

# An Overview of the Chiari Malformation

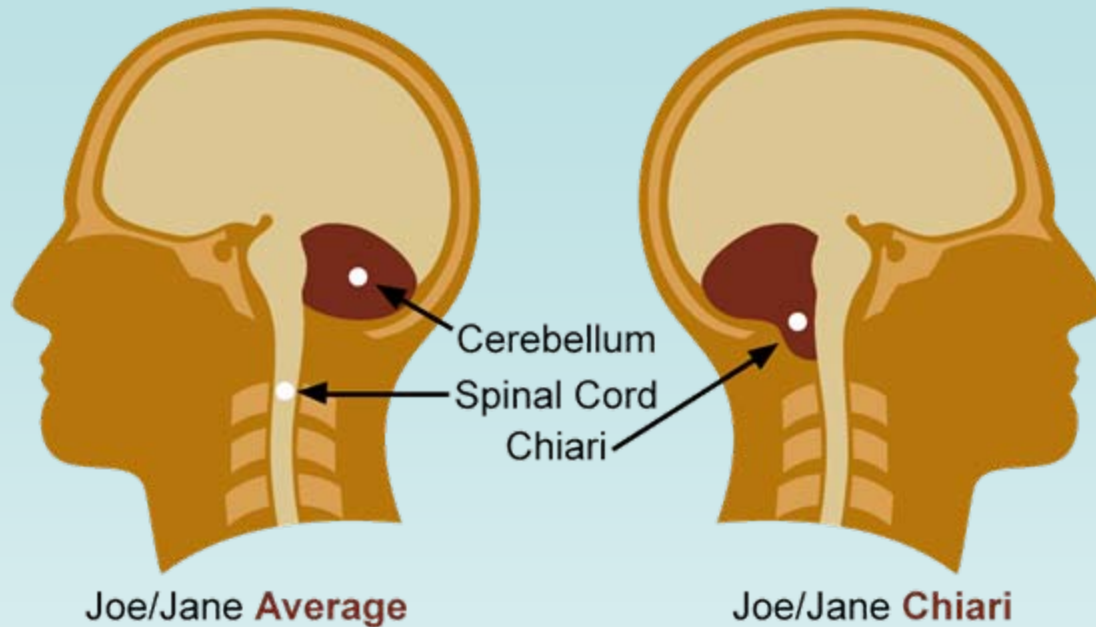


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# What Is Chiari?



**Chiari Malformation** (Arnold-Chiari) 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.

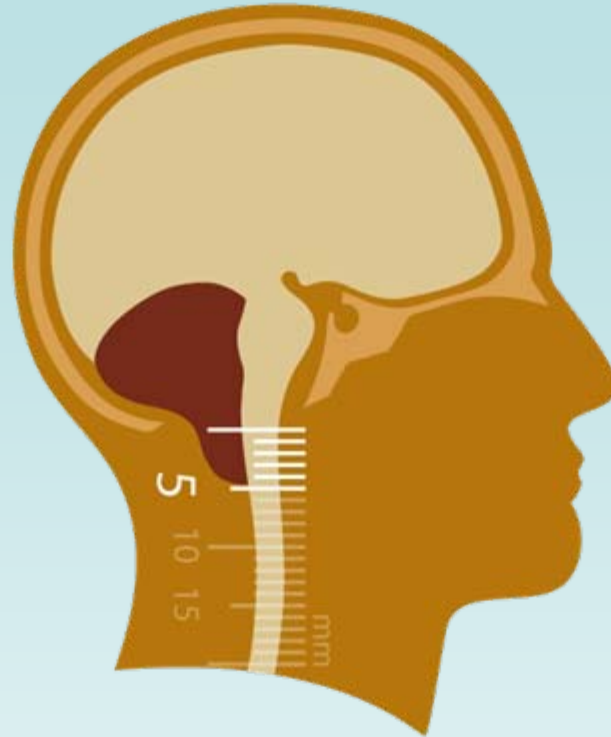
- First identified by Hans Chiari in the 1890's
- Several types, Type I is shown above; Type II is associated with Spina Bifida
- Also known Chiari Malformation (CM), Arnold-Chiari Malformation (ACM), tonsillar herniation, tonsillar ectopia, hindbrain herniation

