

Key Points

1. Study found that 21% of IIH patients had tonsillar herniation of 5mm or more
2. Also found that tonsillar position on average is lower among people with IIH
3. Not clear if IIH causes Chiari; Chiari causes IIH; or they co-exist

Definitions

hydrocephalus - condition in which CSF accumulates in the brain

IIH - idiopathic intracranial hypertension, a condition where, for unknown reasons, the intracranial CSF pressure is chronically elevated

intracranial - inside the skull

obex - point at which the brain – more specifically the 4th ventricle – becomes the central canal of the spinal cord

tonsillar ectopia - term used to describe when the cerebellar tonsils are descended below their normal position (< 5 mm or more)

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

syringomyelia - condition where a fluid filled cyst forms in the spinal cord

Source

[Incidence of Cerebellar Tonsillar Ectopia in Idiopathic Intracranial Hypertension: A Mimic of the Chiari I Malformation.](#) Aiken AH, Hoots JA, Saindane AM, Hudgins PA. *AJNR Am J Neuroradiol.* 2012 Jun 21. [Epub ahead of print]

Incidence of Tonsillar Ectopia in IIH

November 1st, 2012 -- As many readers of this article will know first hand, Idiopathic Intracranial Hypertension (IIH or pseudo-tumor cerebri) refers to chronically elevated pressure of the cerebrospinal fluid in the brain. While IIH can be diagnosed by measuring the pressure using a lumbar puncture, it is often brought into play based on symptomology and a relationship to body mass index. Treatments range from life style changes in an effort to lose weight, to medication to reduce the CSF pressure, to the surgical placement of a shunt.

As the idiopathic in its name implies, the underlying cause of IIH is not at all clear, and neither is its relationship to Chiari. There is enough evidence to support the idea that a subset of Chiari patients also have IIH (or vice versa), but which causes which, or if they simply co-exist is not known. In one sense, it makes sense that constant elevated pressure in the brain could result in the eventual herniation of the cerebellar tonsils. On the hand, it also makes sense that if the outlet for CSF from the brain to the spine is blocked, it could alter the CSF dynamics and result in elevated pressure. It is also not out of the realm of possibility that both elevated pressure and herniated tonsils are manifestations of an underlying development problem in the skull and/or brain. The unfortunate consequence of this state of affairs is that some patients undergo Chiari decompression surgery only to find temporarily relief from their symptoms, and eventually undergo treatment for IIH.

A recent publication from a group at Emory University (Aiken et al) quantified the extent of overlap between IIH and tonsillar ectopia. It is important to note that the authors chose the term tonsillar ectopia to indicate that it is not clear in these cases if the herniated tonsils are the same as Chiari. Regardless, the researchers reviewed the MRIs of 43 patients with established IIH (as determined by pressure readings) and compared them to 44 age matched, healthy controls.

In the IIH group, 21% showed tonsillar herniation of 5mm or more compared to just 2% (one case) in the control group. Similarly, the average position of the tonsils was lower in the IIH group at 2.1mm versus 0.7 in the control group. Of the 9 cases where the tonsils were below 5mm, 5 of them were also peg shaped like a traditional Chiari.

Interestingly, the percentage of people with herniations between 2mm-4mm was not significantly different between the groups. However, the researchers did find that the obex was lower on average in the IIH group. They believe this indicates that in IIH, the brainstem and cerebellum as a whole are actually lower than normal.

Although this work isn't ground breaking, it does highlight a very interesting question. Namely, is tonsillar ectopia associated with IIH Chiari? In some sense it may just be a matter of semantics, but if the treatment is fundamentally different – medication or shunt vs decompression surgery – then the point becomes important. Unfortunately, it seems clear that to answer this question will require more sophisticated analysis than straight MRIs; however, the Conquer Chiari Research Center is actively developing tools which might help shed light on this subject.

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[High Rate Of Chiari Found In Pseudo-Tumor Patients](#)

[Idiopathic Intracranial Hypertension aka Pseudotumor Cerebri](#)

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