



Chiari Academy Video Transcription

Chiari Bootcamp- Diagnosis

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In this lesson we will cover the diagnostic challenge, the diagnostic process missed miss

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and incidental diagnosis. In pediatric issues, not surprisingly the challenges that exist

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in defining what Chiari is carries over to the topic of diagnosis. Recall that Chiari is defined

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radiographically meaning based on Imaging with the criteria being the tip of the cerebellar tonsils

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being located five millimeters or more below the opening in the bottom of the skull. However,

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over the years it has become apparent that many people meet this definition but do not

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and likely will not ever have any Chiari related symptoms. This by itself is enough to say that

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imaging alone is not enough to diagnose someone with symptomatic Chiari. But unfortunately there

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is more. A second problem with defining Chiari based on a measurement is that it assumes that

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measurement can be made with a certain level of accuracy and precision. While MRIs are amazing

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machine means not every MRI is equal in terms of the quality of images it can produce. This

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combined with issues of patients moving during scans can make it difficult to accurately make

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measurements at the millimeter level. Beyond this actually measuring tonsillar position is part

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science and part art this means that different people even experts can come up with different

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tonsil or position values from the same images. Studies have shown that experts can disagree on

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this critical value by five millimeters or more which is the very definition of Chiari. The end

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result of this is that Imaging alone while very important to diagnosing Chiari is not enough by

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itself. In fact while it may be surprising to new patients there is no single objective test

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to diagnose Chiari. Rather than a single test the Chiari diagnostic process involves several factors

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which a doctor often a neurosurgeon will take into account in making an informed intelligent

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decision. These factors include a patient's symptoms and whether they are consistent with and

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likely to become caused by Chiari the result of a neurological exam the result of Imaging and other

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tests and the physician's personal experience and judgment when it comes to Chiari. As we discussed

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in the lesson on symptoms the Hallmark symptom for Chiari is an intense crushing headache in the back

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of the head brought on by straining or coughing. When a patient says they are experiencing this

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type of headache it is a significant clue to a physician that Chiari could be the cause. However

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if a patient has headaches that are more like migraines then it becomes more complicated. While

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migraines are common among Chiari patients they are also common in general and poorly understood.

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In fact many of the most common Chiari symptoms such as neck pain balance issues and brain fog

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can be due to many different things and so by themselves are not specific to a Chiari diagnosis

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but rather one piece of the puzzle. While there is no single definitive test for symptomatic Chiari

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MRI or magnetic resonance imaging is the gold standard. An MRI can create a detailed picture

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of the brain skull spine and spinal cord which the Physician can use to see if the cerebellar

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tonsils are herniated. Whether there is space for cerebrospinal fluid to flow whether there are bony

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abnormalities and whether there is a syrinx or collection of fluid in the spinal cord itself.

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We will discuss syringes more in the lesson on related conditions but for now it is important

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to know that the presence of a syrinx is close to an automatic diagnostic indicator. Depending on

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what type of doctor is doing the investigating and whether the case is complicated or straightforward

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sometimes Chiari patients will undergo further tests these can include additional MRIs of the

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entire spine CT scans a special type of MRI to look at the flow of cerebrospinal fluid.

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balance testing urodynamic testing, cognitive testing, or sleep studies a very important part

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of the diagnostic process is the neurological exam. The neurological exam has been used for

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hundreds of years and is the most basic tool in a neurologist or neurosurgeon's diagnostic toolkit.

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While low-tech this type of exam can reveal a tremendous amount of useful information a

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neurological exam provides a physician with a way to explore the function of the nervous system by

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seeing how different parts of the body respond to different stimulus and tasks. This is because the

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nerves of the human body have been carefully mapped out so we know where nerves that carry

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sensation from and muscle commands to different parts of the body enter the spinal cord. We also

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know what parts of the brain control different things like balance and autonomic reflexes.

