Key Points

1. The natural history of syringomyelia is not well understood
2. However, there is some evidence that over a very long time, symptom progression may stabilize and syrinxes may start to shrink
3. Previous research has identified groups of patients who show signs and symptoms of SM, but do not have a syrinx on MRI
4. This study looked at 168 SM patients in Russia and identified 14 such people (symptoms but not syrinx)
5. Also identified 15 patients with flat, collapsed syrinxes
6. Clinically, the two groups were very similar
7. Authors believe that in some people syrinxes will eventually collapse on their own and call this the Post-syrinx state
8. This may occur because the syrinx expands so much that eventually CSF can drain into the subarachnoid space
9. However, even after collapse, the damage to the spinal cord remains and symptoms do not improve

Definitions

atrophy - the wasting away of a body part

cervical - the upper portion of the spinal cord

communicating - when used in reference to a syrinx, refers to whether CSF can visibly flow into and/or out of the syrinx from another location

dysesthetic pain - pain due to something that is not normally painful

gliosis - an overgrowth of a specific type of nerve cell which tends to occur in a damaged area

Identifying Post-Syrinx Syndrome

March 20, 2006 -- Have you ever wondered what happens to a syrinx if it is left alone over time? The natural history of syringomyelia has not been extensively studied and is not well understood. Because the nerve damage caused by syringomyelia may be permanent and severe, many surgeons in the US recommend surgery if a syrinx related to Chiari is present, so the opportunities to track how syrinxes naturally progress are limited. Despite this, there are indications that for at least a subset of syringomyelia patients, if syrinxes are left alone for many years, they will eventually begin to shrink and even collapse on their own.

One such indication are reports in the literature, although rare, of more than 30 cases of spontaneous resolution of SM. It is ***very*** important to note however, that in these cases, just because the syrinxes collapsed on their own, the patients' symptoms did not necessarily improve.

More prevalent than spontaneous resolution cases, are reports of groups of patients who exhibit the neurological signs and symptoms associated with syringomyelia (as opposed to symptoms caused by Chiari) but do not appear to have a syrinx when examined with MRI. An example of this is Milhorat's landmark study which found a significant group of people who fit this category.

One group who has studied the natural progression of syringomyelia directly is Bogdanov and Mendelevich of Kazan State Medical University, in Russia. In a specific rural area of Russia, there appears to be an unusually high rate of syringomyelia, especially among men. In a previous publication, Bogdanov and Mendelevich reviewed over 100 cases of syringomyelia which were not operated on, but from which people had been suffering from for as long as 46 years. Interestingly, they found that the size of a syrinx tended to decrease at the later stages of the disease. This supports other research findings which have shown that symptom progression stabilizes after 10 years in about one-third to one-half of patients (again, this doesn't mean they got better, just that symptoms stopped getting worse).

Now, in a recent on-line publication of the Journal of Neurology, Bogdanov and Mendelevich - along with Heiss at NIH - extend their work and propose that patients who have signs and symptoms of syringomeylia but no syrinx, may actually be in a post-syrinx state. In other words, they hypothesize that in some cases, syrinxes eventually collapse on their own.

To support their theory, the team looked at 168 Russian Chiari/syringomyelia patients who were seen between 1997 - 2001. Each patient underwent a neurological evaluation and an MRI. From this, the researchers identified two separate groups of patients. The first group, Group A, was comprised of 14 patients who exhibited signs and symptoms of SM, but did not show a syrinx on MRI. The second group, Group B, was comprised of 15 patients who also had signs and symptoms of SM, but who showed flat, collapsed syrinxes on MRI.

When the authors compared the two groups, they found they were very similar clinically (see Table A). Neurological deficits were comparable between the two groups and characteristic of Chiari related SM. In addition, several patients in each group had noticeable atrophy of the spinal cord (their cord was narrower than 8mm). Finally, for the patients in both groups, the progression of symptoms had stabilized for at least 3 years.

The Russian team believes that since the clinical findings of Group A were similar to those patients with collapsed syrinxes which were still visible, it is likely that the patients in Group A had syrinxes which spontaneously resolved at some point in the past. They were, in effect, post-syrinx.

Why would syrinxes resolve on their own in some people? It is important to realize that syrinx growth is a dynamic process and while there are several theories as to its underlying mechanism, none have yet been proven conclusively. One of the current leading theories of syrinx formation is called the piston theory and was developed by researchers at the US National Institutes of Health. The piston theory proposes that with each heartbeat, the cerebellar tonsils are driven down into the crowded spinal area like a piston. This in turn creates a pressure wave in the cerebrospinal fluid, which forces the fluid into the spinal cord, forming a syrinx.

The scientists in this study propose two possible mechanisms for syrinx resolution. First, the natural flow of CSF is somehow restored at the level of the Chiari malformation which results in the syrinx resolving. Second, after an extended period of expansion, the tissue around a syrinx becomes so thin that it eventually ruptures and allows the syrinx to drain into the CSF in the subarachnoid space. In support of this idea, the team identified three patients in whom there was clear communication of fluid - on MRI - between the syrinx and the subarachnoid space.

Based on their findings, the authors define the post-syrinx state as characterized by:

myelopathy - any disease of the spinal cord

posterior fossa - depression on the inside of the back of the skull, near the base, where the cerebellum is normally situated

subarachnoid space (SAS) - space underneath the arachnoid, but above the actual brain and spinal tissue, which contains the cerebrospinal fluid

Common Chiari Terms

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

cerebellum - part of the brain located at the bottom of the skull, near the opening to the spinal area; important for muscle control, movement, and balance

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

Chiari malformation I - 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

decompression surgery - general term used for any of several surgical techniques employed to create more space around a Chiari malformation and to relieve compression

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

Source: Bogdanov EI, Heiss JD, Mendelevich EG.
The post-syrinx syndrome: stable central myelopathy and collapsed or absent syrinx.
J Neurol. 2006 Mar 6; [Epub ahead of print]

1. Stable signs and symptoms of central myelopathy (spinal disease)
2. MRI evidence of Chiari and either an absent or collapsed syrinx
3. No evidence of other diseases which can mimic syringomyelia

While the idea is intriguing, more research is clearly needed to further define the post-syrinx state, how often it occurs, and what the implications are for care. It is also important to note (as the authors do) that the post-syrinx state is just one possible natural outcome of syringomyelia. Today, there is no way to predict for an individual how a syrinx will progress. Given that the damage a syrinx can cause is often permanent, and the cost of not taking action may be extremely high, how to treat syringomyelia is an issue that should be discussed at length with medical professionals.

Table 1
Clinical Features of Post-Syrinx Group (A) vs Flat Syrinx Group (B)

	A (14)	B (15)
Avg. Age	44	55
Symptom Duration (Yrs)	21	28
# w/ dysesthetic pain	3	10
# w/ sensory loss	13	13
# w/ muscle atrophy	4	6
# w/ weakness	13	14
# w/ spinal atrophy	4	6

Note: Group A did not have syrinx on MRI, bud did have syrinx related symptoms; Group B had flat, collapsed syrinx on MRI

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