

Key Points

1. Natural history refers to how a disease progresses without medical intervention
2. Traditionally, the natural history of Chiari has not been well understood, but recently there have been a number of studies published
3. Researchers in Michigan identified 147 pediatric cases of tonsillar herniation >5mm who did not undergo surgery initially
4. Follow-up averaged 3.8 years for MRI and 6.5 years with physicians
5. During that time 14 of the group required surgery for headaches, apnea, or syrinx growth
6. The vast majority of the group did not develop symptoms or require surgery
7. Researchers did find however that individuals had changes in herniation size, CSF flow, and syrinx size over time
8. More research is required to see if any of these children develop symptoms as adults

Definitions

asymptomatic - having no symptoms

natural history - the scientific study of how something evolves naturally, without intervention

prospective - type of research which looks forward in time; for example to follow a group of people over time

retrospective - type of research which looks backward in time at events that have already happened, for example by using medical records

sleep apnea - condition where a person has repeated episodes where they stop breathing for a short period of time during sleep

tonsillar ectopia - another term

Study: Non-Surgical Cases In Children Don't Progress

September 30, 2011 -- For a long time it was common to hear the phrase that the natural history of Chiari is poorly understood, meaning that we don't know much about what happens to Chiari patients who do not get surgery. Recently however, prompted in part by the widespread adoption of MRIs, there is growing evidence that among children at least, many to most non-surgical cases remain that way for at least several years.

In recent years it has become common for pediatric neurosurgeons to evaluate a growing number of children in whom tonsillar herniation is found incidentally and there are few to no obvious Chiari related symptoms. Naturally (pun intended), many of these children are followed medically for at least a couple of years to make sure that symptoms don't develop. In a study published in the *Journal of Neurosurgery: Pediatrics*, a group from the University of Michigan reviews their experience with just such cases in an effort to help elucidate the natural history of pediatric Chiari.

Specifically, the researchers looked retrospectively at over 14,000 MRIs (in a searchable database) that were taken over a 10 year period to identify children with tonsillar herniation of at least 5 mm, but who did not undergo surgery. In addition, the group was looking for cases where there was at least one year of both MRI and clinical follow-up data available. Children with Chiari II, prior surgery for Chiari, or herniation due to a mass in the brain such as a tumor, were excluded. Using these criteria, the group found 147 children that had sufficient follow-up information.

The average age of the group 7.7 years and all were under 18. There were slightly more females than males, and the average follow-up time was 3.8 years for MRI data, 4.6 years for clinical data from the neurosurgery group, and 6.5 years of clinical information from any type of physician. The average herniation size was a substantial 9.5mm and 13 children had syrinxes.

The precise data on the number of symptomatic cases in the group is not clear from the publication but it appears that the vast majority, ~90%, were asymptomatic. While there was no change in the symptom status for the majority of patients, six children who were considered to be symptomatic when first evaluated no longer had Chiari related symptoms at the last follow-up, and 5 who were symptomatic remained that way. In addition, 9 children developed symptoms during the course of follow-up that the doctors believed were related to Chiari.

The most striking finding from following the group was that out of the 147, only 14 ended up requiring surgery at some point. In these cases, the average time between initial diagnosis and surgery was 2.1 years. The most common reasons for surgery were persistent headaches, sleep apnea, and changes in syrinx size. Interestingly, there were no significant differences in initial size of herniation or CSF flow between those who eventually had surgery and those who didn't.

The researchers also noted that over the course of the follow-up period the average amount of tonsillar descent for the group did not change, however individually it did. Not surprisingly, the change was different for different age groups, as it is generally accepted that in normal development the cerebellar tonsils ascend with age. Specifically, for children under 6, the average change was a modest increase in herniation of 0.63mm (Table 1). However, for children between 6-12, the average change was a decrease in herniation of about half a millimeter, and for children over 12, the average change was a decrease of more than a millimeter.

CSF flow studies were available for a subset of patients (74). Among this group, there was no change in the amount of CSF flow around the herniation for the majority of the group, but the flow did increase in 23 patients and decrease in 12 (Table 2).

Similar to herniation size and CSF flow, there were changes in the individual syrinx sizes among patients over time. Of thirteen children who had syrinxes at the initial diagnosis, the syrinx got bigger in 2, didn't change in 6, got smaller in 2, and completely resolved in 3. In addition, new syrinxes developed in 7 children (Table 3). Despite these individual changes, as a group, the average length and width of the syrinxes did not change significantly over time. However, both the children who developed new syrinxes, and whose existing syrinxes got worse, did tend to have larger initial herniations (13.5mm and 14.5mm respectively).

Although the follow-up period for these studies remains frustratingly short, this work does demonstrate that at least for the mid-term the majority of asymptomatic and non-surgical pediatric Chiari cases will remain that way. In addition, the individual changes in herniation size, CSF flow, and syrinx size, also demonstrate the dynamic nature of Chiari in children. However, the study does not shed any additional light on how to identify which children may end up needing surgery and thus require closer monitoring and supervision. What also remains unanswered of course is how many - if any - of these children will develop symptoms as adults, and whether the development of symptoms in previously asymptomatic cases is related to aging, trauma, lifestyle, or some

for tonsillar herniation

tonsillar herniation - term used to describe when the cerebellar tonsils protrude out of the skull, into the spinal area. Sometimes used to distinguish asymptomatic herniations from symptomatic Chiari

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

[Natural history of Chiari malformation Type I following decision for conservative treatment](#). Strahle J, Muraszko KM, Kapurch J, Bapuraj JR, Garton HJ, Maher CO. J Neurosurg Pediatr. 2011 Aug;8(2):214-21.

combination of all three.

Table 1: Changes In Tonsillar Descent By Age Group

Age (yrs)	Avg. Change in Tonsillar Descent (mm)
0-6	+ 0.63
6-12	- 0.53
12-18	-1.24

Table 2: Changes In CSF Flow During Follow-up (74 Total Patients)

Change	Number
Increased Flow	23
No Change	39
Decreased Flow	12

Table 3: Changes In Syrinx Size During Follow-up (20 Total Patients)

Change	Number
Increased	2
No Change	6
Decreased	2
Resolved	3
New Syrinx	7

Notes: Average MRI follow-up was 3.8 years

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