

Key Points

- Study reviewed every pediatric brain and spine MRI over a 10 year period at the University of Michigan, over 14,000 in all
- 2. Researchers searched clinical records for indications of tonsillar herniation and manually reviewed for cases with at least 5mm of descent
- 3. Found 509 children who fit the traditional definition of Chiari
- Only 32% were considered to be symptomatic at the time of their MRI
- Symptomatic children had larger herniations, were more likely to be female, and had more disruption of CSF flow
- Children with larger herniations were more likely to have a syrinx, but syrinx length and width was not related to herniation size
- Study illustrates the limitations of traditional MRI in diagnosing symptomatic Chiari

Definitions

asymptomatic - having no symptoms

cervical - the upper part of the spine, neck area

hydrocephalus - condition where there is an accumulation of CSF in the brain

lumbar – the lower part of the spine

pc-MRI – phase contrast MRI, type of imaging which can show the flow of CSF

scoliosis – abnormal curvature of the spine

thoracic – the middle part of the spine

tonsillar ectopia – another term for the cerebellar tonsils protruding out of the skull

tonsillar herniation - another term for the cerebellar tonsils

Only A Third Of Children With Chiari On MRI Are Symptomatic

March 31, 2012 -- In a finding that highlights the limitations of the current diagnostic technology, a group of researchers has found that only a third of children with 5mm or more of herniation on MRI – the classic definition of Chiari – were symptomatic. This was one finding in a report in the Journal of Neurosurgery: Pediatrics from a group at the University of Michigan.

The Michigan researchers started with every brain and cervical MRI of a child performed at their institution over a 10 year span, 14,116 in total. Next, they searched the associated clinical records for terms such as Chiari, herniation, ectopia, etc. From this subset, they then individually reviewed the films to identify ones with at least 5mm of tonsillar herniation. Children with Chiari II, hydrocephalus, tumors/cysts, or who had previously undergone Chiari surgery were excluded. In the end, they found 509 cases that met the criteria, which represented 3.6% of the total scans.

Within the final study group, there were 260 females and 249 males. Brain scans were available for the entire group, spine images were available for 78%, whole spine images were available for 50%, and CSF flow studies were available for 61% of the group. The average herniation was slightly more than 10mm and 23% (117) had syrinxes (for the purposes of this study a syrinx was defined as at least 3mm in width.)

While many surgeons anecdotally report that they recommend surgery for less than half of the Chiari cases they evaluate, as stated previously, only 32% of the children with 5mm or more of herniation were considered to have Chiari related symptoms by the physician at the time of the MRI. It is important to keep in mind that it is likely that some of the children categorized as not symptomatic may have in fact had symptoms related to Chiari, especially given some of the reasons for the initial MRI (see Table 1). However, even taking these hypothetical cases into account, the symptomatic group would most likely still comprise less than half of the children with 5mm or greater herniations.

Table 1: Reason For Initial MRI In Children Found To Have Chiari (On MRI)

Reason	%
Headache	23
Scoliosis	16
Neurological Change	12
Seizure	12
Hydrocephalus	9
Cranial Nerve Pals	6
Developmental Delay	5
Trauma	5
Pituitary	4
Tumor/Cyst	4
Other	6

The researchers also found that a significantly higher percentage of girls were symptomatic than boys, and that the amount of tonsillar herniation was significantly larger in symptomatic children (13mm) versus asymptomatic children (9mm). In addition, larger herniations were associated with more disruption of the natural CSF flow. On the other hand, while children with syrinxes had larger herniations on average, the syrinx length and width was not found to be related to herniations size.

As MRIs become more prevalent, their limitations in terms of diagnosing symptomatic Chiari are becoming more glaring by the day. If the definition of Chiari is tonsillar herniation of 5mm or greater, but only one third to one half of children who fit that definition actually have symptoms, one has to question either the test, the definition of Chiari, or both.

Some experts have begun to float the idea of changing the naming convention so that cases where there are no obvious symptoms are called something else, such as cerebellar ectopia. While such a semantic debate may have some value, the development of new testing tools and methods, which can objectively and accurately distinguish between symptomatic and asymptomatic people with herniations, would have a much larger impact on patient experiences and outcomes.

protruding out of the skull

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

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

Source

Chiari malformation Type I and syrinx in children undergoing magnetic resonance imaging. Strahle J, Muraszko KM, Kapurch J, Bapuraj JR, Garton HJ, Maher CO. J Neurosurg Pediatr. 2011 Aug;8(2):205-13. In fact, the development of just such diagnostic techniques will be a major focus area of the soon to open Conquer Chiari Research Center at the University of Akron.

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