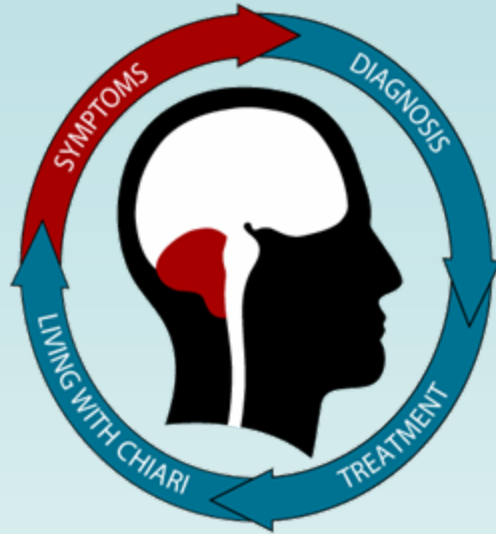




Chiari Malformation: **Symptoms**

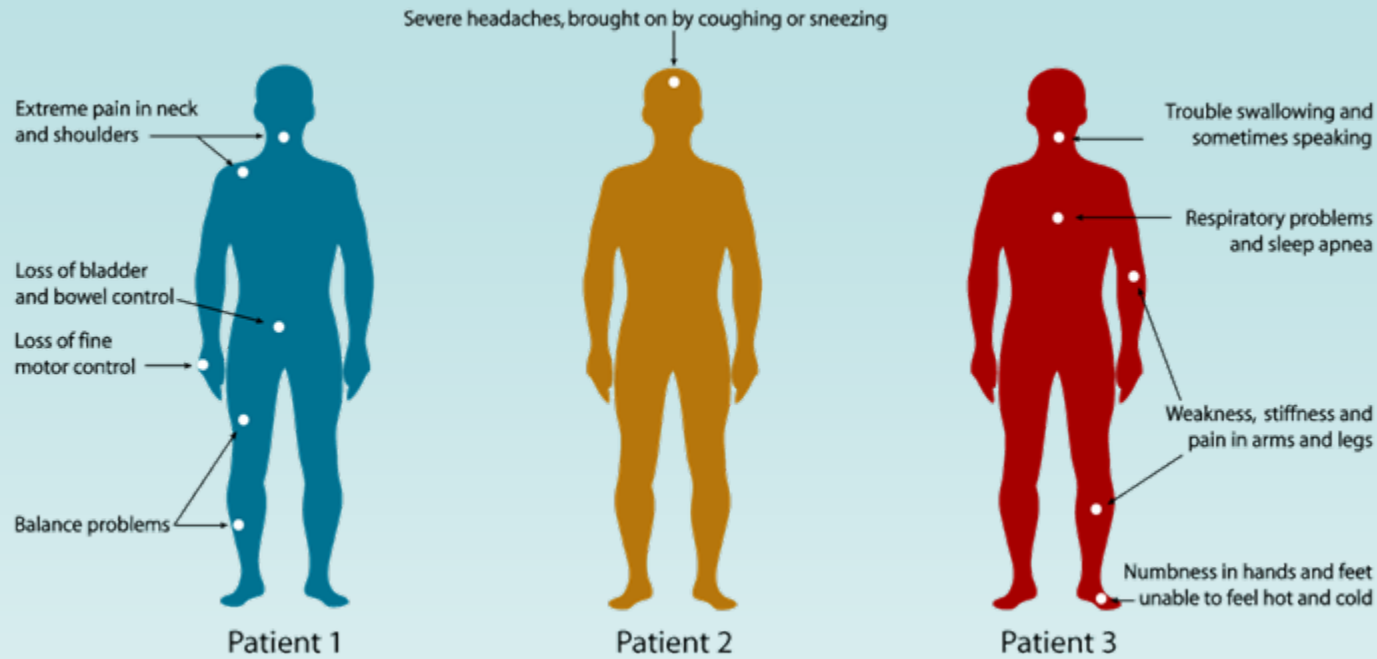


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Every Case is Different



Though there are common symptoms associated with Chiari, the specific set of symptoms and the severity of those symptoms (symptomatology) varies from person to person.

Chiari symptoms are often complicated by related conditions: syringomyelia, tethered cord, intracranial hypertension, bony abnormalities, and genetic disorders, being the most common.

Research has shown that the amount of tonsillar herniation is not strongly related to symptom severity. In other words, patients with relatively small herniations can have severe symptoms, while patients with large herniations can be symptom free.

What Causes Chiari Symptoms?



- Direct compression of the cranial nerves
- Direct compression of the brainstem
- Direct compression of the cerebellum
- Disruption of the natural flow of CSF
- Elevated CSF pressure in the skull/brain
- Damage to nerves in the spine

- ▮ There is anecdotal evidence that head and neck trauma can spark symptoms.
- ▮ Other medical conditions (related and unrelated) may trigger symptoms and/or aggravate existing ones.
- ▮ Simple things like bending over, laughing, coughing and sneezing are common triggers for a “Chiari” headache.

