


Syrinx Symptoms After Surgery; Cognitive Impact; Chiari Anatomy


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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 Syringomyelia, or the abnormal collection of fluid in the spinal cord itself, can be a devastating consequence of Chiari. The syrinx can damage nerve roots leading to loss of sensation, neuropathic pain, weakness, muscle atrophy, and even paralysis. There are several theories on syrinx formation and progression, but in general it is not well understood and it is not known which Chiari patients are at risk for developing a syrinx. When a syrinx is present, the goal of decompression surgery is to stop the progression of syrinx related symptoms, as once the nerves are damaged they may not recover. As with Chiari, clinically the success of surgery is often determined from imaging, meaning did the syrinx shrink significantly or resolve completely? However, a recent publication from the United Kingdom highlights that sometimes even what appears to be a successful surgery may not stop the progression or even the onset of symptoms. A surgical group reviewed 50 of their cases involving Chiari and syringomyelia and identified three cases where the Chiari surgery appeared to be successful, but syrinx related symptoms developed or worsened over time. For example, the one case was a 12 year old boy with severe, progressive scoliosis, 7mm of tonsillar descent with a large syrinx, but no other neurological signs or symptoms. His decompression surgery went well and he recovered quickly. The follow-up MRI four months later showed his syrinx had shrunk almost completely. However, a year later he developed numbness and weakness in his arms and legs. At that point there wasn't really anything else the doctors could offer him and he continues to suffer. The other two cases in the paper are similar in that after surgery the syrinx on imaging improved but the patients' symptoms became worse a few months to a year later. These cases highlight how little is still known about the formation of syrinxes as related to Chiari, but it is also important to keep in mind that in this series these cases represented only 6% of the overall patient group.

SOURCE: *Clinical deterioration despite syringomyelia resolution after successful foramen magnum decompression for Chiari malformation - Case series.* Shetty J, Kandasamy J, Sokol D, Gallo P. *Eur J Paediatr Neurol.* 2019 Mar;23(2):333-337.

 Research into the cognitive effects of Chiari continues to heat up. Recently a group from the University of Chicago reported the results of a study which adds to the rapidly mounting evidence that Chiari has a measurable impact on the cognitive functioning of many adult patients. Specifically, this study compared 18 people with Chiari (before surgery) to 18 volunteers across a variety of cognitive testing. They found that the Chiari group scored significantly lower in the overall categories of immediate memory and language. Specific sub-categories which were affected in the Chiari group included story recall, verbal fluency, copying figures, and visual recall. Interestingly, and in-line with findings from the Chiari1000, the research team also found that a disturbingly high 45% of the Chiari patients were in the clinical range on a depression scale. However, level of depression was not related to cognitive performance. This is the latest in a number of similar studies published over the last two years. While the results of these studies vary in the details – meaning the specific cognitive areas effected – each one has found that the Chiari group performs worse than a control group across a battery of cognitive tests. The next step in cognitive Chiari research is to use advanced imaging such as DTI and resting state fMRI to understand why and how these deficits occur by identifying specific brain regions and connections that may be damaged. Along these lines, Conquer Chiari recently awarded a large grant to the CCRC to do just that. With the increased attention it is receiving, hopefully we are starting to turn the corner on this important area of Chiari research.

SOURCE: *Neurocognitive Functioning in Unoperated Adults with Chiari Malformation Type I.* Lacy M, Parikh S, Costello R, Bolton C, Frim DM. *World Neurosurg.* 2019 Mar 1.

Most of the articles in the Conquer Chiari Research Updates are based on recent publications in the medical and scientific literature. However, it is also important for educated and empowered Chiari patients to have a broad understanding of a number of different topics, including human anatomy. Naturally, much the anatomical focus as related to Chiari concerns the cerebellum, the cerebellar tonsils, and the back part of the skull known as the posterior fossa. What some people may not realize is that many symptoms associated with Chiari can be traced to the brainstem and cranial nerves. The cranial nerves are 12 pairs of nerves that originate in the brain as opposed to the spinal cord (10 of the 12 start in the brainstem). The cranial nerves are responsible for a variety of functions including smell, eye control, chewing, swallowing, balance and more. The chart below indicates the name and general function of each cranial nerve. A quick review reveals how Chiari symptoms such as nystagmus, abnormal gag reflex, fullness in the ears, and others may at least partially arise from some type of damage to or pressure on one or more of the cranial nerves. It is important to note that some of these complex functions, such as balance, also involve the cerebellum as well.

CN Number	Name	General Function
I	Olfactory	Sense of smell
II	Optic	Convey visual information
III	Oculomotor	Eye movement; pupil constriction
IV	Trochlear	Eye movement
V	Trigeminal	Facial sensation; chewing
VI	Abducent	Eye movement
VII	Facial	Facial movement; taste; salivation
VIII	Vestibulocochlear	Balance and equilibrium; hearing
IX	Glossopharyngeal	Oral sensation; taste; gag reflex
X	Vagus	Swallowing; blood pressure; heart rate
XI	Accessory	Head and shoulder movement
XII	Hypoglossal	Tongue movement

SOURCE: Gould, Douglas. (2014). *Neuroanatomy – 5th Edition*. Baltimore, MD: LWW.

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