Research Update | April, 2019

Topics: Complex Chiari (Cranio-vertebral Junction Disorders)

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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Have you ever heard the term Complex Chiari and wondered what exactly it means? Well, like many Chiari terms Complex Chiari does not have a precise definition and is not universally accepted. However, one common way the term is used is to refer to Chiari patients who also have one or more additional cranio-vertebral junction (CVJ) abnormalities. These are well recognized anatomical issues that can also exist without Chiari, but are found in a significant number of Chiari patients. The most common CVJ abnormalities associated with Chiari are basilar invagination (BI), retroflexed odontoid, ventral brainstem compression (brainstem kinking), and cervical instability. To understand these issues, it's important to first identify a key anatomical feature, the odontoid. The odontoid is a bony projection (often described as tooth-like) which extends up from the second vertebra like a post. The first vertebra essentially sits on the odontoid and is able to rotate around it, thus enabling the rotation of the head. With basilar invagination, the top of the odontoid is located too high and pushes into the skull (the opposite of Chiari which pushes out of the skull). The amount of basilar invagination can be measured on MRI and the normal limits are fairly well established. In a large study of Chiari adults, Klekamp found that 14% also had basilar invagination. When BI exists without Chiari, if there is compression on the brainstem it is treated by what is called an anterior decompression. The surgeon approaches the region either through the back of the mouth or endoscopically through the nose. When BI exists with Chiari, patients are often treated with the standard Chiari posterior decompression, but a subset will require both surgeries (and cervical fusion as well).

A retroflexed odontoid refers to when the odontoid does not project straight up from the second vertebra. Rather it is angled back towards the brainstem. Like BI, this angulation, or retroflexion, can be measured on MRI. For some people, the odontoid position will cause the brainstem to bend abnormally, or kink. This can also be assessed on MRI, and normal ranges have been established for clinical use. In a large pediatric series, Brockmeyer reported that 20% of Chiari patients had retroflexed odontoids. The treatment approach for Chiari patients with BI and/or retroflexed odontoid may vary from surgeon to surgeon, with different criteria for when the odontoid should be resected and when cervical fusion/stabilization is required.

These CVJ problems, or other issues such as EDS, can also lead to cervical instability, which means excessive movement between adjacent vertebrae. Unlike BI and odontoid retroflexion, cervical instability is not as easily assessed and may require taking MRIs with the head in different positions. If the instability is severe enough, a surgeon may choose to stabilize the region by fusing vertebra and/or using instrumentation. In the same pediatric series, Brockmeyer found that 10% of the pediatric Chiari patients required additional fusion for stabilization. In an adult series from the Chiari 1000, Eppelheimer found that 7% of female Chiari patients reported they had been diagnosed with cervical instability.

It is important to keep in mind that these issues are not exclusive, and more often than not actually overlap. This is especially true in Chiari patients with EDS. Henderson reported on 22 adult Complex Chiari patients with geneticist confirmed hypermobile connective tissue disorder who all exhibited CVJ abnormalities requiring cervical stabilization.

Interestingly, these CVJ issues may also relate to Chiari 1.5. Some years ago, Chiari 1.5 was a focus of discussion and referred to patients with some features of Chiari 1 and some features of Chiari II. More specifically, it can be defined as when there is also brainstem descent past the foramen magnum (as opposed to just the cerebellar tonsils). Using this MRI based definition, Brockmeyer found that 18% of his pediatric series met the criteria for Chiari 1.5. He also found that these cases were the ONLY ones who required cervical fusion or odontoid resection. In other words, none of his pediatric cases with descent of only the cerebellar tonsils had CVJ issues sever enough to warrant additional surgery. In a different pediatric series of Chiari 1.5 patients, Tubbs found that the major difference between Chiari 1.5 and Chiari 1 was that a higher percentage of the Chiari 1.5 cases required a second posterior fossa decompression for persistent syringomyelia.

While the main features of Complex Chiari are CVJ issues such as BI, retroflexed odontoid, brainstem kinking, and Chiari 1.5, some researchers also include issues such as scoliosis and syringomyelia. Given the overlap with connective tissue disorders, it might also make sense to include EDS in the mix. However it is grouped, Complex Chiari cases are more likely to require surgery beyond the standard Chiari decompression.

Prevalence of CVJ & Related Conditions in Chiari Patients

	Prevalence	Adult/Ped	Source
Basilar Invagination	14%	Adult	Klekamp
Retroflexed Odontoid	20%	Ped	Brockmeyer
EDS	9%	Adult	Eppelheimer
Cervical Instability	7%	Adult	Eppelheimer
Chiari 1.5	18%	Ped	Brockmeyer
Scoliosis	19%	Ped	Brockmeyer
Scoliosis	19%	Adult	Eppelheimer

SOURCES: *The complex Chiari: issues and management strategies. Brockmeyer DL. Neurol Sci. 2011 Dec;32 Suppl 3:S345-7.*
A Retrospective 2D Morphometric Analysis of Adult Female Chiari Type I Patients with Commonly Reported and Related Conditions. Eppelheimer MS, Houston JR, Bapuraj JR, Labuda R, Loth DM, Braun AM, Allen NJ, Heidari Pahlavian S, Biswas D, Urbizu A, Martin BA, Maher CO, Allen PA, Loth F. Front Neuroanat. 2018 Jan 19;12:2.
Chiari I malformation with and without basilar invagination: a comparative study. Klekamp J. Neurosurg Focus. 2015 Apr;38(4):E12.
A critical analysis of the Chiari 1.5 malformation. Tubbs RS, Iskandar BJ, Bartolucci AA, Oakes WJ. J Neurosurg. 2004 Nov;101(2 Suppl):179-83.

Cervical medullary syndrome secondary to craniocervical instability and ventral brainstem compression in hereditary hypermobility connective tissue disorders: 5-year follow-up after craniocervical reduction, fusion, and stabilization. Henderson FC Sr, Francomano CA, Koby M, Tuchman K, Adcock J, Patel S. Neurosurg Rev. 2019 Jan 9

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