

Key Points

1. In recent years, several studies have shown that Chiari patients tend to have small posterior fossas
2. This has led to the belief that Chiari is caused by an underdevelopment of the skull in this region which does not provide enough space for the cerebellum
3. This study looked at PFV and the PFV/ICV ratio in 42 symptomatic Chiari children prior to decompression surgery and compared them to healthy controls
4. Researchers also grouped children based on whether they had syringomyelia
5. Found that for the Chiari only group, there was no significant difference in PFV or ratio to the control group
6. However, the group with both CMSSM did have significantly smaller PFV and PFV/ICV ratios than the controls
7. May imply that Chiari with syringomyelia is fundamentally different than Chiari only

Definitions

intracranial - inside the skull

intracranial volume (ICV) - the volume of the total space inside the skull, can be measured by MRI

occipital - the back of the head

PFV/ICV ratio - a measure of how much of the total skull the posterior fossa takes up, eliminates differences due to head size, age, and gender

posterior fossa - depression in the back of the head where the cerebellum is located

posterior fossa volume (PFV) - volume of the posterior fossa space, as measured by MRI

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

Small Posterior Fossa Linked To Chiari Related Syringomyelia

September 20, 2006 -- In recent years, a number of research studies have shown that Chiari patients, on average, have smaller (and abnormal) posterior fossas than normal. The posterior fossa is the region in the back of the skull where the cerebellum is situated, and the dimensions of this area can be quantified using standard anatomical MRIs.

Using these MRI based techniques, scientists have demonstrated not only that Chiari patients tend to have small posterior fossas, but that specific features of this region - such as the length of certain bone segments - tend to be different with Chiari. These findings have been demonstrated in children, adults, and even in relation to the controversial Chiari 0.

The small posterior fossa finding has become generally accepted to the point that most specialists now refer to Chiari not as a problem of too much brain, but rather as a problem involving the underdevelopment of the bone in the back of the skull, which results in there not being enough space to accommodate the cerebellum.

Now, a publication in the August, 2006 issue of the Journal of Neurosurgery:Pediatrics suggests that the small posterior fossa theory may not apply to all Chiari patients. In the study, a team from the Birmingham Children's Hospital, in England, led by Dr. Spyros Sgouros reports that they found no real difference between the size of the posterior fossa of children with Chiari versus healthy controls; however, they did find that patients with both Chiari and syringomyelia had significantly smaller posterior fossas.

The British team had two goals for their research: first to establish whether Chiari patients did indeed have smaller posterior fossas, and second to investigate whether a small posterior fossa was linked in any way to the presence of syringomyelia.

To investigate this, they looked at 42 children who underwent decompression surgery between 1998-2004. All the children had herniations of at least 5mm and symptoms directly relatable to Chiari, such as exertional occipital headaches, weakness in the arms/legs, and progressive scoliosis. In addition, all had demonstrated abnormal CSF flow, but the flow data was not included in this analysis. On average the group was slightly more than 10 years old and included 17 Chiari only children, and 25 (59%) children with both Chiari and syringomyelia. Children with other skull abnormalities and children who had shunts placed previously were excluded from the study.

A control group was formed from children who had MRIs for different reasons, but the MRIs were found to be normal. In other words, there were no skull or brain abnormalities that would interfere with interpreting the results.

Just prior to surgery, the Chiari children were given MRIs which were used as the basis for the posterior fossa measurements. The researchers used a combination of automatic and manual techniques to calculate three items: the posterior fossa volume, the total intracranial volume, and the posterior fossa volume to intracranial volume ratio. This was accomplished by having a researcher outline the regions of interest on a computer screen, and specialized software would then calculate the underlying area and volume. The team took measurements this way using two independent researchers to minimize human error.

The PFV/ICV ratio is a measure of how much of the total skull space is occupied by the posterior fossa and was used as a way to eliminate the overall effect of head size, age, and gender. In other words, someone with a larger head would naturally have a larger posterior fossa, just as older children would, which might skew the results. By using a ratio this effect can be compensated for.

When they examined the results, the researchers were surprised to find that the children with Chiari only were not significantly different than the healthy controls (see Table 1). Specifically, the Chiari group had an average PFV/ICV ratio of .134, which was nearly identical to the control group average of .135.

However, they also found that the children with both Chiari and syringomyelia were significantly different from their healthy counterparts. The CMSSM group had an average PFV/ICV of only .122, which means their posterior fossas were smaller and took up less of the total skull volume.

The researchers also found that age played a role, with the difference in posterior fossa volume being more pronounced in children under 10. In fact for children under 10 the PFV/ICV ratio was 15% smaller for the CMSSM group, but this dropped to 5% for children older than 10.

In trying to reconcile their findings with previous research which found that Chiari patients on average have a

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

Source

Sgouros S, Kountouri M, Natarajan K. [Posterior fossa volume in children with Chiari malformation Type I.](#) J Neurosurg. 2006 Aug;105(2):101-6.

small posterior fossa, the authors point out that they are the first ones to separate those with syringomyelia from those without, and they believe that prior results are misleading because the subject groups had very high percentages of people with syringomyelia.

If the results of this study are validated by further research, the implications are profound. It might be that Chiari with syringomyelia is fundamentally different than Chiari only, with different underlying causes, and potentially different treatment approaches. It also raises the question of what the underlying cause of Chiari only is if the posterior fossa is indeed of normal size (Ed. note: is it linked more to elevated CSF pressure perhaps?). Finally, it brings into question whether CM/SM is more like Chiari related to spina bifida, where CSF flowing into the spinal canal somehow affects the growth of the posterior fossa region.

While the number of children in this study was relatively small, and the differentiation between the Chiari only group and the Chiari/syringomyelia group was not absolute, the whole research area of linking posterior fossa measurements to more specific aspects of Chiari, syringomyelia, and surgical outcome, would appear to be an important one to pursue.

Table 1
PFV Measurements In Chiari Only, Chiari w/SM, and Controls

	CM Only	CM & SM	Norm
PF Volume (cm ³)	196	171	186
PFV/ICV Ratio	.134	.122	.135

Note: *The PFV and PFV/ICV ratio for the CM only group was essentially the same as the normal controls; however, both the PFV and PFV/ICV ratio for the CM/SM group was significantly lower than the control group*

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