Key Points

1. Over the past 20 years, posterior fossa decompression has become the standard treatment for Chiari and Chiari related syringomyelia
2. Decompression results in significant improvement in about 80% of patients
3. Not sure exactly why decompression surgery works, but it does create more room and restore CSF flow
4. Study proposes that Chiari, SM, and scoliosis are due to a tight filum terminale, which essentially places the spinal cord in traction
5. Propose a simple surgery, cutting the filum terminal
6. Results are encouraging, but much more research is required

Definitions

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

conus medullarus - bottom, bulbous part of the spinal cord; usually located at L!-L2 level

craniectomy - surgical technique where part of the skull is removed

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

New Surgery Proposed For Chiari

The pace of advancement in understanding and treating Chiari and syringomyelia can seem painfully slow at times. The conditions have been known for more than 100 years, and yet so much about them still remains a mystery. What causes a Chiari malformation? Why do syrinxes form? What is the best way to treat them?

While on a day to day basis it seems like little progress is being made, understanding and treatment have advanced over the years. The MRI brought about a significant change in the way Chiari is diagnosed, and many theories on syrinx formation have been proposed, debated, accepted, and then discarded when new insight was gained. As for treating Chiari, over the course of the last 20 years, the posterior fossa decompression surgery has become the standard, and is used - with variations - worldwide.

Although it is not entirely clear why it works, decompression surgery does result in a significant improvement for about 80% of patients. The goal of the surgery is to create more room around the compressed cerebellar tonsils and to restore the natural flow of cerebrospinal fluid. This is accomplished by removing a piece of the skull, removing part of the top vertebra, sewing a patch into the dura, and in some cases removing part of the tonsils themselves.

Given the popularity and relative success of the procedure (between 3,000 - 5,000 Chiari decompressions are performed each year in the US), much of the medical community has focused on refining the technique, trying to minimize patient trauma, and retrofitting theories on Chiari and SM to the surgery. For example, the prevailing theory on Chiari is that for some reason the posterior fossa does not grow large enough to accommodate the normal sized brain, resulting in herniation of the cerebellar tonsils. Similarly, most theories on syrinx formation have focused on the disruption of CSF flow caused by a Chiari malformation, and the resulting dynamics.

Now, against this backdrop of slow progress, Dr. Miguel Royo-Salvador, of the Barcelona Neurological Institute in Spain, and colleagues from the Universtat Autonoma in Barcelona, have proposed a new theory - and treatment - for Chiari, syringomyelia, and scoliosis.

In a paper posted in the on-line, prepublication section of the journal Acta Neurochirurgica, Dr. Royo-Salvador's group revives an idea which was first proposed almost thirty years ago. Namely, that the cause of CM/SM can be found not at the top of the spine near the skull, but rather at the very bottom of the spine.

The spinal cord is a cord-like collection of nerve tissue which extends from the brain down through the spinal column. Normally, the spinal cord proper ends at the top of the lumbar region of the spine in what is called the conus medullarus. Extending down from here is a ligament type piece of connective tissue known as the filum terminale.

In tethered cord syndrome, a well recognized neurological problem, the spinal cord is abnormally attached to the bones of the spine itself. This results in the tissue of the spinal cord being pulled down. Tethered cord is classically diagnosed when the conus medullarus - the bottom of the spinal cord - is positioned lower than L2. A tethered cord causes incontinence and pain and weakness in the legs and is treated by surgically freeing the cord from the spine.

In his recent paper, Royo-Salvador proposes that the cause of Chiari, syringomyelia, and scoliosis is actually a tight filum terminale. Normally elastic, if this string like connection to the spinal tissue is too tight, it would pull the spinal cord down - like it is in traction - and cause the cerebellar tonsils to descend out of the skull. In addition, he claims that this can be the case even if the conus medullarus is in the normal position.

He goes on to propose a simple treatment based on this theory, namely sectioning - or cutting - the filum terminale to relieve the downward pull on the spinal cord, and presents the results of this type of surgery on 20 patients with Chiari, syringomyelia, and/or scoliosis.

The twenty patients were operated on between 1993-2003. Eight had scoliosis, 5 had syringomyelia, 2 had Chiari, and 5 had some combination of the three. Each patient had their filum terminale sectioned, and in 9 patients (the recent surgeries) the dura was not even opened during the procedure. These patients were able to leave the hospital 24 hours after their operation.

Overall, the doctors report good results with their technique (see Table 1). During the operation, they observed a lack of elasticity of the filum terminale in every patient and noted that it was completely inelastic in 50% of the patients. After cutting the filum terminale, they observed that the spinal cord moved up and relaxed, resulting in a space between the two cut ends. Additionally, there was immediate relief of symptoms for most of the Chiari and syringomyelia patients, especially from headaches and pain.

