spine

**scoliosis** - abnormal curvature of the spine

**thoracic** - middle part of the spine, chest area

**cerebellar tonsils** - portion of the cerebellum located at the bottom,

## Features Of Chiari Related Scoliosis

**January 31, 2008** -- Scoliosis is defined as an abnormal curvature of the spine. Naturally, the spine runs essentially straight down the middle of the back. Curves to the right or left are considered to be abnormal and termed scoliosis. Scoliosis is described by the direction of the curve (right or left), the location of the curve on the spine (cervical, thoracic, lumbar, thoracolumbar), and the severity in degrees (the Cobb angle).

In addition to right and left curves, the spine can also curve abnormally to the front and back. To provide structural support, the spine does have natural front-back curves, but when these are outside of the normal range, it is called kyphosis. Scoliosis is commonly found in children and adolescents, often with no underlying cause readily apparent.

Over the years, it has become clear that there is some type of link between scoliosis and Chiari. However, despite significant research the exact nature of this link remains a mystery. Studies have shown that decompression surgery can be effective in halting the progression of scoliosis and that in adults the presence of scoliosis is linked to poorer outcomes.

A challenge for physicians who see children with scoliosis is deciding when an MRI is warranted to look for neurological problems such as Chiari. Since scoliosis is not uncommon among adolescents, and because only a small percentage of those cases are actually related to Chiari, ordering an MRI for every child with scoliosis is not practical. In several studies, researchers have tried to find unique characteristics of Chiari related scoliosis which can alert doctors to when an MRI should be performed. Based on this work, some doctors recommend that Chiari should be checked for if there are any neurological signs and/or severe curves. Others have tried to focus on curve patterns that aren't typically seen, for example certain types of double curves.

To further this type of research, a group from China (Qiu et al.) studied the radiographic features of 87 children with Chiari and syringomyelia related scoliosis. They published their results in the January, 2008 issue of the Journal of Pediatric Orthopedics. Specifically, the scientists wanted to identify any characteristics that were related to the severity of the scoliosis, so they split the children into three groups:

Group 1: Curves between 10 - 30 degrees, 13 children

Group 2: Curves between 30 - 60 degrees, 42 children

Group 3: Curves greater than 60 degrees, 32 children

### For each child, the researchers looked at:

Curve location, direction, and severity

Amount of kyphosis

Amount of tonsillar descent

Syrinx, length, width, and shape

Based on location and direction, the curves were classified as either being typically found in Adolescent Idiopathic Scoliosis (AIS) or atypical (see Table 1). The researchers also classified atypical features of typical curve patterns, which included curves which started or ended at unusual locations on the spine. Finally, the team also classified the syrinxes as distended, moniliform, slender, or circumscribed (see Figure 1).

**Table 1**  
**Typical and Atypical Curve Patterns**

Typical	Atypical
Rt. Thoracic	L Thoracic
Rt Thor/L Lumbar	L Thor/Rt Lumbar
Rt Thor/L ThoraL	L Thor/Rt ThoraL
Thoracolumbar	R&L Double Thor
Lumbar	Triple & Quadruple

**Notes:** Rt = right; L = left; Thor = thoracic; ThoraL = thoracolumbar

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

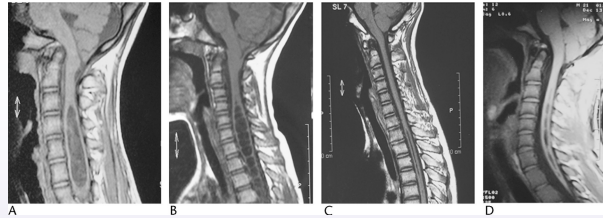
**Chiari malformation I** - condition where the cerebellar tonsils are displaced out of the skull area into the spinal area, causing compression of brain tissue and disruption of CSF flow

**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

Qiu Y, Zhu Z, Wang B, Yu Y, Qian B, Zhu F. *Radiological presentations in relation to curve severity in scoliosis associated with syringomyelia*. J Pediatr Orthop. 2008 Jan-Feb;28(1):128-33.

**Figure 1: Syrinx Classification**



A = Distended; B = Moniliform; C = Slender; D = Circumscribed

Like many researchers before them, the Chinese team failed to find any connection between either the amount of herniation, syrinx size, or syrinx shape and the scoliosis severity. Surprisingly, the percent of atypical curves and atypical features did not differ significantly between the three groups either (see Table 2).

**Table 2**  
**Radiographic Features By Group**

	Group 1 (13)	Group 2 (42)	Group 3 (32)
% with atypical curve	46%	45%	41%
% with kyphosis	0%	14%	60%

**Notes:** Groups are based upon scoliosis severity; only kyphosis is significantly different between groups

One striking difference the doctors did find between the groups involved kyphosis in the thoracic region of the spine. Specifically, the percent of children in Group 3 with kyphosis was 60%, compared to only 14% in Group 2, and 0% in Group 1.

Although not completely in line with their results, the doctors recommend an MRI for scoliosis if there are any neurological signs, if there is a left thoracic curve (which is common with Chiari related scoliosis), and if there is thoracic kyphosis. Interestingly, while researchers have tended to focus on curve features to warrant an MRI, they tend to overlook whether there is a family history of Chiari or Chiari type symptoms. It seems logical that if there is there is a family history of Chiari or any type of headaches or neck pain, that an MRI is probably warranted.

Despite repeated efforts, researchers have for the most part come up empty in trying to link characteristics of Chiari and syringomyelia to the presence and severity of scoliosis. It may be time to look past the X-ray and MRI for clues as to what the real relationship is between the conditions.

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