

Key Points

- Chiari surgery is generally successful about 80% of the time, however a significant number of people (up to 30%) experience symptom recurrence after some period of time
- CSF flow has been examined as a possible way to predict who will benefit from decompression surgery with mixed results
- Study looked at specific features of CSF flow in 44 children to see if it could predict symptom recurrence
- Specifically cine-MRI was used to examine CSF flow both in the back (dorsal) and front (ventral) of the brainspine junction
- Found that patients who had blocked CSF flow both in front and in back were 2.6 times less likely to experience symptom recurrence
- 6. Authors speculate that patients who show markedly decreased flow may tend to have small posterior fossa volumes and thus respond well to surgery
- It is not clear what the underlying symptomology or best treatment options are for patients with adequate CSF flow but Chiari like symptoms.

Definitions

brainstem - part of the brain which connects to the spinal cord and controls basic functions such as breathing

cine MRI - type of MRI (imaging device) which can measure and show the flow of CSF

CSF flow - the natural movement of cerebrospinal fluid between the brain and spinal cord

cranial nerves - twelve pairs of nerves which originate in the brain

dorsal - towards the back

Details Of CSF Flow Used To Predict Symptom Recurrence

March 31, 2008 -- Although decompression surgery significantly improves symptoms about 80% of the time, long term symptom recurrence - meaning symptoms come back - is a serious problem. Chiari & Syringomyelia News has received many reports from patients who after months (or years) of relief are panicked because their symptoms are coming back. Sometimes they are triggered by a trauma, such as a car accident, but other times there is no readily apparent reason or explanation.

There has not been much research on this phenomenon, especially since many patients stop going back to their neurosurgeon after a year, but recently a group from Johns Hopkins (McGirt et al.) examined whether CSF flow prior to surgery could actually predict which patients will experience symptom recurrence. They published their findings in January, 2008 on-line through the journal Child's Nervous System.

Specifically, the surgeons used cine-MRI to evaluate the pre-operative CSF flow both in front of and behind the brainstem in 44 pediatric Chiari patients. On average, the children were 8 years old and all suffered from symptomatic Chiari verified by MRI. The most common symptom was headache (see Figure 1), with many children also showing signs of cranial nerve involvement and brainstem related symptoms. Eighteen percent were being evaluated for a previous failed Chiari surgery and 18% also had syringomyelia.

While the researchers collected a host of clinical information, their primary focus was on CSF flow. Previous research has focused on whether CSF flow can predict surgical outcomes, and overall has produced mixed results.

There are a couple reasons for this. First, CSF flow characteristics vary from individual to individual even among healthy people. With Chiari, it can be even more dramatic. Two people can have the same level of tonsillar herniation, say 5mm, but have completely different flow characteristics. In fact, a certain percentage of patients with Chiari-like symptoms appear to have normal CSF flow on MRI.

Some researchers have begun to use advanced techniques to actually quantify CSF flow in Chiari patients. However, the quantitative measurement of CSF flow is highly dependent on the exact technique used, resulting in different studies producing different results. For this study however, the doctors used a more qualitative approach and grouped the children into three categories:

- 1. Normal CSF flow
- 2. Blocked flow behind the brainstem (dorsal)
- 3. Blocked flow both in front of (ventral) and behind (dorsal) the brainstem

Interestingly, using this criteria the children were almost evenly distributed between the three groups. In other words, there were 14 children with normal CSF flow, 16 with blocked dorsal flow, and 14 with blocked dorsal and ventral flow (see Figure 2).

All the children underwent decompression surgery, with the decision on whether to open the dura made during the procedure itself. After surgery, the children were evaluated 1 month, 3 months, 6 months, and yearly thereafter. At the time the study was written up for publication, the average follow-up duration was 27 months.

Overall, the researchers found that about 30% of the children experienced symptom recurrence at some point during the follow-up period. Interestingly, many occurred after the 3 year mark, indicating that a typical one year follow-up period may be inadequate.

The team also found that size of herniation had no relationship with which children had symptom recurrence. Similarly blocked CSF flow in the back of the cranio-vertebral junction (dorsal) also did not correlate with symptom recurrence. However, children with blocked flow both in front and in back were 2.6 times *less* likely to have symptoms recur. In other words, symptom recurrence rates were about the same in the normal flow and blocked dorsal flow groups, but significantly reduced in the blocked ventral and dorsal flow group. In fact, in the last group, only one child had symptoms come back.

Although this study has a number of limitations - symptom recurrence was not well defined, and it's not clear if the researchers controlled for whether the dura was opened for example - it does raise some very interesting questions. Namely, what is different between the groups of children such that some have long-term success with surgery, while others only experience relief for a period of time.

One possibility which the authors discuss is that the children with blocked flow on both sides may have smaller

dysphagia - trouble swallowing

posterior fossa - region in the back of the skull where the cerebellum is situated

recurrence - reappearance of symptoms after treatment

ventral - towards the 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

Source

McGirt MJ, Atiba A, Attenello FJ, Wasserman BA, Datoo G, Gathinji M, Carson B, Weingart JD, Jallo Gl. <u>Correlation of hindbrain CSF</u> flow and outcome after surgical <u>decompression for Chiari I</u> <u>malformation.</u> Childs Nerv Syst. 2008 Jan 19; [Epub ahead of print] posterior fossa volumes. Research has shown that people with small posterior fossa's (as measured on MRI) often respond well to surgery. But then begs the question as to what is the underlying cause of Chiari in the other cases, and what would the appropriate treatment be?

Towards this end, Conquer Chiari has begun to try to influence researchers to move beyond thinking of Chiari as tonsillar herniation and to figure out ways to classify people according to what is causing the herniation, for example small posterior fossa, or altered hydrodynamics. Once the underlying causes of Chiari are understood and can be readily identified in patients, appropriate treatments can then be developed.

Figure 1 Selected Presenting Symptoms (44 Children)

Symptom	Number With	Percent With
Headache	35	80%
Brainstem, Cranial Nerve	19	43%
Dysphagia	11	25%
Balance Problems	8	18%
Motor Deficits	5	11%

Figure 2 CSF Flow Characteristics (44 Children)

CSF Flow	Number (Percent)
Normal	14 (32%)
Abnormal Dorsal Flow	16 (36%)
Abnormal Dorsal & Ventral Flow	14 (32%)

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