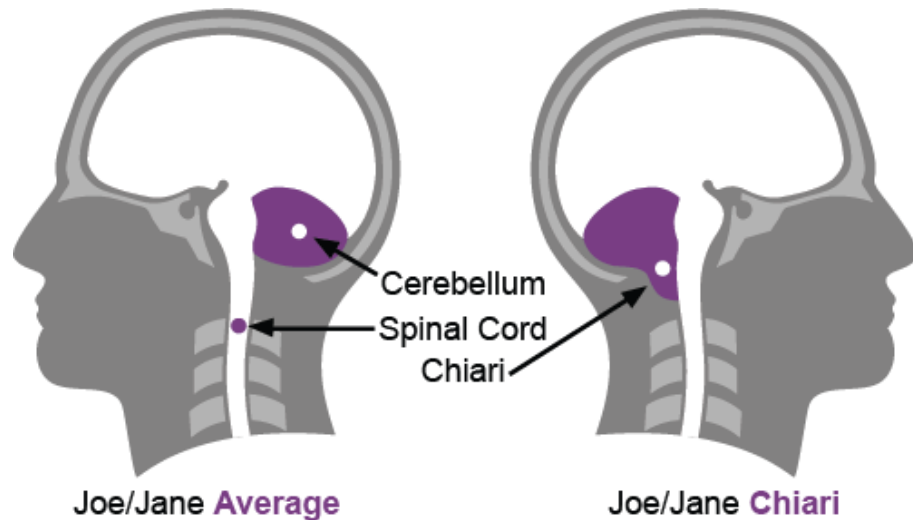


# Conquering Chiari: An Overview

# What is Chiari Malformation?

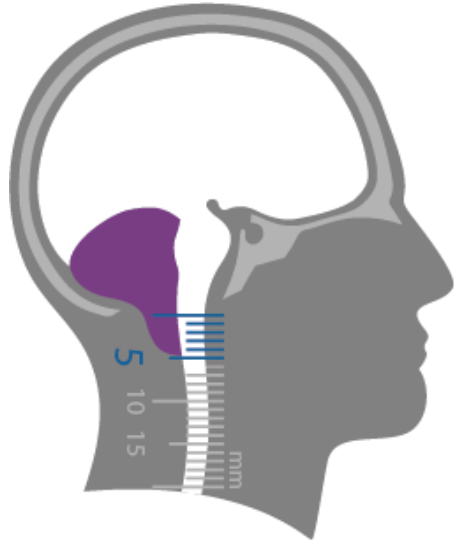
**Chiari Malformation** is a serious neurological disorder where the bottom part of the brain, the cerebellum, descends out of the skull and crowds the spinal cord, putting pressure on both the brain and spine and causing many symptoms.



- ❖ First identified by Hans Chiari in the 1890's
- ❖ In Type 1 the cerebellar tonsils are located outside the skull; Type II involves more of the cerebellum and occurs in association with Spina Bifida
- ❖ Some people have characteristics of each, this is sometimes referred to as Chiari 1.5
- ❖ Also known as: CM, Arnold-Chiari Malformation (ACM), tonsillar herniation, tonsillar ectopia, hindbrain herniation

