

## Key Points

1. Scoliosis is common in children with a syrinx and is often diagnosed before syringomyelia.
2. Non-surgical treatments for scoliosis associated with an active syrinx tend to not work and surgery can worsen the neurological situation.
3. A thorough review of the research literature shows that treating the syrinx benefits the scoliosis as well.
4. If the scoliosis does not stabilize, corrective surgery is safer after syrinx treatment.
5. The exact link between syringomyelia and scoliosis is unknown.

## Scoliosis

- Scoliosis is an abnormal curvature of the spine. A normal spine looks straight when seen from behind, scoliosis is a right-left curve when seen from the rear.
- Scoliosis affects 1-2 percent of the general population.
- Scoliosis can be caused by congenital conditions, neurological conditions (like SM), or can be of unknown origins.
- 80% of scoliosis cases have an unknown cause and are labeled "idiopathic".
- Treatment options include observation, bracing, and surgery.
- Bracing involves wearing an external device for up to 23 hours per day.
- Surgery involves fusing vertebra of the spine using rods, screws, bars, and bone grafts.
- No evidence that exercise, drugs, electrical stimulation, or spinal manipulation helps stop or improve curvature.

## How To Treat Syringomyelia Related Scoliosis In Children

Scoliosis is a common condition related to syringomyelia in children; in fact, some studies have shown that more than 50% of children with a syrinx have abnormal spinal curvature. While many types of scoliosis which develop in children and adolescents will stabilize on their own and not cause any symptoms, scoliosis associated with a syrinx often causes back pain and can progress rapidly.

Compounding the problem is the ineffectiveness of standard treatment. Non-surgical treatments, such as wearing a brace, tend to not work with syrinx related scoliosis. Surgical intervention is even worse, with many reports of worsening neurological symptoms after surgery. Given these facts, it is little wonder that more and more doctors are looking to treat the underlying syrinx before turning their attention to the spine.

In an effort to untangle this difficult situation, Dr. Ken Kontio and a group from the Children's Hospital of Eastern Ontario, in Canada, reviewed nine pediatric cases treated at their facility and combined them with a thorough review of the literature. In reviewing the published literature, studies had to detail curve magnitude, curve progression, and the change with the treatment to be included. Overall, the researchers ended up with a group of 98 cases from the literature, which could be combined with the nine patients treated locally.

The group published their work in the Journal of Pediatric Orthopaedics, in December, 2002, in a paper titled *Management of Scoliosis and Syringomyelia in Children*. Because of differences in the way the studies were conducted, the use of true statistical analysis was not possible, however much can be learned from just looking at the actual data (see Table 1). With the definition of curve progression as a change in 5 degrees or more, the researchers found that among the group of children who were treated for their syrinx (or the syrinx stabilized on its own), 36% experienced continued curve progression, 48% stabilized and experienced no further curve progression, and in 16% of the cases the curve actually improved. In contrast, in the group which did not receive syrinx treatment, 73% of the children's curves continued to worsen, 20% stabilized, and only 7% improved. In another measure of the difference between the two groups, in the group with syrinx treatment, 25% eventually required surgical stabilization of the spine. In comparison, 50% of the children who were not treated for their syrinx required surgical stabilization of their spine.

**Table 1**  
**Scoliosis Progression**

	Worsened	Stabilized	Improved	Surgery
Syrinx Treatment	36%	48%	16%	25%
No Treatment	73%	20%	7%	50%

The researchers also looked at the effect of bracing on curve progression. Interestingly, bracing was not a very effective treatment for either group. Among those children who received syrinx treatment and bracing, 67% experienced further curve progression. The numbers are even worse for the group without syrinx treatment, with 92% of children who received bracing experiencing further curve progression. While bracing appeared ineffective, among the nine children treated locally, for those who needed surgery, fortunately there were no neurological complications. Data was not presented on how many of the children whose syrinx wasn't treated experienced neurological complications after spinal surgery.

The exact link between syringomyelia and scoliosis is unknown. Is scoliosis a direct result of the presence of a syrinx? While this would seem to make sense, research - including this study - has failed to find a statistical link between syrinx size or location, and the type and severity of scoliosis, so perhaps syringomyelia and scoliosis are both the result of an abnormal spinal environment. Despite the research findings, there are several theories on how a syrinx can cause scoliosis. One theory proposes that the formation of a syrinx damages the motor neurons and results in an imbalance of the back muscles, making scoliosis more likely. Another theory proposes that when a fetus is developing, the presence of a syrinx will cause vertebra to form abnormally and lead to scoliosis.

While there are many unknowns surrounding both syringomyelia and scoliosis, it seems clear that the best way to treat the scoliosis is to treat the underlying syringomyelia. Given the frequency of scoliosis in children with syringomyelia - and the potential damage of standard treatments - MRI should be used to identify or rule out the presence of a syrinx in scoliosis cases where there are also neurological symptoms or unusual progression of the curve.

## Definitions

**cervical** - having to do with the upper portion of the spine located in the neck area

**Cobb Angle** - technique used to measure the severity of a spinal curve - in degrees - from spinal images

**lumbar** - having to do with the lower portion of the spine

**motor neuron** - nerve cell that controls muscle activity

**syringomyelia** - neurological condition where a fluid filled cyst forms in the spinal cord

**syrinx** - fluid filled cyst in the spinal cord

**thoracic** - having to do with the middle part of the spine in the chest area

## Sources

Scoliosis Research Society  
www.spineuniverse.com  
Journal of Pediatric Orthopaedics  
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