***Chiari affects everyone differently!!***

# Evolving Definition



## Old Definition



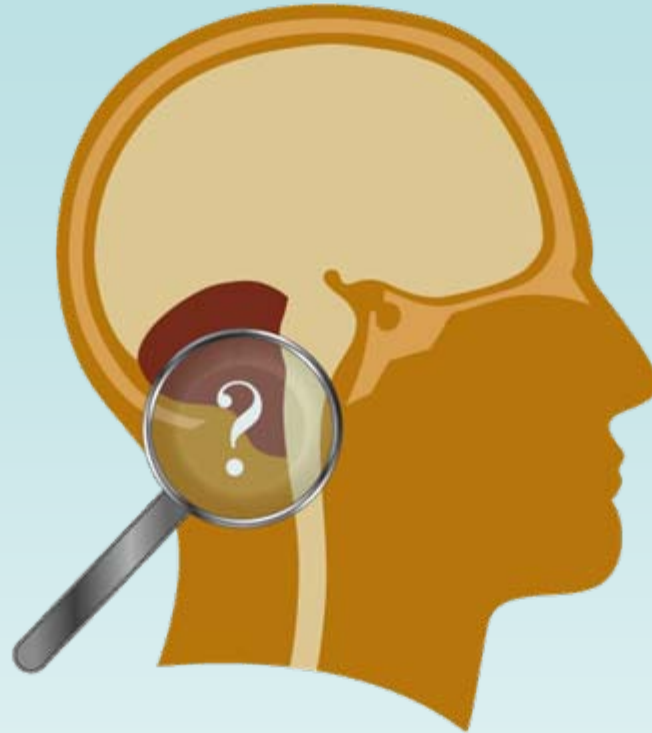
- Chiari malformation is traditionally defined as the cerebellar tonsils being located 3mm-5mm or *more* below the foramen magnum (opening at the bottom of the skull) as measured on an MRI
- However, research has shown that the size of a herniation is *NOT* strongly related to how severe symptoms are or how people respond to treatment

***Some people have large herniations with no symptoms; while others have small herniations with severe symptoms***

# Evolving Definition



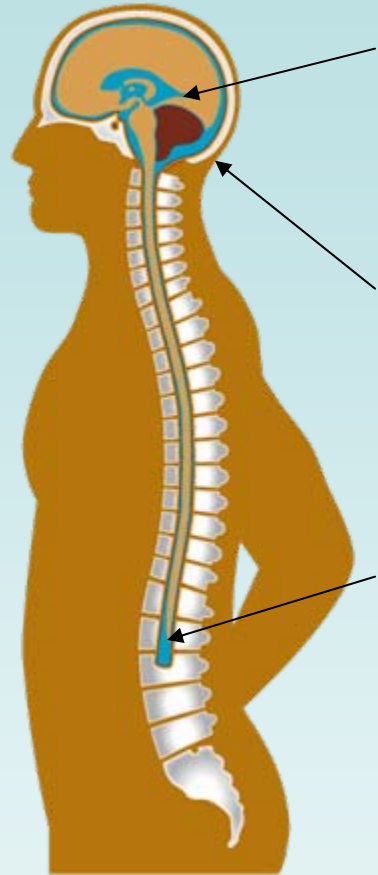
**New  
Definition?**



- Researchers are looking for a new way to measure Chiari severity
- Focus areas include advanced MRI and engineering techniques to quantify cerebrospinal fluid (CSF) flow, crowding, and compliance
- Some preliminary results are encouraging, but it will take time to develop

***Conquer Chiari is funding research to develop a quantitative, objective test of symptomatic Chiari***

# What Causes Chiari?



## **Cerebrospinal Fluid Issues**

Altered CSF dynamics “push” brain out of skull. For example, hydrocephalus, intracranial hypertension.

## **Small Posterior Fossa**

Back of the skull is too small for the normal sized brain. May be most common.

## **Tethered Cord (very controversial)**

Abnormal anchoring of the spinal cord at the base leads to herniation, also causes symptoms of its own.

## **Other Causes:**

- Acquired due to a mass in the brain
- Spina Bifida (Chiari II)
- Genetic conditions such as an Ehlers-Danlos variant
- Incidental; found when an MRI is done for a different reason, such as an accident

***It is believed Chiari can be due to several things. However these are only theories and exact mechanisms are not understood. It is not known how common each type may be.***

# Common Questions



- **How many people have Chiari?**

*Chiari is more common than many people think.*

*Conquer Chiari estimates that 1 in 1,000 people have 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. There are some indications that it affects more women than men. 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 – cont'd



