

## Key Points

1. Study reports two cases where syrinxes improved - reduced in size - on their own
2. There have been 37 reported cases, confirmed by MRI, of spontaneous resolution since 1990
3. As with syrinx formation, spontaneous resolution is not well understood, though there are several theories
4. Implications for patients is unclear; waiting before surgery risks permanent nerve damage; the number of cases that resolve on their own is very low compared to the overall number of CM/SM cases
5. A deeper understanding of syrinx dynamics may enable predictions for who will resolve spontaneously, who is a good candidate for surgery, etc.

## Definitions

**brainstem** - base of the brain which connects to the spinal cord

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

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

**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

**cisterna magna** - CSF filled space below the cerebellum

**foramen magnum** - opening at the base of the skull, through which the spinal cord passes

**idiopathic** - of unknown cause

**intubation** - insertion of a tube into a body canal or organ; in this case the trachea prior to surgery

## Can Syrinxes Resolve On Their Own?

It seems miraculous; after being diagnosed with a terrible condition and suffering through worsening symptoms, the symptoms start to go away, you start feeling better, and then after an MRI a doctor confirms you are getting better. You avoided the surgery you were dreading and you feel close to your old self.

When a syrinx reduces in size without intervention, it is called spontaneous resolution. Given the nature of corrective surgery, it may be tempting to hold out hope for this type of mini-miracle, but is it realistic? There is no disputing there are well documented cases of spontaneous resolution, but how often does it occur, and what are the risks of adopting a wait and see approach?

In the September issue of the journal *Neurosurgery*, Dr. Kazuhiko Kyoshima from Shinshu University in Japan, and Dr. Enver Bogdanov from the Kazan State Medical University in Russia, report two more cases of spontaneous resolution and provide a comprehensive overview of the literature on the subject.

The researchers' first case involved a 10-year old Japanese girl with a history of scoliosis and facial palsy who had developed neck pain over the prior 6 months. With an MRI, the doctors found a syrinx that extended from C4 - T11 and a tight cisterna magna - although the fluid flow at the foramen magnum was not completely blocked. Decompression surgery was planned, but never happened because of problems intubating the young girl. MRI scans 22 months later revealed that the syrinx was smaller and the girl reported her neck pain was not as severe. A follow-up MRI 10 months later showed a further reduction in syrinx size and the girl's neck pain had completely disappeared along with an improvement in other symptoms. A third MRI follow-up, 21 months later, showed no further change in syrinx size.

The second reported case involved a 39-year old Russian man. He was an agricultural worker who was suffering from progressive weakness in his legs. A neurological exam revealed some sensory deficits and an MRI showed a malformation with crowding at the foramen magnum and a syrinx from C2-T2. The man did not want surgery and after suffering through worsening symptoms for a month, his symptoms stabilized over the next month and then began to slowly improve. Six months later, the man reported a substantial improvement in his symptoms and an MRI revealed that the cerebellar tonsils had actually moved up - so there was less crowding - and the syrinx was smaller.

In addition to their two cases, the authors review 37 cases of spontaneous resolution that have been well documented since 1990 (see Side Bar). While most of the cases are Chiari related syringomyelia, spontaneous improvement has been documented in cases of trauma, Multiple Sclerosis, and syrinxes of unknown origin as well. Interestingly, in the cases involving Chiari related syringomyelia, children were much more likely to show improvement in the actual Chiari malformation as well, compared to adults. This finding implies that different mechanisms may be responsible for improvement in children than in adults.

Some researchers believe that a Chiari malformation develops as a result of an abnormally small posterior fossa. An extension of this theory is that a child with a Chiari malformation whose posterior fossa grows faster than the brain will have less crowding and may experience spontaneous resolution of CM, SM, and/or symptoms. However, some cases of spontaneous resolution occur very quickly, and can not be accounted for by typical childhood growth. The authors in this study speculate that the Japanese girl experienced spontaneous improvement in CSF flow either at the foramen magnum or out of the fourth ventricle possibly due to the disruption of scarring that may have been blocking normal flow.

As for spontaneous resolution of a syrinx in an adult, one theory holds that as a syrinx continues to expand, it eventually ruptures out of the spinal cord, allowing for drainage into the surrounding CSF. Essentially things get worse before getting better. While there is some evidence to support this, the idea is not yet universally accepted and in the case of the Russian farm worker, his improvement is more likely due to stopping activities - namely hard, manual labor - which were aggravating his condition.

So what is a patient to make of all this? Clearly there are well documented cases where syringomyelia, and even Chiari malformations, stop progressing or improve on their own, however there is no clear understanding of how or why this occurs. In addition, noted Chiari expert Dr. Thomas Milhorat points out in a comment published in the same issue that very long-term follow up of these types of cases would be required to determine if the improvement is permanent or just temporary. In another comment, Dr. Edward Benzel points out that relative to the total number of Chiari and syringomyelia cases, the number of spontaneous resolutions is very low.

There are no easy answers, but it is clear that spontaneous resolution is a rare occurrence and much more work is required to understand how, when, why, and to whom it may happen in order to benefit patients beyond the lucky few.

1. Study reports two cases where syrinxes improved - reduced in size - on their own

**Multiple Sclerosis (MS) -**

autoimmune disease which affects the nervous system; causes weakness, loss of coordination, etc.

**posterior fossa** - portion of the skull that houses the cerebellum and portions of the brainstem

**spontaneous** - occurring without external causes, self-generating

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

**trachea** - the windpipe; tube which carries air to the lungs

**ventricle** - CSF filled space in the brain

**Source**

Kyoshima K, Bogdanov E; [Spontaneous resolution of syringomyelia: Report of two cases and review of the literature.](#) Neurosurgery 53(3) Sept, 2003. pg 762-9.

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**The Lucky Few:  
Profile of Spontaneous Resolution Cases (37)**

- Age range: 19 mo - 61 yr
- 21 females, 16 males
- Time between diagnosis and confirmed improvement by MRI ranged from 2 mo - 11 yrs.
- 30 cases were Chiari related
- Other causes included trauma, Multiple Sclerosis, and idiopathic
- Syrinx reduction was accompanied by an improvement in the Chiari malformation in 24 of the 30 cases
- The improvement in Chiari was more common among pediatric cases than adults
- Symptoms completely disappeared in 6 cases

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