

Research Update: January 2018

Topics: Morphometrics, Symptomatic vs Asymptomatic, Scoliosis, Heart Related Symptoms, Fibromyalgia, Cerebrospinal Fluid Pressure

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.

- 🎧 A study from the University of Michigan and researchers from the Conquer Chiari Research Center (CCRC) found no real differences in looking at 10 morphometric measures in comparing 51 children with symptomatic Chiari and 51 children with tonsillar herniation greater than 5mm, but with no symptoms. The CCRC and others have previously focused on using morphometrics – measurements of the brain and skull – to compare Chiari patients to healthy controls. These studies have produced some interesting results; however, identifying why some people with tonsillar herniation show symptoms while others don't remains elusive. This study specifically looked at 6 two-dimensional measures and 4 volume based measures but found no statistical differences between the groups. Estimates for the number of people with tonsillar herniation seem to keep going up, with some saying that as many as 3% of children have tonsillar descent of 5mm or more. However, the large majority of these individuals does not, and probably won't, experience Chiari type symptoms. Thus, the important question of why? What makes one person suffer from symptoms while another doesn't? The CCRC will continue to investigate using more morphometrics, but there is a possibility that the answer is not in MRIs and that other avenues of investigation will need to be pursued. **SOURCE:** *Morphometric and volumetric comparison of 102 children with symptomatic and asymptomatic Chiari malformation Type I. Khalsa SSS, Geh N, Martin BA, Allen PA, Strahle J, Loth F, Habtzghi D, Urbizu Serrano A, McQuaide D, Garton HJL, Muraszko KM, Maher CO. J Neurosurg Pediatr. 2018 Jan;21(1):65-71.*
- 🎧 Chiari related scoliosis (an abnormal curvature of the spine) is one area that has actually received a significant amount of research attention. Among adults, the presence of scoliosis has been shown to be a predictor of poorer outcomes. Among children, Chiari related scoliosis often presents with atypical curve patterns. In addition, studies have shown that Chiari decompression surgery, in the short-term, can be successful in halting the progression of the scoliosis and prevent the need for spinal fusion. Unfortunately, a recent study from Utah found that the long-term success of Chiari decompression surgery in treating scoliosis is not as promising. Specifically, among 23 children who had Chiari surgery for Chiari related scoliosis, nearly half (11) eventually required spinal fusion anyway, some as long as 5 years later. Statistically, the extent of the Chiari herniation and the presence of syrinxes did not predict who would later require fusion surgery. However, the initial severity of the scoliosis did, which may help inform treatment decisions regarding Chiari related scoliosis moving forward. **SOURCE:** *Chiari-related scoliosis: a single-center experience with long-term radiographic follow-up and relationship to deformity correction. Ravindra VM, Onwuzulike K, Heller RS, Quigley R, Smith J, Dailey AT, Brockmeyer DL. J Neurosurg Pediatr. 2017 Nov 24:1-5.*

👤 A medical team from California reported an interesting case study. A 31 year old woman suffered from minor headaches (her classification), but repeatedly went to the ER for heart related issues such as a rapid heart beat and palpitations. Heart medication didn't work and neither did a heart procedure known as ablation (part of the heart muscle is scarred to disrupt abnormal electrical signaling). An MRI then revealed a significant Chiari. Subsequent decompression surgery resolved all of her heart issues and her headaches. Heart related issues, in terms of POTS, have been discussed in the Chiari literature before, and in fact about 3% of patients in the Chiari 1000 report heart related symptoms. However, it is striking to see a case where the heart issues resolved so quickly and completely after decompression surgery.

SOURCE: *Resolution of tachyarrhythmia following posterior fossa decompression surgery for chiari malformation type I: A case report.* Elia C, Brazdzionis J, Tashjian V. *World Neurosurg.* 2017 Dec 27.

👤 Medical Hypotheses is a journal which provides a forum for scientists and clinicians to speculate. While most journals are focused on experimental results or case studies, this journal allows researchers to submit articles where they lay out their ideas about different diseases. In a recent issue, a group from Belgium published an interesting theory on Fibromyalgia where they link the widespread body pain to abnormal cerebrospinal fluid (CSF) pressure. In their theory, the abnormal CSF pressure results in CSF flooding into the sheaths of nerve roots and causing damage, which in turn results in the characteristic pain. Some of the evidence they cite to support their theory are studies which have shown that Fibromyalgia patients show definitive indications of neurological deficits upon exam, studies which show that Fibromyalgia patients lose muscle strength, and studies of idiopathic intracranial hypertension (IIH or pseudotumor) patients who show indications of nerve sheath damage. It is of course well recognized that Chiari leads to a disruption in the regulation of CSF pressure, and both IIH and Fibromyalgia diagnoses are commonly reported among CM patients. Data from the Chiari 1000 indicates that 22% of adult women with CM also report a Fibromyalgia diagnosis and 10% report an IIH diagnosis, so this theory may be an interesting one to explore for the Chiari research community. The authors, however, acknowledge that testing their theory will be quite challenging. **SOURCE:** *Fibromyalgia and unexplained widespread pain: The idiopathic cerebrospinal pressure dysregulation hypothesis.* Hulens M, Dankaerts W, Stalmans I, Somers A, Vansant G, Rasschaert R, Bruyninckx F. *Med Hypotheses.* 2018 Jan;110:150-154.