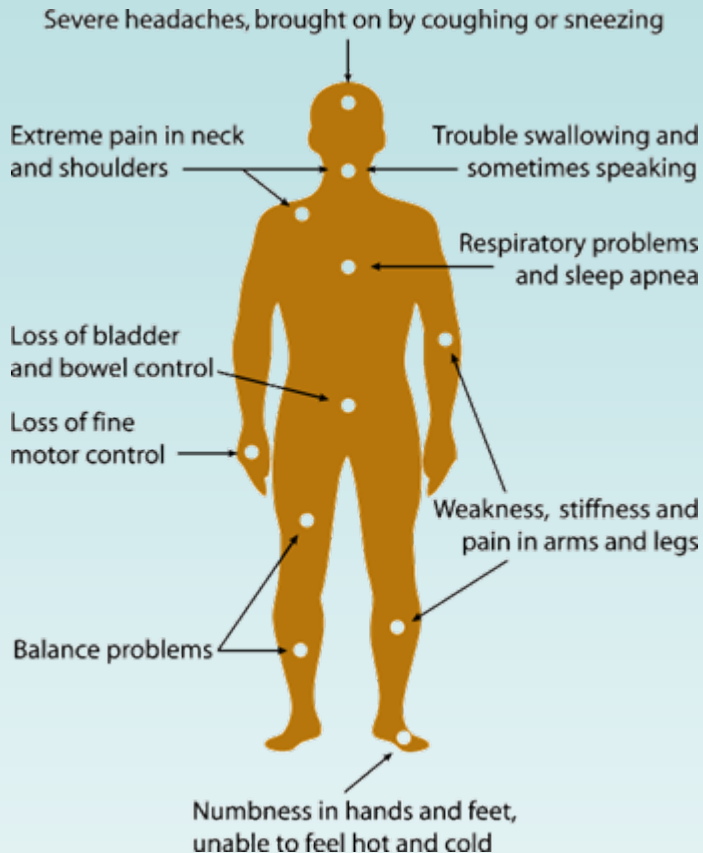
- **Is Chiari genetic?**

*Research has identified more than 100 families with multiple affected members, implying that at least some cases are genetic in origin. However, it is not known what percent of cases are, what genes are involved, and there is no genetic test for Chiari.*

- **Is Chiari fatal?**

*In general Chiari I is not considered fatal. For example 5 year survival rates are not calculated or discussed. However, there are case reports of sudden death associated with Chiari I due to breathing problems, although this is likely very uncommon. Also, the impact that long-term symptoms may have on life expectancy has not been studied. Finally, severe cases of Chiari II in infants and children can be fatal.*

# 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 due to compression of brain and spinal tissue, disruption of the natural flow cerebrospinal fluid, and increased pressure in the brain
- Some people with malformations are asymptomatic, meaning they have no symptoms
- The cognitive effects of Chiari are being researched, but are not well understood

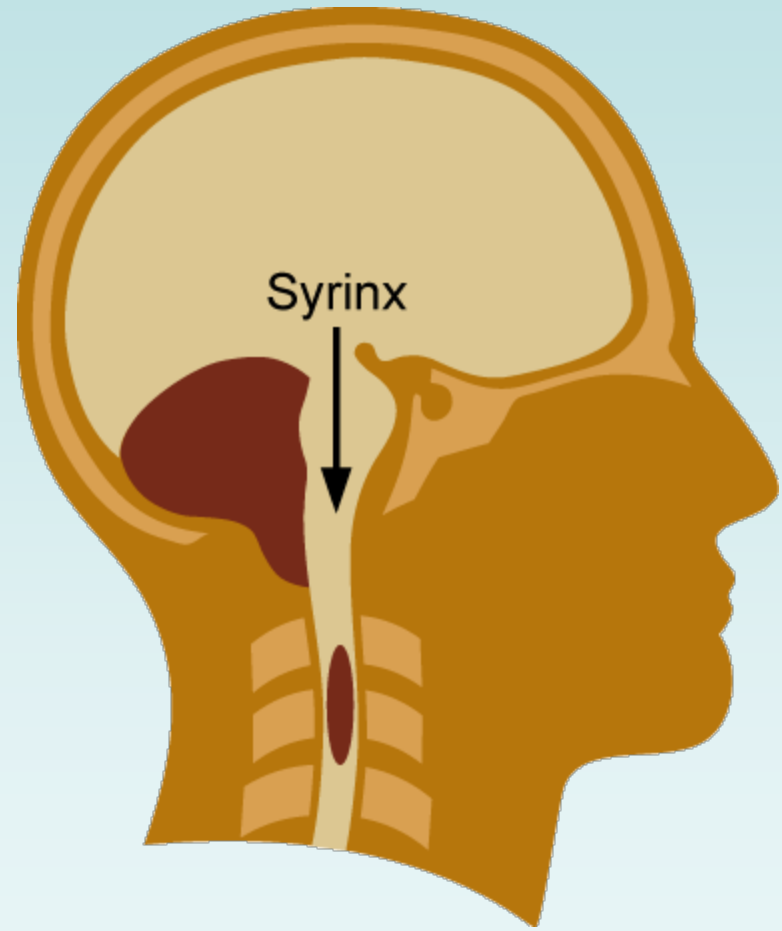
***Chiari symptoms can range from mild, to severe, to completely debilitating***

