Chiari Academy Video Transcription
Beyond Tonsillar Position – A New Chiari Theory

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In the previous modules we discussed a number of static and dynamic measures that on average

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are significantly different in Chiari patients compared to healthy people. However, none of these

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on their own fully explain how symptomatic Chiari arises, why many people with herniations don't

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experience symptoms, or why most Chiari patients aren't diagnosed until their early thirties.

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In 2022 Conquer Chiari researchers published a new theory to address these questions that focused on

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something called compliance. Compliance is a technical term for how easily a material

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expands in response to pressure. In this case the material is the dura covering the spinal cord in

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the cervical region and the pressure comes from the spinal fluid underneath. Specifically, when

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the heart beats blood is pumped into the brain which forces a certain amount of spinal fluid

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out of the brain and into the spinal area. In a healthy person, the fluid flows freely across this

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junction and back, but in a person with Chiari, the herniated tonsils block some of this flow.

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However, the researchers also hypothesize that in a healthy person the cervical dura is somewhat

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flexible, or compliant, and expands outwards in response to the inrush of spinal fluid.

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In contrast, in Chiari patients they believe the cervical dura is stiffer, or less compliant, and

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does not expand as much during the cardiac cycle. The result of the stiffer dura is an increase in

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the pressure of the spinal fluid, similar to how hardening of arteries is linked to high blood

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pressure. Therefore, in symptomatic Chiari, the combination of the tonsils blocking spinal fluid

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and the reduced cervical compliance at the same level due to a stiff dura creates a situation

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where there are abnormal pressure spikes of the spinal fluid during the cardiac cycle.

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These pressure spikes are made worse by activities that naturally increase pressure

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such as coughing, sneezing, and physical exertion. Unfortunately, researchers have not yet figured

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out how to directly measure compliance using MRIs, so this idea can't be directly tested. However,

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research from Europe that involved continuous pressure monitoring of Chiari patients showed

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that while the average spinal fluid pressure of Chiari patients was normal, there were high

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spikes of pressure during the cardiac cycle. If shown to be true, compliance explains why

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herniation alone in most cases does not lead to problems. In these cases there is enough

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compliance in the dura to compensate for the blockage. It also explains why some people
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have small herniations and severe symptoms – because there compliance is low - and
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others have large herniations with minimal symptoms – because their compliance is high.
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The next logical question is why do symptomatic Chiari patients have stiffer, less compliant
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duras in this region? One possibility is that the cause is genetic and that Chiari patients
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are born that way and over time this leads to problems. Supporting this idea is a genetic
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study which found that nearly half of Chiari have an alteration to a collagen, or connective tissue,
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related gene. However, another possibility involves a little known connection between the
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suboccipital muscles of the neck and the cervical dura known as the myodural bridge complex or MDBC.
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The MDBC was first identified in 1995 and is a collection of fibrous links between the
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suboccipital muscles and the dura. Microscopic studies have shown that the MDBC doesn't simply
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connect to the outside of the dura, but rather it penetrates and merges with it. This has led
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some researchers to speculate that the function of the MDBC is to control the tension of the dura
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during head and neck movements and promote the flow of spinal fluid across this junction. While
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this has not been definitively established, animal studies do support the idea that the

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MDBC influences the flow of spinal fluid. So, if the MDBC is capable of passing tension
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from the suboccipital muscles to the cervical dura then that means the compliance of the
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cervical dura in a healthy person is dynamic in nature and changes in response to activation
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of specific neck muscles. Based on this, the researchers speculate that in Chiari patients,
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the MDBC experiences mechanical failure and stops working properly. This then changes the
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properties of the cervical dura with causing it to become stiffer and less compliant. Preliminary
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studies of MDBC pieces removed during Chiari surgery show that the material composition is
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different than the MDBCs of healthy people. Of course this leads to another question,
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why would the MDBC fail in Chiari patients? At this point the theory becomes more speculative,
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but one possibility has to do with instability, specifically at the atlanto-occipital joint
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which is where the skull rests on the spine. Different research studies have shown that
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Chiari patients have smaller stabilizing muscles and shorter stabilizing ligaments,
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both suggestive of instability in this region. It is possible that subclinical instability of
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the atlanto-occipital joint causes repeated activation and overwork of the suboccipital
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muscles as they try to compensate. Over time, this repeated overactivation leads to mechanical

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failure of the MDBC which then causes the dura to become stiff and reduces cervical compliance.
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The reduced compliance, in combination with the herniated tonsils then leads
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to abnormal pressure spikes which are made worse by straining and physical activities.
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The final question then is how does this abnormal pressure environment lead to symptoms? Here is
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where we circle back to our earlier discussion, specifically the strain placed on the cerebellum
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and brainstem in Chiari patients. In this theory the abnormal pressure environment caused by the
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combination of tonsillar blockage and reduced compliance leads to abnormal pulsations which
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in turn cause microstructural damage to the cerebellum, brainstem, and the upper cervical
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spine. Damage in these regions of Chiari patients has been found using a special type of MRI.
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It is very important to keep in mind this is just a theory and parts, or even all of it,
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are very likely to be wrong. While it is based on existing evidence, many research studies will
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need to be undertaken to verify each part and the theory as a whole. It is also important to realize
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that the theory it is not all encompassing. For example, how it applies to pediatric cases is
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not known. Even among adults, it is likely that Chiari comprises several sub-groups of patients.
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This theory is most likely applicable to what is sometimes referred to as ‘classic Chiari’;

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how it applies to patients who also have Ehlers-Danlos or pseudotumor cerebri is not clear.

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A comprehensive theory of Chiari, even if parts of it are wrong, is an important step forward in

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our journey. Conquer Chiari has already launched a research project to test certain aspects of this

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theory and in the future will commit whatever resources are necessary to develop it fully.