

### Key Points

1. Despite being studied for many years, the precise mechanism whereby syringes form is not known
2. The recent Piston Theory focuses on the Chiari malformation creating a pressure wave in the subarachnoid space which forces CSF into the cord
3. This article introduces a new theory, the Intramedullary Pulse Pressure Theory which posits that a syrinx expands, and fills from the inside
4. The theory states normally pressure waves are in unison between the cord tissue and CSF space
5. However, when there is a small narrowing of the CSF spaces, the pressure waves are no longer synchronized and the cord tissue starts to distend and fills with extracellular fluid
6. This theory can also be used to explain post-traumatic SM, and SM due to tumors, scarring, etc.
7. While there is some evidence to support this theory, it will take time for other scientists to review it and determine if it is valid

### Definitions

**arachnoid** - thin, veil-like, middle layer of the coverings of the brain and spinal cord

**distend** - expand abnormally

**dura** - tough, outer covering of the brain and spinal cord

**edema** - swelling due to an accumulation of fluid

**extracellular** - fluid which exists just outside of cell membranes

**fluid dynamics** - the quantitative study of how fluids behave

**hydrocephalus** - condition involving an abnormal

## New Theory Attempts To Explain All Types Of Syringomyelia

June 20, 2006 -- Syringomyelia, defined by the presence of a fluid-filled cavity, or syrinx, in the spinal cord was first identified hundreds of years ago. Yet despite the intervening years and scrutiny of many brilliant minds, it remains largely a mystery. In particular, the mechanism by which the namesake, fluid-filled cavity forms, to this day, is not fully understood.

Naturally, there have been many theories as to how and why syringes form (many of which are detailed in other C&S News articles), but none have withstood the tests of time and ongoing scientific scrutiny, and ultimately failed to solve the syringomyelia riddle.

To understand the complexity of the problem, it is important to understand, at least at a basic level, the relevant anatomy (see Figure 1). At the heart of the matter, so to speak, is the spinal cord itself. The spinal cord is an extension of the brain, and is comprised of similar tissue, which extends down the back through the bony vertebra.

Bundles of nerve fibers branch out at various levels and run throughout the body, carrying sensory information to the brain, and command signals to various body parts. The spinal tissue itself is covered with varying layers of protective coatings. Between the tissue of the spinal cord and the outer coating layers (the arachnoid and the dura) is a region called the subarachnoid space (SAS). Thus, a simplified, cross-sectional view of the spinal cord could be represented by concentric circles, where the inner circle is the spinal tissue, the outer circle is the arachnoid/dura, and the space in between is the subarachnoid space

The SAS is where cerebrospinal fluid (CSF), flows, bathing and cushioning the brain and spine. A Chiari malformation (Figure 1, letter D) blocks this natural flow of CSF.

Historically, it has been assumed that the fluid in a syrinx is comprised of CSF, so the question has always been why and how does it get there.

For example, recent attention has focused on the so called Piston Theory, put forth by doctors at the National Institutes of Health. The Piston Theory states that with Chiari, the cerebellar tonsils are driven into the SAS with each heartbeat, like a piston. This, in turn, creates a pressure wave in the CSF filled SAS, which drives CSF into the spinal tissue through small openings called perivascular spaces.

While this appears to make sense, and align with some clinical observations, some researchers have pointed out that filling a syrinx with CSF is like blowing a balloon up by forcing air into it from the outside; in other words impossible. They point out that a balloon expands because we use a hole to get air inside, which then pushes the surface of the balloon out and expands it.

In the end, it's all a matter of pressure. For a syrinx to expand, the pressure inside the syrinx has to be higher than the pressure in the SAS outside of it. However, if this is the case, then fluid from the SAS can not get into the syrinx.

In an effort to get around this problem, Dan Greitz, a Swedish researcher who has studied syringomyelia for some time, decided to focus his attention inside the spinal cord, rather than at the subarachnoid space. In an article aptly titled, "Unraveling the riddle of syringomyelia", published on-line recently in Neurosurgical Review, Greitz describes a new, comprehensive theory of syrinx formation.

The new theory (really an extension and expansion of his previous work), called the Intramedullary Pulse Pressure Theory, differs from most earlier theories in that it states that a syrinx is not filled with cerebrospinal fluid, but rather extracellular fluid which is formed from repeated pressures on the spinal tissue itself.

According to the Intramedullary Pulse Pressure Theory, the heart beating creates a natural pressure wave which propagates through the CSF of the subarachnoid space and the tissue of the spinal cord itself. In a healthy person, the pressure waves in the CSF and the tissue are in sync, so the pressures balance themselves out.

However, if there is a blockage (like from a Chiari malformation or trauma) or narrowing of the subarachnoid space, the pressure wave in the SAS is disrupted and is no longer in sync with the pressure wave in the spinal tissue. The result is higher pressure in the spinal cord, which when repeated over time, causes sections of the cord near the obstruction to expand and fill with extracellular fluid.

Although the publication is fairly technical, Greitz discusses how the new theory works for all types of syringomyelia, not just Chiari related SM. Specific to Chiari, he points out how the tonsils do act as a piston, but

accumulation of CSF in the brain

**perivascular space** - small space just outside of veins and arteries which feed the spinal cord

**spinal cord** - extension of the brain which runs down the back

**subarachnoid space (SAS)** - CSF filled space between the spinal cord and the arachnoid/dura covering

**Valsalva maneuver** - any straining type activity, such as coughing, holding breath, etc.

**ventricle** - any of several CSF filled spaces in the brain

**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

he believes this does not force CSF into the cord, but greatly amplifies the disruptive effect of any narrowing of the CSF space. Similarly, in this theory, Valsalva type maneuvers, such as coughing and straining can be shown to have a dramatic effect on the forces driving the spinal cord distension. Greitz also invokes some advanced fluid dynamics principles to demonstrate how CSF flowing through a narrowed SAS can actually create a suction force which will pull the spinal cord out and contribute to syrinx formation.

In support of his theory, Greitz cites recent hydrocephalus research which has shown that brain tissue will expand - or bulge out - and fill with extracellular fluid when exposed to repeated pulse pressures. This occurs even when the pressures are relatively small. Since spinal tissue has similar mechanical properties to brain tissue, Greitz believes a similar effect occurs in syrinx formation.

But perhaps the strongest evidence supporting the Intramedullary Pulse Pressure Theory comes from experiments on rats. In one study, the spinal cords of rats were loosely constricted with a ligature (thus narrowing the space where CSF could flow). Repeated MRI's showed that three weeks later every single rat showed edema (swelling with fluid) both above and below the constriction point. Eleven weeks later, every single rat had developed a syrinx both above and below the constriction. Contrast agents used with the MRI's indicated that the fluid which formed the syrinxes was likely extracellular fluid from the spinal tissue itself, and not CSF. The extracellular origin of syrinx fluid is further supported by studies which have shown that fluid drawn from a syrinx does not exactly match, chemically, CSF.

While the Intramedullary Pulse Pressure Theory is original and broad reaching, it is too soon to say whether the riddle of syringomyelia is truly solved. The scientific method will ensure that this theory is put to the test by skeptics and either proven to be true, or shown to be yet another in a long line of failed attempts to explain syringomyelia.

**Figure 1**  
**Line Diagram Representing Spinal Cord/SAS Anatomy**



A = arachnoid membrane/dura

B = sub-arachnoid space (SAS), filled with CSF

C = spinal cord tissue

D = Chiari malformation; cerebellar tonsils block the SAS

### Source

Greitz D. [Unraveling the riddle of syringomyelia](#). Neurosurg Rev. 2006 May 31; [Epub ahead of print]

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