# Syringomyelia

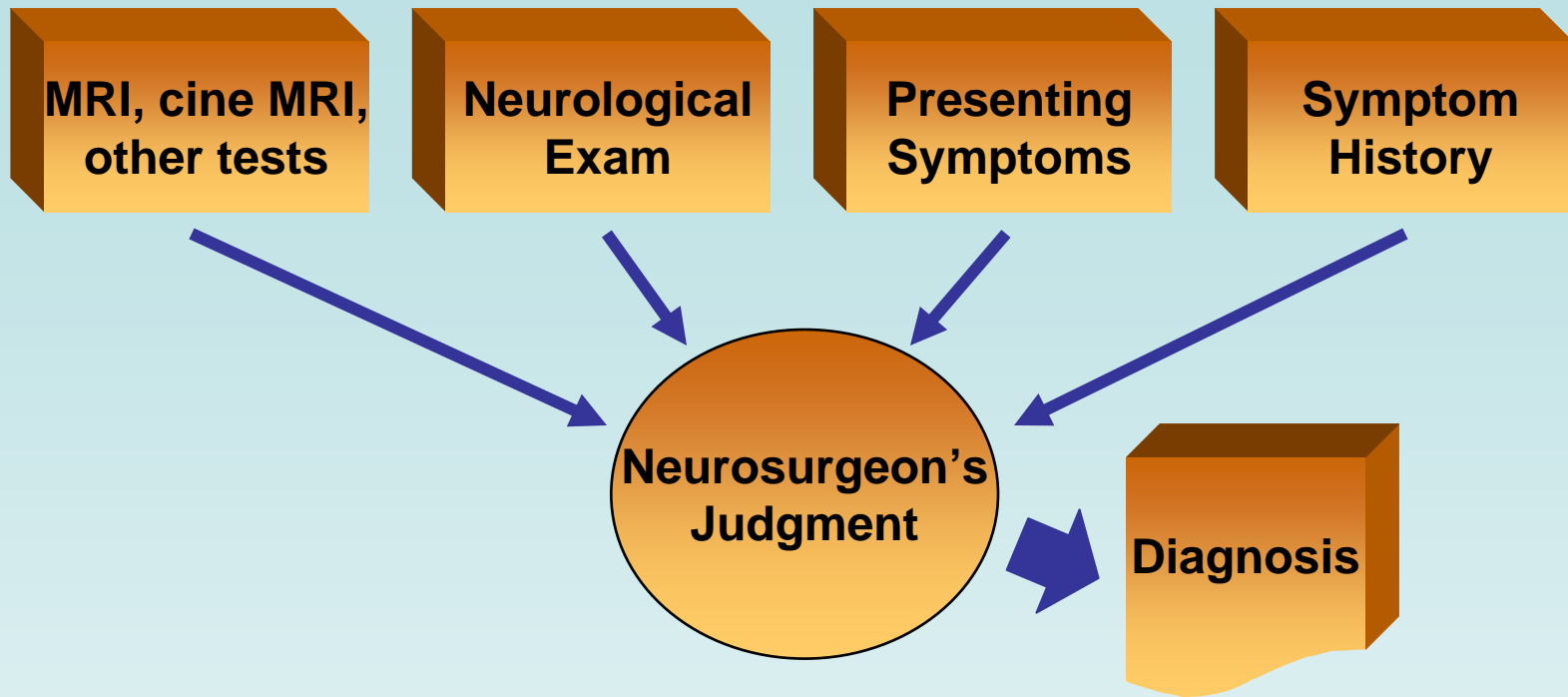


**Up to 50% of Chiari patients also develop syringomyelia (SM):**

- 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
  - Likely due to disruption of the natural flow of cerebrospinal fluid (CSF) between the brain and spinal cord
- Can cause permanent nerve damage and paralysis
- Other symptoms include neuropathic pain, loss of sensation, muscle weakness, and bladder/bowel problems



# 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 mis-diagnosed 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
- Alternative is to monitor situation and treat individual symptoms (Wait & See)
- Since surgery is the primary treatment option, most patients see a neurosurgeon for treatment opinions

