

Syringomyelia FAQ's

These FAQ's are for informational purposes only and in no way represent an attempt to provide medical advice. This information may or may not apply to your case and anyone with a question or concern about their health is strongly encouraged to consult with a medical professional.

1. What is syringomyelia?

Syringomyelia (SM) is a neurological disorder where a fluid-filled cyst - or syrinx - develops inside the spinal cord. This cyst can grow over time, causing the spinal cord to expand and stretch nerve tissue. Eventually, the syrinx can cause permanent nerve damage and paralysis.

2. What causes syringomyelia?

One researcher noted that syringomyelia is not really a disease unto itself, because it is always the result of something else. By far, the most common cause of syringomyelia is a Chiari malformation. Although there are no strong statistics, about 30%-50% of people with Chiari also have syringomyelia. Syringomyelia can also form after a spinal cord injury (SCI), such as from a car accident or fall; this is called post-traumatic syringomyelia (PTS). PTS can form months or even years after such an injury. Syringomyelia can also be the result of a tumor or mass in the spinal cord.

3. Why does a syrinx form?

Over the years there have been several theories as to why a syrinx forms (mostly dealing with the effects of a Chiari malformation on cerebrospinal fluid flow), but none have been universally accepted or proven. One current theory states that the cerebellar tonsils act like a piston and beat down into the spinal area with every heartbeat. This piston motion then forces cerebrospinal fluid (CSF) into the spinal cord itself, where it forms a syrinx. However this theory does not account for evidence that the pressure inside a syrinx is higher than outside and that syrinx fluid does not exactly match CSF.

4. What is idiopathic syringomyelia?

In rare cases, there is no discernable cause for why a syrinx forms; this is referred to as idiopathic syringomyelia. Recent research has shown, however, that even in some cases of idiopathic syringomyelia there is crowding at the skull-spine junction, even though there appears to be no Chiari malformation. In these cases, the surgery used to treat Chiari has been successful in treating idiopathic syringomyelia.

5. What are the symptoms of syringomyelia?

Because the syrinx is putting pressure directly on nerves, the number one symptom associated with syringomyelia is pain. Many patients report severe pain in the neck, upper back, and shoulders. Doctors refer to this as the "cape effect" of syringomyelia - meaning pain in the area where a cape is draped over the shoulders. For patients with a syrinx located in the thoracic region of the spine, the pain may be in their chest, stomach, or lower on the back. In addition, many people with syringomyelia lose strength in their arms and legs and develop numbness in their hands and feet. Additional symptoms include trouble regulating body temperature, abnormally stiff muscles, and loss of bladder and bowel control.

6. Does syringomyelia cause paralysis?

Yes, an active syrinx can eventually lead to paralysis.

7. How is syringomyelia diagnosed?

An MRI (Magnetic Resonance Imaging) exam of the spine can clearly show the presence of a syrinx. If a Chiari malformation is found on an MRI, usually an MRI of the entire spine will be ordered to look for syrinxes.

8. I have a syrinx but don't have any symptoms, what does this mean?

For reasons that are not well understood, some people can have a syrinx - even a large one - and not have any symptoms. While some people will stay symptom free, cases have been noted where people become symptomatic suddenly and deteriorate rapidly.

9. How is syringomyelia treated?

Because of the risk of permanent nerve damage, if symptoms are significant or progressing, most surgeons will recommend surgery of some type. The treatment for cases where there is a syrinx but no real symptoms is more controversial. Some surgeons will recommend surgery anytime there is a syrinx, while others will take a wait and see approach and monitor the situation with routine MRI's and neurological exams.

10. What is the surgery like?

For Chiari related syringomyelia, the most common procedure is a Chiari decompression see (Chiari Q14) Alternative procedures include placing a shunt (a tube like device) to divert cerebrospinal fluid around a Chiari malformation, or trying to drain the syrinx by placing a shunt directly into it.

11. Will I get better after surgery?

Unfortunately, there is not a lot of good, long-term, surgical outcome research which adequately measures a patient's quality of life after surgery. For Chiari related syringomyelia, surgery will reduce the syrinx, or at least stop it from growing, up to 80% of the time. However, this does not always translate into a significant improvement in symptoms, and many people still experience some pain - and other symptoms - after surgery. There is currently no way to identify beforehand who will get better with surgery and what symptoms will improve. However, some research has indicated that the longer someone has symptoms before they receive surgery, the less chance they have of a successful outcome. It is important for every patient to discuss the possible surgical outcomes, in detail, with their doctor, so they know what to expect.

12. What happens if I don't have surgery?

The natural progression of syringomyelia is not well understood. For people with symptoms and an active syrinx, the syrinx may eventually lead to paralysis. For people without symptoms, however, the future is less clear. A recent study followed 11 people with syrinxes, but no symptoms, for more than 10 years. Of the 11 subjects, only 1 eventually developed symptoms and required surgery. The decision to have or not have surgery should be carefully discussed with your doctor. If surgery is not performed, the doctor may recommend closely monitoring the situation with routine MRI's and neurological exams.

13. How long will it take to recover after surgery?

Recovery will vary for each individual, will depend in a large part on whether there is permanent damage from the syrinx, and in some cases can take years [Ed Note - I still feel like I am getting better and stronger 5 years after my surgery]. What many people don't realize is that even with a successful surgery, the syrinx may take up to a year to collapse and may not go away completely. In addition, many people will have had symptoms for years before surgery, so while some things will improve quickly, others may take years to improve. Often recovery can be a series of ups and downs, with long periods of improvements punctuated by temporary setbacks.

14. What is neuropathic pain?

Neuropathic pain refers to pain that is caused by damage to the nervous system. Unfortunately, neuropathic pain is all too common among syringomyelia patients and can be very difficult to treat. There are many articles on this website about different ways to cope with pain.

15. Am I eligible for disability?

Many people with Syringomyelia have qualified for government disability. Some people have been able to get disability easily, while others have had to fight for it.

16. How is post-traumatic syringomyelia different from Chiari related syringomyelia?

Up to 25% of spinal cord injury patients will develop a syrinx months - or even years - after the initial injury. The mechanisms underlying the syrinx formation are not well understood and PTS is difficult to treat. Surgical options include correcting any bone deformities in the area, creating more space around the syrinx, and draining the syrinx with a shunt. Unfortunately, less than half of PTS patients show long-term improvement after treatment.

17. How many people have syringomyelia?

Syringomyelia was once thought to be extremely rare with early estimates of only 20,000 people in the United States having it. Increased use of MRI has since shown that syringomyelia is more common and, while there has not been a rigorous study to determine how many people have it, a Duke University analysis estimates that between 100,000 - 200,000 people in the US may have syringomyelia.

18. Is there a support group for syringomyelia?

Yes, the American Syringomyelia Alliance Project www.asap.org offers an on-line message board and several regional support groups.



For More Information Visit: www.conquerchiari.org