Chiari Academy Video Transcription
Chiari Bootcamp- What is Chiari?

Introduction
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Hello in this lesson we will cover several important aspects of Chairi including:

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the different definitions of Chiari, the anatomy of Chiari, a brief history of Chiari,

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and how the concept of Chiari is evolving. Since you are taking this course it is likely

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that either you or someone you care about has been affected by Chiari. It is also likely that before

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this happened you had never heard of Chiari, had any idea what it is or even how to pronounce it.

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That's okay you're not alone in this regard and that's why we're here specifically to learn about

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Chiari. The first thing to understand is that Chiari can mean different things to different

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people. To a patient Chiari is a difficult to understand disease with many symptoms

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and manifestations. To a neurosurgeon Chiari is a structural defect to be corrected with

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surgery. To an uninformed physician Chiari might be considered a harmless variant from normal and

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nothing to worry about. To a family member Chiari is often an invisible affliction hurting their

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loved one. Now let's turn our attention to how Chiari is defined. Unfortunately Chiari is not

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always easy to define even in medical terms this is because the definition of Chiari is evolving

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both as technology improves and as we learn more about it. However for the purposes of this lesson

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understanding what Chiari is will first require deciphering some medical jargon and learning

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a little bit about human anatomy. Let's start by looking at what Chiari is in general terms. Chiari

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is a serious often chronic neurological disorder that involves abnormalities of the craniovertebral

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junction and compression of the brain and spinal cord. So what does this mean? Let's break it down:

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First, chronic means that for some patients the effects of Chiari are long lasting and Chiari

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is something they will deal with for their entire life. Second, the term neurological refers to the

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nervous system which is comprised of the brain spinal cord and the many nerve fibers which run

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through our body. Next, Chiari is often referred to as a disorder because it is not a disease in

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the sense of a virus or bacterial infection. The craniovertebral junction is where the skull, or

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cranium, meets the spine. Internally the junction is also where the brain stem becomes the spinal

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cord and in Chiari where the herniated cerebellar tonsils are located. And finally, abnormalities in

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this case refers to anatomical structures which aren't normal. So in broad terms Chiari patients

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have unusual anatomy in the region where the skull meets the spine which then causes compression or

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squeezing of the cerebellum brain stem and spinal tissue inside. It is said a picture is worth a

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thousand words so let's look at a simple picture of what Chiari Anatomy is. Here we see that in an

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average or healthy person. The bottom part of the brain called the cerebellum is completely

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contained in the skull. However, in a person with Chiari the very bottom of the cerebellum,

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known as the tonsils, are protruding out of the skull through the opening where the brain meets

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the spinal cord. Like a cork being jammed into a wine bottle this protrusion then squeezes or

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compresses these parts together. Unfortunately there's more. In the human body the brain and

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spinal cord are bathed in a clear liquid called cerebrospinal fluid or CSF this fluid plays a

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vital role in that it supports the weight of the brain acts as a shock absorber delivers

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vital nutrients to the central nervous system and removes waste products. With every heartbeat blood

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is pumped into the brain which then pushes CSF out of the brain and into the spinal area and back

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again. In a Chiari patient this natural sloshing of CSF from brain to spine and back is blocked and

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disrupted by the protruding cerebellar tonsils the effects of this are not clearly understood but are

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a major focus of Chiari research. So at its core Chiari is about unusual anatomy. Chiari patients

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tend to have skulls spines and brains which are not like the average persons. Now that we've gone

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over what Chiari is in general and learned a little bit about the anatomy of Chiari let's

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look at the history of Chiari. Chiari like many medical diseases and disorders was named after

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the person who first described it which in this case was Hans Chiari an Austrian pathologist in

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1891 more than 130 years ago Chiari published a paper describing the case of a 17 year old woman

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in whom he observed after she died from typhoid fever and elongation of the cerebellar tonsils.

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Later he published on more cases and classified his observations of the malformations into several

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types which serve as the basis for the classic definition of Chiari. The most common are: Chiari

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I where the cerebellar tonsils are herniated through the opening in the bottom of the skull.

