Key Points

1. For reasons that are not clear, Chiari surgery fails to provide any symptom improvement 15%-20% of the time
2. In 2003, Dr. Bejjani hypothesized that some Chiari patients also have pseudotumor cerebri (PTC), which is why symptoms came back quickly after surgery
3. This study reviewed 192 Chiari surgery patients and identified 36 patients who did not improve
4. 15 of those 36 were found to have signs/symptoms of (PTC)
5. These patients were treated with shunts to relieve the pressure in the skull
6. Shunts did little to help the adults, but the children improved significantly
7. The link between Chiari and PTC is not yet understood

Definitions

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

hydrodynamics - the study of how fluids move

idiopathic intracranial hypertension - another term for pseudotumor cerebri

intracranial pressure (ICP) - the pressure of the spinal fluid inside the skull

lumbar puncture - technique where a needle is inserted into the spine, near the bottom; can be used to deliver medicine, drain CSF, or take a pressure reading

posterior fossa - region of the skull where the cerebellum is normally situated

pseurdotumor cerebri (PTC) - condition where intracranial pressure is elevated above normal for an extended period of time, for no readily identifiable reason

Pseudotumor Cerebri Linked To Failed Chiari Surgery

January 20, 2006 -- For many Chiari patients, decompression surgery provides at least some level of symptom relief. However, the medical literature has consistently revealed that between 15%-20% of Chiari patients do not improve with surgery (or their symptoms come back shortly after).

The reasons for this are not clear. One study (Trying to identify why surgeries fail) looked at failed pediatric surgeries and identified complex anatomy and failure to ensure CSF flow out of the 4th ventricle as major reasons for failure.

In 2003, Dr. Ghassan Bejjani (a scientific advisor to the Foundation and this publication) hypothesized that for at least a subset of patients, the problem was not the surgery itself, but rather a second condition, known as pseudotumor cerebri (PTC).

PTC, known formally as idiopathic intracranial hypertension is a poorly understood condition which involves chronically elevated pressure of the cerebrospinal fluid in the skull. Pressure in the skull can be elevated from a tumor, or hydrocephalus, but in PTC, it is not really known why the pressure is raised. Interestingly, the symptoms can be very similar to Chiari, and include pressure headaches and vision problems.

Now, a study out of the University of Chicago by Fagan, Ferguson, Yassari, and the well known neurosurgeon Frim, adds substantial evidence to the hypothesized PTC-Chiari connection. Published in the January, 2006 issue of the journal Pediatric Neurosurgery, the Chicago team reviewed the records of 192 Chiari surgeries and identified 15 patients for whom decompression surgery failed, and who appeared to have PTC.

Specifically, they looked back at 192 surgeries they had performed (they excluded patients with abnormal skull shapes and hydrocephalus) and found that surgery had failed in 36 cases (19%). They then identified PTC among the 36 failures with the following criteria:

1. Cine MRI showed adequate CSF flow out of the 4th ventricle and across the skull-spine junction
2. Chiari-like symptoms came back after decompression surgery
3. Elevated CSF pressure as measured by a lumbar puncture (see Tables 1,2)
4. Symptoms were temporarily relieved after draining a large amount of CSF (through the lumbar puncture) and reducing the CSF pressure in half

Using these criteria, the researchers found 15 patients, 6 adults and 9 children, who were classified as having PTC after their decompression surgery. The symptoms they suffered from included head pain, body aches, balance problems, and visual disturbances. The fifteen PTC-Chiari patients accounted for nearly half of the failed surgeries (42%) and 8% of the entire surgical group.

Five of the six adults and all of the children underwent a second operation to insert a shunt in order to drain CSF and lower their intracranial pressure. Unfortunately, only one of the adults showed any improvement after this procedure. However, 7 of the 9 children did improve significantly with the shunting. These results are not as good as those from Bejjani's study, but the number of patients in each study is too small to draw any conclusions from regarding treatment.

While this study provides strong evidence of a link between Chiari and PTC, what exactly that link is remains a mystery. Does Chiari, or even decompression surgery lead to PTC? Is PTC another cause of Chiari? Or, are the conditions just different manifestations of a deeper problem?

Although they didn't include them in their study, the authors also reported on 3 patients who were initially diagnosed with PTC. MRI then showed they had asymptomatic Chiari. Interestingly, all three eventually developed Chiari symptoms and required decompression surgery. In these patients, the PTC developed first, so it might be possible that the increased pressure associated with PTC eventually pushes the cerebellum out of the skull and leads to Chiari.

However, it is much to soon to say for sure. In most of the patients in this study, it appeared that PTC developed after the decompression surgery. So it may be that the blockage associated with a Chiari somehow leads to PTC; or that decompression surgery itself may alter the hydrodynamics of the CSF system and lead to PTC.

Finally, it may be possible that both conditions are a result of something else, like a small posterior fossa. If the skull is too small, it may lead to PTC in some people, Chiari in other people, or even both for yet other people.

shunt - tube like device which is implanted in the body to drain, or divert, CSF

ventricle - one of several CSF filled spaces in the brain

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

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 (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 measured in mm

Source

Fagan LH, Ferguson S, Yassari R, Frim DM. The Chiari pseudotumor cerebri syndrome: symptom recurrence after decompressive surgery for Chiari malformation type I.
Pediatr Neurosurg. 2006;42(1):14-9.

Currently, the only way to measure CSF pressure is invasively (usually through a lumbar puncture); however several groups have tried developing a non-invasive method over the years. Perhaps soon technology will enable us to easily measure pressure in Chiari patients and help shed more light on the PTC-Chiari connection. Until then, it is worth noting that in this study, nearly half of the failed surgeries showed indications of pseudotumor cerebri.

Figure 1
PTC/Chiari In Adults

Patient #	Time to PTC (months)	Max CSF Presssure (cm H2O)	Symptom Resolution
1	0.7	52	None
2	9	24	None
3	5	18	None
4	12	16	None
5	2	28	None
6	6	18	Minimal

Figure 2
PTC/Chiari In Children

Patient #	Time to PTC (months)	Max CSF Presssure (cm H2O)	Symptom Resolution
1	8	30	Significant
2	8	28	Significant
3	12	19	Mild
4	48	01	Mild
5	8	23	Significant
6	21	13	Significant
7	1	27	Significant
8	7	34	Significant
9	3	24	Significant

Note: Normal CSF pressure is 15-20 cm of water; CSF pressure varies throughout the day and due to a number of factors

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