Research Update | July, 2019

Topics: "Experts" Agree 5mm Rule Not Valid; Origins of the 5mm Chiari Rule

Conquer Chiari's monthly research updates highlight and summarize interesting publications from the medical literature while providing background and context. The summaries do contain some medical terminology and assume a general understanding of Chiari. Introductory information about Chiari, plus many more research articles, can be found at www.conquerchiari.org.

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"Experts" Agree 5mm Rule Not Valid

For patients who have seen several doctors looking for Chiari answers it can often seem like no one agrees on anything about Chiari; however, a recent survey of Chiari "experts" found a surprising amount of agreement on a number of topics. Specifically, 100 clinicians with published research on Chiari were contacted for the survey. Sixty-three of them, from four continents and representing an experience base of over 15,000 patients, responded. The bulk of the survey consisted of 51 statements about Chiari and the clinicians were asked if they agreed or disagreed. For 15 of these statements, 90% or more of the experts indicated they agreed, and more than 75% agreed with 29 of the 51 statements. On the other end of the spectrum, the majority of experts disagreed with 13 of the statements. The headline for patients is that nearly 90% of these experts do NOT agree with the old 5mm Chiari rule (meaning that herniations less than 5mm are not Chiari). From the responses, the authors put together several consensus statements which they believe at least 75% of Chiari experts currently agree with (note the wording of the statements has been modified for this update):

- Typical Chiari I is characterized by a small posterior fossa which results in tonsillar herniation
- Tonsillar herniation can also be caused by other factors, but these cases are not the same as Chiari I
- The old radiological axiom that tonsillar herniation less than 5mm is not Chiari I is NOT valid
- There is likely an as yet unknown genetic component to Chiari I, but it is heritable in less than 25% of cases
- Tonsillar herniation can cause a mass effect on surrounding tissue and disrupt CSF flow
- Symptoms are not limited to cough related headache
- Symptoms can be triggered by head/neck trauma
- Chiari I is not a Rare Disorder as defined by the Rare Disease Act
- Chiari I is more prevalent in females
- Comorbidities include connective tissue disorders and pseudotumor cerebri
- Complex Chiari I involves additional craniovertebral abnormalities such as basilar invagination or cervical instability

The authors plan on reporting on the second part of the survey, which involved surgical technique in a later publication.

SOURCE:*Chiari I Malformation: Opinions on Diagnostic Trends and Controversies from a Panel of 63 International Experts. Bolognese PA, Brodbelt A, Bloom AB, Kula RW. World Neurosurg. 2019 May 20.*

Origins of the 5mm Chiari Rule

If you're a Chiari veteran, you've probably heard it over and over…Chiari is 5mm or more of tonsillar herniation. But do you know where this definition came from? It didn't come Hans Chiari, the pathologist who first described several cases (which incidentally were not representative of what we think of as Chiari I today), and it didn't come from Arnold, whose students somehow got his name attached to the disorder for a period of time. In fact, like far too many things that are taken as given in science and medicine, the origins of the 5mm rule are a bit murky and sadly do not have a solid scientific basis. In the 1980s, the advent of MRIs provided an opportunity to study Chiari malformation non-invasively and also quantitatively. While it is likely not the whole story, a publication in 1986 (Barkovich) by a group of radiologists used MRI to measure the tonsillar position and is often cited as the origin of the 5mm rule. The study involved 25 Chiari cases and 200 healthy controls. The Chiari group was selected as having definite Chiari clinical signs and symptoms. The MRIs by today's standards were low powered and low resolution. The researchers used hard copies of the images for their measurements. They did find that in the control group, the tonsillar position ranged from 8mm above the foramen magnum to 5mm below and there was overlap between the groups in the 3-5mm range. The researchers then calculated the accuracy of using different cut-offs between 1-5mm. From this data, over time people interpreted that Chiari should be at least 3-5mm of herniation. For reasons that are not clear, this later evolved to 5mm. Interestingly, the authors in the Barkovich paper stress in their discussion of the results that it is better to err on the side of inclusion and to not rule out Chiari too early, and therefore suggest a cut-off of 2mm. Of course, the problems and limitations with these cut-offs in general have been discussed extensively in past research updates and indeed were laid bare by Milhorat in the late 90s with his seminal article showing that symptomatic Chiari does exist with smaller herniations. More recently, studies by Meadows and Maher have shown that 1-3% of the general population may meet the 5mm rule for Chiari, but that only a small fraction of these will ever be symptomatic. As the survey discussed above shows, the Chiari community is now at the point where experts the Chiari community is now at the point where experts discount the 5mm rule and researchers are looking aggressively for a new way to define Chiari malformation.

SOURCE: Significance of cerebellar tonsillar position on MR. Barkovich AJ, Wippold FJ, Sherman JL, Citrin CM. AJNR Am J Neuroradiol. 1986 Sep-Oct;7(5):795-9.

Conquer Chiari is a 501(c)(3) public charity dedicated to improving the experiences and outcomes of Chiari patients through education, awareness and research.