Research Update | May, 2019

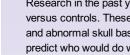## Topics: Digging Deeper Into Cognitive Effects; Can Morphology Predict Outcome

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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Now that several studies have found varying types of cognitive deficits in Chiari patients, researchers are beginning to dig deeper into the cognitive impact of Chiari. Previous studies have relied on either extensive or modified testing batteries to assess cognitive performance. While these were successful, they may not be practical to use in a clinical setting. Therefore, researchers at the CCRC wanted to see if the same cognitive effects could be found using a faster, widely accepted clinical tool known as RBANS (Repeatable Battery for the Assessment of Neuropsychological Status). They also wanted to determine if the cognitive impact of Chiari was caused by the indirect effects of the pain that Chiari causes – chronic pain is known to affect cognition – or is tied more directly to the Chiari pathology. In a recent publication in the journal Neuropsychology, the CCRC team reported they found significant differences in 18 adult Chiari patients compared to carefully matched healthy controls in the areas of attention, immediate memory, delayed memory, and overall RBANS score. Interestingly, when they statistically controlled for self-reported pain, the deficit in the attention score remained, but the others did not. This implies that the memory issues Chiari patients have could be a result of the chronic pain caused by Chiari, but that other cognitive issues are more directly tied to the Chiari pathology. To explore this further, the research team quantified the brain and skull morphology of the participants' MRIs (these morphometric measurement techniques have been reported on in previous Research Updates). They found that people with larger posterior fossa bone structure did score better overall and on the attention sub-scale of the RBANS test. Surprisingly, they also found a correlation between the extent of tonsillar herniation and the level of self-reported pain. While all these results are interesting, the main takeaway from this work is that the RBANS, which takes only 30 minutes to administer, can be used to identify cognitive deficits in Chiari patients in a clinical setting and over time may become a standard part of the Chiari diagnostic process.

SOURCES: *Type I Chiari malformation, RBANS performance, and brain morphology: Connecting the dots on cognition and macrolevel brain structure. Houston JR, Allen PA, Rogers JM, Lien MC, Allen NJ, Hughes ML, Bapuraj JR, Eppelheimer MS, Loth F, Stoodley MA, Vorster SJ, Luciano MG. Neuropsychology. 2019 May 16.*

Research in the past year or so (in large part from the CCRC) has found significant differences in the skull and brain morphology of Chiari patients versus controls. These differences go well beyond tonsillar position and some of the most significant include a shortened clivus, lower hindbrain, and abnormal skull base angles. Given that, a group from China wanted to see if any of these types of morphometric measures could be used to predict who would do well with Chiari surgery and who wouldn't. The Chinese team looked at 39 adult surgical Chiari patients and took more than 20 linear, angular, and area measurements from their MRIs. They used the pain and non-pain symptom subscales of the Chicago Chiari Outcome Scale (CCOS) to measure outcome. Based on the modified CCOS scores, the patients were divided into two groups: improved/successful and not improved/unsuccessful. Interestingly, the researchers found NO significant morphometric differences between the successful and unsuccessful outcome groups, meaning that the morphometrics did not predict who would do well with surgery. It should noted that while interesting, this study does have several limitations. First, the failure rate for their patients was nearly 40%, which is about double what is reported in the US and Europe. This could be because nearly 90% of their patients also had a syrinx, which is not representative of the general Chiari population. Second, only about one third of the surgical patients they saw over the given time span were included in the study (for various reasons). This is a fairly low participation rate and again means the sample may not be representative of a larger group. Despite these, the work raises an interesting question, namely can static geometry be used to predict who is a good candidate for decompression surgery, or do dynamic factors need to be looked at as well.

SOURCES: Predictive value of posterior cranial fossa morphology in the decompression of Chiari malformation type I: A retrospective observational study. Liu Z, Hao Z, Hu S, Zhao Y, Li M. Medicine (Baltimore). 2019 May;98(19):e15533.

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

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