

Key Points

1. Idiopathic syringomyelia accounts for very few of the overall SM cases
2. Since there is no tonsillar herniation, it is usually not treated with decompression surgery; sometimes the syrinx is shunted directly
3. Japanese researchers report 4 cases of idiopathic SM which were successfully treated (3 out of 4) using suboccipital decompression surgery
4. In all 4 cases, the cerebellar tonsils were impacting on the cisterna magna - tight cisterna magna - and likely disrupting normal CSF flow
5. There are other reports of surgeons using decompression surgery in the absence of a herniation
6. This has been termed Chiari 0, but remains somewhat controversial

Definitions

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

cisterna magna - CSF filled space below the cerebellum

Cine MRI - Type of MRI which shows CSF flow instead of a static image

craniocervical junction - the area where the skull and spine meet

dura - thick outer layer covering the brain and spinal cord

duraplasty - surgical procedure where a patch is sewn into the dura

foramen magnum - opening at the base of the skull, through which the spinal cord passes

idiopathic - of unknown cause or origin

laminectomy - surgical removal of part (the bony arch) of one or

Tight Cisterna Magna May Account For Idiopathic Syringomyelia

While most syringomyelia cases are caused by Chiari malformations, trauma, and tumors, there a small, but important, number of cases where the cause is unknown. These cases are referred to as idiopathic syringomyelia. Since there is no real tonsillar herniation, historically these cases have been treated by shunting the actual syrinx as opposed to a Chiari type decompression.

In the last several years however, some surgeons have started to evaluate whether a Chiari type decompression can help with non-hindbrain related syrinxes. As imaging technologies continue to improve, doctors are realizing that even when the tonsils aren't grossly herniated there can obstruction of CSF flow around the craniocervical junction. This can cause Chiari like symptoms and in many theories, may lead to syrinx formation.

Some very well known, and respected, neurosurgeons have reported successful outcomes in employing a decompression surgery on cases where there is little or no tonsillar herniation, but the CSF flow may be blocked. These cases are sometimes referred to as Chiari 0. Unfortunately, the concept of Chiari 0 is controversial. Some of the controversy stems from the difficulty in displacing established ideas, but a lot of it stems from a well publicized case where a surgeon was performing many decompression surgeries for people without any identifiable malformation and as a way to treat Chronic Fatigue Syndrome.

Despite the controversy, in cases of idiopathic syringomyelia, obviously there is something wrong even if the cause is unknown. In work that builds on the previous reports of using decompression surgery for cases without tonsillar herniation, surgeons from Japan have reported good results in treating 4 cases of idiopathic syringomyelia with decompression surgery. Dr. Kyoshima, et al, described their cases in the March, 2002 issue of the Journal of Neurosurgery (Spine).

The four patients included three women and one man ranging in age from 16-52 years old. All four had syrinxes - in different locations - without any tonsillar herniation. The group suffered from standard syringomyelia symptoms such as weakness and loss of sensation in their extremities. Even though the group did not have a classic Chiari malformation, the surgeons found that in each case the cerebellar tonsils were impacting - or crowding - the cisterna magna (fluid filled space below the cerebellum). The researchers refer to this condition as a tight cisterna magna. The results of CSF flow studies using Cine MRI were mixed, with some patients showing clear obstruction and some showing reasonable flow.

All patients underwent a suboccipital craniectomy, C1 laminectomy and duraplasty. Additional techniques - such as removing adhesions - were employed as needed to ensure adequate CSF flow out of the fourth ventricle [Ed. Note: Previous articles in this publication have shown the importance of the free flow of CSF out of the fourth ventricle. Failing to ensure this during surgery can lead to a recurrence of symptoms and the need for more surgery].

The surgeries were fairly successful with 3 out of the 4 patients experiencing long term (9-11 years) improvement in symptoms and a decrease in syrinx size. Unfortunately, after an initial improvement, the fourth patient experienced worsening symptoms and two and a half years after the initial surgery, underwent a second surgery to shunt the syrinx directly. While her syrinx did decrease in size, her symptoms did not improve.

The researchers believe that even if the cerebellum isn't herniated past the foramen magnum in the classical sense, if the tonsils impact on the cisterna magna, they can interfere with the flow of CSF out of the fourth ventricle and eventually cause a syrinx to form. It is not clear how common this situation is as the four cases reported here were observed over a span of more than a decade.

What is clear is that as imaging tools continue to improve, and surgeons look beyond how far the cerebellar tonsils are descended and characterize the whole region, the number of idiopathic syringomyelia cases will likely decrease even further and decompression surgery may be able to help even more people.

more vertebrae

shunt - technique to divert -or
drain - CSF through an inserted
tube

suboccipital craniectomy -
surgical removal of part of the
skull, or cranium, in the back of the
head, near the base

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

ventricle - a cavity in an organ,
the fourth ventricle is a space in
the brain where CSF collects

[Home](#) | [About Us](#) | [Email](#) | [Donate](#) | [Get Involved](#) | [Privacy Policy](#)

Disclaimer: This publication is intended for informational purposes only and may or may not apply to you. The editor and publisher are not doctors and are not engaged in providing medical advice. Always consult a qualified professional for medical care. This publication does not endorse any doctors, procedures, or products.

© 2003-2020 C&S Patient Education Foundation