Key Points

1. Chiari and syringomyelia are complicated disorders
2. There are likely several different reasons why syrinxes form
3. The term communicating refers to whether a path exists for fluid to flow from one place to another in the body
4. Case highlights the complexity of syrinxes; cine MRI revealed patient had two adjacent, but distinct syrinxes which did not communicate
5. Surgery resolved one, but not the other
6. MRI's also showed that surgery dramatically improved the CSF flow in the subarachnoid space at the syrinx level, and reduced the amount of spinal cord movement

Definitions

caudal - towards the tail; in this case CSF flow from the brain to the spine

central canal - tubelike center of the spinal cord; often closes off in adults

cephalad - towards the head; in this case CSF flow from the spine to 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 - 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

Case Highlights The Complexity Of Chiari And Syringomyelia

[**Ed. Note:** *Dr. Terry Lichtor, the lead author of the paper discussed in this article, is a Scientific Advisor to the C&S Patient Education Foundation.*]

It's been said many times and in many ways: Chiari and syringomyelia are complex diseases. While Chiari is largely thought to be congenital - meaning you are born with it - the exact cause is not known. In fact, it is believed that the underlying mechanism for Chiari II (associated with Spina Bifida) is probably different than that for Chiari I. In addition, there are clearly documented cases of acquired Chiari, meaning while the end result may be the same, i.e. cerebellar tonsils jamming into the spinal area, it appears there are many ways to get there, none of which are clearly understood.

Not to overstate the complexity of the issue, but the situation is strikingly similar for syringomyelia, meaning there appear to be several mechanisms at play and not all syrinxes are alike. Clearly there is a difference between Chiari related syringomyelia and post-traumatic syringomyelia, but even within the Chiari family of SM, the picture is somewhat murky.

Some researchers have tried to classify different types of syrinxes by location and/or shape. Sometimes doctors will refer to syrinxes within the central canal (sometimes referred to as hydromyelia) or outside of the canal in the actual tissue of the spinal canal (extracanalicular); while others will focus on the shape, using terms such as central, enlarged, and deviated. The problem with classifying syrinxes in this way is that human anatomy is not always that precise. It is not always possible to determine whether a syrinx lies within the central canal, especially since the central canal tends to disappear as we age.

Another term which is commonly used to describe syrinxes is 'communicating'. In reference to the human body, the term communicating is used to mean there is a connection, or path, between two parts. In reference to syringomyelia, the term is used most commonly to refer to a syrinx which is somehow connected to the 4th ventricle, such that CSF can flow easily between them.

One of the early theories of syrinx formation held that the blockage created by a Chiari malformation meant that the natural flow of CSF was diverted and would back up into the 4th ventricle and then enter the spine through the central canal. Problems with this theory arose, however, when it became clear that in most adults the central canal closes, thus preventing this from occurring. Thus, it was thought that most syrinxes associated with Chiari are actually non-communicating.

Unfortunately, as often happens when trying to describe phenomena that are poorly understood, there is still controversy surrounding the communicating issue. One of the current leading theories regarding syrinx formation, is the so called 'Piston Theory' put forth by researchers at the National Institutes of Health (NIH). The Piston Theory states that with every heartbeat, the cerebellar tonsils act like a piston and push down into the spinal area. This in turn creates a pressure wave in the CSF which is driven into the spinal cord through tiny spaces outside of blood vessels. So if this theory is true - which is far from proven - are these syrinxes communicating? According to the theory they communicate with the subarachnoid space (SAS) outside the tissue of the cord itself, but this is different than the original intent of the descriptor.

To confuse things further, at least one research team has recently proposed that most syrinxes do in fact communicate with the 4th ventricle via the central canal, but that current imaging technology is not sensitive enough to see it.

A unique case published in the June 1, 2005 edition of the journal Spine, highlights the complexity currently confronting doctors and researchers. Dr. Terry Lichtor, a neurosurgeon at Rush University Medical Center, along with colleagues from Cook County Hospital and the University of Illinois Chicago, report their findings regarding a patient with two distinct syrinxes, which although they were next to each other, did not appear to communicate.

The patient in question was a 30 year old woman with Crouzon Syndrome, a genetic condition which affects the development of the skull and face, plus Chiari and syringomyelia. The woman had been experiencing progressive symptoms for a number of years, and a neurological exam revealed a number of other problems as well.

The researchers used phase contrast MRI (cine MRI) to measure both the movement of the spinal cord and the amount of CSF flowing at different levels of the spine, both before and after surgery (see Figure 1).