***Chiari Affects Everyone Differently!!***

# Evolving Definition



**Traditional Definition**



**New Definition**

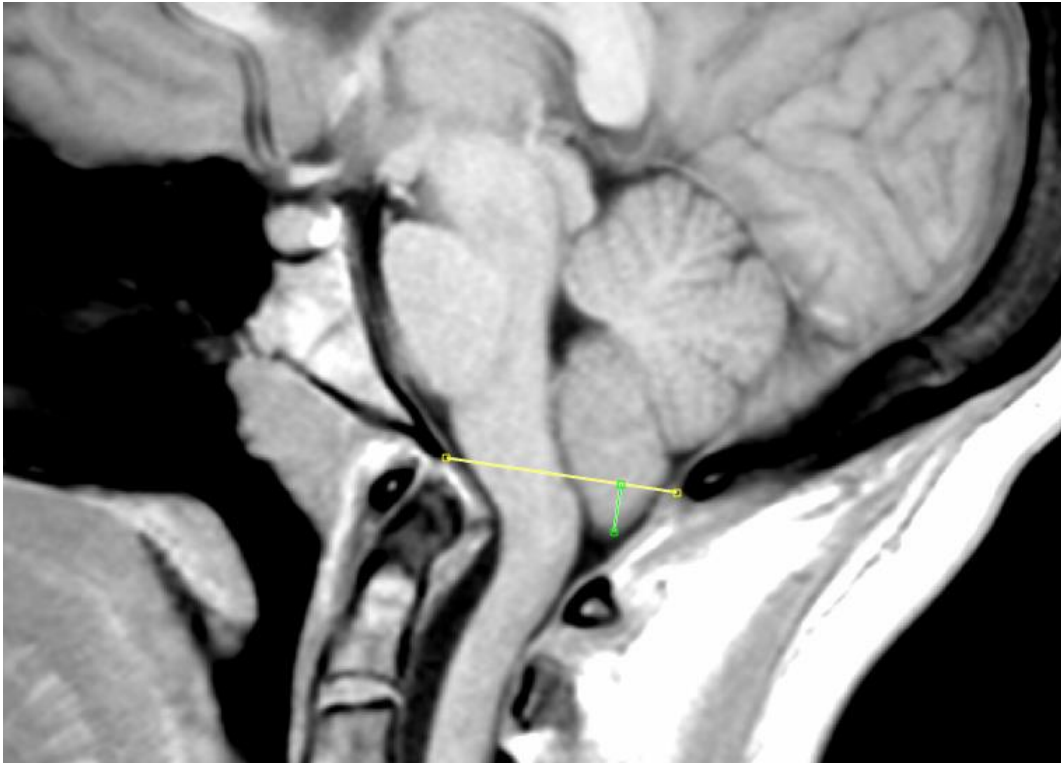
- ❖ Chiari malformation is traditionally defined as the cerebellar tonsils being located 5mm or more below the foramen magnum (opening at the bottom of the skull) as measured on an MRI.
- ❖ However, there are several issues with the traditional definition which has led some to question if Chiari should be defined differently

# Issue #1: Too Many Healthy People



Studies have shown that between 1-3% of the general population (depending on age) have tonsillar herniation of 5mm or more, yet over 90% of these people will NEVER experience any problems or symptoms, so do they really have Chiari?

