

Key Points

1. In many SM patients, pain persists even after surgery reduces the size of a syrinx
2. Researchers looked at whether age, duration of symptoms, and/or shape of a syrinx was related to which SM patients had pain and whose pain would improve post-operatively
3. Age was not related to either pain or pain improvement, but duration of symptoms was; namely, people in pain and people whose pain did not improve had had their symptoms for a longer time
4. Study also found that people with a deviated syrinx (see below) were not likely to have their pain improve, even after a successful surgery
5. Researchers believe that deviated syringes may damage nerves in the dorsal horn which do not recover their function even after surgery
6. Study highlights the critical need for early and accurate diagnoses

Definitions

afferent pathways - nerve paths that carry signals from the periphery - like the hand - to the spinal cord and brain

anterior - at or near the front of something

central canal - very center of the spinal cord, so named because it starts as a hollow tube which closes in most people as they age

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 (CM) - condition where the cerebellar tonsils are displaced out of the skull area into the spinal area,

Does The Shape Of A Syrinx Predict Improvement?

One of the more frustrating aspects of syringomyelia is that even after successful surgery - meaning restoration of CSF flow and even a reduction in the syrinx - symptoms, especially pain, often don't improve. In fact, if you look at descriptions of the disease from years ago, surgery is described as a way to stop the progression of symptoms, not as a cure.

While it is well known that nerve damage from a traumatic injury doesn't heal well, the exact mechanisms underlying pain associated with a syrinx are not well understood. Some people have syringes that go virtually the length of their spine before painful symptoms appear; others suffer from unrelenting pain from what looks like a much smaller syrinx. This lack of knowledge makes it very difficult to predict, on an individual basis, whether pain symptoms will improve after surgery. [Ed note: In making his recommendation for me to have surgery, my neurosurgeon pointed out that if I waited, I wouldn't know at what point the damage to my nerves would be permanent.]

In an attempt to identify factors that influence pain, and pain improvement after surgery, a group from Keio University in Japan, led by Dr. Masaya Nakamura, looked at 25 Chiari related syringomyelia patients they had treated over the past 15 years. The group wanted to know whether age, duration of symptoms (prior to surgery), and/or syrinx shape were related to pain and post-surgical pain improvement. They published their results in the March, 2004 issue of the Journal of Neurosurgery.

The 25 patients ranged in age from 13 to 57 and there were 4 men and 21 women. Eleven patients underwent decompression surgery, 12 patients were treated with shunts (placed directly into the syringes) and 2 patients received both. The group was followed for an average of 5 years after surgery.

The researchers divided the patients into two groups: those with pain directly attributable to a syrinx, and those without such pain. They further divided the pain group into those whose pain improved after surgery and those whose pain didn't. In addition, the researchers classified each person's syrinx as either central, enlarged, or deviated (see Fig 1). In all, 17 patients had pain, and 8 had no pain. Of the 17 pain patients, 6 improved after surgery, and 11 did not. Prior to surgery, there were 2 central, 15 enlarged, and 8 deviated syringes.

In looking at their data, the group found that age had no relation to whether a person had pain or whether their pain improved after surgery. They did find however, that duration of symptoms was significantly related to both. The average duration of symptoms for the pain group was more than 30 months. In contrast, the average duration for the no pain group was only 15 months. After surgery, the difference was just as striking. The average duration of symptoms for the improved group was about 20 months, while the average for the no improvement group was much higher at about 40 months.

In addition to how long people had had symptoms, syrinx shape also appeared to influence pain. All 8 patients with deviated syringes had pain, while only 9 of the 15 enlarged syringes, and neither of the two central syringes caused pain. Post-surgically, only 1 out of the 8 patients with a deviated syrinx showed any pain improvement. In addition, in three patients, an enlarged syrinx transformed into a deviated syrinx, and in all three cases the patients showed no pain improvement. In total, 10 out of 11 patients who had deviated syringes either before or after surgery still suffered from pain after surgery. Unfortunately, the authors did not say whether the type of treatment (decompression, shunt, or both) influenced the pain outcome, so it is not known if there is another variable to account for.

The researchers believe that the deviated syringes cause so many problems because their shape means they occupy part of the dorsal horn. Research has shown that damage to this area of the spine causes spontaneous pain and does not heal on its own.

While the authors don't speculate on this in their paper, it is interesting to note that many researchers believe that given enough time, a syrinx will expand to a maximum and then essentially rupture and begin to reduce in size as the fluid in the syrinx finds a path back into the normal CSF spaces. While not stated explicitly by the authors, it seems likely that the patients with deviated syringes also were the ones who had had symptoms for the longest time. One has to wonder if in these cases, the syringes were starting to rupture, and it is this process that causes the most damage. Either way, the obvious implication from this work is the importance of early, accurate diagnoses. If these conditions can be identified and corrected early enough, maybe the pain can be stopped before it even begins.

Fig 1
Syrinx Classification

causing compression of brain tissue and disruption of CSF flow

dorsal - towards the back, or posterior, of something

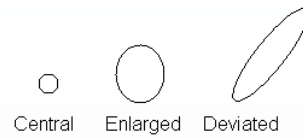
dorsal horn - also known as the posterior column, part of the spinal cord located in the back part of the cord where many afferent nerve pathways are located, note there is a right and left dorsal horn

duraplasty - surgical technique where a patch is sewn into the dura, the tough covering of the brain and spinal cord

posterior - at or near the back of something

shunt - a surgically implanted tube used to divert, or drain, CSF

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



Central Syrxinx - Contained within the central canal

Enlarged Syrxinx - One that enlarges the central canal

Deviated Syrxinx - A syrxinx that bulges in one direction more than others; often into the dorsal horn area of the spine

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