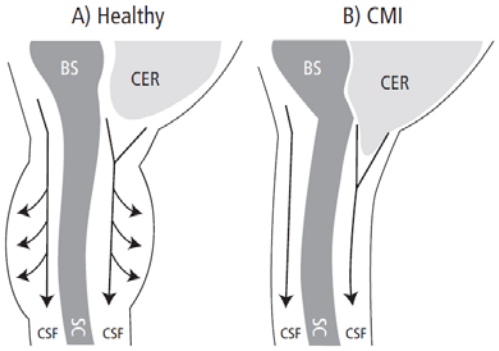
CCRC Proposes New Theory for Adult, Symptomatic Chiari – Part 1

CCRC researchers and some of their partners have published a novel theory on the underlying processes involved in symptomatic Chiari in adults. The theory is based on existing research literature combined with preliminary experiments undertaken by the researchers themselves. The theory is comprised of several key elements, is fairly broad in scope, and addresses some of the major issues that have puzzled Chiari researchers for quite a while. Long-time readers will recall that the prevailing theory for Chiari for many years has been that an undersized skull results in a normal sized brain herniating into the spinal cord area. In this scenario, symptoms arise from the physical pressure this puts on the brain and spinal tissue and from blocking the natural flow spinal fluid across the craniovertebral junction. The problem with this theory is that there a lot of people walking around with herniation of the cerebellar tonsils that meets the definition of Chiari but who have absolutely no symptoms. In fact, the ratio is could be as high as 10:1 of people with no symptoms to people with symptoms. Research has shown that very few of these cases, if any, are ever likely to develop problems associated with their herniations. In addition, the CCRC has shown, using an NIH database of more than 10,000 subjects, that while adolescents with such herniations look like Chiari cases in terms of MRI measurements, their performance on a wide variety of behavioral and health assessments is completely normal. All of this raises the question of what is different between these asymptomatic cases and those suffering from Chiari symptoms?

The researchers think the difference has to do with something called *compliance*. Compliance is a technical term for how easily a material expands in response to pressure. In this case the material is the dura covering the spinal cord in the cervical region and the pressure comes from the spinal fluid underneath. Specifically, when the heart beats blood is pumped into the brain which forces a certain amount of spinal fluid out of the brain and into the spinal area (spinal fluid is contained in the subarachnoid space between the dura and the tissue of the brain and spine). In a healthy person, the fluid flows freely across this junction and back. As we know, in a person with Chiari, the herniated tonsils block some of this flow. However, the researchers also hypothesize that in a healthy person the cervical dura is somewhat flexible (compliant) and expands outwards in response to the inrush of spinal fluid. However, in Chiari patients they believe the cervical dura is stiffer (therefore less compliant) and does not expand as much during the cardiac cycle (see diagram below).



The result of the stiffer dura is an increase in the pressure of the spinal fluid, similar to how hardening of arteries is linked to high blood pressure. Therefore, in symptomatic Chiari, the **combination** of the tonsils blocking spinal fluid and the reduced cervical compliance at the same level due to a stiff dura creates a situation where there are abnormal pressure spikes of the spinal fluid during the cardiac cycle. These pressure spikes are made worse by activities that naturally increase pressure such as coughing, sneezing, and physical exertion.

The researchers have collected preliminary data using advanced MRI that indicates that Chiari patients do in fact exhibit lower compliance in this region. In addition, research from Europe that involved continuous pressure monitoring of Chiari patients showed that while the average spinal fluid pressure of Chiari patients was normal, there were high spikes of pressure during the cardiac cycle.

To summarize, the theory states that it's the combination of herniated tonsils and reduced compliance at the same level that leads to an abnormal pressure environment in Chiari patients. If this is shown to be true (more research is required), it explains why herniation alone in most cases does not lead to problems; in these cases there is enough compliance in the dura to compensate for the blockage. It also explains why some people have small herniations and severe symptoms (compliance is very low) and other have large herniations with minimal symptoms (compliance is high).

The next logical question is why do Chiari patients have stiffer, less compliant duras in this region? We will address this in Part II.

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.