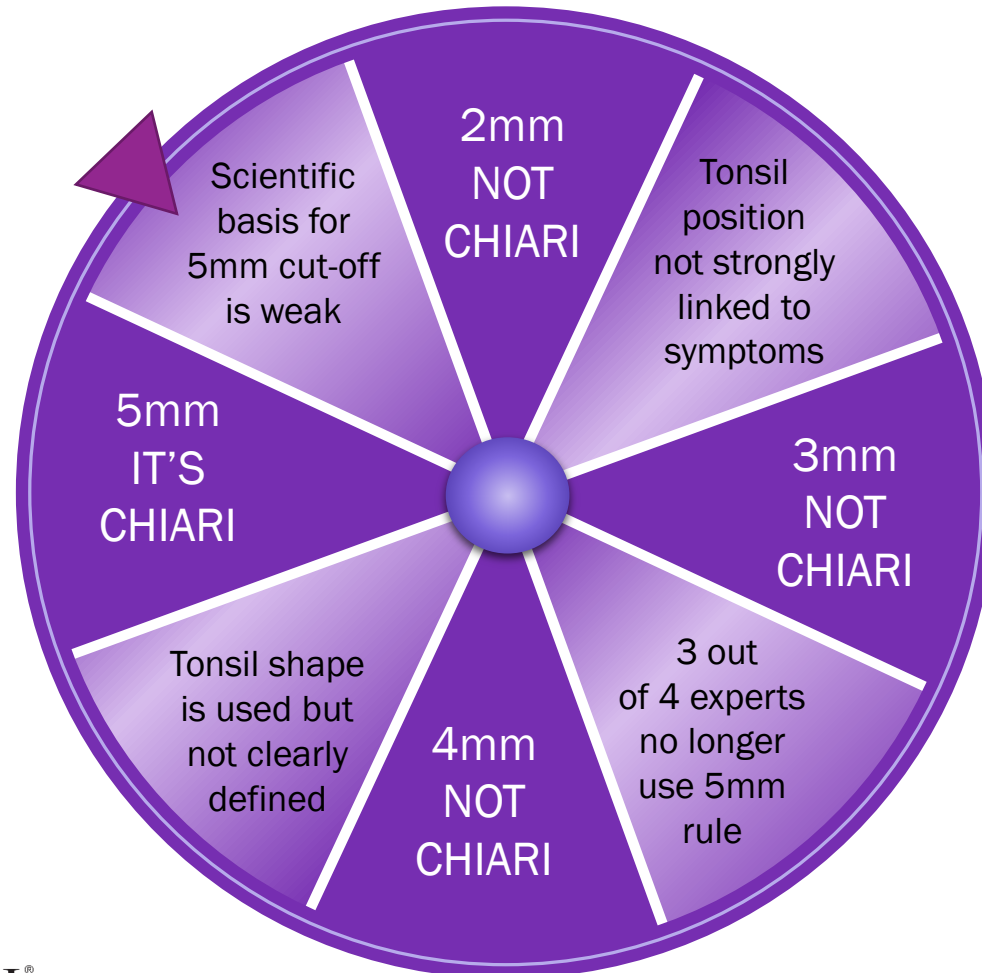
# Issue #2: TP Is Difficult To Measure Accurately



- ❖ Research has shown significant variation in measurements even between experts
- ❖ The specific MRI machine and settings can impact the quality of the images and make anatomical landmarks difficult to precisely mark
- ❖ Even a healthy person's tonsillar position changes over time as they age
- ❖ Anecdotally, the extent of herniation on MRI does not always reflect what is found during surgery

