CCRC Proposes New Theory for Adult, Symptomatic Chiari – Part 3

In the first two parts of this research update, we reviewed a new theory proposed by CCRC researchers that explains the underlying causes of adult, symptomatic Chiari. In summary, the theory states that adult Chiari patients have bone and ligament abnormalities that affect how the skull sits on the spine and causes subtle instability of this joint. This instability, in turn, leads to overwork of the suboccipital muscles as they continuously try to compensate for the instability. Over time, this leads to mechanical failure of the myodural bridge complex (MDBC), which is a set of connections between the suboccipital muscles and the cervical dura (the covering of the spine). The failure of the MDBC causes the cervical dura to become stiff and less compliant. This, combined with the tonsillar blockage at the same level, creates abnormal spinal fluid pressure spikes during the normal cardiac cycle. The pressure spikes are made worse by activities that naturally raise the heart rate and/or lead to rapid pressure changes, such as coughing, sneezing, and physical activity.

The final question then is how does this abnormal pressure environment lead to symptoms? Here again, there is evidence suggesting certain processes. Specifically, the CCRC has discovered that Chiari patients have increased movement of the cerebellum, brainstem, and cervical spine during the cardiac cycle, which puts those regions under increased strain. The CCRC has also shown, using advanced imaging, that Chiari patients have microstructural damage to those same regions. Based on this evidence, the researchers hypothesize that the abnormal pressure environment leads to strain on the cerebellum, brainstem, and cervical spine which causes microdamage to these areas, and in turn many of the symptoms common to Chiari.

If this theory is shown to be valid it explains several important aspects of Chiari, including:

- Why there are more asymptomatic cases than symptomatic
- Why the size of the herniation is not strongly linked to symptom severity
- How trauma can lead to a sudden onset of symptoms (via a rapid mechanical failure of the MDBC)
- Why symptoms may not become noticeable until adulthood in some people

However it is not all encompassing. For example, how it applies to pediatric cases is not known. Even among adults, it is likely that Chiari comprises several sub-groups of patients. This theory is most likely applicable to what is sometimes referred to as ‘classic Chiari’; how it applies to patients who also have Ehlers-Danlos or pseudotumor cerebri is not clear.

It is very important to keep in mind this is just a theory and parts, or even all of it, could be wrong. While there is evidence to support every link in the sequence of events that have been laid out, much more research is needed to both test and refine it. For example, the evidence supporting this theory comes from many different projects involving different subjects undertaken at different locations and points in time. Tying all the pieces of the theory together will require having the same Chiari subjects undergo a wide array of imaging and testing in order to find statistical connections between things like the microscopic structure of the MDBC and measured compliance, or linking strain in the brainstem or cerebellum to actual tissue or fiber damage.

Having said this, the implications of this theory on diagnosis and treatment are profound. If shown to be true, imaging protocols may expand to evaluate compliance and instability of the atlanto-occipital joint. Patients may be evaluated to assess the individual contribution of tonsillar blockage and cervical compliance to their symptoms. Treatments may evolve that focus on restoring compliance in addition to – or instead of – creating more space around the herniated tonsils.

Developing a comprehensive theory of Chiari, even if parts of it are wrong, is an important step forward in our journey. Conquer Chiari has already launched a research project to test certain aspects of this theory and in the future will commit whatever resources are necessary to develop it fully.

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.