Key Points

1. Mathematical model, or analysis, of the CSF system in Chiari and syringomyelia was created
2. It models the system as two, co-axial tubes filled with fluid (CSF)
3. New theory says that during a cough or sneeze, a pressure wave travels up the spine and bounces off the tonsils; when this occurs, for a short period of time, the pressure inside the spinal cord is greater than outside and could form a syrinx
4. Same model identifies problems with current theories on syrinx formation
5. Model makes a number of assumptions which may or may not affect whether it is accurate
6. Model need to be proven experimentally

Definitions

arachnoid - middle-layer of the the membranes which cover the brain and spinal cord

central canal - the very innermost part of the spinal cord; starts as a hollow tube in children, but closes off in adulthood

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

cervical - the upper part of the spinal cord; neck area

Chiari malformation - condition where the cerebellar tonsils are displaced out of the skull area into the spinal area, causing compression of brain tissue and

New Theory On How Syrinxes Form

Many theories on syrinx formation have come and gone over the years. While they vary in their details, most modern theories focus on the dynamics of how CSF flows and under what conditions it would create a syrinx. Cerebrospinal fluid bathes the brain and spinal cord and acts as a protective cushion and shock absorber. In a healthy person, CSF flows freely from the brain to the spinal region (around the cerebellar tonsils) and back again in concert with a person's heartbeat. When the cerebellar tonsils descend out of the skull and crowd the spinal cord, one of the main passages for this CSF can be blocked. For decades now, doctors and scientists have been trying to understand how this blockage can lead to a syrinx forming in the spinal cord..

Gardner kicked it all off with a theory that stated if a Chiari malformation blocks the natural flow of CSF out of the brain, it will be redirected and flow down the central canal of the spinal cord instead and create a syrinx. This theory may work for a subset of cases, but it has since been shown that in most adults, the central canal closes off and would not allow this type of CSF flow.

One of the more popular theories of today is the so-called piston theory. Developed by surgeons at the National Institutes of Health (Oldfield and Heiss), this theory holds that in a case of Chiari, the cerebellar tonsils act like a piston. With every heartbeat, they drive down into the spinal area and create a pressure wave of CSF. This pressure wave crashes into the spinal cord and somehow forces CSF inside. While there is some evidence to support this theory - tonsils have been shown to move on MRI, some type of pressure wave does flow down during the cardiac cycle, and dye studies have shown that CSF can flow into the spinal cord along the outside of veins and arteries - many engineers have expressed some doubts.

They point out that for a syrinx to form - and bulge out - the pressure inside the cord itself should be higher than in the subarachnoid space outside the cord (where the CSF is). In other words, you blow up a balloon by creating higher pressure inside the balloon, not by forcing air into the balloon from the outside. In fact, some research has indeed shown that the pressure inside a syrinx is higher than outside the spinal cord. In addition, if the spinal cord is somewhat soft - which it tends to be - than an outside force should crush it as opposed to forcing CSF into it.

Yet another theory - put forth by Greitz - tries to take this into account. This theory holds that if the normal pressure wave that forces CSF from the brain into the spine (due to the heartbeat) is blocked, the pressure wave itself is transmitted down the spinal cord proper instead of the subarachnoid space. This results in a force pushing out - from the spinal cord - below the tonsils. It is interesting to note that many Chiari related syrinxes due form just below the blockage.

One reason there are so many theories is that the CSF system is extremely complicated and difficult to observe (although advances in MRI technology are changing this). A second reason is that doctors and engineers look at the problem differently. While surgeons may tend to base their ideas on their experiences with real cases; engineers, on the other hand, faced with a complex situation tend to create simplified models - or mathematical representations - which can be used to study a system and make predictions.

Professor P.W. Carpenter, Director of the Fluid Dynamics Research Center at the University of Warwick, UK, and his colleagues created just such a model (see Fig 1) and published their results in a two part paper in the December, 2003 issue of the Journal of Biomechanical Engineering. The group started by equating the spinal CSF system to two co-axial tubes filled with a fluid (CSF). They made additional assumptions to simplify the math, such as that the area between the tubes is uniform, that fluid only flows up and down, and that there is no resistance to the fluid flow.

The team then set about defining the equations that describe how fluid - meaning CSF - would flow in this situation. Once they had a general description worked out, they focused in on what would happen when a person coughs. In their model, a cough creates a pressure wave in the CSF that travels up the spinal column until it hits the cerebellar tonsils which are blocking the passage (see Longitudinal View).