# Issue #3: 5mm is Somewhat Arbitrary

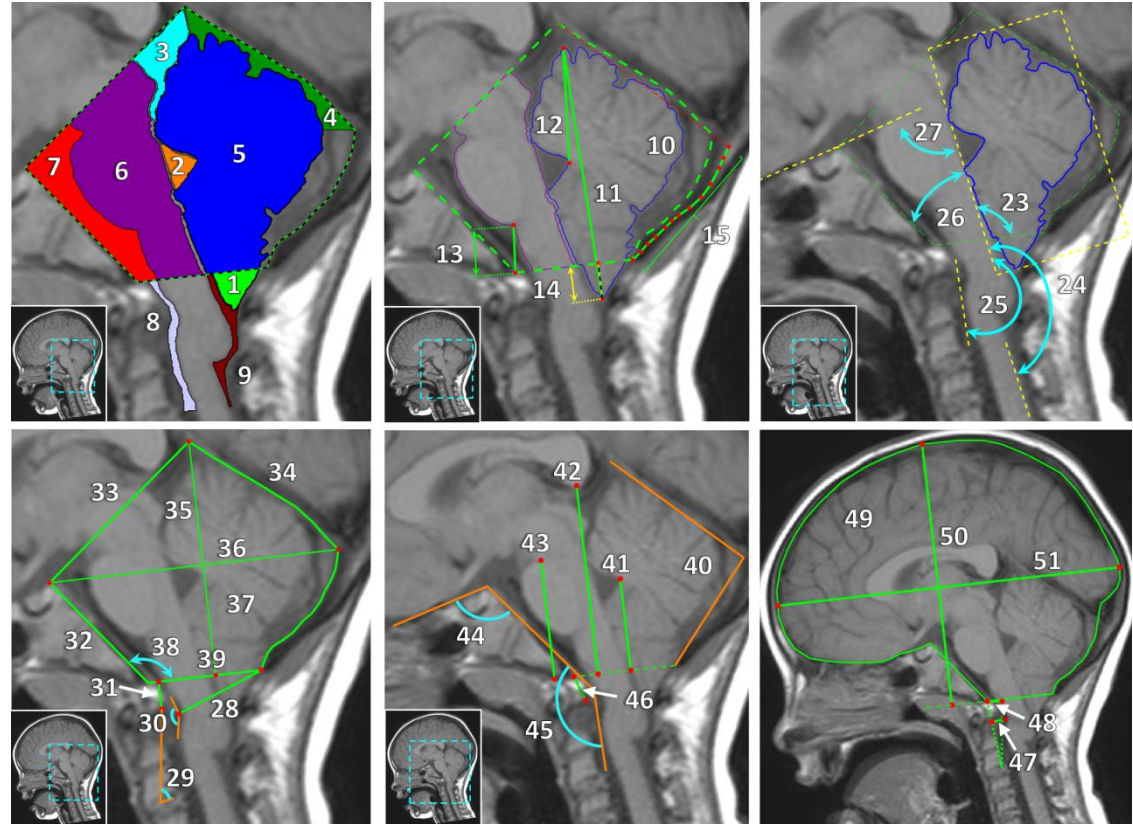
**SPIN THE CHIARI  
WHEEL!!**



*Despite growing evidence, some patients are told they do not have Chiari if an MRI report indicates 3 or 4mm of tonsillar herniation.*

# Looking Beyond Tonsillar Position

- ❖ The Conquer Chiari Research Center is a world leader in studying brain & skull morphometrics (angles and distances)
- ❖ They have found that Chiari patients have many anatomical differences from healthy controls
- ❖ Several studies indicate the clivus bone (part of the skull base) may play a key role and be useful in diagnosing Chiari
- ❖ It also appears that more than just the cerebellar tonsils are “lower” in Chiari brains



# What Causes Chiari?

## Small Posterior Fossa:

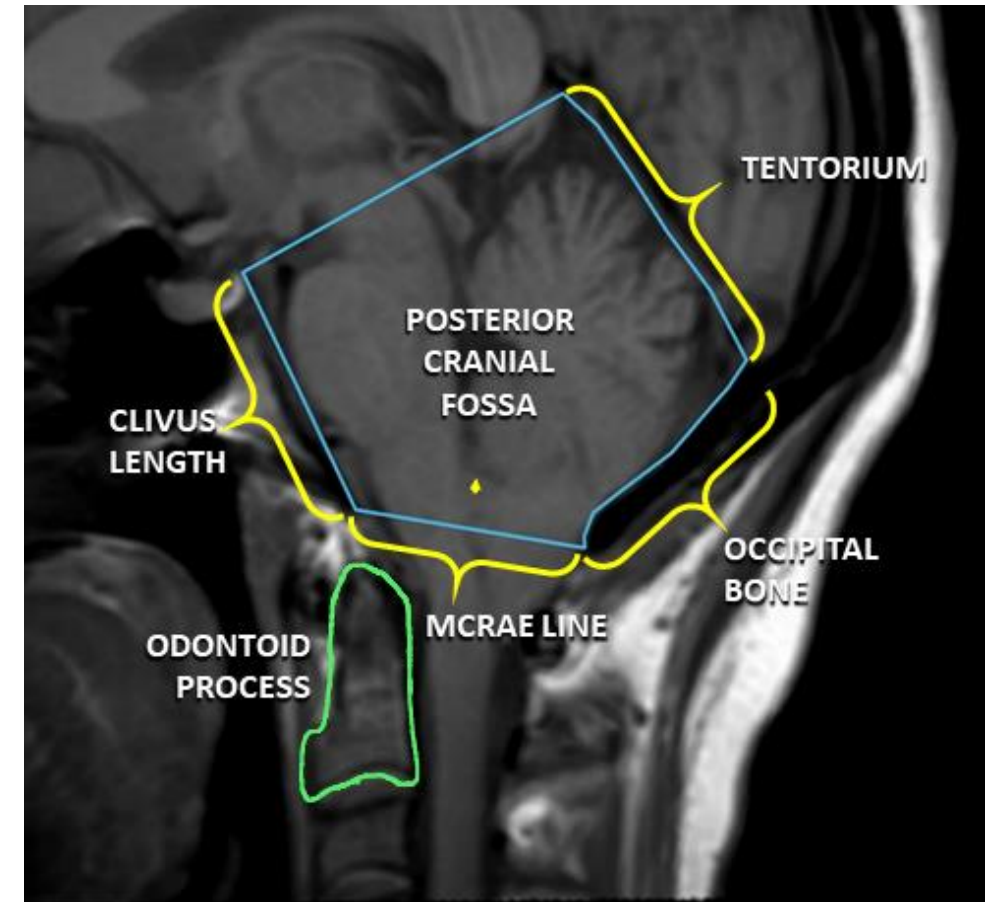
- ❖ The Posterior Fossa is the region of the skull in the back where the cerebellum is located. People have speculated that this region is too small in Chiari patients and so the cerebellum grows outside of the skull
- ❖ However, not all Chiari patients have small PFs and this theory doesn't account for why so many healthy people have tonsillar herniations

