

Key Points

1. Headache is the most common symptom reported by Chiari patients
2. For years, headache researchers have classified provoked headaches as due to cough, exertion, and sexual activity and either primary or secondary in nature
3. Study looked at provoked headache patients and used imaging for every patient
4. Provoked headaches accounted for 1.5% of all headache cases seen
5. Among provoked headaches, 70% were cough headaches
6. Among the cough headaches, half were due to Chiari; Chiari also accounted for one third of all the provoked headaches
7. Chiari headaches differed from primary cough headaches in location, duration, age of onset, and years with headache
8. Strong evidence that people with cough headaches, and possibly everyone with a provoked headache, should get an MRI

Definitions

bilateral - affecting both the left and right side

CT - computerized tomography; type of X-ray which uses computers to take detailed pictures from different angles

foramen magnum - opening at the base of the skull through which the brain and spine meet

hemispheric - on one side of the head

indomethacin - an anti-inflammatory drug used to treat migraines

intracranial - inside the skull

lesion - an area of abnormal tissue in the body

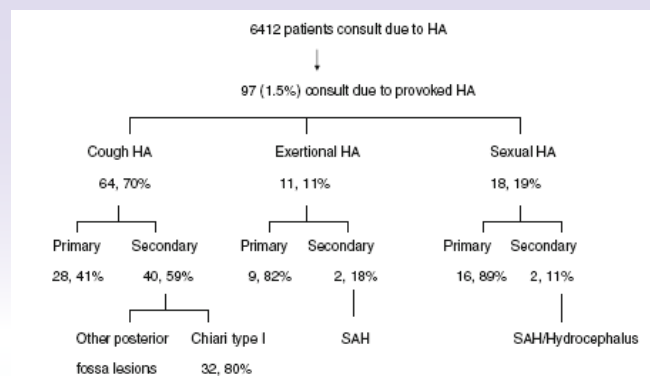
Chiari Accounts For Half Of All Cough Headaches

November 30, 2008 -- A new study out of Spain reports that Chiari accounted for only one half of one percent of headache cases seen in a major neurology clinic, but accounted for one half of all cough provoked headaches. The study (Pascual et al.), published in the October, 2008 issue of the Journal of Headache & Pain, also found that headaches due to Chiari have some unique characteristics which could help in reducing the time to diagnosis. [Ed Note: As an aside, a couple of years ago I was at a presentation being held by a neurosurgeon who was speaking on Chiari. The person sitting next to me was a neurologist, who like many, came in somewhat skeptical about Chiari in general. In talking with him, I made the statement that I thought the Chiari headache was somewhat unique, but he disagreed and told me that many people, especially men, develop exertional type headaches in middle age. Well, as you will read in this article, I may have been correct. To his credit though, after he heard the presentation on Chiari, he changed his views on Chiari in general.]

The Spanish research was not focused specifically on Chiari, but rather was a broad, prospective look at provoked headaches in general. Provoked headaches, which are brought on by something, have been studied since the 1920's. Since that time the medical community has moved towards classifying them as being provoked by cough, prolonged physical exercise, and sexual activity. These types of headaches are further classified as being primary, meaning not related to a physical lesion inside the head, or secondary, meaning they are related to an intracranial lesion. Historically, this classification scheme has been developed based mostly on symptoms. What the Spanish researchers wanted to do was look at the characteristics of provoked headaches combined with modern imaging.

To accomplish this, they enrolled patients seen at their clinic for provoked headaches over a 10 year period. Each patient was followed for at least one year and evaluated through structured interviews. Headache characteristics, location, duration, quality, and treatment effectiveness were recorded. In addition, anyone with a cough headache was given a cranio-cervical MRI (note, those found to have Chiari were also given a CSF flow study); patients with exercise and sexual activity headaches were initially given a CT and most were given an MRI as well.

