Key Points

1. Researchers used the Kaiser Permanente database to study Chiari in children
2. Database contains records of 740,000 people under the age of 20
3. Looked at head and neck MRIs over a two year period to identify Chiari patients (>5mm herniation)
4. Found that 0.7 out of 10,000 children over that time period had identified Chiari
5. This finding supports the Conquer Chiari estimate that 1 in 1,000 people have Chiari.
6. Found 51 children with Chiari but only 32 were symptomatic
7. Found additional 19 with borderline (2-4mm) Chiari
8. Followed the group for an average of 6.4 years
9. Only 4 of the 19 asymptomatic children developed symptoms over that time
10. Only 15% of the children underwent surgery over that time period.

Definitions

ataxia - trouble walking

blinded - in research, when a rater or scorer does not know the purpose of the study, helps to prevent bias

herniation - when a body part is located out of its natural position; with Chiari refers to the cerebellar tonsils being located out of the skull\

natural history - refers to the studying the natural state of something, in this case Chiari; for example looking at symptoms and progression without intervention

tonsillar ectopia - another name for when the cerebellar tonsils are herniated

vertigo - dizziness

Studying The Natural History Of Chiari In Children

May 31st, 2009 -- In the medical literature, Chiari publications are dominated by single patient case reports; basically a physician writes about the details of an interesting case where Chiari may just be one.htmlect of the case. In fact, an analysis by Chiari & Syringomyelia News (*The Year In Research 2008, The Song Remains The Same*) found that over the past several years such case studies have comprised upwards of half of all published research involving Chiari. Unfortunately, while case studies can be interesting, since they only involve one (or a couple) patient, they do little to add to the scientific knowledge base of Chiari.

Beyond case studies, most of the rest of the published research involving Chiari emerges from the individual patient experiences of one doctor or university based group. While these reports are valuable, and sometimes involve hundreds of patients, the fact that all the data comes from one facility limits how broadly the results can be applied.

In scientific terms, the sample of patients is potentially biased. What this means is that one specific doctor may over time attract a specific type of Chiari patient. Therefore any results based on that group of patients, may not be representative of the general Chiari population. Another way to think of the potential for bias is to consider that if a Chiari patient goes to a neurosurgeon, there is a good chance their symptoms are more severe than a Chiari patient who does not seek a surgical opinion.

In a recent publication in Pediatric Neurology, a group of researchers from the University of California, San Francisco and the Kaiser Research Division (Aitken et al.), got around this limitation by using the Kaiser Permanente Medical Program's massive electronic database to explore the natural history of Chiari in children.

Kaiser Permanente provides comprehensive care to more than 30% of the population of Northern California and their database has records for more than 740,000 children (age 20 or less). The researchers wanted to use this database to characterize pediatric Chiari in a way that would be more representative of the general population.

To do this, they used keywords such as Chiari, ectopia, herniation, tonsils, low-lying, etc. to search the database for children under 20 who had either brain or spine MRIs with the diagnosis of Chiari over a two year period (1997-1998). The scientists chose to use a strict definition of Chiari as a minimum of 5mm of tonsillar herniation. Recognizing the potential limitations of this definition however, they also categorized children with 2-4mm of herniation as borderline. Children with Chiari II were excluded from this study.

Once MRIs were identified that met the criteria they had established, the scans were reviewed by an expert blinded to the purpose of the study. The reviewer remeasured the amount of herniation, categorized the shape of the tonsils, and evaluated the amount of space behind the tonsils (for CSF flow). In addition, the associated medical records were reviewed for symptoms, treatment outcomes, and other relevant information.

In total, the team found 51 children with Chiari (greater than 5mm) out of 5248 head and neck MRIs. While this translates to Chiari being found in 1% of the head and neck MRIs, it represents a frequency of only 0.7 cases per 10,000 children in the overall database.