Most Common Symptoms



A 2004 study of 265 adult patients (Mueller,Oro) documented the wide range of symptoms that can accompany Chiari. **13 symptoms** were reported by more than half of the patients.

Headache was the most commonly reported symptom with 98% of the group claiming they suffered from them.

20%-50% reported depression, body weakness, balance problems, memory problems, leg/foot numbness, hoarse voice, chest pain, facial numbness, anxiety, slurred speech, arm pain, abdominal pain, and photophobia.

Less than 20% reported tachycardia, trouble hearing, vomiting, double vision, vision loss, blackouts, apnea, vertigo, loss of peripheral vision, nystagmus, earache, snoring, thoracic pain, hypotension, wake up choking, leg pain, palpitations, hypertension, gag reflex, and face pain/tingling.

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

95% of patients experienced at least 5 symptoms

Headache



Chiari headaches are usually described as starting in the back of the head (suboccipital) and sometimes radiating forward to behind the eyes.

The pain is described as a feeling of intense pressure and is brought on or aggravated by straining (known as a Valsalva maneuver), coughing, sneezing, posture, singing, laughing, etc.

Although most Chiari experts agree there appears to be some type of connection between Chiari and migraines, the nature of the connection is unknown.



Headache, in the back of the head, is the defining Chiari symptom

Neck/shoulder/upper back pain



Pain in the neck, shoulder, and upper back is another common problem with Chiari due to several causes:

- nerves are compressed by herniated tonsils
- unusual anatomy in the upper spine
- nerve damage due to syrinx (Syringomyelia)

Once a muscle is affected by nerve damage in the neck and shoulders, it can affect how the rest of the muscles work.

Instead of working together to perform a task, muscles start to work against each other which in turn leads to pain and loss of function.

For reasons that are not entirely clear, it appears that the right side of the neck and shoulder is affected more often than the left side.

Research indicates a high rate of scoliosis among Chiari patients.

Eye Issues



Chiari can affect the eyes in several ways: blurred vision, double vision, sensitivity to light and near-sightedness.

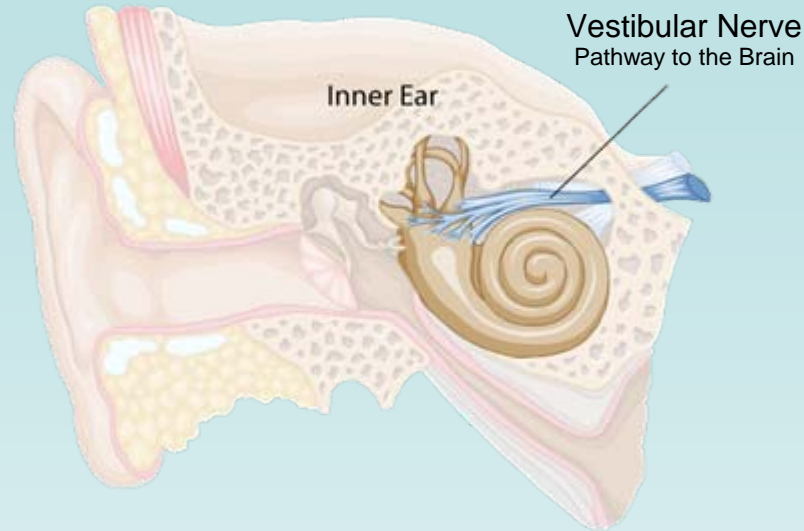
One of the most common eye problems is a rapid, involuntary movement of the eyes known as **nystagmus**.



There may be a connection between Chiari and **strabismus**, when the eyes don't align properly (example: cross-eyed, lazy eye).

- One study found that strabismus related to Chiari can be treated effectively with decompression surgery, rather than the standard strabismus eye surgery
- In these cases Chiari may affect the muscles which position the eyes properly, or it may be that Chiari patients are just more prone to developing strabismus

Ears/Balance Issues



||| The inner ear has fluid-filled structures which send balance information to the cerebellum.

||| If the cerebellum is compressed, like with Chiari, the human balance system or (vestibular system) may not work properly, which may cause tinnitus (ringing in the ears), depth perception, running into walls and tripping.

Sleep Apnea



A published study in France (Dauvilliers et al.) showed that 73% of adults and 60% of children with Chiari suffer from some form of sleep apnea, a disorder characterized by repeated incidents where a person stops breathing, partially wakes up, then starts breathing again.

There are three types of sleep apnea:

Obstructive sleep apnea occurs because something physically blocks, or obstructs, the airway.

Central sleep apnea, the problem lies with the respiratory control center itself, which fails to signal the body to breathe.

Mixed apnea refers to someone who suffers from episodes of both types.

Untreated, sleep apnea can cause high blood pressure and other cardiovascular disease, memory problems, weight gain, headaches and in serious cases, death.

