Key Points

1. Study looked at 6 Chiari patients for whom decompression surgery failed
2. Symptoms came back around 6 months after initial surgery
3. Follow-up MRI's showed CSF flow behind the cerebellar tonsils
4. Eye exam showed signs of elevated ICP
5. Lumbar puncture relieved symptoms for about 1 week
6. Shunt was placed in 4 patients
7. 2 patients chose periodic lumbar punctures combined with medicine to control ICP
8. All patients showed improvement for at least 18 months after follow-up procedures
9. Researchers speculate that at least some of the 6 suffered from idiopathic intracranial hypertension in addition to having Chiari malformation
10. Exact relationship between IIH and Chiari is not thoroughly understood

Definitions

acetazolamide - also known as Diamox; medicine used to lower elevated ICP

cerebellar tonsils - portion of the cerebellum located at the bottom, so named because of their shape

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

Chiari malformation - 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

dura - thick outer layer covering the brain and spinal cord

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

Treatment Options After Failed Surgery

A neurosurgeon from the University of Pittsburgh believes that some cases of failed Chiari decompression surgery may be due to a partial misdiagnosis and co-existence of another condition, idiopathic intracranial hypertension (see Side Bar). Dr. Ghassan Bejjani along with neuro-opthamologist, Dr. Kimberly Cockerham, and their colleagues reported on 6 cases of failed decompression surgery in the February, 2003 issue of the European journal Acta Neurochirurgica.

Bejjani's study looked at Chiari patients whose primary symptoms returned after decompression surgery. The six cases included 5 women and 1 man ranging in age from 19 to 43 years. Each patient had a Chiari malformation verified by MRI and neurological symptoms consistent with Chairi and severe enough to warrant intervention. As to be expected, the most common symptom was headache along with neck pain, visual disturbances, and weakness and numbness in arms and legs. None of the six had a syrinx.

The patients all underwent similar decompression surgeries involving a suboccipital craniectomy, C1 laminectomy, and duraplasty. For all six patients there was some symptom relief initially, but symptoms began to return 1 to 9 months later. The patients underwent follow-up MRI's, including cine-MRI to look at CSF flow. The MRI's were reviewed by a neuroradiologist, who did not know the purpose of the study or the clinical reports of the patients, to assess whether there was adequate decompression. The radiologist looked at whether the tonsils had moved up or down after surgery, the shape of the tonsils, and whether there was CSF flow behind the tonsils. There was very little movement of the tonsils as compared to before surgery, but in 4 of the patients the tonsils were more rounded in shape. In addition, there was CSF flow behind the tonsil in all 6 patients. While not conclusive, these findings are suggestive that surgically, the decompression was adequate and should have relieved the symptoms.

In addition to the radiological review, 4 of the 6 patients underwent an eye exam by Dr. Cockerham. All four patients showed signs of elevated intracranial pressure (ICP). Specifically, the patients lacked what are known as spontaneous venous pulsations (SVPs). In most people, the veins in the retina (in the back of the eye), fluctuate in size periodically. In a person with elevated ICP, these fluctuations do not occur. Besides the absence of SVP's, two of the patients showed additional signs of elevated ICP.

Following the MRI's and eye exams, all six patients underwent a lumbar puncture to both measure their ICP and relieve it if it was elevated. Most of the patients were near or above the pressure level considered normal and all patients reported a temporary (about one week) improvement in symptoms following the lumbar punctures. Because of this, four patients chose to have ventriculo-peritoneal shunts placed in an effort to drain CSF and lower their ICP. Two patients chose to undergo periodic lumbar punctures along with taking acetazolamide, a drug which can lower ICP. Following these treatments, all patients reported significant improvement in their symptoms and were stable at least 16 months later.

Why did CSF drainage - using shunts or lumbar punctures - work where decompression surgery failed? The researchers cite several possibilities, including surgical scarring disrupting CSF flow and inadequate decompression, but speculate that at least some of the patients also suffered from idiopathic intracranial hypertension - a condition where ICP is abnormally high for unknown reasons. Bejjani points out that in the case of surgical scarring, the lumbar punctures should not have relieved symptoms as the procedure is below the level of CSF blockage. While it is difficult to define an adequate decompression, the MRI's for most patients in this study showed signs that are typical of an adequate decompression.

In support of the idiopathic intracranial hypertension (IIH) theory, Bejjani points out that IIH symptoms, as reported, are similar to Chiari and could be difficult to distinguish. In addition, the eye exams and elevated pressure readings after surgery are suggestive of IIH. Adding to this theory is that some of the patients were overweight, a predisposing factor for IIH. Perhaps the strongest piece of evidence that supports the IIH theory however, is that the patients responded to treatments designed to drain cerebrospinal fluid and didn't respond to the decompression surgery.

An association between IIH and Chiari has been identified and discussed previously in the medical literature, but the exact nature of the relationship is not known. Does a Chiari malformation cause elevated ICP, or can sustained high pressure actually cause the cerebellar tonsils to herniate? Much more research will be required to sort out the link between the two conidtions.

Failed decompression surgery can be devastating to a patient and happens much too frequently. While there are likely many reasons why surgeries fail - as reported elsewhere in this issue - for one group of patients, even if its a small group, the reason may be because of a second, often treatable, condition.

ICP - intracranial pressure; pressure of the CSF inside the skull

Idiopathic Intracranial Hypertension (IIH) - condition where ICP is elevated for unknown reasons; see other side bar

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

lumbar puncture (LP) - procedure where a hollow needle is inserted into the spinal area (near the base) in order to measure/relieve pressure or withdraw CSF for testing

papilledema - swelling/bruising of the optic nerve due to increased ICP

pseudotumor cerebri - another name for IIH; so named because the symptoms mimic the presence of a tumor

spontaneous venous pulsations (SVP) - periodic changes in size of veins in the retina (in the back of the eye); absence indicates elevated ICP

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 - CSF filled spaces in the brain

ventriculo-peritoneal shunt - neurosurgical procedure where a tube-like device is inserted into a ventricle to divert and drain CSF into the peritoneal space in the abdomen

- Condition where intracranial pressure (ICP) is abnormally high for unknown reasons; normal ICP is considered <20cmH2O (water)
- Also called pseudotumor cerebri; first identified in the late 19th century
- Most common symptoms are severe headache and visual disturbances
- Can lead to blindness from pressure on the optic nerve
- Diagnosed when an MRI is normal and lumbar puncture shows elevated ICP
- Eye exam may reveal papilledema, a sign of increased ICP
- Number of people affected is not well established, but some estimates are as high as 1 in 100,000
- More common among overweight women of childbearing age
- Usually treated medically with drugs that lower ICP
- Some patients require surgery around the optic nerve to prevent blindness
- In some patients, a shunt is placed to drain CSF out of the brain

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For More Information Visit:
The Intracranial Hypertension Research Foundation - www.ihrfoundation.org

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