The prevalence of Chiari, meaning how many people suffer from it in the general population has not been established. Conquer Chiari has estimated (based on different data points, such as number of surgeries performed annually) that about 1 in 1,000 people have Chiari, or roughly 300,000 in the US. Interestingly, while the authors of this study did not try to calculate prevalence (because of statistical limitations), using their data and a few assumptions yields a result very close to the Conquer Chiari estimate.

First, it is important to note that while the research team identified 51 children, only 32 of them were symptomatic. For the purpose of this exercise, we will stick with symptomatic patients only. So, let's assume the following:

1. Prevalence is roughly equivalent to the incidence of a disease times the disease duration. Think about a pool of people with new cases coming in each year and some people dying each year either due to the disease or other causes.

2. The average age of diagnosis is 11 (based on this study). Clearly this is only for those patients who are diagnosed as children rather than adults, but it is likely that with increased use of MRIs that more and more people are being diagnosed in childhood.

3. Average duration is 60 years. This assumes that Chiari has only a minimal impact on lifespan in general.

Using these numbers produces a prevalence estimate of 1.2 per 1,000. However, as noted the average age of

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

Chiari type I malformation in a pediatric population. Aitken LA, Lindan CE, Sidney S, Gupta N, Barkovich AJ, Sorel M, Wu YW. Pediatr Neurol. 2009 Jun;40(6):449-54

diagnosis for both children and adults is probably more likely to be around 20 so this would bring the number down somewhat. On the other hand, this only counted patients with at least 5 mm herniation, and since we know some patients have smaller herniations, this would bring the number back up. Also, there are likely to be missed and mis-diagnoses in the population that was studied which would also increase the prevalence number. In the end, this is an unscientific analysis, but it is interesting that the results from this study tend to support the idea that 1 in 1,000 people have Chiari.

Turning back to the UCSF study, the researchers were able to track, using medical records, the children for an average of more than six years. They found, not surprisingly, that headache and neck pain were the most common symptoms (Fig 1). Other common symptoms included vertigo, numbness in the hands/arms, and ataxia. Interestingly, in contrast to the typical Chiari headache in adults, very few children reported pain in the back of their head, or that the headaches were brought on by straining.

In looking at the MRIs, the average herniation was 7 mm, and nearly 1 in 4 had herniations greater than 10 mm (Fig 2). Slightly more than half showed pointed tonsils and reduced space for CSF flow. It should be noted that these two findings went hand in hand, meaning that almost every child with pointed tonsils also had reduced space behind them.

In a disturbing finding, only half of the children were diagnosed within 14 months of the onset of symptoms. However, this data was from 10 years ago, so one would hope that this number would improve if more recent years were looked at.

As mentioned before, only 63% of the children were symptomatic. This means that nearly 4 in 10 children with herniations greater than 5mm did not originally have any symptoms associated with Chiari. Over time however, 4 of these 19 children did develop some symptoms. Overall however, only 15% of all the children ended up requiring surgery.

The group also noted that while 75% of the borderline (2-4mm) children suffered headaches, none of them showed pointed tonsils or reduced CSF space on MRI. This again raises the notion that perhaps patients with small herniations represent a different sub-group of patients and may fare better with a different treatment.

Overall this study presented a lot of interesting data without providing much context. However, it does serve as a strong reminder that results arising from surgical patient series may be inherently shifted towards the worse end of the spectrum (patients with more severe cases opt for surgery), so it is valuable to see data from a more general group.

Figure 1: Common Symptoms Among Identified Chiari Children (51 Total)

	At Diagnosis	Developed During Follow-Up	Total
Headache	55%	6%	61%
Neck Pain	12%	10%	22%
Vertigo	8%	6%	14%
UE Numbness	6%	8%	14%
Ataxia	6%	4%	10%

Note: UE - upper extremity; headaches were rated as severe, moderate, or mild

Figure 2: Selected Characteristics of Identified Chiari Children (51 Total)

Symptomatic	63%
Avg. Herniation	7 mm
>10mm Herniation	22%
Pointed Tonsils	55%
Reduced Space Behind Tonsils	57%
Syrinx	12%
Diagnosed In Less Than 14 Months	50%

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