

### Key Points

1. While the average time to diagnosis is years for adults, in some cases symptoms come on rapidly and can be severe
2. Group identified 6 such pediatric cases representing 3% of surgical cases and less than 1% of total Chiari cases
3. 5 of the 6 had syrinxes
4. In three children the symptoms were sparked by a minor trauma
5. All underwent surgery within a week and symptoms were completely resolved 12 months after surgery
6. The connection between trauma and Chiari symptoms is not known or understood
7. There is no general guidance on whether kids with Chiari (both pre and post surgery) should be restricted from contact sports, sports in general, or other activities

### Definitions

**acute** - in terms of symptoms, of short duration but often severe

**chronic** - ongoing

**hemianesthesia** - inability to feel touch sensations on one side of the body

**paraparesis** - partial paralysis of the lower limbs

**parasthesias** - abnormal sensations due to nerve issues

**quadraparesis** - partial paralysis of upper and lower limbs

**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,

### Some Chiari Cases Develop Acute Symptoms

September 30th, 2011 -- While the common perception of the Chiari diagnostic process is one involving years of frustration and many doctors, there is a subset of patients for whom symptoms develop suddenly and are severe in nature. Because of the symptom severity, Chiari is usually found fairly quickly and surgery is performed on an urgent basis. In a recent publication in the Journal of Neurosurgery: Pediatrics, a group of doctors from the St. Louis Children's Hospital describe their experience with 6 such cases.

In a retrospective study, the group reviewed all the Chiari cases seen between 1990 - 2008 for cases involving a sudden onset of significant neurological deficits. The six cases they found - 3 boys, 3 girls, ranging in age from 3-14 years - represented only 3 % of the 189 surgical cases during that time period. Unfortunately, complete records for non-surgical Chiari evaluations were only available starting in 1994, but since there were close to 500 cases during that shorter period of time, it seems reasonable to deduce that the 6 rapid onset cases represent 1% or less of the total Chiari cases seen.

Five of the six children had syrinxes and the sixth had changes to their spinal cord which were evident on MRI (referred to some as a pre-syrinx). Three of the children reported that their symptoms started immediately after a minor trauma, including a blow to the head during football, a fall from the standing position, and after doing a flip on a trampoline (Table 1).

**Table 1: Selected Characteristics of 6 Sudden Onset Cases**

Age	Syrinx?	Trauma	Deficit
12	Yes	No	quadriparesis
13	Yes	No	parasthesias
3	No	No	vocal cord paralysis
14	Yes	Yes	parasthesias
10	Yes	Yes	paraparesis
13	Yes	Yes	hemianesthesia

The quickly developing symptoms included partial paralysis of the arms and legs, loss of sensation, abnormal sensations, and in one case vocal cord paralysis. Chiari was diagnosed quickly in all cases and the average time to surgery was about a week after symptom onset.

Interestingly, one child's symptoms had resolved before surgery, but the doctors decided to proceed with surgery. In the rest of the cases, the symptoms were significantly improved by 2 weeks after surgery and completely resolved by 1 year. Follow-up MRIs were only available for 4 of the children, but in those 4 the syrinx size was significantly reduced.

Although there is only a limited amount of information that can be gleaned from this report, it does touch on a number of key questions that the Chiari community is currently wrestling with.

First, what is the role of trauma in either sparking or aggravating symptoms? Milhorat found that a significant number of patients reported some type of trauma as sparking their symptoms, but to date there are no good theories on what mechanism might link trauma to sparking Chiari symptoms. In addition, a number of Chiari cases are found after motor vehicle accidents, but in those cases it can be difficult to sort out whether symptoms such as neck pain are due to Chiari or whiplash.

Another question is whether people who are found to have Chiari incidentally and have no symptoms, should do anything about it. While there are some reports that the vast majority of asymptomatic cases stay that way for at least several years, it could be that if these people experience a trauma, they may become symptomatic. However, given that there is no good theory on what sparks Chiari symptoms and what role trauma might play, combined with the fact that the percentage of sudden onset cases is likely very low, makes it impossible to predict which people with incidental Chiari are at risk of developing symptoms.

Finally, this leads to a topic which generates considerable debate among the medical community, namely whether there should be any sports restrictions on children both with untreated herniations and/or after decompression surgery. The authors discuss this issue and correctly point out that there is little research and no generalized guidance in this regard, which leads to individual doctors giving their own - often differing - advice to families. The senior author notes that he counsels families that if there is a syrinx there may be an increased risk for problems associated with contact sports even after surgery. But the question remains, how much risk?

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

Is it worth limiting a child's experience? Unfortunately, these questions remain problematic and difficult to address

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

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[Effects Of Minor Head Trauma](#)

#### Source

[Patients with Chiari malformation Type I presenting with acute neurological deficits: case series.](#)  
Yarbrough CK, Powers AK, Park TS, Leonard JR, Limbrick DD, Smyth MD. J Neurosurg Pediatr. 2011 Mar;7(3):244-7.

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