

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

decompression surgery
- common term for any of several variations of a surgical procedure to alleviate a Chiari malformation

duraplasty - surgical procedure where a patch is sewn into the dura, the covering of the brain and spinal cord

foramen magnum - opening at the base of the skull, through which the spinal cord passes

intrathecal pressure - pressure of the CSF within the spinal area

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

lumbar puncture - procedure where a hollow needle is inserted into the spinal area to withdraw CSF; can be used to measure CSF pressure

pathophysiology - the physical or functional changes associated with a disease

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

What causes the Chiari cough headache?

Headache is one of the most frequently reported symptoms associated with Chiari, and for many people the most troublesome. A particularly unpleasant variation of the Chiari headache is the one associated with coughing. I speak from experience. I underwent corrective surgery during the winter, and I remember dreading every cough brought on by the dry winter air leading up to the surgery.

Some researchers have speculated that headaches are aggravated during a cough by tonsillar movement compressing nerve roots in the spine. Another headache theory is based on a difference in pressure between inside the skull and inside the spinal area. However, neither of these theories has substantial data to back them and they are not well accepted by the research community.

Now, a new theory has emerged involving a sharp increase in pressure in the spinal area during a cough. Dr. Sansur, Dr. Heiss and their colleagues at NIH (National Institute of Health) reported on their theory in the March, 2003 issue of the Journal of Neurosurgery in a study titled, *Pathophysiology of headache associated with cough in patients with Chiari I malformation*.

The NIH group hypothesized that the peak intrathecal pressure during coughing would be higher in Chiari patients who suffer from cough headaches than in Chiari patients who don't get cough headaches and in healthy people. As part of an ongoing NIH study, the researchers evaluated a group which included: 26 adults with Chiari I and syringomyelia, 4 adults with just Chiari I, and 15 adult, healthy volunteers. Of the 30 patients, 11 suffered from cough headaches (interestingly, all 4 Chiari only patients had cough headaches, but only 7 of the CM/SM patients had cough headaches).

At the start of the study, the participants were evaluated by measuring their internal spinal pressure using lumbar puncture. First, a baseline pressure was established, then measurements were taken during coughing, jugular compression [Ed. Note: I can't even stand to button the top buttons of my shirt and these poor people had a cuff placed around their neck and inflated for 10s!], and while blowing into a tube (Valsalva maneuver).

Patients subsequently underwent a suboccipital craniectomy, C-1 laminectomy, and duraplasty. Six months after the surgery, the patients intrathecal pressure was measured again at baseline, during cough, during jugular compression and during Valsalva maneuver.

What the researchers found is that before surgery, the intrathecal pressure in Chiari patients who suffered from cough headaches was significantly higher during coughing than in both Chiari patients without headaches and the healthy volunteers. In addition, the Chiari cough patients had the highest baseline pressures. The pressure during jugular compression and the Valsalva maneuver was not that different among the groups.

After corrective surgery, both the baseline pressure, and pressure during cough, for the group with headaches had come down to essentially the same level as the other groups. Perhaps more importantly, the cough headaches completely went away in 10 of 11 patients and improved in the last patient. Despite strong evidence that supports their original theory, Dr. Heiss believes this is the true value of the study for patients, "For patients with headaches that are made worse with coughing, [surgical decompression] was shown to be very effective for relieving or improving the headaches. Neurosurgeons and neurologists tend to focus on neurological deficits such as paralysis, weakness, and loss of sensation, and pay less attention to symptoms of headache, which are very frequent in Chiari patients...[patients] are thankful when their headaches are relieved after surgery."

As for why some people with Chiari get cough headaches and some don't, Dr. Heiss says, "The narrowness of the CSF pathways at the foramen magnum varies among patients with Chiari I and syringomyelia...people with cough headache have more narrowing of the CSF pathways than those without. In addition, some people are just more prone to headaches in general."

While the NIH research revealed a possible cough headache mechanism, i.e. a spike in pressure during cough, testing pressure this way is impractical in a clinical setting. But the research also showed that having the symptom of cough associated headache is a strong predictor of CSF blockage and should be considered by physicians when evaluating patients. Unfortunately, many Chiari patients go years suffering from these types of headaches before being properly diagnosed; maybe now physicians will be more aware of the Chiari-cough headache connection.

Editor's Note: While at NIH, Dr. Sansur was a senior level medical student participating in a clinical research training program. He is now a neurosurgical resident at the University of Virginia Neurosurgery Department.

Meet The Surgeon:

Valsalva maneuver - breathing out in a straining manner

vertebra - segment of the spinal column

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- **Neurosurgical Residency**

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Selected Publications:

- Heiss JD, Oldfield EH: **Pathophysiology and Treatment of Syringomyelia**. Contemporary Neurosurgery 25(3):1-8, 2003
- Heiss JD, Oldfield EH: **Syringomyelia and related diseases**. In: Weatherall D, Nathan D, editors. Encyclopedia of Life Sciences (Nature, Scientific American). London: Macmillan Reference Limited, 2001.
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