Decompression surgery reduces the occurrences of Chiari related sleep apnea by 90%

Cognitive Issues



Cognitive: *Of or pertaining to the mental processes of perception, memory, judgment, and reasoning*

What We Know:

- Many adult Chiari patients report problems with thinking, memory, and processing information
- Some pediatric Chiari patients have trouble in school and experience developmental delays
- While originally believed to be involved mainly in coordination of movement, the cerebellum is now believed to be involved in many higher order cognitive functions

Possibilities:

- Chiari directly causes cognitive issues
- Chiari indirectly causes cognitive issues: chronic pain, medications, lack of sleep, depression....
- Chiari does not cause cognitive issues either directly or indirectly

While to date there have been few publications on the cognitive impact of Chiari, Conquer Chiari has funded research in this area

Emotional Issues



Emotional: *associated with mood, temperament, personality, and disposition*

What We Know:

- Mueller found that close to half of Chiari patients in a large series reported depression
- Research has also shown that mood and anxiety disorders are more common with chronic disease and pain
- Chronic pain depletes the levels of key neurotransmitters in the brain which are known to affect mood
- Pain and depression share the same neurochemical pathways in the brain; so pain can influence mood at the most basic level and vice versa

What We Don't Know:

- If Chiari directly causes depression and mood problems
- The effect of a doctor telling a Chiari patient their symptoms are all in their head has emotional well being
- The best treatments for Chiari patients with depression or mood/anxiety disorders

Feelings of depression or anxiety should be reported to your physician

Pediatric



Presenting symptoms for children are often different than adults.

■ A 2002 study (Greenlee, Menezes) found that oropharyngeal problems – having to do with the throat – were the most common presenting symptom among very young children (3 and under).

- Common symptoms included: choking, trouble swallowing, stridor, chronic cough, and poor weight gain



■ Among older children, scoliosis - an abnormal curvature of the spine - is a common symptom.

- Although scoliosis among teens is not unusual, Chiari related scoliosis often involves unusual curvature types and locations

■ While some Chiari children experience seizures, the connection between Chiari and seizures is not clear.

For more information on pediatric Chiari, visit www.conquerpediatricchiari.org

All Is Not Chiari



Excerpted from “All is Not Chiari” by Ray D’Alonzo

“Chiari patients often experience a wide array of symptoms and have a natural tendency after a while to assign all of their symptoms and problems to Chiari. Care must be taken however not to assign all symptoms to Chiari. It is certainly possible to have two diseases at once. When this occurs we call it a concomitant condition. Over the years, I have read numerous posts of Chiari e-group sites where patients wonder if Chiari can be responsible for abnormal thyroid function or blood sugar levels. It is critical for Chiari patients when new symptoms or changes in lab test result values emerge to discuss these changes with their doctor and resist the tendency to ignore them by assigning their origin to Chiari. While Chiari is known to be associated in rare cases with serious heart and respiratory problems, it is not known to be associated with thyroid disease or diabetes. And, even if the Chiari patient is having heart or respiratory symptoms, he/she should not ignore them just because there is a known association. Remember, all is not Chiari.”

Consult a doctor before assuming a symptom is related to Chiari, it may be a sign of another illness that needs immediate attention

Strange Names



Words you may hear during your doctor visit and what they mean.

| Name | What it is | Name | What It Is |
|-------------|--|---------------|---|
| Allodynia | pain in response to something that should not cause pain | Malaise | a feeling of discomfort or uneasiness, "general feeling of being unwell" |
| Aphasia | trouble finding the right words to use when speaking | Nystagmus | rapid involuntarily eye movements |
| Ataxia | difficulty walking due to loss of muscle coordination | Photophobia | sensitivity to bright lights |
| Atrophy | wasting away of muscle | Strabismus | cross-eyed |
| Cephalgia | headache | Stridor | A harsh, high-pitched sound in inhalation or exhalation. |
| Diplopia | double vision | Syringomyelia | neurological condition where a fluid filled cyst forms in the spinal cord |
| Dysesthesia | unusual, unpleasant sensations | Tachycardia | rapid heart beat |
| Dysphagia | trouble swallowing | Tinnitus | ringing in the ears |
| Kyphosis | hunchback | Vertigo | dizziness |

Conquer Chiari – Learn More



Articles:

- [Large Study Reveals Wide Range of Chiari Symptoms](#)
- [What causes the dreaded Chiari cough headache?](#)
- [Minor Head/Neck Trauma Sparks Chiari Symptoms In Some](#)
- [Looking Back: Milhorat Redefines Chiari...](#)
- [Study Identifies Features Of Chiari Related Scoliosis...](#)
- [Treating Chiari Related Eye Problems](#)
- [Looking To The Ear For Treatment Guidance](#)
- [Majority Of Chiari Patients Suffer From Sleep Apnea](#)
- [Chiari Related Memory & Learning Problems](#)
- [Trouble Swallowing May Signal Chiari In Very Young Children](#)
- [More Than Half Of Patients With Chronic, Disabling Spinal Problems Suffer From Major Depression](#)

Books:

- [Conquer Chiari: A Patient's Guide](#)
- [Contents Under Pressure](#)

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