CCRC Proposes New Theory for Adult, Symptomatic Chiari – Part 2

In Part 1, we discussed the hypothesis that the cervical dura of symptomatic Chiari patients is stiffer and less compliant than it should be. This, in combination with the blockage of the herniated cerebellar tonsils, results in abnormal spinal fluid pressure spikes during the cardiac cycle. Now the question is, why do Chiari patients have stiffer duras in this region? One possibility is that the cause is genetic and that Chiari patients are born that way and over time this leads to problems. However, the authors of this paper believe the situation is a little more complicated than that. They believe the answer lies in a little known connection between the suboccipital muscles of the neck and the dura in this exact region known as the myodural bridge complex (MDBC). The MDBC was first identified in 1995 and is a collection of fibrous links between the suboccipital muscles and the dura. Microscopic studies have shown that the MDBC doesn't simply connect to the outside of the dura, but rather it penetrates and merges with it. This has led some researchers to speculate that the function of the MDBC is to control the tension of the dura (how stiff it is) during head/neck movements and promote the flow of spinal fluid across this junction. While this has not been definitively established, animal studies do support the idea that the MDBC influences the flow of spinal fluid.

So, if the MDBC is capable of passing tension from the suboccipital muscles to the cervical dura then that means the compliance of the cervical dura in a healthy person is dynamic in nature and changes in response to activation of specific neck muscles. Based on this, the researchers speculate that in Chiari patients, the MDBC experiences mechanical failure and stops working properly. This then changes the properties of the dura the MDBC merges with causing the dura to become stiffer and less compliant. In fact, preliminary studies of MDBC pieces removed during Chiari surgery suggest just this.

Of course, this leads to the question, why does the MDBC fail in Chiari patients? In this theory, the answer is instability. Cervical instability in Chiari patients (meaning between the top two spinal vertebrae) has been widely discussed in the Chiari literature. However, only a modest percentage of Chiari patients exhibit cervical instability on imaging and it is usually associated with the subset of patients who also suffer from Ehlers-Danlos syndrome. Receiving less attention is the atlanto-occipital joint, or where the skull rests on the top vertebrae (the atlas). The skull has protrusions, known as the occipital condyles, which sit is depressions on the surface of the atlas allowing for normal movement of the head. Recently, researchers from China found that in Chiari patients these joints are abnormal. Specifically, they found that the occipital condyles are smaller and the depressions on the vertebrae are shallower, indicating the potential for subtle instability. In fact, this finding is similar to what is seen in Down syndrome where atlanto-occipital instability is well recognized. In addition, other research has shown that the stabilizing ligaments of this joint are abnormal in Chiari patients again indicating the potential for instability.

Bringing this back to the MDBC, the authors hypothesize that subclinical (meaning not currently recognized during standard clinical assessment) instability of the atlanto-occipital joint causes repeated activation and overwork of the suboccipital muscles as they try to compensate. Over time, this repeated over activation leads to mechanical failure of the MDBC which then causes the dura to become stiff and reduces the cervical compliance. The reduced compliance, in combination with the herniated tonsils, leads to abnormal pressure spikes which are made worse by straining and physical activities.

In Part 3 we will look at how the abnormal pressure spikes cause symptoms, what this theory may or may not explain in terms of Chiari, and its implications for diagnosis and treatment.

Source: A new hypothesis for the pathophysiology of symptomatic adult Chiari malformation Type I. Labuda R, Nwotchouang BST, Ibrahimy A, Allen PA, Oshinski JN, Klinge P, Loth F. Med Hypotheses. 2022 Jan;158:110740.

Note: *The full text of the journal paper is available for free at:*
https://www.sciencedirect.com/science/article/pii/S0306987721002590

Note: *Rick Labuda is the author of this research update and is an author of the journal paper it is based upon.*

Conquer Chiari's 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 and many more research articles can be found at www.conquerchiari.org.

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