Before surgery, the MRI scans showed that there was a good deal of spinal cord movement at the C1 level with

cine MRI - type of MRI which can show CSF flow, also known as phase contrast MRI

communicating - refers to whether a path, or connection exists between two places in the body; most often used to refer to whether a syrinx "communicates" with the 4th ventricle, meaning that CSF can flow between them

cranium - the skull

craniectomy - surgical technique where part of the skull is removed

Crouzon Syndrome - genetic condition which involves abnormal development of the skull and face

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

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

duraplasty - surgical technique where the dura is opened and expanded by sewing a patch into it

flow - the amount of a fluid which moves across a space in a given amount of time; for example one milliliter per second

laminectomy - surgical technique where part of a vertebra is removed

magnetic resonance imaging (MRI) - diagnostic device which uses a strong magnetic field to create images of the body's internal parts

subarachnoid space (SAS) - space in between the tissue of the spine and brain, and their covering, through which CSF flows and circulates

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

syrinx - fluid filled cyst in the spinal cord

ventricle - cavities in the brain which hold CSF

voxel - a single point in an MRI image; like a pixel on a TV or computer screen

Source

Lichtor T, Egofske P, Alperin N.

each heartbeat. In addition, at the level of the syrinx (C3-C4) the cord was swollen, resulting in reduced CSF flow outside the cord in the SAS. However, the most interesting finding before surgery was that what appeared at first to be one syrinx, was actually two distinct syrinxes (see Figure 2), which although next to each other, did not 'communicate'.

After surgery (a standard decompression with duraplasty), the movement of the spinal cord, which was so prominent before surgery, was reduced by more than 60% (see Table 1). In addition, at the level of the syrinx, the cord was much less swollen, allowing the amount of CSF to flow in the subarachnoid space to increase by 50 %. Interestingly, the amount of CSF flowing above the syrinx was virtually unchanged.

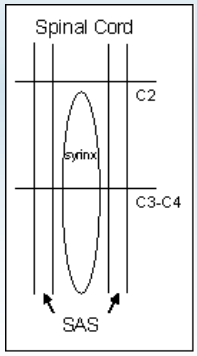
The most striking finding after the surgery was the fact that what used to be two syrinxes was now one. One syrinx had completely resolved, but it's neighbor hadn't. The remaining syrinx did, however, demonstrate only a tiny fraction of the CSF flow inside of it as compared to before the surgery.

The implications of this case as they pertain to current theories on syrinx formation are not immediately obvious. The authors believe this is the first reported case of two adjacent, noncommunicating syrinxes. According to Dr. Lichtor, the lead author, "This the first time CSF flow was carefully measured at two points inside and outside the syrinx cavity before and after surgery. This should lead to a better understanding regarding the natural history of syringomyelia, which patients should undergo surgery and whether or not further surgery is indicated. In particular in most cases after foramen magnum decompression the cyst has decreased in size but is still present. However if the CSF flow studies document, as in this case, that the cyst is not longer active and growing, there is no indication for further surgery."

In science, the true test of a theory is its ability to make predictions. And while imaging techniques continue to evolve, as this and other cases have shown, the current theories on Chiari and syringomyelia have a long way to go in this regard. At this point in time, there is no single theory which can accurately predict who will become symptomatic, who will develop a syrinx, and who will benefit from surgery.

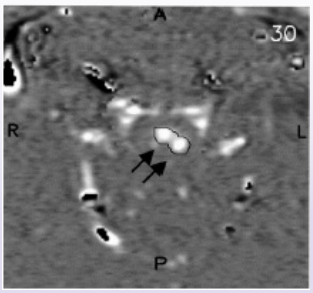
Fortunately, our understanding of these complex diseases continues to evolve and advance, and with each step forward we move close to a day where our understanding will translate into advances in the experiences and outcomes of Chiari and syringomyelia patients.

Figure 1
Location Of CSF Flow Measurements Using MRI



Note: SAS refers to subarachnoid space; upon examination of MRI results, it became clear that the syrinx was actually two distinct syrinxes

Figure 2
MRI Scan Showing Two Distinct Syrinx Cavities



Note: The white in the scan represents CSF flow from the cranium to the spine; black the reverse; arrows show two adjacent, but distinct syrinx cavities

Table 1

Noncommunicating cysts and cerebrospinal fluid flow dynamics in am patient with a Chiari I malformation and syringomyelia--part I. Spine. 2005 Jun 1;30(11):1335-40.

Selected MRI Parameters, Pre and Post Surgery

Parameter	Pre	Post
Cord movement at C2 (mm)	1.2	0.45
SAS CSF volume at C2 (ml)	0.53	0.56
SAS CSF volume at C3-C4 (ml)	0.3	0.45
Syrinx CSF volume at C3-C4 (ml)	0.121	0.023

Note: SAS refers to subarachnoid space; CSF volume refers to the amount of CSF which flows through the region of interest during one heartbeat cycle

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