Key Points

1. There is controversy over when to perform surgery for Chiari, especially if headaches are the only symptoms
2. Study looked at 33 pediatric Chiari patients with headaches as only symptom
3. Headaches were classified as frontal, occipital, or generalized; cine MRI was used to also classify CSF flow as obstructed or normal
4. Patients with occipital headaches were 10 times more likely to have obstructed CSF flow and 8 times more likely to have tonsillar herniation greater than 7mm
5. 17 patients underwent decompression surgery
6. Surgery was successful for all 10 with occipital headaches

Definitions

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 - 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

cine MRI - type of MRI which is capable of showing CSF flow

craniectomy - surgical technique where part of the skull is removed

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

Type Of Headache May Indicate If Surgery Is Required

With no objective test to say when decompression surgery is necessary, it is no surprise that there is little agreement on this topic in the medical community. A recent, worldwide survey (Survey Shows How Doctors Worldwide Treat Chiari) of neurosurgeons highlighted the controversies surrounding the management of this disease. While some cases are easy - a syrinx with progressive symptoms - cases where headaches are the only symptom are particularly troublesome. The question boils down to, are the risks of the highly invasive surgery worth the benefit in these cases?

According to Dr. Mathew McGirt of Johns Hopkins Medical Center, and his colleagues at Duke University, the answer may depend on the type of headache the Chiari patient has. This was the finding of a study the team performed on 33 pediatric Chiari patients whose only symptoms were headaches. They reported their results in the April, 2005 issue of the journal Neurosurgery.

Between 1998 and 2002, the researchers identified children who had a Chiari malformation greater than 5mm and who were suffering only from headaches. Children with syringomyelia, hydrocephalus, scoliosis, or other abnormalities in the region of the skull/spine junction were excluded from the study. Thirty-three children were identified, with an average age of 14. There were 14 girls and 19 boys with an average tonsillar herniation of 8.8mm

Each child's headaches were classified as being frontal, generalized, or occipital, based on where they started. In addition, each child underwent a cine-MRI to examine their CSF flow. From this test, each child was also classified as having obstructed flow (if flow was completely blocked in one of several areas), or having non-obstructed flow. Seventeen of the children then underwent a standard decompression surgery, including lamiectomy and duraplasty, and were evaluated again one year later.

Of the 33 patients, 11 suffered from frontal headaches, 7 from generalized headaches, and 15 from occipital headaches. When the researchers compared headache types to CSF flow, they found a significant relationship between occipital headaches and both obstructed CSF flow, and a greater degree of tonsillar herniation (see Table 1). In fact, patients with occipital headaches were ten times more likely to have obstructed CSF flow and 8 times more likely to have herniations greater than 7mm, than patients with either frontal or generalized headaches. In contrast, there was no association between headache type and age, sex, or length of symptoms.

Table 1
Characteristics of Headache Only Chiari Patients (33)

Characteristic	Frontal or Generalized Headache (18)	Occipital Headache (15)	Significant?
Avg. Age	14	15	N
Female	39%	46%	N
Male	61%	54%	N
Length of symptoms (mo's)	14	15	N
Herniation >7mm	33%	80%	Y
Obstructed CSF flow	17%	66%	Y

As stated previously, 17 patients were selected to undergo decompression surgery. They included 7 patients with frontal headaches and 10 patients with occipital headaches. The surgical group was evaluated one year later to determine if the surgery had eliminated the headaches. The researchers found that the surgery was completely successful for all 10 of the patients with occipital headaches, but was only successful for 3 out of 7 of the frontal headache group (see Table 2). In looking at the frontal headache group more closely, the two patients who also had obstructed flow improved with surgery. This means that in the surgical group as a whole, 12 out of 12 (100%) of patients with obstructed CSF flow improved with surgery, whereas only 1 patient out of 5 (20%) with normal flow improved.

The authors interpret these results as indicating that occipital headaches - which are associated with obstructed CSF flow - are likely a direct result of the Chiari malformation; whereas frontal and generalized headaches may have several causes which may not be linked to the tonsillar herniation at all. They go on to say that this means that surgery should be considered to treat occipital headaches (when Chiari is demonstrated), but other treatments should be considered for frontal and generalized headaches.

In comments published in the same journal, Dr. Thomas Milhorat - a Chiari pioneer - points out that the study has several limitations. Age and sex matched controls were not used to identify the general rate of headaches

dura - tough, outer covering of the brain and spinal cord

duraplasty - surgical technique where the dura is opened and expanded by sewing a patch into it

laminectomy - surgical technique where part of a vertebra is removed

magnetic resonance imaging (MRI) - diagnostic device which uses a strong magnetic field to create images of the body's internal parts

occipital - referring to the back of the skull

spinal cord - thick cord of nerve tissue which extends from the brain down through the spinal column, and from which nerves branch off to different parts of the body

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 measure in mm

Source

McGirt MJ, Nimjee SM, Floyd J, Bulsara KR, George TM. Correlation of cerebrospinal fluid flow dynamics and headache in Chiari I malformation. Neurosurgery. 2005 Apr;56(4):716-21.

and the classification of headaches based solely on location is of limited value.

Despite it's limitations, this study does support previous research findings on a number of fronts. A recent study by Heiss (What causes the dreaded Chiari cough headache?) showed that a cough related headache is a strong predictor of CSF blockage. In addition, a recent study from India (Studying CSF Flow To Predict Surgical Outcome) demonstrated that Chiari patients with blocked CSF flow improved more with surgery than Chiari patients with normal CSF flow.

While it is likely that the management of Chiari cases - especially ones where headaches are the only symptom - will remain controversial for quite some time, it is encouraging for patients to see research which attempts to link symptoms to objective tests. Given recent imaging advances, and a focus on the importance of parameters beyond simple CSF flow (Intracranial Compliance Linked To Surgical Success), perhaps the elusive goal of an objective "Chiari" test is at last on the horizon.

Note: Significant refers to whether there is a statistically significant correlation between the characteristic and the type of headache

Table 2
Surgical Outcome One Year Post-Op (17 Patients)

	Headache	No Headache
Occipital, Obstructed Flow	0	10
Frontal, Obstructed Flow	0	2
Frontal, Normal Flow	4	1

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