Chiari Academy Video Transcription
Beyond Tonsillar Position – Tonsillar Position as a Chiari Diagnostic

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In this module we will examine how good tonsillar position is as a test

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for symptomatic Chiari and the impact it can have on patients.

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Chiari is classically defined as tonsillar herniation of 5mm or more,

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yet only a fraction of people with herniations of this size will ever experience symptoms. So

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what does this mean scientifically? If we think of the 5mm rule as a test for Chiari,

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like a COVID-19 or diabetes test, we can evaluate that test quantitatively. To assess

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how good a test is at identifying a disease, scientists create a simple table showing the

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number of people who test positive and actually have the disease; the number who test positive

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but don't have the disease; the number who test negative but actually have the disease,

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and finally the number who test negative and don't have the disease. The table then also

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includes row and column totals, plus a grand total in the lower right corner. For our case,

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the table will show how many people have tonsillar position greater and less than 5mm and how many

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people in each of those categories have symptomatic Chiari and how many don't.

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How do we get the numbers to fill out the table for Chiari? Let's start by taking 10,000 random

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people of any age. Therefore, our total in the lower right cell is 10,000. It is estimated that

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symptomatic Chiari affects about 1 in 1,000 people, so if we start with 10,000 people

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that means in total, we will have 10 people with Chiari. Now, Conquer Chiari research has

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found that among Chiari patients about 20-25% have a tonsillar position of less than 5mm.

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Since we can't have half of a person, let's say that out of the 10 symptomatic people,

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8 have tonsillar position of 5mm or more and 2 have less than 5mm.

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You may say, wait a minute! This is only because doctors dismiss patients with less

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than 5mm of herniation as not having Chiari, so the actual percentage is higher. Well,

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you are very likely correct. However, these are the best numbers we have at this time,

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so for now we will use them and factor that possibility in later.

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Moving on, we also know from research that depending on age and gender 1-3% of the

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general population has a tonsillar position of 5mm or more, so let's pick a number in the

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middle and assume that 1.5% percent of the total group has a tonsillar position of 5mm or more.

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That means that 150 people out of our total group have tonsillar position of at least 5mm,

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and 9,850 people out of our total group have tonsillar position less than 5mm.

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Now, we can fill in the rest of our table using simple arithmetic and we find that

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142 people have tonsillar position greater than 5mm but don't have symptomatic Chiari

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and 9,848 people have tonsillar position less than 5mm and DON'T have symptomatic Chiari.

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From this table, scientists then look at 4 different measures, two of which are focused

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on how well the test performs among a population, and two which are focused on what the test means

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for an individual. Starting at the population level, Sensitivity refers to how well a test

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identifies people who have a disease. In our analysis, 10 people actually have Chiari,

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and the 5mm test correctly identifies 8 of them, so the test's sensitivity is 80%. However,

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here's where your earlier objection comes in. Our data is only as good as the current standard in

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identifying Chiari, so if a certain percentage of doctors do not consider anyone with tonsillar

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position less than 5mm as having Chiari, then the data could be inaccurate. Therefore, it is

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probably more accurate to say that the sensitivity of the 5mm test is at MOST 80%, but likely lower.

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The next measure is Specificity, which refers to how well a test

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identifies people who don't have the disease. In our analysis,

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out of 9,990 people who don't have symptomatic Chiari, the 5mm test correctly

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identifies 9,848. This means the Specificity of the 5mm test is 98.6% which is pretty good.

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Sensitivity and specificity relate to how well a test performs among a population,

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but there are two more measures that are specific to the individual:

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positive and negative predictive value. Positive predictive value is the probability

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that a person with a positive test result actually has the disease. In our analysis,

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a total of 150 tested positive with the 5mm test, but out of that group only eight people actually

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have symptomatic Chiari. That means the Positive Predictive Value of the 5mm test is only 5.3%!

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Finally, negative predictive value is the probability that someone with a negative

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test result is actually disease free. For Chiari, out of 9.850 people who test negative,

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9,848 of them will not have symptomatic Chiari, meaning the Negative Predictive Value is 99%.

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To summarize, at the population level, the 5mm test does a good job of screening out people who

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don't have symptomatic Chiari but misses 20% or more of the people who do have symptomatic Chiari.

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At the individual level, a negative test result means it is very likely you don't have Chiari,
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but a positive test result doesn't mean much because only 5% of those who test
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positive with the 5mm rule are symptomatic. Although these specific numbers aren't discussed
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in the Chiari literature, the limitations of the 5mm test are well known. That's why
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among clinicians a Chiari diagnosis is made through a combination of imaging – meaning
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tonsillar position – plus patient-reported symptoms, and a neurological exam. However,
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the lack of an objective, valid test for Chiari is problematic in two ways.
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First, some physicians still rely on the 5mm rule, which of course means that a significant group of
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patients who actually have symptomatic Chiari are being blocked in the diagnostic process.
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The other problem is that without an objective test, the diagnosis becomes inherently subjective,
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which in turn means that different doctors may disagree on what is and what isn't Chiari.
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This is especially problematic when it comes to trying to define Chiari based on symptoms. Many
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clinicians consider the cough associated or Valsalva headache to be the signature symptom
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of Chiari. However, a 2023 meta-analysis that included nearly 2,000 pediatric and adult Chiari
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patients found that while 78% of patients reported headaches, only 48% of those with headaches

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experienced typical Valsalva type headaches, while 29% suffered from atypical headaches and
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23% from both. So, if a Chiari test were comprised of tonsillar position plus the presence of typical
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headaches, it still would not be very accurate. Interestingly, the same study also found that
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typical headaches improved more with decompression surgery than atypical headaches. This raises the
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possibility that Chiari is being defined by doctors' current ability to treat it. In other
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words, if you have tonsillar herniation and Valsalva headaches, or a syrinx, then
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surgery may help, so we'll say you have Chiari. Obviously, from the patient's point of view this
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is not an ideal way to define or test for Chiari. It would be better to understand
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what Chiari is at a fundamental level and then develop tests from that knowledge.
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Over the rest of this course, we will explore various efforts that have been made to move beyond
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tonsillar position and the theories that have evolved to explain different aspects of Chiari.
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In this module we learned: • The 5mm tonsillar position
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rule for Chiari can be evaluated quantitatively • In the population, the 5mm rule does good job of
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identifying who doesn't have symptomatic Chiari, but it is not as good at identifying
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people with symptomatic Chiari • At the individual level,

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only about 5% of people who have tonsillar position of

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5mm or more actually have symptomatic Chiari • Clinicians are aware of the limitations of the

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5mm rule, but a better understanding of Chiari is needed to develop a better diagnostic test