Key Points

1. Research has shown that the classic definition of Chiari, based on length of herniation, is not very useful
2. Based on the theory that Chiari results from a small posterior fossa, one area of interest is measuring posterior fossa volume and skull dimensions
3. Previous research has found that Chiari patients tend to have smaller posterior fossa regions than normal
4. This study looked at 61 pediatric Chiari patients as compared to normal controls
5. Found that overall Chiari group had smaller posterior fossa as measured by the PFV/ICV ratio
6. Also found that for children age 0-9, both symptomatic and asymptomatic subjects had small posterior fossas
7. However asymptomatic older children had skulls similar to the controls
8. Long-term studies would be required to see if skull size/shape is useful for predicting who will develop symptoms

Definitions

asymptomatic - not having any symptoms

herniation - the protrusion of a body part into an area where it isn't normally located

intracranial volume (ICV) - a measure of the total amount of space inside the skull

morphometric - referring to the physical structure and size of something, in this case the skull

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

poseterior fossa volume (PFV) - measure of the total amount of space occupied by the posterior fossa region

Can Posterior Fossa Volume Indicate Symptom Severity?

September 30, 2007 -- Chiari is traditionally defined as the cerebellar tonsils herniating at least 3mm-5mm out of the skull into the spinal area. Although this definition has been used for a long, long time, research in the last 10 years or so has consistently shown that the amount of herniation is not a good indicator of symptom severity or clinical outcome.

In other words, there are many people with small herniations who have severe symptoms and there are probably even more people with large herniations (greater than 5mm) who have no symptoms. The bad news is that many physicians still rely on the old definition of Chiari, which means that time and again patients with small herniations are told their symptoms are not related to Chiari. The good news is that most, if not all, Chiari experts are aware of the problem and are looking for new ways to define Chiari, objectively measure severity, predict who will develop symptoms and identify good candidates for surgery.

There are several active areas of research in this regard, one of which is morphometric skull analysis. Morphometric is a fancy word which simply refers to the physical size and shape of something, in this case the human skull. Chiari is thought to be a result of the underdevelopment of certain parts of the skull (which are then too small for the brain) and researchers have begun to use MRIs to quantitatively measure skull dimensions.

Studies using this technique have shown that on average Chiari patients have small posterior fossas as compared to healthy people. Research has also shown that skull size may be linked to the development of a syrinx and that people with very small herniations, but Chiari-like symptoms, tend to have abnormal skulls as well.

Now, in a study posted in July on the Child's Nervous System website, a group from Canada (Trigylidas et al.) revealed that even some asymptomatic people with herniations have abnormally small posterior fossas. The Canadian team looked at the skull shapes of 61 pediatric Chiari cases from the Eastern Ontario Children's Hospital and compared them to 20 controls who had no history of intracranial problems.

In the Chiari group, 34 children were considered symptomatic with headaches, scoliosis, balance problems, and muscle and sensory problems being the most common. To assess the size of the posterior fossa, the researchers used MRIs to measure the total intracranial volume (ICV) and the volume of the posterior fossa (PFV). They then calculated the ration of PFV to ICV as a measure of how large the posterior fossa is relative to the total skull. This ratio method is a technique other researchers have used to account for the variability in skull sizes of children of different ages.

Not surprisingly, the data showed that on average the Chiari group (both symptomatic and asymptomatic) had a smaller PFV/ICV ratio than the control group (see Table 1). However, the Canadian researchers went further and analyzed the data by comparing symptomatic to asymptomatic and by age. Since the average age of the Chiari group was 10, the scientists split the data into two group, 0-9 years and 10-18 years.

The results of this analysis were intriguing. In the younger group (0-9), both asymptomatic and symptomatic Chiari patients had smaller PFV/ICV ratios. However, in the older group (10-18) while the symptomatic children again had small ratios, the asymptomatic children had normal PFV/ICV ratios. In other words among the asymptomatic children, the younger ones had abnormal skull shapes, while the older ones had normal skull shapes.

Based on this, one has to wonder if some young children with small posterior fossas become symptomatic in time, while in others the skull growth catches up and they remain symptom free. What would be really useful is a longitudinal study which identified young children with asymptomatic herniations and small posterior fossas and followed them over time as they grew up.

As an interesting side note, the researchers also looked at CSF flow data for a subset of the children. They found that both symptomatic and asymptomatic children demonstrated abnormal flow characteristics. While some surgeons have begun to rely on flow studies, others have questioned their utility and this result explains why.

It appears that what makes people with herniations (even small ones) symptomatic is either very complicated or due to something that for the time being remains unknown. What is clear is that identifying that unknown would not only provide a way to objectively measure Chiari, but may lead to a redefinition of the entire problem.

Table 1

PFV/ICV ratio - measure of how much of the entire skull the posterior fossa takes up; used as a way to eliminate the variability of skull sizes in children of different ages

tonsillar herniation - refers to the cerebellar tonsils extending below, and out of, the skull into the spinal area

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

Source

Trigylidas T, Baronia B, Vassilyadi M, Ventureyra EC. Posterior fossa dimension and volume estimates in pediatric patients with Chiari I malformations. Childs Nerv Syst. 2007 Jul 27; [Epub ahead of print]

PFV/ICV Ratio By Age and Symptomology

Age	Asymptom	Symptom	Control
0-9	.105	.103	.127
10-18	.118	.106	.127
Total	.115	.103	.127

Note: *PFV = posterior fossa volume; ICV = intracranial volume; asymptom = no symptoms; symptom = symptoms attributable to Chiari*

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