

## Key Points

1. Nomenclature surrounding Chiari and syringomyelia can be very confusing; not all doctors use terms to mean the same things
2. Chiari I, Chiari II, Arnold-Chiari are not used consistently
3. It is thought that Chiari I and Chiari II are caused by two different things
4. However, some people have features of each
5. Study examined the MRI's of 22 Chiari 1.5 children
6. No single symptom or sign was found specific to Chiari 1.5
7. Surgery was not as successful for children with syrinxes as compared to Chiari I cases
8. Not clear whether Chiari 1.5 is related to Chiari II.

## Definitions

**brainstem** - part of the brain, at the base, which connects the rest of the brain to the spinal cord; controls basic functions such as breathing and reflexes

**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 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

**Chiari malformation 1.5** - name used in this study to refer to people who have Chiari I, but also have brainstems which have descended out of the skull and do not have spina bifida

**Chiari II** - more serious type of Chiari malformation where parts of the brainstem, in addition to the

## Between Chiari I & Chiari II

What's in a name? With the name Chiari, quite a bit. When Hans Chiari first wrote about the malformations (in the 1890's) that would later be named for him, he identified different types based upon what brain structures were displaced out of the skull. Type 1 was defined as the cerebellar tonsils descending past the foramen magnum; Type II included parts of the brainstem as well.

Unfortunately, as time has passed, what started as a something fairly straightforward has proven to be anything but. Today, a newly diagnosed patient is confronted with a confusing litany of "names", such as Arnold-Chiari, tonsillar herniation, tonsillar ectopia, etc. To make matters worse, doctors - and researchers - do not always use the terms to mean the same things. For example, some people use the term Arnold-Chiari in referring to Chiari Type II, while others use it as a general term. Even Chiari Type II is not clearly defined; some doctors use it only in reference to Chiari related to spina bifida, although this definition is not universal.

The confusion doesn't stop with the brain, it extends to the spine as well. The terms hydromyelia, syringomyelia, and hydrosyringomyelia are even more loosely defined. Some doctors will use the term syringomyelia only to refer to a syrinx in the tissue of the spine and use hydromyelia to refer to one in the central canal. However, since the central canal closes in most adults, and where a syrinx is located exactly is not always clear, nomenclature that splits hairs like this is hardly productive.

One result of this confusion is that overwhelmed patients often feel like they have been transported through the looking glass. While it is easy to wonder why everyone can't agree on something so basic as a name, the current state of affairs is actually a reflection of two things: the complexity of the Chiari/syringomyelia complex and the lack of true understanding regarding the causes and mechanisms of each.

The reality is that what is generally referred to as Chiari can vary quite a bit from person to person. There are a virtual smorgasbord of bony abnormalities that often accompany the tonsillar herniation, and any given person may have from one to many of them.

The many possible manifestations of Chiari is one reason it has been difficult to achieve consensus on the naming. Should Chiari I and Chiari II be considered two instances of the same disorder, and the difference just a matter of degree; or are they distinct entities?

On the one hand, most researchers believe that the underlying cause is different for Chiari I and Chiari II. Chiari II - as defined in association with spina bifida - is believed to occur because the opening in the lower part of the spine which defines the disorder allows CSF to leak. The CSF leak in turn causes a pressure difference between the skull and spine areas and results in the cerebellar tonsils and the brainstem descending out of the skull. In contrast, the current theory on the origin of most Chiari I malformations is that the bony space of the posterior fossa (the part of the skull where the cerebellum rests) does not grow large enough to accommodate the growing brain, and so the tonsils end up outside the skull.

Unfortunately, human anatomy and disease are rarely simple and straightforward. There are some people who have both the cerebellar tonsils and part of their brainstem located below the skull (just like Chiari II), but do not have spina bifida. So what do we call these people?

A recent publication in a November, 2004 supplement to the Journal of Neurosurgery used the term "Chiari 1.5" to refer to cases such as this. A group of doctors from the University of Alabama and the University of Wisconsin reported (Dr. Tubbs was the lead author) on their experience with 22 children who fall somewhere in between Chiari I and Chiari II.

In an attempt to describe the Chiari 1.5, the researchers studied the patient's MRI's and measured the amount of tonsillar herniation, brainstem herniation, and the angle of the odontoid process (a structure which projects up from the 2nd vertebra, through the top vertebra) (see Table 1). They then compared the MRI characteristics to the symptoms and neurological signs of the group.

On average the group was 12.5 years old and 50% of them had syrinxes (see Table 2). Interestingly, their symptoms were nearly identical to what is found in Chiari I and the researchers could not identify a single symptom or sign that was peculiar to Chiari 1.5. Similarly, when they looked at the MRI measurements the only meaningful relationship they could find was between the amount of tonsillar and brainstem herniation. They could not tie any of the symptoms back to any of the MRI findings.

Surgically however, the authors did note that the Chiari 1.5 group did not fare as well as a comparative Chiari I group. In the Chiari 1.5 group 18 patients improved with surgery, but 13.6% of the group required a second

cerebellar tonsils, are displaced out of the skull; associated with spina bifida

**ectopia** - when a body part is out of position; tonsillar ectopia is another name for Chiari malformation

**foramen magnum** - opening at the base the skull where the brain and spine connect

**holocord** - relating to the entire length of the spinal cord

**obex** - considered to be the top of the spinal canal

**odontoid process** - small, toothlike, upward projection from the second vertebra around which the first vertebra rotates

**posterior fossa** - depression on the inside of the back of the skull, near the base, where the cerebellum is normally situated

**scoliosis** - abnormal curvature of the spine

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

**tonsillar herniation** - descent of the cerebellar tonsils into the spinal area; often measured in mm

surgery for a persistent syrinx. This rate was twice as high than the authors' experience with Chiari I patients.

Aside from the surgical outcome, this study of the so called Chiari 1.5 would appear to only add to the complexity and confusion surrounding Chiari malformations. Until the true underlying causes and symptomatic mechanisms are understood, it is unlikely that the confusion regarding terminology will clear up. For now, the best a patient can do is to NOT get caught up in the nomenclature, see past the fancy words, and try to understand clearly what their individual anatomy is and how it will be treated.

**Table 1**  
**Selected Anatomical Characteristics of Chiari 1.5 Patients**

Characteristic	Average
Age (yrs)	12.5
Tonsillar Herniation (mm)	12.7
Brainstem Herniation (mm)	14.4
Odontoid Angle (degrees)	84.4

**Note:** Brainstem herniation measure by position of the obex below the foramen magnum.

**Table 2**  
**Location of Syrinxes**  
**(Total = 11)**

Location	Number
Holocord	6
Cervical	2
Thoracic	1
Cervico-Thoracic	2

### Source

**Source:** Tubbs RS, Iskandar BJ, Bartolucci AA, Oakes WJ. [A critical analysis of the Chiari 1.5 malformation](#). J Neurosurg. 2004 Nov;101(2 Suppl):179-83.

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