## Cervical Instability:

- ❖ Recent theory that Chiari type symptoms are actually due to cervical instability
- ❖ However, a review of the literature found no real evidence to support this and some contrary evidence

## Established, but less common causes:

- ❖ A mass in the brain, such as a tumor or cyst can push out the cerebellum
- ❖ A myelomeningocele (spina bifida) can lead to a pressure drop and herniation
- ❖ Rare genetic conditions that severely affect the skull shape





# Common Chiari Questions

## How many people have Chiari?

- ❖ Symptomatic Chiari is more common than many people think. Conquer Chiari estimates that 1 in 1,000 people have symptomatic Chiari, which translates to 300,000 people in the US alone. According to the American Association of Neurological Surgeons, there are over 10,000 Chiari surgeries each year.

## Who gets Chiari?

- ❖ It is generally believed that Chiari affects people of all races. Among adults, women are affected more often than men, but among children it is an even split. Symptoms can develop at any age, but people are usually diagnosed as children or as adults in their late 20's or early 30's.

# Common Questions - continued

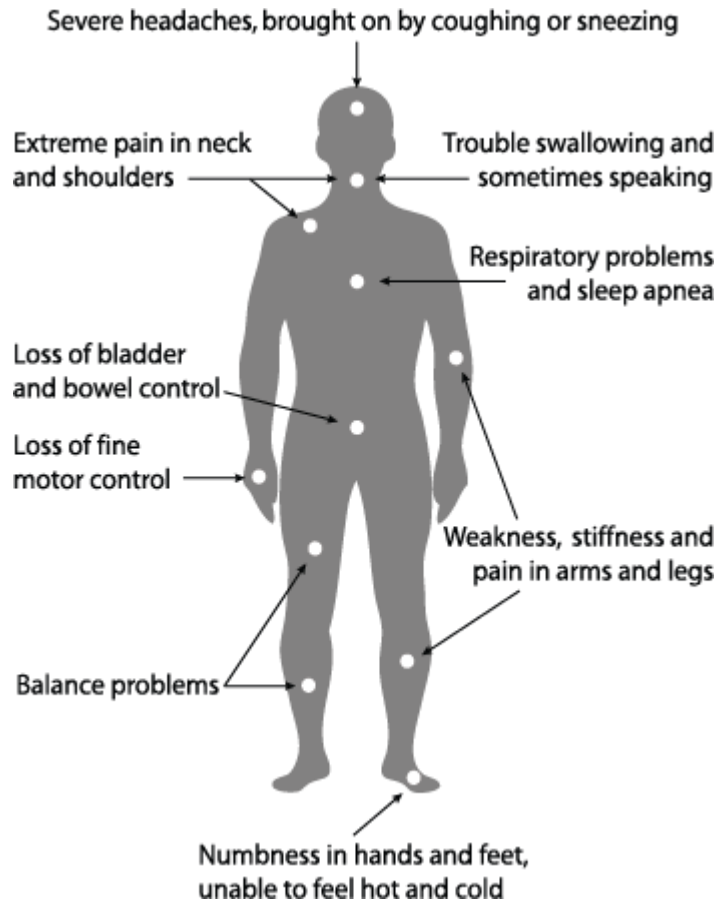
## Is Chiari genetic?