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Chiari I is the focus of this course. Chiari II where additional brain structures such as

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the brain stem or main body of the cerebellum are also herniated. Today most people use Chiari II

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to refer to cases that coincide with spina bifida. Although these definitions appear simple at first

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the history of Chiari especially for Chiari I is full of confusion and disagreement. A few years

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after Chiari published his paper Julius Arnold published his observations of malformations in

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newborns similar to chiari's type II and in the early 1900s people started using the term Arnold

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Chiari. Some people used this only for type 2 while others referred to all cases as Arnold

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Chiari. Adding to the confusion over the years other names and terms have been introduced in

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the literature in reference to Chiari but for our purposes when we refer to Chiari we mean Chiari

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Malformation Type I. it's important to keep in mind that during these early years and the

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decades that followed there was no medical imaging available to look at people's brains, so while

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Chiari had been identified, until the introduction of MRIs in the 1980s it was very difficult to

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diagnose. However MRIs changed everything for the first time doctors could get a detailed picture

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of a patient's skull and brain which led to the modern or clinical definition of Chiari based on

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tonsillar position. This technological medical and scientific progression has led to a more detailed

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clinical definition of Chiari let's dive into what this means. Tonsillar position refers to

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how far the tip of the cerebellar tonsils are located below the opening at the bottom of the

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skull known as the foramen magnum. To understand how tonsillar position is measured it's important

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to understand that an MRI creates a complete three-dimensional representation of the brain and

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skull which are then viewed in individual slices. To measure tonsil or position first we select what

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is called the mid-sagittal slice of the MRI which basically splits the head in half running from the

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nose to the back of the head. Next in this video an imaginary line is drawn across across the skull

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opening this is known as the McRae line. Finally another line is drawn perpendicular to this one

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down to the tip of the cerebellar tonsil the length of this line in millimeters represents

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the tonsillar position. Now that they were able to quantify Chiari malformations researchers

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started studying them in a different way. In 1986 Barkovich and a team published a study

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which measured the tonsillar position of 25 patients with what they determined to be clear

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signs and symptoms of Chiari and compared them to 200 healthy people they found that the tonsillar

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position of the two groups overlapped between three to five millimeters. In other words the

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healthy people had tonsillar positions from above the McRae line to five millimeters below and the

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Chiari patients had tonsillar positions of at least three millimeters below the McRae line from

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this the radiological definition meaning based on imaging of Chiari emerged as tonsil or herniation

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of at least three to five millimeters. Over time this evolved to five millimeters and became known

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as the five millimeter rule. However as time went on and MRIs became more prevalent it became clear

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there were some problems with the five millimeter rule this ultimately led to the evolution of the

Problems with the 5mm Rule

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definition of Chiari. The first problem that came to light was that tonsillar position is

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not strongly related to symptom severity in 1999 Thomas Milhorat and colleagues published

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a landmark paper which provided a detailed report on over 300 Chiari patients they found

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that tonsillar position was not related to symptom severity they also identified patients with clear

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indications of Chiari who had less than three millimeters of herniation. The second problem

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that emerged is that as MRIs became more and more common it became clear that a certain number of

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people meet the five millimeter MRI definition of Chiari but did not have any symptoms. In fact

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large studies have shown that symptomatic Chiari is the exception not the rule specifically 1 - 2%

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of the adult population and up to 3% of kids have been found to have tonsillar position

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greater than five millimeters. Yet only about .1% of people have symptomatic Chiari therefore there

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are many many more people without symptoms than with. It has become clear that tonsil

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or position varies across the population much like height and weight do. In addition,

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studies have shown that tonsillar position even in healthy people is not fixed but rather varies

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over time. All of this results in confusion about what Chiari actually is while some people use the

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term Chiari to refer to anyone with tonsil or herniation of five millimeters or more others

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reserve the term Chiari to only refer to people who are symptomatic. Because of these issues

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many medical experts have now begun to question the five millimeter rule. However, finding a new

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measure of Chiari has proven elusive so although it is not perfect tonsillar position is still the

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best marker for symptomatic Chiari available, but it is important to keep in mind that it is

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not the whole story. To summarize, Chiari can mean different things to different people. Chiari is a

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serious, often chronic, neurological disorder that involves compression of the cerebellum

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brain stem and spinal cord. In Chiari patients the cerebellar tonsils protrude or herniate out of the

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skull. The current definition of Chiari is based on measuring tonsil or position on MRI. However,

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this is not a great definition and many experts are looking for a better one. [Music]