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A complete neurological exam is an extensive procedure that involves testing the strength

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of different muscles, the response to different kinds of touch, tests of coordination and gait,

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testing balance autonomic reflexes and more. The neurological exam shows if different parts of the

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nervous system such as the brain stem and cranial nerves are functioning properly and can provide

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strong objective clues as to whether a herniation found on MRI is causing problems. However it's

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important to keep in mind that many doctors will only check enough things until they are satisfied

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they know what is going on sometimes this can be disconcerting to a patient who may feel the

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doctor is not being thorough but from the doctor's point of view they have enough information and

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they don't need to perform every test. The final piece of the puzzle is the individual

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Physician's own experience and judgment. This can be difficult for patients to accept but the

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reality is that doctors must pull together all the information and make an informed decision

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based on their experience as to what is causing a person's symptoms. In some cases this decision

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vision is straightforward while in other cases it is not so clear. The practical implication for a

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patient is that if they have symptoms that could be from Chiari but could also be due to something

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else and if the MRI is not conclusive they are likely to get different opinions from different

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doctors. With this in mind Conquer Chiari advises patients who ask about getting second opinions

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to be prepared to hear different opinions. Some people want to hear all the opinions and options

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but for others it can be frustrating when they don't get a definitive answer. Because of the

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ambiguities that come with Chiari patients end up taking many different paths to a final diagnosis.

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For some people diagnosis can come quickly after a sudden onset of symptoms but for others it can

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take years and many different doctors to finally get a diagnosis. For example a survey of over

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1,300 adult female Chiari patients showed that 1/3 identified a specific event often some type

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of trauma that triggered their symptoms and started their search for a diagnosis but of

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course this means that for the rest there wasn't a specific event. Similarly people may start their

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diagnostic journey with different types of doctors some people may start with a sleep doctor due to

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apnea. While others may see a neurologist from migraines and still others may see an

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ear nose throat doctor due to balance problems but in the end since the primary treatment for

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symptomatic Chiari is surgical. Conquer Chiari recommends that anyone with an MRI indication

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of cerebellar herniation and symptoms be evaluated by a neurosurgeon. While some people are diagnosed

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quickly far too often a Chiari patient's symptoms are dismissed and the diagnosis is missed. This

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young woman's symptoms became so bad that she was essentially in crisis doctor after doctor brushed

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off what was happening to her and blamed it on stress or other factors only when her symptoms

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were crippling did she get an accurate diagnosis. Unfortunately this is a major problem for the

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Chiari Community especially for adult patients the lack of an objective test probably combined

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with the fact that most adult patients are women result in many Chiari patients being told their

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symptoms are psychological in nature. In fact a study of over 300 Chiari patients found that over

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half had been told by a doctor they suffered from a mental problem in addition to missed

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diagnosis. Chiari is also sometimes misdiagnosed as something else because of overlapping symptoms

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Chiari patients have been wrongly diagnosed with multiple sclerosis, chronic fatigue syndrome,

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cluster headaches, ALS and even carpal tunnel syndrome. At the other end of the spectrum are

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incidental diagnoses this is when imaging is ordered for some other reasons such as after

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a head injury and a herniation is found but the person does not have a history of Chiari type

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symptoms and was not looking for a diagnosis of any kind this type of incidental finding is more

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common among kids who may be routinely imaged after a sports related head injury. Recall that

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up to 3% of children actually have cerebellar herniation of five millimeters or more so if

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someone gets an MRI due to concussion related issues then if they are one of those three percent

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it will likely be noted by the radiologist this can be worrisome for patients who aren't sure what

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to make of it. We will discuss this more in depth later in the course but fortunately among children

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very few incidentally found cases end up becoming symptomatic in the near to medium future that

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is not the only difference between pediatric and adult. Chiari in terms of diagnosis although there

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is no definitive evidence for this in general it seems like Chiari is more readily diagnosed

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among children in fact a survey of pediatric neurosurgeons found that most thought that

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pediatricians in general had the proper training and education to diagnose and refer Chiari

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you have to wonder what a similar survey among adult neurosurgeons would say about Primary Care

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Providers. However not all the differences are positive for young children especially pre-verbal

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diagnosis can be difficult if a child cannot express what they are feeling their symptoms may

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result in changes in behavior attitude and affect which may be dismissed as developmental phases

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or diagnosed as something else also an MRI on a younger child is more complicated and likely

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to require sedation. This may make a physician more reluctant to order the test. To summarize

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there is no single objective test to diagnose Chiari diagnosis is a process that involves

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symptoms Imaging and neurological exam and a physician's experience in judgment. Up to 1/3

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of patients can identify a specific event often some type of trauma that sparked their symptoms

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Chiari diagnoses are often missed especially among adults or misdiagnosed as something else.

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Sometimes a herniation is found incidentally meaning when there are no symptoms. Diagnosing

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Chiari in young children can be difficult if they can't verbalize what they are feeling.