

## Key Points

1. Chiari is usually thought of as a chronic condition and people often go years before being diagnosed
2. Authors report two cases where symptoms came on suddenly and the patients were in an emergency situation
3. 16 year old boy with worsening headaches, neck pain, trouble swallowing and breathing
4. After Chiari was found on MRI, immediate decompression surgery stabilized his symptoms
5. 7 year old boy with no history of problems became rapidly weaker on one side and showed abnormal neurological signs
6. Decompression surgery was performed immediately and symptoms improved gradually
7. Few reports of rapid onset of symptoms in literature; however several reports of sudden death
8. Compared to the total number of Chiari cases; rapid onset and sudden death are rare events

## Definitions

**anisocoria** - refers to when the pupils of the eyes are different sizes; an indication of injury or disease

**brain stem** - part of the brain that connects to the spinal cord and controls basic, critical functions like breathing and heart rate

**dysphagia** - trouble swallowing

**hemiparesis** - weakness on one side of the body

**hypopnea** - slow or shallow breathing

**intubation** - procedure where a tube is inserted into the trachea of someone who can't breathe

## Urgent Surgery Is Sometimes Necessary For Chiari

July 31, 2007 -- Chiari Type I is generally thought of as being chronic in nature. In fact, the average patient suffers with symptoms for years before being diagnosed. However occasionally (perhaps rarely is a better word) a person's symptoms can come on suddenly, progress rapidly and turn into an emergency situation.

Wellons et al. report on two such cases in the July, 2007 issue of the Journal of Neurosurgery: Pediatrics. Their first case involved a morbidly obese 16 year old boy who went to the emergency room with rapidly worsening headaches, neck pain, weakness on one side of his body, trouble swallowing and shallow breathing. In fact, he was having so much trouble breathing, the ER doctors had to intubate him. All of his problems had started less than two days earlier and his doctors initially suspected he had suffered a stroke.

Because of his size, he had to be sent to a different facility for an MRI which did not show any signs of a stroke, but did reveal a large Chiari malformation but no syrinx. Given his deteriorating condition and his inability to breathe and swallow, he underwent decompression surgery less than 24 hours later.

After surgery, his arm and leg strength improved, but he still had trouble swallowing and a feeding tube was inserted. He was transferred to a rehab facility and eventually the feeding tube was removed. When he was finally sent home, his only symptom was mild weakness on his right side.

Their second published case was a 7 year old boy with no significant medical history who experienced rapid onset weakness on the right side of his body. There was no indication of a head trauma, but his neurological exam showed significant abnormalities, including his left pupil being a different size than his right. As with the first case, stroke was initially suspected, but an MRI revealed an 11mm herniation with a syrinx the size of his entire spinal cord. Interestingly, the boy said he did not have any headaches or neck pain.

An emergency decompression was performed and the boy gradually recovered his strength over the course of a couple of months. After one year, his strength was near normal, and the syrinx was reduced in size, but still present.

The authors point out that there are very few reports in the medical literature of rapid onset of symptoms, and the few reports that do exist tend to be adults and involve respiratory distress. They also stress that Chiari was found in these patients only because stroke was suspected by the ER doctors so MRIs were ordered. In a sense, the patients were lucky.

Although not discussed in the article, that raises the question of what happens if someone isn't lucky enough to have the Chiari found during an emergency situation, or can symptoms come on so quickly that nothing can be done? A PubMed search by Chiari & Syringomyelia News revealed several case reports of sudden death attributed to Chiari (see Table 1).

The case reports include:

- Three people, one child and two adults, who died suddenly after a minor trauma (to the head); Chiari was found during autopsy
- Several cases of sudden respiratory arrest, including two children. It should be noted that recently a strong association between Chiari and sleep apnea has been found.
- An adult with a history of headaches and fainting. The headaches were diagnosed as migraines, but the person died suddenly and Chiari was found after they died.
- Two 25 year old men

Sudden death due to Chiari is thought to be related to brain stem malfunction, likely due to the pressure placed on it by the herniation.

While sudden death, and even rapid onset of symptoms, can be frightening, it is important to put these events into perspective.

The PubMed search revealed about 10 reports of sudden death related to Chiari since 1984 (the earliest report). During that time there have been thousands and thousands of people with Chiari who have lived with their symptoms, or no symptoms, and did not require emergency care. Assuming conservatively that about 10,000 people per year are found to have Chiari on MRI (in the US alone), that would translate to 230,000 people since 1983 with Chiari. Thus, 10 cases out of more than 200,000 is a miniscule number.

Of course, the one unknown in an analysis like this is that we don't know how many deaths might be due to

**respiration** - breathing

**respiratory arrest** - when someone stops breathing

**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

## Source

Wellons JC 3rd, Tubbs RS, Bui CJ, Grabb PA, Oakes WJ. [Urgent surgical intervention in pediatric patients with Chiari malformation type I. Report of two cases.](#) J Neurosurg. 2007 Jul;107(1 Suppl):49-52.

Chiari which are not discovered and we don't know how many deaths, even if they are attributed to Chiari, are not reported in the medical literature. In fact, Conquer Chiari has received a couple of emails from family members whose relatives were found to have Chiari after their unexpected death.

But even accounting for these unknowns, it is likely that sudden death and even the emergency onset of symptoms is a rare event compared to the total number of Chiari cases.

**Ed Note:** *Have you, or do you know someone, who experienced a rapid onset of severe symptoms? If so, we'd like to hear from you. Send us a short description of what happened to [director@conquerchiari.org](mailto:director@conquerchiari.org).*

**Table 1**  
**Selected Published Reports of Sudden Death Related To Chiari 1**

Year	Age	Notes
1984	3	Minor Trauma
1993	Two Children	Respiratory Arrest
1995	25	--
1998	71	Minor Trauma
1998	21	Minor Trauma
1999	Adult	Headaches, Fainting
2003	25	--

**Notes:** Taken from a PubMed search with the terms "Chiari sudden death"

### Related C&S News Articles:

[Chiari Patients May Be At Greater Risk With Head Trauma](#)

[Study Looks At Abnormal Gag Reflex In Chiari Children](#)

[Effects Of Minor Head Trauma](#)

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