- ❖ Research continues to grow that indicates that some percentage of cases are hereditary in nature. However, it is not known what this percent is, and what genes are involved. There is no genetic test for Chiari at this time.

## Is Chiari fatal?

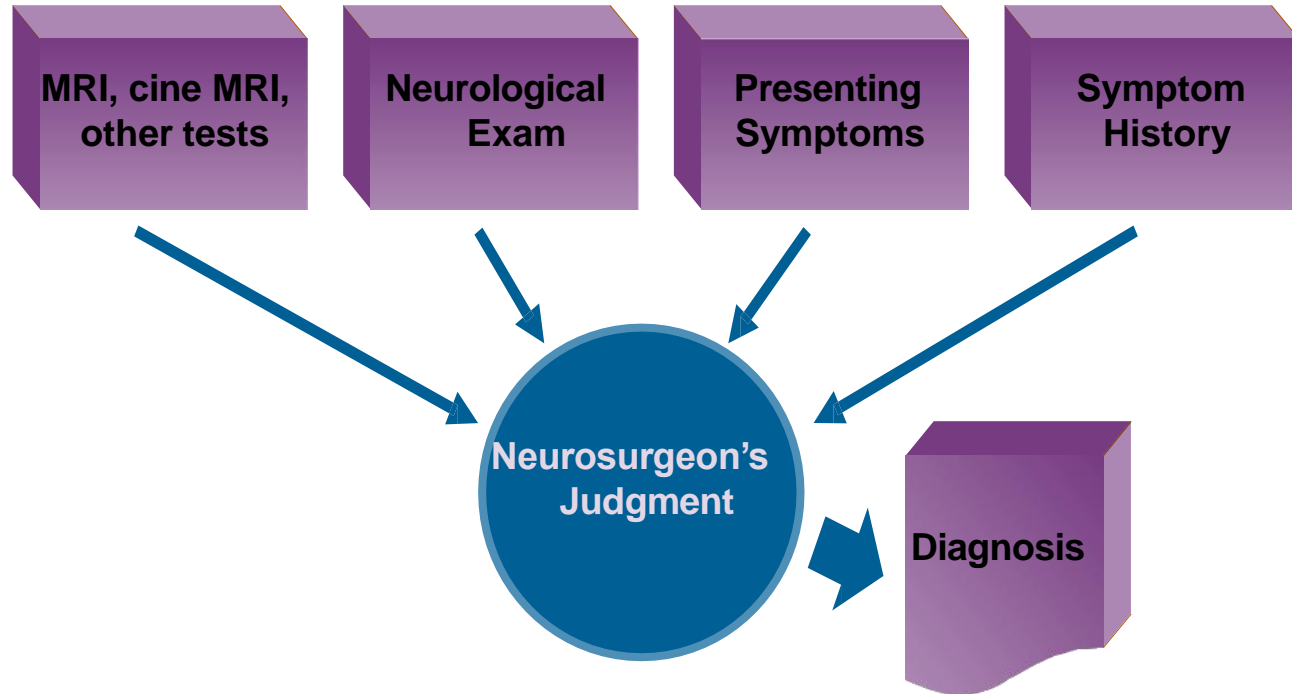
- ❖ In general Chiari 1 is not considered fatal and is not tracked in terms of 5 or 10 year survival rates. However, there are case reports of sudden death associated with Chiari 1 due to breathing problems and mortality associated with surgical complications, however both of these are uncommon. Although not studied extensively, Chiari patients are at higher risk for self-harm behaviors and anecdotally there are cases of suicide each year. Finally, the impact that long-term symptoms may have on life expectancy has not been studied at all.

