## Key Points

1. There are case reports linking rickets to Chiari; meaning Chiari is more common in children with rickets
2. Study measured the posterior fossa volume of 7 children with rickets and compared them with a control group
3. Found that the rickets group had significantly smaller posterior fossas than the control group
4. The two children with the smallest posterior fossas also had Chiari (29% of the group)
5. Authors believe that rickets causes a bony overgrowth in the posterior fossa region, which doesn't leave as much room for the developing brain and predisposes people to Chiari

## Definitions

**Apert's Syndrome -** condition where the sutures of the skull close prematurely in infants, causing a misshapen head and face; other problems may also be present

**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

**control group -** in a study, a group of subjects who are used as a basis for comparison; the control group is usually healthy or does not receive the treatment the experimental group does

**Crouzon's Syndrome -** condition where the sutures of the skull close prematurely in infants, causing a misshapen head and face; other problems may also be

# Rickets Provides Chiari Clues

If you liken the effort to fully understand Chiari to putting together a 1,000 piece jigsaw puzzle, the hard truth is that we barely have all the pieces out of the box. Some of the pieces are right side up, and some are even joined together to form pockets of understanding, but for the most part we are faced with a table full of individual clues that we know must somehow fit together to form a complete picture.

The other hard truth is that unlike working a jigsaw puzzle with a group of kids, where everyone is crowding around and fighting for pieces, there are relatively few people trying to solve the Chiari puzzle. Thankfully, one team that is working hard on the puzzle is a group out of the University of Alabama at Birmingham (UAB) which includes Dr. R. Shane Tubbs, a researcher and physician's assistant, and Dr. Jerry Oakes, a pediatric neurosurgeon with an extensive Chiari patient base. Over the years, the UAB team has used their Chiari experience to explore many different aspects of the disease, and in a report published in the August, 2004 issue of the journal Neurosurgery, the team tries to link several puzzle pieces by exploring the relationship between Chiari and the childhood bone disease rickets.

One such puzzle piece is the idea that the cause of at least some Chiari cases is not too big of a brain, but rather, too small of a skull. Recently, some researchers have shown that the posterior fossa - the part of the skull where the cerebellum is situated - is smaller in people with Chiari; the implication being that the skull is not big enough to accommodate the growing brain, resulting in the cerebellar tonsils extending down into the spine.

Similarly, there are reports of a high incidence of Chiari among children with Crouzon's and Apert's syndrome - conditions where the sutures of the skull close prematurely and result in skull deformities. Finally, there are also case reports indicating there might be a link between rickets - a bony disease caused by a lack of Vitamin D or an inability to process it - and Chiari.

In order to put these pieces together, the UAB team decided to study whether the posterior fossa is smaller in children with rickets than in normal children. The researchers used a database to identify 7 children who had been diagnosed with rickets and for whom MRI or CT (computed tomography) images of the head were available. The researchers also created a control group of similar ages to compare the rickets group with.

Next, the researchers used the MRI and CT images to calculate the volume of the posterior fossa for each subject. This was done by overlaying a grid of dots onto a series of images. By counting the number of regularly spaced dots inside the posterior fossa for several images per subject, they were able to quantify the posterior fossa volume.

Using this technique, the team found that the average volume of the posterior fossa was significantly smaller in the rickets group versus the control group (see Table 1). In addition, two of the seven children in the rickets group also had Chiari (29% of the group), and interestingly, the two with Chiari had the smallest posterior fossas. There was no significant difference between the volumes of girls versus boys.

While cautioning that the number of subjects in the study is small, the researchers speculate that bony overgrowth due to rickets reduces the size of the posterior fossa and predisposes these people to Chiari. If this turns out to be true, it not only connects one more piece of the puzzle, but adds to the evidence that a number of Chiari cases may be due to small posterior fossas.

It is almost assured that there are several "causes" of Chiari, with a small posterior fossa being one; but hopefully as the pieces start to fall into place, we will gain a better understanding of the true, underlying reasons people have Chiari, and thus be better able to address - and even prevent - the problem.

**Table 1**
**Average Posterior Fossa Volume By Age Group**

| | Volume By Age Group (cm3) | | |
|---|---|---|---|
| | **1-4** | **5-10** | **11-15** |
| **Control Group** | 87 | 138 | 148 |
| **Rickets Group** | 126 | 68 | 102 |

present

**dura -** thick outer covering of the brain and spinal cord; beneath the dura are the arachnoid and the pia

**duraplasty -** surgical technique where a patch is sewn into the dura

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

**rickets -** skeletal disease in children, caused by a lack of - or inability to process - Vitamin D, resulting in bony deformations

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

## Source

**Source:** Tubbs RS, Webb D, Abdullatif H, Conklin M, Doyle S, Oakes WJ. Posterior cranial fossa volume in patients with rickets: insights into the increased occurrence of Chiari I malformation in metabolic bone disease.
Neurosurgery. 2004 Aug;55(2):380-3