**Tight Cisterna Magna May Account For Idiopathic Syringomyelia**

While most syringomyelia cases are caused by Chiari malformations, trauma, and tumors, there is 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 be obstruction of CSF flow around the cranio cervical junction. This can cause Chiari like symptoms and in many theories, may lead to syrinx formation.

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