Key Points

1. Spontaneous resolution of Chiari in children has been reported several times in the literature
2. It is not known how often or why this occurs
3. Doctors encountered a young boy with a 13mm asymptomatic Chiari
4. Due to his other condition, he had MRIs over a course of years which showed the herniation resolved on its own
5. Researchers used morphometric analysis to measure the volume of the posterior fossa and the cerebellum
6. All the volumes were within normal ranges at age 3 and 7, the posterior fossa grew significantly more than the cerebellum
7. This provides some evidence to the theory that a growth differential between the brain and skull can lead to Chiari
8. It would be interesting to apply this research to a broad group of children with Chiari and track them over time.

Definitions

morphometric - in this article, refers to measuring dimensions of the skull and brain

posterior fossa -region in the back of the skull where the cerebellum is situated

posterior fossa volume - space available in the posterior fossa region; precise definition can vary

seizure - sudden, uncontrolled electrical activity in the brain which causes involuntary movements and loss of consciousness

spontaneous resolution - in this article, refers to a Chiari malformation improving with no intervention; the cerebellar tonsils ascend back into the skull

Can A Child Grow Out Of Chiari?

March 31st, 2009 -- Spontaneous resolution - meaning Chiari that goes away on its own - has been documented in the medical literature, but is poorly understood. Though it is certainly every patient's (and parent's) highest hope, how often it occurs, why it occurs, and who it occurs to is largely unknown. Now, a report in the Journal of Neurosurgery: Pediatrics from a group of doctors at Duke University (Waldau et al.) offers some clues as to one possible mechanism underlying the near miracle of spontaneous resolution.

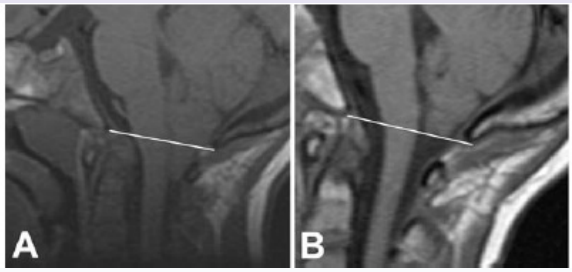
Chiari & Syringomyelia News has reported on cases of spontaneous resolution, primarily among children, and this journal article cites several such examples (Figure 1). Despite these reports it is very difficult to estimate how often this might occur. Certainly, a handful of case reports compared to hundreds of thousands of confirmed cases implies that it is a very rare event; however, it is impossible to know how many cases might have resolved on their own without anyone knowing about it.

Further, it is not known how or why Chiari can resolve on its own, although there are several logical possibilities. First is if the underlying cause of the herniation resolves or heals by itself. For example, Chiari can be caused by a spinal fluid leak which lowers the pressure in the spine compared to the brain and essentially pulls the cerebellum down. Some such CSF leaks are known to heal by themselves over time, so some cases could be due to an initial CSF leak which heals.

A different possibility involves the relative growth of the skull and the brain. A leading theory for a major cause of Chiari is a small posterior fossa (the part of the skull where the cerebellum is situated) which forces a normal sized brain out of the skull. There is a significant body of research which supports this theory in part and researchers have begun to focus on the potential dynamic nature of Chiari in children by looking at the relative growth rates of the skull versus the brain. In other words, if for a period of time the brain grows faster than the skull, then a problem can develop. However, if at a later time the skull catches up with its growth, then perhaps the problem, or the herniation, could resolve itself. Given the tremendous growth which children go through, this is certainly a reasonable theory.

It was this theory that the Duke researchers decided to explore when they encountered a case of spontaneous resolution in a young boy. The child was first seen at the age of 3 for seizures. The doctors determined that he had a genetic disease known as tuberous sclerosis which was causing the seizures. However, during their testing, they also found that he had a 13mm Chiari malformation (Figure 2).

Figure 2: MRIs Showing 13mm Chiari at Age 3 (A) and Complete Resolution at Age 7 (B)



Despite its size and shape, the doctors could find no symptoms or problems directly attributable to the Chiari and there was no syrinx. When the boy was seen again at age 5, it was noted that the tonsils had ascended partially, and by the time he was 7, they had risen back into the skull (Figure 2 B).

In order to understand how this may have occurred, the Duke team used morphometric analysis to measure the volume of both the posterior fossa and the cerebellum from the MRIs at 3 years of age and 7 years of age. Techniques to calculate these values have been published previously. To minimize error, the researchers took the average of the results from five different experts who performed the measurements.

Interestingly, they found that all four volume measurements - posterior fossa and cerebellum at age 3 and 7 - were within published normal ranges for children that age (Figure 3); however over that period of time the posterior fossa grew 11.5% compared to only 4% for the cerebellum (Figure 4). In other words, at age 3 the cerebellum was much larger relative to the space available than at age 7. The authors of this study did not do so, but other publications have focused on looking at just such a ratio. Regardless, the data is suggestive that the relative growth rates of the posterior fossa skull and cerebellum may have played a role in the Chiari resolving naturally.

Due to his genetic condition, which involves masses in the brain, this patient is not a good one to generalize

tuberous sclerosis - rare genetic disease which causes benign tumors to grow in the brain

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

syringomyelia - condition where a fluid filled cyst forms in the spinal cord

Source

Spontaneous resolution of a 13-mm Chiari malformation Type I in relation to differential growth of the posterior fossa volume.Waldau B, Domeshek LF, Leigh FA, Lum KC, Fuchs HE, Marcus JR, Mukundan S, Grant GA. J Neurosurg Pediatr. 2009 Feb;3(2):110-4

from; however, the approach the researchers took in tracking morphometric changes over time is interesting and applying it to more and different Chiari patients may provide some valuable insights.

It is also worth noting that cases such as this paint a picture of Chiari as a dynamic condition, in which not only symptoms can change, but the underlying structure can change as well. This is contrast to the traditional view that a person is born with Chiari and remains that way.

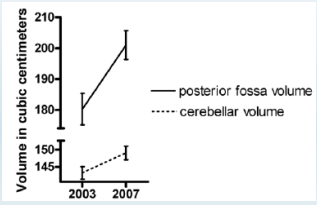
Figure 1: Published Cases of Spontaneous Resolution Cited In This Study

Author	Age at Diagnosis	Age at Resolution or Improvement
Avellino	5	10.5
Avellino	5	16
Castillo	9	13
Guillen	6	12
Sun	11	13
Jatavallabhula	1.5	6
Sudo	13	16
Sun	7	13

Figure 3: Posterior Fossa and Cerebellum Volumes at Age 3 and 7

	Age 3	Age 7	Growth
PFV (cm3)	180	201	11.5%
CV (cm3)	143	149	4.08%

Figure 4: Chart of PFV and CV Growth From Age 3 To 7



Note: The difference in growth between the posterior fossa and cerebellum was statistically significant

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