

## Key Points

1. Association between Chiari/SM and scoliosis in children is well documented
2. MRI is expensive and with children often involves sedation, so MRI is not used routinely on scoliosis patients.
3. Study examined atypical scoliosis characteristics to predict when an MRI would show a problem
4. Patients with severe curves despite a young age and abnormal neurological changes are most likely to have a positive MRI
5. Study proposes a clinical test which agrees with MRI results 75% of the time

## Definitions

**atypical** - not usual or normal

**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

**idiopathic** - due to an unknown cause

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

**scoliosis** - abnormal curve of the spine

**sensitivity** - the ability of a

## Study Identifies Types Of Scoliosis That Indicate Chiari

While the link between Chiari, syringomyelia, and scoliosis is well established, when to order an MRI for a scoliosis patient (to rule out Chiari) is not as clear. There are over 500,000 people in the US with scoliosis and over 50% of them are idiopathic (meaning the scoliosis is due to an unknown cause). To order an MRI to check for Chiari for every one of these cases would be cost prohibitive. In addition, an MRI for children often involves sedation and can be a traumatic event for both the child and their family.

Since ordering blanket MRI's for every scoliosis patient is not reasonable, doctors must rely on their judgment and experience to determine when an MRI is necessary. The good news is that there is general agreement in many cases. Research has shown that MRI's rarely reveal anything in what are considered typical scoliosis cases, so most doctors will not order an MRI if there is nothing unusual about the patient's scoliosis. At the other end of the spectrum, patients with severe neurological deficits in combination with scoliosis, are very likely to have Chiari or syringomyelia, so MRI's are routinely ordered. However, much like the question of when to recommend surgery for Chiari, there is less agreement in the middle. What if there are only minor neurological signs? What if the scoliosis isn't quite typical?

In an effort to help doctors decide what to do with these middle ground cases, Dr. Jose Morcuende, an orthopedic surgeon at the University of Iowa, along with colleagues from Spain and Thailand, tried to identify characteristics that would predict a positive MRI finding (meaning the presence of Chiari, syringomyelia, or some other problem) in children with scoliosis. The team published their findings in the January 1, 2004 issue of the journal Spine.

The researchers decided to focus on six characteristics that they consider to be atypical in scoliosis patients:

1. Early onset, meaning before the age of 10
2. Atypical curve pattern, a single left thoracic curve is one such example
3. Severe curves despite immaturity, defined as a Cobb angle of greater than 45 degrees
4. Rapid curve progression, defined as an increase in curvature of more than 1 degree per month
5. Abnormal neurological changes, such as altered or absent reflexes
6. Chronic, disturbing back pain or headache

To see if any of these factors can predict a positive MRI result, the team reviewed the records of 1,168 patients identified as having idiopathic scoliosis and found 72 that had one or more of the six atypical characteristics. They then compared the MRI results of the patients with atypical characteristics to those with typical characteristics (see Table 1).

**Table 1**  
**Atypical Scoliosis Characteristics & MRI Findings**

Characteristic	# With Char.	% Of People w/Char & Positive MRI	% Of People without Char & Positive MRI
Early Onset	29	10%	18%
Severe Curve	9	56%	10%
Atypical Curve Pattern	24	17%	15%
Rapidly Progressing Curve	17	12%	16%
Abnormal Neurological Findings	20	40%	6%

Overall, 11 of the patients (15%) had positive MRI's for either Chiari, syringomyelia, or both. The two most suggestive characteristics were abnormal neurological findings and a severe curve. Forty percent of the patients with abnormal neurological findings had a positive MRI result, compared with only 6% with normal neurological exams. Of the nine patients with severe curves, 56% had a positive MRI test, while only 10% of the patients without severe curves did. Interestingly, none of the patients with confirmed Chiari or SM had headaches or back pain.

Based on these results, the researchers then used statistical modeling to develop a diagnostic test that can predict when Chiari or syringomyelia will be found on MRI. The model showed that patients without a severe curve or abnormal neurological findings only had a 3% probability of having a positive MRI result. Patients with

diagnostic test to detect a disease or condition that is really there

**specificity** - the ability of a diagnostic test to exclude a disease or condition when it is really not there

**syringomyelia - (SM)** 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

Morcuende JA, Dolan LA, Vazquez JD, Jirasirakul A, Weinstein SL. *A prognostic model for the presence of neurogenic lesions in atypical idiopathic scoliosis*. Spine. 2004 Jan 1;29(1):51-8.

abnormal neurological findings, but no severe curve had a 29% chance of having a positive MRI. Similarly, patients with a severe curve, but normal neurological findings, had a 32% chance of a positive MRI. Finally, patients with both a severe curve and abnormal neurological findings, had an 86% chance of finding CM and/or SM on MRI.

The authors believe that this means that any patient with either severe curves or abnormal neurological changes (or both) should be tested with an MRI. If this rule is applied to their data, the rule agrees with the MRI finding - meaning it accurately predicted the presence or absence of CM/SM - 75% of the time (see Table 2).

**Table 2**

**Rule:** Presence of severe curve and/or abnormal neurological findings is indicative of a neurological problem and patients should be tested with an MRI

### Agreement With MRI (Positive and Negative)

75%

### Sensitivity -

82%

### Specificity -

74%

While the authors stress that this type of test - or rule - only deals in probabilities and should not be used by itself to determine treatment options, it does appear that in the proper context, it can be a significant clinical aid to doctors treating idiopathic scoliosis. And if it can help even one child get an earlier, accurate Chiari diagnosis, it is definitely worth using..

--Rick Labuda

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