

Key Points

1. TCI used their large database of patients to study morphological features of different Chiari subtypes
2. Over 700 patients' images were analyzed
3. Patients were broken into groups such as classical Chiari, TCS, cranial settling, etc.
4. Occipital bone size, PCF volume, and foramen magnum size were compared to age controlled healthy normals
5. The Classical Chiari and craniosynostosis groups had significantly smaller measurements in all three categories
6. The TCS group had normal sized occipital bones and posterior fossa volume, but had significantly larger foramen magnums
7. Classifying Chiari patients in a system such as this could have positive clinical and research implications

Definitions

CT - computerized tomography, imaging machine which uses computer reconstructions of x-rays

craniosynostosis - condition where the sutures in the skull close too early resulting in a misshaped head

etiology - the cause of a disease

filum terminale - thread like structure at the end of the spinal cord

foramen magnum - opening at the base of the skull through which the brain and spine connect

intracranial hypertension - elevated CSF pressure in the brain region

intracranial hypotension - abnormally low CSF pressure in the spinal area

Milhorat Weighs In On Underlying Causes of Chiari

March 31st, 2011 -- [Ed Note: It remains Conquer Chiari's longstanding and stated policy to not endorse any specific doctors, hospitals, procedures, or products. As such, we will not comment on the legal actions and controversy regarding The Chiari Institute. However, I can assure you that Conquer Chiari requires research projects that it funds to have IRB approval (when necessary), comply with all regulations, and meet generally accepted ethical research guidelines. Conquer Chiari did not provide any funding for the work described in this article.]

For some time now, Conquer Chiari has speculated - both in this publication and in calls for research proposals - that in Chiari research it may be useful to try to identify subgroups of patients based on the underlying cause of their Chiari. Now in a recent publication, The Chiari Institute (Milhorat), leveraging their large database of patients, has published a study proposing just such a classification.

Over the past several years, it has become more apparent that Chiari is likely due to a number of different etiologies, for example a small posterior fossa, CSF issues, or the more controversial tethered cord. Nearly all Chiari research simply uses a standard definition of Chiari for their subjects and does not attempt to separate, or exclude, patients by the underlying etiology. However, it would seem reasonable that if there is a different underlying cause, that patients in the different groups would display differences in their clinical findings and how they respond to treatment such as surgery.

Therefore, the strength of certain findings may be diminished when the subject group is taken from across all Chiari subtypes. For example, if posterior fossa decompression works better for classical Chiari cases involving a reduced skull size, then any study which includes patients who have Chiari due to other issues, would present results that are misleading.

The TCI researchers hypothesized that different subgroups of Chiari patients could be identified by morphometric analysis (measurements of their anatomical features). To test this, they pulled information from their database of over 3,000 patients. Specifically, they found 741 patients between the age of 15-69, who had complete imaging records, and had not yet had any type of Chiari surgery prior to the images being taken. To be conservative, all the patients had herniations of greater than 5mm.

Based on the clinical records, the patients were classified into general categories. For example, cranial settling was identified by signs of joint hypermobility at the top of the spine, and tethered cord was identified by either a low lying conus on MRI, a thickened or fatty filum terminale, or symptoms consistent with TCS. Classical Chiari was defined as the absence of any other indications such as the ones cited above. All told, the researchers developed 7 such categories of patients (Table 1).

Table 1: Selected Morphological Findings for Different Chiari Types (755 Total Patients)

Type	# of Patients	Occipital Bone Size	PCF Volume	FM
Classical	388	Small	Small	Small
Craniosynostosis	5	Small	Small	Small
CM-II	11	Small	Small	Large
TCS	55	Normal	Normal	Large
Cranial Settling	225	Normal	Normal	Normal
Intracranial Lesions	30	Normal	Normal	Normal
Shunt	28	Normal	Normal	Normal

The largest group was termed Classical Chiari with 388 patients. The second largest group was Cranial Settling with 225 patients involving both trauma and connective tissue disorders. There were 55 cases of Tethered Cord Syndrome and 11 cases of Chiari II.

Next, one of the researchers, who was not aware of the patient categorizations, performed a morphometric analysis using the images of the patients. Specifically, bones in the skull base area were measured, as was the posterior fossa volume, and the size of the foramen magnum. All the measurements were compared to age matched, healthy controls who had also been scanned.

The researchers found, not surprisingly, that the Classical Chiari cases had significantly smaller bones in the occipital region, smaller posterior fossa volumes, and a smaller foramen magnum. Similar in this regard were a

morphology - measurements of the skull

occipital bone - bone in the lower back of the skull

posterior fossa - area in the back of the skull where the cerebellum is situated

Tethered Cord Syndrome (TCS) - term which refers to a condition where the spinal cord is put under abnormal tension, resulting in symptoms

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

[Mechanisms of cerebellar tonsil herniation in patients with Chiari malformations as guide to clinical management.](#) Milhorat TH, Nishikawa M, Kula RW, Dlugacz YD. Acta Neurochir (Wien). 2010 Jul;152(7):1117-27.

handful of cases where the patients had conditions which affected the shape of their skull.

In contrast, patients who the researchers suspected had Tethered Cord Syndrome, had normal occipital bone size and posterior fossa volume, but had significantly larger foramen magnums (as compared to healthy controls). Patients with cranial settling, hydrocephalus, cysts, tumors, and shunt related Chiari all had normal measurements.

Based on the morphometric results, the researchers propose five categories of Chiari etiology: cranial constriction, spinal cord tethering, cranial settling, intracranial hypertension, and intracranial hypotension (Table 2). While this is just a first pass at identifying Chiari subgroups, the authors point out the clinical implication is that a quick morphometric measurement can be done to determine if a patient falls into the cranial constriction category, and if not, that further investigation may be required to identify the underlying cause.

It should be pointed out that this work is not without criticism, especially regarding the controversial link between tethered cord and Chiari. A published comment after the article points out the lack of a theory as to how a thickened filum can actually lead to herniation of the cerebellar tonsils, and also criticizes the researchers for combining Chiari II into this same group.

Regardless, it would be in the best interests of the Chiari community as a whole for researchers to begin thinking about some type of classification of patients in their studies, so that significant results for one subgroup are not lost by including other subgroups.

Table 2: Proposed Chiari Classifications

- Cranial Constriction -
 - Classical
 - Craniosynostosis
- Spinal Cord Tethering
 - TCS
 - Chiari II
- Cranial Settling
 - Trauma
 - Connective Tissue Disorders
- Intracranial Hypertension
 - Hydrocephalus
 - Tumors, cysts
- Intracranial Hypotension
 - Prolonged shunting
 - CSF leaks

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