Key Points

1. Study prospectively looked at 16 patients with persistent syringomyelia after decompression surgery
2. Pointed tonsils, disrupted CSF flow, reduced CSF pathways, and elevated CSF pressure in the cervical region indicated that there was still compression around the herniations
3. All patients underwent revisions surgery where surgeons found not enough bone had been removed in some and many had adhesions that were interfering with CSF flow
4. 15 of 16 patients improved long term after revision surgery
5. Syrinx size was reduced, CSF pathways were enlarged, and flow and pressure were normalized in all but one patient
6. Authors believe that many persistent syrinxes may be due to continued blockage of CSF and that if there is evidence of this that additional decompression surgery may be helpful

Definitions

dysesthesia - condition where a normal sensation, such as a light touch, causes pain

dorsal - medical term for upper side or back

foramen magnum - opening in the base of the skull through which the brain and spine meet

jugular compression - a cuff is put around the subjects neck and inflated to create pressure

pc-MRI - phase contrast MRI, type of imaging which can show the flow of CSF

valsalva - activity that involves straining

ventral - medical term for

Study Examines Persistent Syringomyelia

March 31st, 2012 -- While Chiari is the most common cause (if that word is appropriate) of syringomyelia, the exact mechanism involved is not completely understood. The vast majority of researchers believe that it has something to do with the disruption of the natural flow of cerebrospinal fluid (CSF), back and forth between the brain and spine and driven by the cardiac cycle, caused by the herniated tonsils. Indeed, phase contrast MRI consistently shows disrupted CSF flow around the tonsils, but how fluid collects in the spine to form a syrinx has yet to be completely established. And perhaps of more direct importance to patients, scientists do not know why some people develop syrinxes, while others do not.

Despite these gaps in the underlying theory, surgeons have fairly quickly converged on decompression surgery as the treatment of choice in cases with Chiari and syringomyelia. Unfortunately, maybe due in part to a lack of complete theoretical understanding, as with Chiari, syrinxes do not always resolve after surgery.

A short time ago, a group of physicians from the National Institutes of Health and the University of Virginia published a report analyzing sixteen cases of just such persistent syringomyelia. All sixteen were adults who had had prior decompressions surgery 1-5 years previously. None of the patients had shunts of any kind.

The research group prospectively looked at symptoms, MRI features such as syrinx width and length, CSF flow, and even pressure readings of the spinal fluid in the high neck and lumbar areas during rest, Valsalva maneuver, and during jugular compression. Data was collected before revisions surgery, one week after surgery, 3-6 months after surgery, and annually thereafter, with an average follow up time of more than six years. Eighteen healthy volunteers were used as a comparison group.

Prior to revision surgery, more than half the patients had moderate to severe dysesthesia, sensory loss and headaches (Table 1). The average syrinx width was a hefty 7.5mm and the average length was 17.5cm (Table 2). The average herniation size was almost 12mm, with 11 patients showing pointed tonsils indicative of compression. Perhaps most strikingly on MRI, the CSF pathways both in front of and behind the tonsils were significantly smaller compared to the healthy controls. Specifically, behind the tonsils measured only .8mm in the patients, but 8.2mm in the controls. Internal CSF pressure readings in the high cervical area were obtained in 6 patients, and also showed abnormally high pressure. Finally, phase contrast MRI showed disrupted CSF flow around the herniated tonsils. In summary, there was clear evidence that the CSF flow in the patients, despite the prior surgery, was still compromised.

Table 1:

Severity of Selected Symptoms, Pre/Post Revision (16 patients)

	Absent	Mild	Moderate	Severe
Headache	6/13	2/2	6/0	2/1
Dysesthesia	4/7	4/6	8/3	0/0
Subj Weakness	5/11	6/1	0/2	5/2
Sensory Loss	2/1	0/6	9/7	5/2
Exam Weakness	7/11	4/2	3/2	2/1

All patients underwent a second decompression surgery and the surgeons noted that in several cases not enough bone had been removed and in other cases there were extensive adhesions and some other issues. In general, 15 of the 16 patients improved in varying degrees, but even with the second surgery (and a third for shunt placement) one patient did not improve over the long term.

Many symptoms improved after the revision surgery, and follow up imaging showed that the syrinx shrank for 15 of the 16 patients. On average, the width was reduced 2.6mm and the length to 10.7 cm (Table 2). In addition, the tonsils took on a rounded shape for all but one patient. The CSF pathways also increased, with the dorsal ones expanding on average to a sizable 6.8mm. Finally, both the CSF flow and internal pressure returned to closer to normal for most patients.

underside or front

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 I - 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

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

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

Source

Pathophysiology of persistent syringomyelia after decompressive craniocervical surgery. Clinical article. Heiss JD, Suffredini G, Smith R, DeVroom HL, Patronas NJ, Butman JA, Thomas F, Oldfield EH. J Neurosurg Spine. 2010 Dec;13(6):729-42.

Table 2:

Selected Radiographic Features, Pre and Post- Revision (Avg of 16 patients)

	Pre-Revision	Post-Revision
Syrinx diameter (mm)	7.5	2.6
Syrinx length (cm)	17.5	10.7
Herniation (mm)	11.8	7.5
Pointed tonsils (#)	11	1
Rounded tonsils (#)	5	15
Ventral CSF pathway diameter (mm)	1.2	2.1
Dorsal CSF pathway diameter (mm)	.8	6.8

The authors believe that these findings indicate that many syrinxes that fail to resolve are likely due to continued obstruction of CSF flow around the herniation, and that if there is evidence of such, that a second decompression can result in an improved outcome.

Given that fact that the damage from a syrinx can become permanent at any time, the community can hope that our understanding of the connection between Chiari and syringomyelia will continue to evolve and translate to improved outcomes from initial surgeries.

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