

Key Points

1. It often takes years to diagnose Chiari, which results in poorer outcomes
2. Study prospectively analyzed the self-reported symptoms of 265 Chiari patients (with or without SM) before any surgery was performed.
3. 3 patients were asymptomatic, but 95% reported 5 or more symptoms
4. Average duration of symptoms was 6.3 years
5. 49 separate symptoms were reported by 2 or more patients (see Tables below)
6. Most common symptom was headache (98%)
7. Other common symptoms authors believe were related to CM/SM included neck pain, burning pain in cervical/thoracic region, dizziness/balance problems, extreme fatigue or general body weakness
8. Chiari headache is described differently than typical tension and migraine headaches

Definitions

apnea - temporary stop in breathing, sleep apnea

asymptomatic - having no symptoms

cephalgia - headache

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 (CM) - 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

hypertension - abnormally high blood pressure

Large Study Reveals Wide Range Of Chiari Symptoms

A study published in the March, 2004 issue of the Journal of the American Academy of Nurse Practitioners has verified what Chiari patients have known for years; namely, that while some symptoms are common to many Chiari sufferers, the malformation also causes an array of symptoms which can vary widely from person to person. Indeed, one of the more common email questions this publication receives is asking whether X, Y, and Z symptoms are related to Chiari.

This plethora of Chiari related symptoms is not only a source of frustration for patients, but it is also a contributing factor to the all too often missed and delayed diagnoses that many patients endure. Both statistical (see Key Points #4) and anecdotal evidence show that many patients go through doctor after doctor, and years of suffering, before being correctly diagnosed. The varied, vague, and sometimes fleeting symptoms can make an early diagnosis difficult. This not only takes an emotional toll on the patient is who being given ineffective treatments, or being told it's all in their head, but some research has shown that the longer someone is symptomatic before treatment, the less chance they have of a good outcome.

In an attempt to help Primary Care Providers (PCPs) diagnose - or at least suspect - Chiari, Dr. Diane Mueller, a Neurosurgery Nurse Practitioner, and neurosurgeon, Dr. John Oro, undertook a study to examine what symptoms patients with MRI verified Chiari first talked about (or to use the medical term, presented). What they found was both encouraging for PCP's making earlier diagnoses and validating for every patient who has a laundry list of symptoms.

In the study, Dr. Mueller analyzed the self-reported symptoms of 265 Chiari patients who ranged in age from 12-78. The group included 221 women and 44 men. The size of the malformations ranged from 3mm-30mm, with most patients falling in the 5mm-10mm range. Thirty-seven patients had syrinxes, although not every patient underwent a full spine MRI. Interestingly, 3 patients had thoracic syrinxes, without the more common cervical syrinx. Also of note is that 3 patients were asymptomatic.

The encouraging part of what the researchers found is that 13 symptoms were reported by more than half the patients (see Table 1), with 98% of them saying they suffered from headaches. While the headache finding is not surprising, Dr. Mueller points out that the way Chiari headaches are usually described by patients is different than standard tension and migraine headaches. Chiari headaches are usually described as starting in the back of the head and sometimes radiating forward to behind the eyes. The pain is described as a feeling of intense pressure, or even explosive in nature, and is brought on or aggravated by straining (Valsalva maneuver), coughing, sneezing, posture, singing, laughing, etc. The study also noted that some patients had modified their lifestyles to avoid situations, such as singing or laughing, which would bring on the headaches. As PCP's become aware of the distinct nature of the Chiari headache, they should be able to make a diagnosis (or at least order an MRI) earlier in the disease progression.

The validating part of the research - at least from a patient's perspective - is the tremendous variation among symptoms. Forty-nine distinct symptoms were reported by two or more people and 95% of the patients reported 5 or more symptoms (see Table 2). As demonstrated by the extensive list, Chiari can affect many different parts of the body in many different ways. And of course, once the body is compromised for an extended period of time, secondary factors, like depression, come into play.

Often patients look for validation from their doctors that the symptoms they are experiencing are real and want to know if they're related to Chiari. Unfortunately, all too often doctors ignore this need and casually dismiss patient reports as if they were wielding a line item veto; related to Chiari, not related to Chiari, all in you head, etc. Given the results from this study, perhaps both doctors and patients should take a more holistic approach when dealing with symptoms. Patients should look inward for validation and doctors should take into account the complex nature and varied presentations of this disease and not try to categorize every symptom.

Ed Note: *I thought it might be useful for some people to read about the varied symptoms that I enjoyed just prior to surgery, so that they have a real example of what this study demonstrates. Here are the ones I can remember:*

Extreme headache especially with coughing, climbing stairs, etc; extreme neck pain; shoulder pain; weakness in right hand (hand started to curl up); leg weakness; balance problems aka walking into doorways; fullness and ringing in ears; hoarseness, trouble finding the right words (brain knew what to say, but voice didn't listen); trouble swallowing aka gagging on many types of foods; light sensitivity; frequent urination... You get the idea.

Table 1
Symptoms Reported By At Least 50% Of Patients

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hypotension - abnormally low blood pressure

nystagmus - involuntary, rapid eye movements

photophobia - sensitivity, sometimes painful, to light

prospective - type of study where the experiment is designed before the data is collected

syringomyelia (SM) - neurological condition where a fluid filled cyst forms in the spinal cord

tachycardia - rapid heart beat

thoracic - relating to the middle part of the spine, or chest area

tinnitus - ringing in the ears

Valsalva maneuver - straining

Symptom	%
Headache	98
Dizziness	84
Difficulty sleeping	72
Weakness in arms/hands	69
Neck pain	67
Numbness/tingling in arm, hand	62
Fatigue	59
Nausea	58
Shortness of breath	57
Blurred Vision	57
Tinnitus	56
Difficulty swallowing	54
Leg weakness	52

Table 2
Symptoms Reported By Less Than 50% Of Patients

Source

Source: Mueller DM, Oro' JJ. Prospective analysis of presenting symptoms among 265 patients with radiographic evidence of Chiari malformation type I with or without syringomyelia. J Am Acad Nurse Pract. 2004 Mar;16(3):134-8.

- (20%-50%) - depression, body weakness, balance problem, memory problems, leg/foot numbness, hoarse voice, chest pain, facial numbness, anxiety, slurred speech, arm pain, abdominal pain, photophobia
- (<20%) - tachycardia, trouble hearing, vomiting, double vision, word-finding problems, vision loss, blackouts, apnea, vertigo, loss of peripheral vision, nystagmus, earache, nosebleeds, snoring, thoracic pain, hypotension, wake up choking, leg pain, palpitations, hypertension, abnormal gag reflex, face pain/tingling

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