

Key Points

1. The natural history of Chiari is not well understood
2. People who are found to have Chiari but have no or few symptoms are not sure how to proceed or whether they will develop symptoms over time
3. Study followed 22 children with herniations of at least 5mm for an average of 6 years
4. Children were evaluated yearly with MRIs and clinical exams
5. Out of the entire group, only 3 ended up needing surgery, and 2 of those were for worsening hydrocephalus
6. The majority of patients became, or remained, symptom free
7. MRIs showed that most of the children did not get worse in terms of amount of herniation and some actually improved. However, 3 children did develop cervical syrinxes
8. A conservative approach seems warranted in treating Chiari with few or no symptoms, but it is still impossible to predict who may develop symptoms later in life

Definitions

asymptomatic - having no symptoms

hydrocephalus - condition where there is an unusual build up of CSF in the brain

incidental - a medical finding, such as herniated cerebellar tonsils, which was not being looked for and may or may not be relevant

natural history - how a disease or medical condition will progress if there is no intervention

neuropsychiatric testing - a series of tests designed to identify cognitive deficits associated with neurological injury or disease

vertigo - sensation of spinning

Study Explores The Natural History of Chiari

November 30, 2008 -- As MRIs become more prevalent, more cases of Chiari (for lack of a better term) are being found where there are few, if any, symptoms. Generally, these cases fall into two main categories. First is the incidental diagnosis, where an MRI was ordered to evaluate a patient for something other than Chiari, such as a head trauma. Second, are cases where the symptoms are commonly associated with Chiari, such as headache and neck pain, but are mild and/or infrequent.

These types of cases can pose a challenge to the treating surgeon, especially if the malformation is significant or if there is a syrinx. A published survey of neurosurgeons showed that while very few would recommend surgery based on tonsillar herniation with no symptoms, more than a quarter would recommend surgery if there is a syrinx, even if there are no symptoms.

In addition, doctors do not know what to tell patients, and their families, in regards to what to expect, because the natural history of Chiari is not known. In other words, very little research has been done on whether a person who is found to have a tonsillar herniation but does not have symptoms will develop symptoms over time. This of course leaves patients in the position of not knowing whether there is really anything that is wrong with them, and what, if anything, they should do about it.

Now, a report from a group of Italian doctors (Novegno et al.) published in the September, 2008 issue of the Journal of Neurosurgery: Pediatrics, suggests that only a small percentage of such cases will eventually need surgery. The researchers, based out of the Catholic University Medical School in Rome, followed 22 children who were found to have tonsillar herniation of at least 5 mm, but who were either asymptomatic or had mild symptoms which did not necessitate surgery.

The children represented 23% of the total number of Chiari related cases seen over a period of several years. Interestingly, exactly half (11) were found to have the tonsillar herniation incidentally and were completely asymptomatic, while the other half were initially seen for mild, Chiari related symptoms, such as headache and vertigo. The children ranged in age from 1-16 years and were given complete MRIs and clinical exams six months after the initial evaluation and annually thereafter. A subset of the patients (10) were also given neuropsychiatric evaluations on a routine basis to look for cognitive deficits and behavioral disorders.

As mentioned previously, all of the children had at least 5mm of herniation, and a number of them, even asymptomatic, had herniations greater than 10mm (see Figure 1). Only one child had a syrinx, but five were found to have hydrocephalus as well. Each individual was followed for at least 3 years and on average for 5.9 years.

The researchers found that among those patients who were originally asymptomatic, 7 (64%) remained symptom free during the entire follow-up period. Of course, that means that 4 children (36%) did develop symptoms, such as headaches, neck pain, and vertigo, three to five years after the initial evaluation. Interestingly, one patient who developed symptoms, later became symptom free again (Figure 2). Overall, only two patients out of this group eventually required surgery, and in both cases the surgery was related to worsening hydrocephalus and the emergence of a syrinx.

In the mildly symptomatic group, 7 patients became, and remained, symptom free; 2 patients improved slightly; and two got worse. However, only one child out of this group required surgery to treat her symptoms (a posterior fossa decompression).

In evaluating the MRI results, the doctors found a mild improvement in the amount of herniation in 4 patients and complete resolution of the herniation in one child. On the flip side, there was a demonstrated increase in the amount of herniation in two patients, and three patients developed cervical syrinxes.

Finally, although it was a small group, of the children who underwent neuropsychological testing, 8 showed a normal level of intelligence, while two were at the lower borderline of normal. Despite the relatively normal intelligence scores, the testing did reveal naming speech delays in half of the children tested. Additional problems included visual attention disorders, and visual memory deficits.

To completely characterize the natural history of Chiari is a monumental, very long term task, but this study does seem to reinforce the current thinking of the surgical community in terms of conservatively managing patients with mild or no symptoms. However, this study involved only a small number of patients, and while the follow-up period was long relative to other Chiari research, it certainly is not long enough to say that these children will never develop symptoms. In fact, it would be interesting to see what percent of these cases might develop symptoms as adults, which is when many people become strongly symptomatic and are diagnosed. It

and dizziness

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

Source: Novegno F, Caldarelli M, Massa A, Chieffo D, Massimi L, Pettorini B, Tamburrini G, Di Rocco C. *The natural history of the Chiari Type I anomaly.* J Neurosurg Pediatrics. 2008 Sep;2(3):179-87

could be that if the adults who are diagnosed now with Chiari were given MRIs as children, they would look like the cases described in this study.

One issue not addressed by the authors was whether children found to have tonsillar herniation but no symptoms should have any activity restrictions. This question was raised recently at the Conquer Chiari Research Conference and generated the most spirited debate of the event. There were strong opinions on both sides, but unfortunately not much strong research to back them up.

Finally, it is worth noting that this study once again highlights the limited utility of current MRI technology as a diagnostic tool for Chiari and its complete inability to predict which patients may eventually need surgery.

Figure 1: Extent of Tonsillar Herniation (22 Patients)

Group	# of Patients	Herniation (mm)
Asymptomatic (11 Patients)	2	5
	6	5-10
	3	>10
Mild Symptoms (11 Patients)	4	5
	1	5-10
	6	>10

Figure 2: Symptom Development Over Time (22 Patients)

	Asymptomatic (11 Patients)	Mild Symptoms (11 Patients)
Asymptomatic	7	7
Improved	-	2
Developed Symptoms or Got Worse	4	2

Notes: In the Asymptomatic Group, one of the patients who developed symptoms became asymptomatic again over time; average follow up was 5.9 years

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