Key Points

1. Recently, several research publications have focused on the link between Chiari and PTC
2. The exact relationship between the two conditions is not known
3. Study looked at the prevalence rate of Chiari among PTC patients prior to any surgery
4. Reviewed the MRI's and charts of 68 PTC patients
5. Found that 24% had some form of tonsillar descent; 10% had Chiari and 13% had cerebellar ectopia
6. This group was all female and 14 of the 16 were either overweight or obese as measured by BMI
7. Authors believe that Chiari (or CE) can be easily missed on MRI when looking at PTC
8. Also believe that in some people PTC leads to Chiari and in others Chiari leads to PTC

Definitions

body mass index (BMI) - number which is used to generally assess whether someone is overweight, calculated as weight divided by height squared

cerebellar ectopia - in this study, defined as the cerebellar tonsils being located 2mm-4mm below the foramen magnum

ectopia - when an organ or body structure is located in an abnormal position

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

idiopathic intracranial hypertension (IIH) - condition where ICP is chronically elevated for no known reason

inferior tonsillar displacement (ITD) - term used to describe when the cerebellar tonsils are

High Rate of Chiari Found In Pseudo-Tumor Patients

May 20, 2006 -- Pseudotumor Cerebri (PTC), also known as idiopathic intracranial hypertension (IIH), is a condition where a person's intracranial pressure is chronically elevated for unknown reasons. As Chiari & Syringomyelia News has previously reported, there is growing evidence of a link between Chiari and PTC.

Because both conditions involve elevated ICP, there is a good deal of overlap between the two, and their exact relationship is murky. For example, it is not known if the two are just coincidentally related, if PTC causes Chiari, if Chiari causes PTC, or if both are a result of a more fundamental problem. Despite our limited knowledge, some Chiari researchers are beginning to believe that a subset of Chiari patients also have PTC, and that this co-existing condition is a possible explanation for failed Chiari surgeries.

Now, rather than looking at PTC among Chiari patients, a group of researchers from Johns Hopkins and Albert Einstein College (Banik, Lin, Miller) recently studied the prevalence of Chiari among patients initially diagnosed with PTC. They published their results this month in the on-line section of the Journal of Neurological Sciences.

To determine the rate of Chiari among PTC patients, the researchers reviewed the MRI's and medical records of every patient diagnosed with PTC at the two institutions between 1993-2005 and recorded demographic information, height, weight, co-existing conditions, and medications being used.

In addition, a neuroradiologist reviewed MRI's (if they were avialable) to determine if and how much the cerebellar tonsils were descended. For the purposes of this study, the group defined Chiari as the tonsils being located 5mm or more below the foramen magnum, and cerebellar ectopia (CE) as the tonsils being between 2mm - 4mm. They used the term inferior tonsillar displacement (ITD) as a catch-all to refer to both Chiari and CE.

Finally, because there is such a strong link between PTC and obesity, the team calculated the Body Mass Index for each patient and established a scale for normal, overweight, and obese. (see Table 1)

Table 1
BMI Classification of Patients With PTC & ITD

BMI Classification	#
Normal	1
Overweight	2
Obese	12
Not Available	1

Out of 130 patients diagnosed with PTC at the two facilities, MRI's were available for 68, and this group formed the basis for the study. The group of 68 was predominantly female (85%) and their average age was 34 years. Only 8% had a Body Mass Index in the normal range, 20% were considered overweight, and more than 70% were obese.

When they looked at the original MRI reports, they found that only 8 of the patients were noted as having any degree of tonsillar displacement (four with Chiari and four with cerebellar ectopia). However, when the neuroradiologist reviewed the MRI's for this study, he found something much different.

Fully 24% of the group had some level of their tonsils descending out of position (see Table 2). Of these 16 patients, 7 had Chiari (meaning the amount of herniation was at least 5mm) and 9 had cerebellar ectopia. This subgroup - with PTC and ITD - was entirely composed of women, and 14 out of the 16 were either overweight or obese as determined by BMI.

Table 2
Prevalence of ITD in PTC Patients (68 Total)

	#	%
Chiari	7	10
CE	9	13
Total	16	24

descended, regardless of how much

intracranial pressure (ICP) - the pressure of the CSF in the skull

pseudotumor cerebri (PTC) - common name for IIH

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

Source

Banik R, Lin D, Miller NR. *Prevalence of Chiari I malformation and cerebellar ectopia in patients with pseudotumor cerebri.* J Neurol Sci. 2006 May 5; [Epub ahead of print]

Note: *Chiari is defined as cerebellar tonsils at least 5mm below foramen magnum on MRI; CE refers to cerebellar ectopia, defined as the tonsils being descended 2mm-4mm*

The authors note that the significant rate they found in their study (24%) is higher than previously published research, which tended to find single digit rates of Chiari/cerebellar ectopia among PTC patients. They point out, however, that this may be due to missed diagnoses on MRI. With their own study as evidence of how often cerebellar herniation is missed, it is evident that unless problems with the cerebellum are the focus, they are often overlooked.

The researchers also speculate that the link between PTC and Chiari might be two-way. In other words, they believe that some people first have PTC, and the sustained increase in pressure forces the cerebellum out of the skull. But the reverse can also happen; some people are born with a Chiari malformation, which blocks the natural flow of CSF, and leads to a chronic increase in intracranial pressure.

For patients, it is not clear if which condition came first is important for treatment or not. The authors of this study suggest that patients with PTC and Chiari might benefit from Chiari surgery followed by PTC treatment; however, the Chiari literature has shown that decompression surgery for patients with Chiari and PTC is often unsuccessful in relieving symptoms.

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