Key Points

1. Scoliosis is commonly associated with Chiari and syringomyelia; 50% or more of Chiari/SM patients also have some degree of scoliosis
2. Previous research on children with Chiari related scoliosis has tried to identify the types of scoliosis which indicate Chiari and the benefits of performing Chiari surgery to stabilize the scoliosis progression
3. This study looked at a group of pediatric Chiari patients and identified those with Chiari, SM, and scoliosis
4. All children underwent decompression surgery and then were monitored for curve progression
5. Found that children under 10 and with scoliosis less than 30 degrees did not need additional corrective surgery
6. This is in line with previous research which found that age and amount of scoliosis were critical
7. Should be noted that decompression surgery does not usually help adults with Chiari related scoliosis

Definitions

cervical - the upper part of the spinal cord; the neck area

Cobb Angle - technique used to measure the severity of a scoliosis curve, in degrees, from an image of the spine

levoscoliosis - a scoliosis curve which bends to the left

lumbar - the lower part of the spinal cord; the lower back

scoliosis - abnormal curvature of the spine

thoracic - the middle part of the spinal cord; the chest area

cerebellar tonsils - portion of the cerebellum located at the bottom, so named because of their shape

Surgery Helps Chiari Related Scoliosis

April 20, 2006 -- Scoliosis, an abnormal curvature of the spine, is very common among Chiari and syringomyelia patients. In fact, it may be that more than one half of Chiari patients also suffer from some degree of scoliosis. Fortunately, scoliosis related to Chiari is one of the few areas that has received a fairly significant amount of research attention, especially from the pediatric community.

Previous research has focused on the types of scoliosis which indicate the need for an MRI to check for Chiari, and the best way to treat Chiari related scoliosis. Scoliosis not related to Chiari can be treated with braces or surgically corrected with rods and bolts. However, research has shown that trying to correct Chiari related scoliosis in this manner is often unsuccessful. Instead, although the underlying connection between Chiari and scoliosis is not understood, research has shown that for some patients, Chiari decompression surgery is actually enough to stop to the progression of scoliosis and eliminate the need for direct surgical correction of the spine. This has led many doctors to suggest that in cases with Chiari related scoliosis, it is best to treat the Chiari first, and then address the scoliosis if it doesn't improve.

In a study published in March, 2006 in the on-line section of the journal, Child's Nervous System, two surgeons from the Birmingham Children's Hospital in the UK provide further support for this notion, and identify characteristics of patients for whom decompression surgery alone is likely to be an adequate treatment for Chiari related scoliosis.

Dr. Ranjeev Bhangoo and Dr. Spyros Sgouros prospectively looked at 36 children they treated surgically for Chiari between 1998 - 2003. To be included in the study, the children had to have symptomatic Chiari, syringomyelia, and scoliosis, with no other spinal abnormality which could affect the progression of scoliosis.

Using these criteria, 10 of the 36 children qualified. For the group of 10, the researchers collected data including: age, gender, duration of symptoms, length of syrinx, age at decompression surgery, initial Cobb angle (a measure of the severity of scoliosis), and the need for additional corrective surgery or bracing after the decompression surgery.

The Chiari surgery for all the children was similar and involved a craniectomy (removing a portion of the skull), opening the arachnoid, and ensuring good flow of CSF out of the 4th ventricle. Interestingly, the surgeons chose to leave the dura wide open rather than insert a patch. The cerebellar tonsils themselves were not removed or reduced in size. The children were followed for an average of close to three years, and all but one had a successful outcome in terms of Chiari and syringomyelia symptoms.

During the follow-up period, six of the ten children required no further treatment for their scoliosis, while the other four required corrective surgery. When the researchers looked at the characteristics of the children who required additional scoliosis surgery versus those who didn't, two thing jumped out.

First, the average severity of the scoliosis - as measured by the Cobb angle - was much lower for the group that did not require additional surgery, than it was for the group that did (see Table 1). Specifically, the average Cobb angle for the group that didn't require corrective surgery was 29 degrees, whereas the average curve for the group which did need additional surgery was 76 degrees.

Table 1
Age and Degree of Scoliosis of Children Who Required Corrective Surgery vs Those Who Didn't

	No Scoliosis Surgery	Scoliosis Surgery
Number of Patients	6	4
Avg. Cobb Angle (degrees)	29	76
Avg. Age (months)	125	158

Note: *All patients underwent Chiari decompression surgery; this table highlights the significant differences between those who also required scoliosis surgery and those who didn't*

Second, the group of children who didn't require the scoliosis surgery tended to be younger when they underwent decompression surgery as compared to the other group. On average, the children who only underwent Chiari surgery were about 10 years old at the time of surgery. The children who continued to have problems with their scoliosis were on average a little older than 13 years at the time of the decompression surgery.

However, these were the only two significant differences between the two groups. Symptom duration, syrinx

cerebrospinal fluid (CSF) - clear liquid in the brain and spinal cord, acts as a shock absorber

MRI - magnetic resonance imaging; large device which uses strong magnetic fields to produce images of soft tissue inside the human body

syringomyelia (SM) - neurological condition where a fluid filled cyst forms in the spinal cord

syrinx - fluid filled cyst in the spinal cord

Source

Bhangoo R, Sgouros S. *Scoliosis in children with Chiari I-related syringomyelia*. Childs Nerv Syst. 2006 Mar 16; [Epub ahead of print]

length, and location of syrinx were not factors in determining whether additional surgery would be required.

The results from this study, despite including only a small number of children, line up very closely with previous research. Other studies have also found that both age and degree of curvature at the time of decompression surgery are important factors in determining whether additional procedures will be required for scoliosis treatment. In fact, one prior study also found that 10 was the key age for success, and a separate study found that 30 degrees of curvature or less predicted whether decompression surgery alone would be enough. Based on their results, and the results of previous research, the authors of this study propose that when it comes to Chiari related scoliosis, decompression surgery when the child is less than ten years old and the scoliosis is less than 30 degrees may prevent the need for additional corrective surgeries. It should be pointed out that the authors note that further research should follow children into adulthood to make sure scoliosis treatments are not needed later in life.

While it is certainly gratifying to see research begin to produce consistent results for at least one.htmlect of Chiari, the actual link between Chiari, syringomyelia, and scoliosis remains a mystery. Study after study has found no link between physical characteristics of the syrinx - such as size and location - and the type or severity of the accompanying scoliosis.

In addition, there is one negative implication of age being a critical factor in Chiari related scoliosis. Namely, what happens if the decompression surgery is performed too late. Unfortunately, research has shown that decompression surgery does little when it comes to adults with Chiari related scoliosis. Let's hope that the researchers in this area continue their efforts to untangle the mysterious relationship between Chiari, syringomyelia, and scoliosis.

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