Figure 1: Distribution of Headache Type (6412 Total)



Out of 6,412 patients seen for headaches over that time period, only 97 were determined to have provoked headaches (1.5%, Figure 1). Among the 97 with provoked headaches, cough headache was the most common (70%), followed by sexual headache (19%) and exertional headache (11%) respectively. Within the cough headache group, 59% were found to be secondary to a lesion, which the majority of the time was a Chiari malformation. In total 32 patients out of the provoked headache group were found to have Chiari malformation on MRI.

Interestingly, this represents only 0.5% of the total number of cases seen for headaches. Patients often wonder why Chiari is hard to diagnose, and this may be one reason. While headache is the cardinal symptom of Chiari, headaches are so common, that Chiari only accounts for a small fraction of the patients evaluated. However, as these researchers also found, there are characteristics of Chiari related headaches, which should make it easier to spot.

First, it is important to note that while Chiari only made up a tiny fraction of the total headache population, Chiari cases actually accounted for one third of the provoked headache group. This is a significant number and should be a strong indication that an MRI might be warranted in evaluating any patient with a provoked headache. This number is also much higher than has been previously reported in the literature; however, the authors point out that previous studies did not involve MRIs for every patient, so it is likely that many Chiari cases were missed.

MRI - magnetic resonance imaging; device which uses magnets to create detailed pictures of internal organs and body parts

occipital - referring to the back of the head

primary headache - headache not associated with an intracranial lesion

provoked - brought on by something

secondary headache - headache associated with an intracranial lesion

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

syringomyelia - condition where a fluid filled cyst forms in the spinal cord

Before jumping to an MRI, the study also uncovered some additional clues that a cough headache may be due to a problem like Chiari as opposed to primary in nature. On average, the secondary cough headache group was much younger than the primary cough headache group (44 vs 60, Figure 2). In addition, 70% of the secondary cough headaches were located in the back of the head, while only 11% of the primary cough headaches were. Secondary cough headaches on average lasted longer and the patients had endured them for an average of 5 years versus only 1 year for the primary headaches. While most of the primary cough patients did not have other symptoms, more than 80% of the secondary cough patients had additional symptoms, such as dizziness and vertigo, indicative of a problem in the posterior fossa. Finally, in terms of non-surgical treatment, every case of primary headache responded to the drug indomethacin, while the same drug was not effective in treating the secondary cough patients.

The CSF analysis of the Chiari patients clearly showed there was flow blockage at the level of the foramen magnum. Interestingly, it also showed that in patients who required surgery, there were intracranial pressure spikes which disappeared after surgery.

This study presents powerful evidence that anyone with a cough induced headache should be evaluated for Chiari, especially if the headache is in the back of the head, have been occurring for more than a year, and are accompanied by other symptoms. The argument could also be made, based on this research, that anyone with a provoked headache should undergo imaging as nearly half of the cases were due to a physical lesion.

Even though not every Chiari patient suffers from headaches, disseminating findings such as these throughout the medical community is an important step in reducing the amount of time it takes for Chiari patients to be diagnosed. It also points out the importance of patients being clear and precise in describing their symptoms to doctors.

Figure 2: Characteristics of Primary vs Secondary Cough Headache

		Prim.	Sec.
Total #		28	40
Male/Female %		36/64	30/70
Avg Age of Onset		60	44
Location	Hemicranial	50%	14%
	Bilateral	39%	14%
	Occipital	11%	72%
Quality	Electrical	22%	21%
	Explosive	17%	21%
	Pressing	17%	31%
	Mix	44%	24%
Duration of Pain	Seconds	78%	45%
	Minutes	11%	10%
	Variable	11%	45%
Years w/Headache		<1	5

Notes: 80% of secondary headaches were associated with a Chiari Malformation

Source

Headaches precipitated by cough, prolonged exercise or sexual activity: a prospective etiological and clinical study. Pascual J, González-Mandly A, Martín R, Oterino A. J Headache Pain. 2008 Oct;9(5):259-66

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