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

filum terminale - piece of connective tissue which is attached to the bottom of the spinal cord and connects to the bottom of the bony part of the spine

laminectomy - surgical technique where part of a vertebra is removed

lumbar - lower part of the spine

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

scoliosis - abnormal curvature of the spine

section - to cut

spinal cord - thick cord of nerve tissue which extends from the brain down through the spinal column, and from which nerves branch off to different parts of the body

tethered cord - condition where the tissue of the spinal cord is abnormally attached to the bony spine

traction - the act of drawing, or pulling, with force, usually elastic

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

syrinx - fluid filled cyst in the spinal cord

tonsillar herniation - descent of the cerebellar tonsils into the spinal area; often measure in mm

Source

Source: Royo-Salvador MB, Sole-Llenas J, Domenech JM, Gonzalez-Adrio R. Results of the section of the filum terminale in 20 patients with syringomyelia, scoliosis and Chiari malformation. Acta Neurochir (Wien). 2005 Feb 24

All patients were evaluated at a follow-up in 2004, where 5 showed 90% or more clinical improvement, 3 showed 50%-90% improvement, and 6 patients showed 20%-50% improvement. Most patients rated the surgery as either very useful or useful, with only person reporting the surgery was not useful. Five patients were unavailable for follow-up.

In science, any new theory - especially a revolutionary one - should be met with skepticism and evaluated critically. In looking at this new theory, a number of factors should be considered. First, the standard definition of a tethered cord involves the bottom of the spinal cord being located below L2. Yet, the spinal cords of the patients in this study were essentially in a normal position. Thus, the first question to ask is: is it possible to have a tethered cord with the spinal cord in a normal position.

There is indeed evidence that the definition of tethered cord may have to change. Some neurosurgeons are beginning to recognize that cords can be tethered even when in a normal position. In a review of the subject, Tubbs and Oakes (who publish widely Chiari) concluded that there is likely a subset of patients with tethered cords for whom the bottom of the spinal cord is in a normal position. They go on to say that it may be better to interpret tethered cord as meaning tautness of the cord and not the cord being pulled out of position. In further support of this, Selcuki reported that the filum terminale from 8 patients with symptoms of tethered cord, but whose spinal cord was in normal position, were abnormal compared to healthy ones. They found the filum terminale contained dense fibers and tougher connective tissue.

The second question to address is then: can a taught spinal cord cause Chiari? The simple answer here is yes. Sustained downward traction of the spinal cord could cause the cerebellar tonsils to herniate out of position. In addition, as reported in this publication, some researchers are now focusing on the importance of compliance - how easily something expands - in Chiari symptoms. If the dura of the spinal cord is pulled taught, clearly compliance would be reduced and could cause problems.

The final question to address is then: are all Chiari and syringomyelia cases due to a tight filum terminale? The answer here is not nearly as clear. There is no way to draw this conclusion from a single study, and indeed the authors themselves concede this and cite the need for more research - perhaps on animals. In addition, it is not clear how the researchers selected which patients would undergo the new surgery. So if not every Chiari patient has a tight filum terminale, how are patients selected for this surgery? Dr. Terry Lichtor, a neurosurgeon at Rush-Presby in Chicago doubts that this is the case for every Chiari patient, "Patients with a tethered spinal cord may have other associated abnormalities including a Chiari malformation and/or syringomyelia ... the first line of treatment in this patient population should be to release the tethered cord. I think that some of these patients may need other procedures addressing their associated Chiari or syrinx, but in some patients it may be sufficient to just release the tethered cord. However the majority of patients with a syrinx and/or syringomyelia do not have a tethered spinal cord."

While it is exciting just to have a new theory, and a less traumatic treatment, proposed for CM/SM, much more research will be needed to evaluate it's merits. Still it's simplicity, in both cause and treatment, is appealing. It will be interesting to see if there is a day when Chiari patients are treated with sectioning of the filum terminale, posterior fossa decompression, or both.

Editor's Note: *I wanted to include one thought, but was reluctant to put it in the article because it is completely speculative in nature and not based on anything. When I first read this study I was struck by the similarity of what they are proposing - a connection at the bottom of the spinal cord is not elastic - to a finding that most neurosurgeons have made during decompression surgery. Most surgeons have reported that there is a dense, fibrous band which runs from top vertebra to the skull. This membrane is usually cut during the decompression surgery. I think it is interesting that there might be something at both the top and bottom of the spine which is supposed to act like a bungee cord, but in some cases might lose its elasticity and cause problems.*

Table 1
Outcome After Sectioning of Filum Terminale Surgery (10/04)

Patient #	Surgery Date	Clinical Improvement	Patient Description of Surgery
1	10/93	?	?
2	2/94	90%	Very Useful
3	6/94	?	?
4	6/94	50%	Useful
5	7/96	?	?
6	12/97	95%	Very Useful
7	4/98	20%	Not Useful
8	7/98	?	?

9	9/98	90%	Very Useful
10	10/98	10%	Very Useful
11	6/99	100%	Very Useful
12	10/00	60%	Useful
13	6/01	?	?
14	9/01	80%	Very Useful
15	9/01	40%	Useful
16	12/01	40%	Useful
17	2/02	30%	Useful
18	12/02	30%	Useful
19	3/03	40%	Useful
20	9/03	100%	Very Useful

Notes: Follow-up data was not available for 5 patients; patients were asked whether they thought the surgery was very useful, useful, not useful, or harmful