

Topics: Cognitive Effects; Spontaneous Resolution, Syrinx, Pediatric; Novel MRI

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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The team at the Conquer Chiari Research Center has demonstrated fairly convincingly that Chiari often has a negative impact on cognitive function; however, much work remains to be done to fully characterize and understand this effect. In a recent publication, researchers from Spain contributed to this effort by looking at the cognitive impact of Chiari on a group of adults who had not had surgery. Specifically, 39 patients underwent an extensive battery of neuropsychological testing and were compared to 39 controls matched for age, gender, and education levels. The researchers found that the Chiari patients showed deficits in many areas, including executive function, verbal fluency, copy accuracy, visual memory, naming ability, verbal memory, processing speed, and facial recognition. Since pain, anxiety, and depression are known to negatively impact cognitive function, the group used a statistical technique to control for this and found that the Chiari group still showed deficits beyond what could be attributed to those factors. This implies that damage to the cerebellum itself, or damage to pathways to other parts of the brain, are playing a role in the cognitive deficits of Chiari patients. It is not clear when clinicians will start including neuropsychological testing as part of the standard Chiari diagnostics, but the evidence keeps mounting that it may provide valuable information, especially for cases without classic Chiari symptoms.

SOURCE: *Cognitive Functioning in Chiari Malformation Type I Without Posterior Fossa Surgery.* García M, Lázaro E, López-Paz JF, Martínez O, Pérez M, Berrocoso S, Al-Rashaida M, Amayra I. *Cerebellum.* 2018 May 15

An interesting case report from India highlights the dilemma that parents face when deciding whether a child with Chiari should undergo surgery. Research has shown that most children who are found to have tonsillar herniation, but are not symptomatic, are unlikely to develop symptoms over the next several years. Most surgeons do not recommend any type of treatment in these cases, and in fact, case reports have shown the amount of herniation may improve over time. However, when there are symptoms and a syrinx, the vast majority of doctors do recommend surgery since a syrinx can cause permanent nerve damage. But what about when parents go against this advice? The doctors from India report on two children, an 8 year old boy and a 9 year old girl, with Chiari and syringomyelia, whose parents declined surgery. The boy had a 14mm herniation and syrinx extending from C3 to T4. He had issues swallowing when drinking fluids. After declining surgery, he was seen every 6 months for 7 years. At his most recent visit, the herniation was down to 3mm, the syrinx had shrunk significantly, and he had no symptoms. The girl suffered from headaches and neck pain and had a 7mm herniation with a sizable syrinx. She too was observed for 7 years and like the boy her herniation improved over time, the syrinx resolved, and she became symptom free. It is difficult to interpret what these case reports mean because the data simply is not there as to how often this occurs and whether it can be predicted based on certain anatomical or other factors. It is also important to note that there are case reports of sudden and rapid neurological deterioration after trauma in patients who had not yet been diagnosed with Chiari and syrinxes.

SOURCE: *Spontaneous Resolution of Chiari 1-Associated Syringomyelia: A Report of Two Cases.* Ramnarayan R, Ganesh CVS, Kumar R. *Pediatr Neurosurg.* 2018 May 7:1-5.

 MRI's play an important role in the diagnosis of Chiari and are an active subject of ongoing research, as shown in two recent publications. First, in a project funded by Conquer Chiari, a Harvard researcher used a new MRI technique, called real-time pencil beam MR to measure how much CSF flowed across the craniovertebral junction in thirteen Chiari patients. The patients were divided into two groups: those with cough related headaches and those other types of headaches or no headaches at all. The patients were measured at rest and then were asked to cough as hard as they could. The MRI showed that for those with cough headaches, the volume of CSF flow (as a percentage) dropped significantly after coughing compared to the non-cough headache group. In addition, the percentage drop in CSF volume was correlated with symptom severity as determined by two neurosurgeons blinded to the MRI results. Although this study was small, if developed and validated further, it would provide clinicians with a way to quantify the severity of the signature Chiari symptom, the cough headache.

In the second study, led by Dr. Bryn Martin formerly of the CCRC, an existing technique, PC-MR, was used to examine the motion of the spinal cord in Chiari patients versus controls. PC-MR has previously been used to look at CSF flow with mixed results, but in this project the researchers used it to measure the motion of the spinal cord at the level of the foramen magnum in 20 surgical Chiari cases and 15 controls. They found that in the pre-surgical Chiari cases there was significantly more spinal tissue motion than in the controls. They also found that after surgery, the spinal cord motion decreased significantly among the patients. The researchers point out that this motion, driven by the heartbeat, occurs about 30 million times a year and may explain some of the cord related symptoms associated with Chiari. Similar to the pencil beam technique, if this data is validated further, spinal cord motion could also provide clinicians with additional quantitative data about Chiari.

SOURCE: *Relationship between Cough-Associated Changes in CSF Flow and Disease Severity in Chiari I Malformation: An Exploratory Study Using Real-Time MRI.* Bezuidenhout AF, Khatami D, Heilman CB, Kasper EM, Patz S, Madan N, Zhao Y, Bhadelia RA. *AJNR Am J Neuroradiol.* 2018 May 10.

Cardiac-related spinal cord tissue motion at the foramen magnum is elevated in Type I Chiari malformation patients and decreases post-decompression surgery. Lawrence BJ, Luciano M, Tew J, Ellenbogen RG, Oshinski JN, Loth F, Culley AP, Martin BA. *World Neurosurg.* 2018 May 4.

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