# The Chiari Experience: Symptoms



- ❖ Signature symptom is a severe headache in the back of the head brought on by straining, coughing, etc.
- ❖ It is not known why but symptoms can develop at any age
- ❖ Trauma may play a role in sparking symptoms
- ❖ Symptoms are believed to be due to compression of brain and spinal tissue, disruption of the natural flow cerebrospinal fluid, and increased pressure in the brain
- ❖ Recent and ongoing research is beginning to characterize the cognitive and psychological effects of Chiari

# The Chiari Experience: Diagnosis



- ❖ There is no single, objective, definitive test for symptomatic Chiari. Diagnosis is based on a combination of factors and is an informed opinion on whether symptoms are due to herniated cerebellar tonsils
- ❖ Chiari is often missed and misdiagnosed as other diseases

*It can take years and many doctors for a patient to get an accurate diagnosis*

# Treatment

- ❖ Only treatment which addresses underlying problem is surgery
- ❖ Treatment decision is based on whether symptoms are clearly due to Chiari and bad enough to warrant surgery
- ❖ No objective test or criteria to say when a patient should have surgery
- ❖ Since surgery is the primary treatment option, most patients see a neurosurgeon for treatment opinions
- ❖ Only about half (or less) of patients evaluated are recommended for surgery

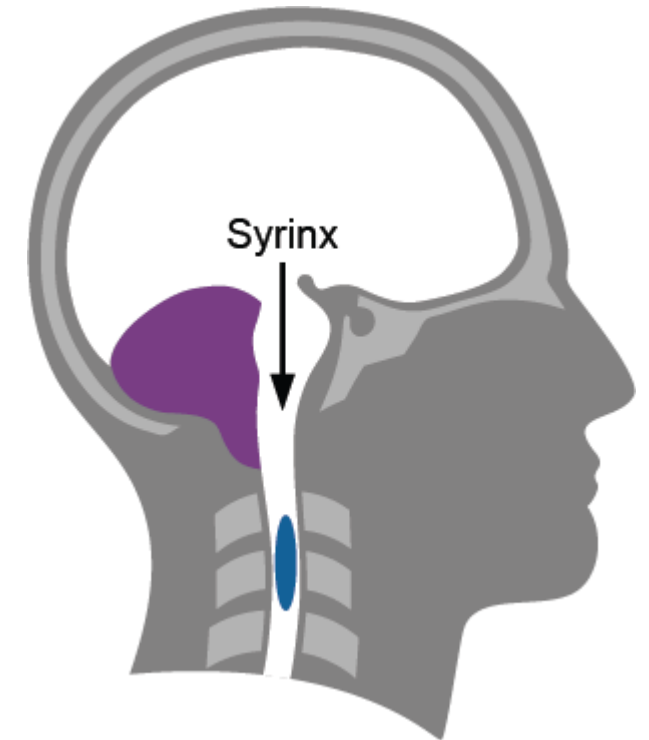


*It's common for patients to receive different opinions from different neurosurgeons*