When this happens, the wave of high pressure is reflected back and creates an interesting situation. If their math and model are correct, for a very short period of time - about 1/10 of a second - the pressure inside the spinal cord is higher than outside the cord. This is just the type of environment that some feel is necessary for a syrinx to form.

The group also used their model to examine Greitz's theory and the piston theory. They do not believe the Greitz idea is valid because in their model most of the energy of a pressure wave is reflected back when it hits the malformation, and very little is transmitted down the spinal cord itself.

As for the piston theory, the group does believe that the tonsils can create pressure waves which hit the spinal cord. They don't think, however, that this can account for a syrinx forming because they believe syrinx formation requires a force pushing out from the spinal cord, not pushing in on it. So while the piston theory may be valid for disruption of CSF flow

co-axial - two tubes which have the same axis (see Fig 1)

elastic - something that returns to it's original shape after being stretched, compressed, or deformed; a rubber band is elastic

lumbar - the lower part of the spine

rigid - something that isn't flexible and won't bend

spinal cord - the actual column of nerve fibers and tissue that connects with the brain and runs down the back

stenosis - narrowing or blockage of a passage

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

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

syrinx - fluid filled cyst in the spinal cord

thoracic - the middle part of the spine; chest area

vertebra - segment of the spinal column, noted as region plus number (C = cervical, T = thoracic, L = Lumbar)

Source

Source: Carpenter PW, Berkouk K, Lucey AD. Pressure wave propagation in fluid-filled co-axial elastic tubes. Part 2: Mechanisms for the pathogenesis of syringomyelia. J Biomech Eng. 2003 Dec;125(6):857-63.

a syrinx growing in size, in their view, it is not valid for the initial formation.

The co-axial tube model, and subsequent syrinx theory, developed by the Warwick group have a number of limitations. First, the assumptions they made to simplify their model may not be valid assumptions to make. For example, cine MRI has shown that CSF flows in many directions around herniated tonsils, not only up and down. They also assume that the area between the tubes is uniform which also is not really true in the case of a real person. Whether these assumptions lead to false conclusions is not clear.

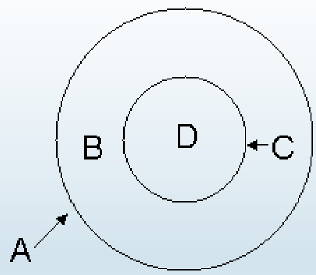
Second, their theory only takes into account the action of a cough (and maybe a sneeze). The regular CSF motion produced by the heart beating does not play a role in this model. While coughing does seem to play a big role in headaches and symptoms, it seems a bit of a stretch for it to be the only triggering mechanism. What about blocking the natural flow of CSF from the brain to the spine? What about straining, and exertion, and minor traumas?

Additionally, the model predicts only a localized area of pressure difference that could form a syrinx. How then would it account for Chiari related syrinxes in the thoracic or lumbar regions of the spine? The author of this article has a cervical syrinx close to the tonsils, but also a thoracic syrinx much lower down. Decompression surgery reduced them both, implying a similar mechanism was responsible for each one.

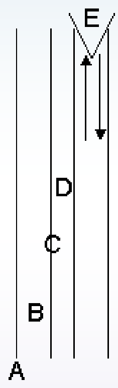
Finally, and most importantly, at this stage the model is just math. There is no real evidence to support the idea that the reflected wave creates a high pressure situation in the spinal cord. Even if this were true, there is no data to say how high the pressure in the spinal cord gets; is it high enough to create a syrinx?

For now it appears the surgeons will develop their theories, the engineers will develop theirs, and the patients will have to wait a little bit longer for true understanding.

Figure 1 Fluid-filled Co-Axial Tube Model
(Cross-sectional view)



(Longitudinal View)



A = arachnoid membrane; the outer tube
B = sub-arachnoid space (SAS), filled with CSF
C = spinal cord; the inner tube in the model
D = central canal; the inside part of the inner tube
E = Chiari malformation; cerebellar tonsils block the SAS

Theory: When a person coughs (or sneezes) a pressure wave flows up the SAS and hits an obstruction - the malformation. When this happens, the pressure wave reflects off, but creates a temporary situation where the pressure in the spinal cord (C/D) is significantly higher than the SAS (B). This could lead to syrinx formation.