

Research Update: January, 2019

Topics: Natural History; Measuring Tonsillar Position Consistently is Difficult

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.

- Several recent publications have shown that the vast majority of people who are found to have herniations over 5mm will not need surgery and are unlikely to develop symptoms. Now, a large pediatric study from the University of Alabama provides more evidence that kids who are found to have Chiari on imaging, but do not have classic Chiari symptoms or CSF issues, are unlikely to require surgery in the near to mid-term. Specifically, the researchers examined 10 years of records at their institution and identified over 1600 children who were noted to have tonsillar herniation of at least 5mm. They then removed all the patients who had surgery within the first nine months, syringomyelia, hydrocephalus, Valsalva headaches, or other symptoms traditionally associated with Chiari. This left close to 700 patients, but a large group of these (over 250) did not return for follow-up exams. The rest, 427, were followed for an average of about 2 years, with some for more than 10 years. Out of this group, 15 ended up undergoing Chiari surgery at some point in time, representing only 3.5% of the conservatively managed group. Of the fifteen, 5 had surgery for Valsalva headaches, 5 because a syrinx formed, and 5 for other types of headaches. Interestingly, all 5 of the Valsalva headaches resolved with surgery, but only 3 of the syrinxes did, and only 3 of the general headaches improved. The researchers looked at a number of variables, but could not identify any that predicted who went on to develop symptoms and need surgery. It should be noted that the outcomes of the large number of children who did not return after the initial visit could dramatically alter the results of this study. If a large percentage of them eventually required surgery and went elsewhere, then of course the risk would be higher than the authors found. On the other hand, if they all stayed symptom free, the percentage of kids who required surgery would drop even further.

SOURCE: Patients with "benign" Chiari I malformations require surgical decompression at a low rate. Leon TJ, Kuhn EN, Arynchyna AA, Smith BP, Tubbs RS, Johnston JM, Blount JP, Rozzelle CJ, Oakes WJ, Rocque BG. J Neurosurg Pediatr. 2019 Jan 4:1-9.

- Chiari malformation has traditionally been defined based on imaging showing the cerebellar tonsils 3-5mm or more below the foramen magnum (the opening at the bottom of the skull). However, there are several problems with this definition. First, research has shown that 1-3% of the general population will show tonsillar herniation of at least 5mm on imaging, but have no symptoms. Second, even among symptomatic Chiari cases, the extent of the tonsillar herniation is not strongly correlated with symptom severity. Now, a new report from the Conquer Chiari Research Center adds a third problem to the mix; namely that measuring the tonsillar position on MRI varies a good deal, even among experienced clinicians. For the study, 33 sets of MRIs were given to 7 experts (neurosurgeons, neurologists, radiologists) who were asked to measure the tonsillar position of each case using the standard McRae line methodology. The MRI set included 10 controls with no herniation, 11 surgical Chiari cases, and 12 non-surgical Chiari cases. The experts used their own choice of software and had to select their own image view from each set. While statistically the overall correlation of those measuring was considered clinically good, there were some striking findings in the details. For example, the average range of measurements for the entire set was over 7mm, and the range of measured positions for 30 out of the 33 cases (90%) was 5mm or more, the very definition of the Chiari. In addition, 8 of the 33 cases had false negative measurements, meaning at least one person measured the tonsillar position at less than 3mm while others put

it at over 5mm. Similarly, there were even more false positives where an image was classified by one person as having tonsillar position at more than 5mm but others measured it at less than 3mm. This study highlights the challenges some people face when an initial MRI report shows a herniation of say 4mm and they are told that is not enough to cause symptoms. At this point they essentially become stalled in the diagnostic process. This is why the CCRC is putting so much time and energy into morphometrics research and trying to identify additional, quantifiable data beyond tonsillar position to help with Chiari diagnosis.

SOURCE: Cerebellar tonsil ectopia measurement in type I Chiari malformation patients show poor inter-operator reliability. Lawrence BJ, Urbizu A, Allen PA, Loth F, Tubbs RS, Bunck AC, Kröger JR, Rocque BG, Madura C, Chen JA, Luciano MG, Ellenbogen RG, Oshinski JN, Iskandar BJ, Martin BA. *Fluids Barriers CNS*. 2018 Dec 17;15(1):33.