# Related Conditions: Syringomyelia

**Some Chiari patients may also develop syringomyelia (SM):**

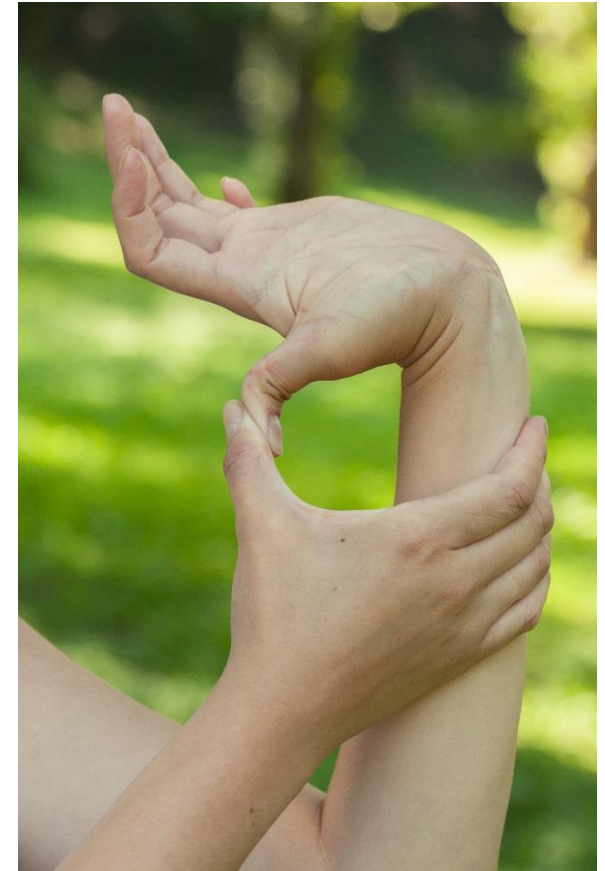
- ❖ Neurological condition where a fluid filled cyst, or syrinx, forms in the spinal cord
- ❖ The syrinx expands the spinal cord, sometimes to more than twice its normal diameter, and stretches the nerve tissue paper thin
- ❖ Exactly how Chiari causes a syrinx is not completely understood
- ❖ Can cause permanent nerve damage and paralysis
- ❖ Other symptoms include neuropathic pain, loss of sensation, muscle weakness, and bladder/bowel problems



# Related Conditions: EDS

A subset of Chiari patients also have hypermobile EDS (Ehlers-Danlos Syndrome) which is characterized by joint hypermobility (joints that stretch further than normal), skin hyper-extensibility (skin that can be stretched further than normal), and tissue fragility.

- ❖ Data from the Chiari1000 shows that 11% of over 1100 patients in the database report being diagnosed with EDS by a physician.
  - ❖ Because the diagnosis of EDS is fairly involved, it is possible this number underestimates the true rate of EDS among Chiari.



# Related Conditions: PTC & Hydrocephalus

Research from the Chiari1000 shows more than 10% of adults with Chiari have been diagnosed with Pseudotumor Cerebri (PTC)

According to the Mayo Clinic, **Pseudotumor cerebri** (SOO-doe-too-mur SER-uh-bry) occurs when the pressure inside your skull (intracranial pressure) increases for no obvious reason. Symptoms include:

- ❖ Headaches behind the eyes
- ❖ Nausea/vomiting
- ❖ Dizziness
- ❖ Blurred Vision
- ❖ Brief Blindness to one or both eyes
- ❖ Difficulty with side vision
- ❖ Neck, shoulder or back pain

Research from the Chiari1000 shows more than 7% of Chiari patients have been diagnosed with Hydrocephalus, a buildup of fluid in the ventricles (cavities) inside the brain. Informally known as water on the brain

Symptoms of Hydrocephalus include:

- ❖ Headaches
- ❖ Nausea
- ❖ Difficulty focusing
- ❖ Unsteady walk
- ❖ Sudden falls
- ❖ Leg weakness
- ❖ Drowsiness

*It is not clear if Chiari causes these conditions, these conditions lead to Chiari, or if they are both the result of something else*



# Three Keys To Understanding Chiari



**Each person's Chiari experience is unique**

It is impossible for others to completely understand what you are going through; look for validation within yourself



**Not every question has an answer**

Don't waste time searching for absolutes or a magic bullet



**There are two options: take control or be controlled**

Be proactive, educate yourself, and trust your instincts

# Summary

- ❖ Chiari is a serious neurological condition that can have a severe impact on patients and their families
- ❖ As research advances, limitations with the traditional definition of Chiari are becoming clear
- ❖ Chiari causes a variety of symptoms
- ❖ The only real treatment is surgery to create more room and restore the natural flow of spinal fluid
- ❖ Syringomyelia, EDS, PTC, and hydrocephalus are commonly associated with Chiari
- ❖ Chiari patients should educate themselves and be proactive in managing their healthcare

Learn More in the [Conquer Chiari Library](#)