Key Points

1. More and more patients, especially children, are being found to have Chiari malformation with no symptoms or symptoms not usually attributed to Chiari
2. It is not always clear what, if anything, should be done in these cases
3. Study looked retrospectively at 124 children with greater than 5mm tonsillar herniation but who did not receive surgery
4. Researchers grouped children into asymptomatic, symptoms not likely due to Chiari, and symptoms likely due to Chiari
5. Children were followed an average of close to 3 years
6. In the asymptomatic group, only one child developed symptoms, but they were not due to Chiari
7. In the symptoms not due to Chiari group, 12 out of 67 got worse over time
8. In the last group, 4 out of 14 got worse
9. Study has many limitations, but only 17 children total out of 124 children got worse over several years

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

cerebellar tonsils - portion of the cerebellum located at the bottom, so named because of their shape

Study Follows Non Surgical Chiari Children

June 30th, 2011 -- While the widespread adoption of MRIs has revolutionized the diagnosis of Chiari, it has also raised questions regarding the traditional definition of Chiari which have proven resistant to an easy answer. Specifically, as more and more people are scanned, both to look for Chiari and for other reasons, it is becoming clear that simply measuring the amount of tonsillar herniation is not a good indicator of symptomatic Chiari.

Not only has research shown that among symptomatic Chiari patients the size of herniation does not correlate well with symptoms severity or outcomes, but clinically, doctors are now facing two new types of patients. First are those for whom a Chiari malformation is found completely incidentally, meaning that the CT or MRI was not initially performed to look for Chiari, but rather for a different reason, such as after a head injury.

Incidental Chiari cases are usually asymptomatic and most doctors do not recommend any type of surgery or other treatment. However patients, and especially parents when a child is involved, are left wondering what it means. Will they develop symptoms in the future and require surgery? Should they restrict their activities at all? Should children found to have incidental Chiari be allowed to participate in contact sports? Although research has indicated that very few cases that are found incidentally will become symptomatic in the short term, because of the cost and logistical challenges, no one has studied over the long to very long term how many, if any, incidental cases become symptomatic.

The second group which doctors are confronted with today are patients who have symptoms and are found to have a Chiari malformation, but the symptoms are not necessarily traditionally associated with Chiari. While it is tempting from the patient point of view to associate every ache and pain with Chiari, and far too many patients who are clearly symptomatic have been told incorrectly it is not due to Chiari, research has shown that symptoms not traditionally associated with Chiari are less likely to improve with surgery.

Conquer Chiari has advocated for years that what is needed is a quantitative, objective diagnostic test for symptomatic Chiari malformation. But until such a test is developed (Conquer Chiari has funded research to do just that), doctors and patients face a dilemma when an MRI shows significant tonsillar herniation, but there are no symptoms or symptoms are not usually associated with Chiari.

Another way to shine some light on this murky situation would be to have a better understanding of the natural history of Chiari. In other words, what happens to people over time who do not have surgery? The ideal research study would recruit people as they are found to have the tonsillar herniation and follow them yearly for 30 - 40 years; however this type of study is not likely to occur given the general funding constraints associated with Chiari research.

While scientifically not as rigorous, another way to look at the problem is for medical groups to review their records retrospectively and see how patients that have not had surgery have progressed (or not progressed) over time. A recent publication from a group of pediatric neurosurgeons at the University of Miami did just that (Benglis et al).

Specifically, the group reviewed their experience with 179 pediatric patients evaluated for Chiari over a ten year period. Of the total evaluated, 54 were recommended for and underwent surgery. The other 124 did not receive surgery, but were followed for an average of close to three years and for as long as 8 years. Each child in the non-surgery group had an MRI which showed tonsillar herniation of at least 5mm (avg was more than 8mm). Seven children in the group had small syrinxes.

Of the 124, 43 were completely asymptomatic and the Chiari had been found incidentally. In this group, only one child developed any type of symptoms, but the surgeons did not feel the symptoms were related to Chiari.

Of the remaining 81 patients, 67 had symptoms which the evaluating surgeon did not feel were likely related to Chiari. In this group, 29 improved over time, 26 remained stable, and 12 got worse (Table 2). None developed symptoms that the surgeon thought were treatable by Chiari, and perhaps more importantly, none developed any objective neurological signs or deficits.

The last group of 14 patients had symptoms that the surgeons felt were due to Chiari but either were not severe enough to warrant surgery (9) or surgery was recommended but the parents would not consent (5). In the group of nine, 4 improved over time, 4 remained stable, and 1 got worse. In the group of 5 for whom surgery was recommended, 2 improved and 3 got worse. None of the children in the group of 14 developed any new neurological signs or deficits and none of the syrinxes in any group showed progression.

Beyond the scientific limitations of this study, experienced patients may point out that the classification of

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

Outcomes in pediatric patients with Chiari malformation Type I followed up without surgery. Benglis D Jr, Covington D, Bhatia R, Bhatia S, Elhammady MS, Ragheb J, Morrison G, Sandberg DI. J Neurosurg Pediatr. 2011 Apr;7(4):375-9.

symptoms as being related to Chiari or not by the surgeons is a matter that can be easily disputed and in one sense represents one of the major problems Chiari patients face. Namely that doctors don't believe their symptoms are related to Chiari. While there is no way to tell in these cases whether the children whose symptoms worsened would have benefited from surgery, it is interesting to note that out of 124 children with significant herniations who did not receive surgery, symptoms only got worse for 17 of them and no new neurological signs developed in the short to mid term.

The authors acknowledge some of the limitations of their study and do not draw any strong conclusions. They do however recommend that surgery should not be considered for cases without clear symptoms, a significant syrinx, or scoliosis.

Table 1: Most Common Symptoms Among Patients With Symptoms (81 Total)

Symptom	# With
Non posterior headache	34
Nausea/vomiting	20
Sensory deficits	15
Posterior headache	14
Neck pain	14

Table 2: Outcomes For 81 Symptomatic Children With Chiari

Symptoms Not Likely Due to CM = 67	Improved = 29
	Stable = 26
	Worse = 12
Symptoms Likely Due to CM = 14	Improved = 6
	Stable = 4
	Worse = 4

Notes: Average follow up was 2.8 years; Symptoms Likely Due to CM category contained 5 children who were recommended for surgery but parents declined

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