*A 2004 survey of neurosurgeons (Schjiman) found a wide range of thinking on when to operate for Chiari.*

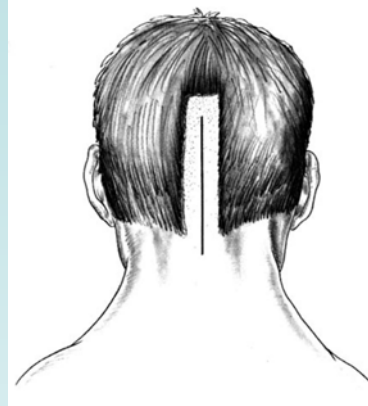
Case #	Symptoms/Diagnosis	% Would Operate
1	7 yr old with no symptoms 12 mm tonsils- <u>no</u> syrinx	8
1A	w/ 2mm wide syrinx	28
1B	w/ 8mm wide syrinx	75
2	9 yr old with headaches 10 mm tonsils- <u>no</u> syrinx	46
2A	w/ 2mm wide syrinx	64
2B	w/ 8mm wide syrinx	90
3	11 yr old w/progressive scoliosis 12 mm tonsils- <u>no</u> syrinx	58
3A	w/ small syrinx	85
3B	w/ 6mm wide syrinx	97

***It is common for patients to receive different opinions from different neurosurgeons***

# Surgery



*Most common surgery is posterior fossa decompression. The goal is to create more room around the malformation and restore the flow of spinal fluid. There are many variations:*



## Surgical Steps

1. Craniectomy – A piece of the skull is removed
2. Laminectomy – Part of one or more vertebrae are removed
3. Duraplasty – The covering of the brain is opened and a patch is sewn in to make it bigger
4. Tissue removal – Cerebellar tonsils are sometimes cauterized (*controversial*)
5. Plate insertion – Mesh is inserted where skull was removed

## Surgical Variations/Controversies

1. How much bone is removed
2. Whether the dura is opened at all
3. What type of material is used for a dural graft (patch)
4. Whether any brain tissue is removed
5. Whether a mesh is put in to take the place of the removed piece of skull

\* *Images courtesy of Dr. Ghassan Bejjani*

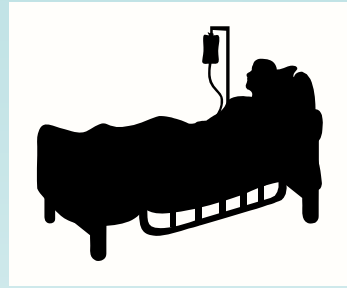
# Surgery – The Experience



*Every case is unique, but a typical experience might be:*



Surgery: ~ 4 hrs.



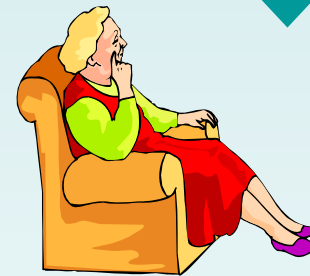
ICU: One night



Hospital: 3-4 days



Normal Activity: Variable



Rest at home: 3 weeks to 3 months

# Outcomes



*Precise numbers are not available, but in general:*

**30-40%**

**Excellent  
Symptom Free  
No Real Limitations**

**30-40%**

**Good  
Some Improvement  
Varying Limitations**

**10-20%**

**Poor  
No Improvement  
Significant Limitations**

- Success is very difficult to objectively define and quantify
- Factors that may affect outcomes include: age, syringomyelia, bony abnormalities, scoliosis, duration of symptoms, and concurrent conditions
- Common residual symptoms include pain, muscle weakness, and loss of sensation; patients will adapt their lifestyle accordingly
- Symptoms may fluctuate over time; patients report that trauma can cause symptoms to come back
- Long term recurrence of symptoms is a recognized problem

***A large study of adults found that 80% reported significant improvement in symptoms after surgery***

# Why Do Surgeries Fail?

*Surgery can fail for a number of reasons*



## ***Surgery Related***

- Inadequate decompression
  - Dura was not opened
  - Not enough bone removed
- Recurrent obstruction
  - Scarring disrupts spinal fluid flow
  - Bone regrowth in children
  - Spinal cord retethers
- Surgical complications
- Altered CSF dynamics
- Cervical instability

## ***Patient Related***

- Other, concurrent conditions still cause symptoms
  - Pseudotumor cerebri; elevated pressure in the brain
  - Basilar invagination; compression of the brainstem from the spine
  - Genetic disorders
- Symptoms were not due to Chiari in the first place
  - Chiari was actually asymptomatic

*It is important to try to understand the reason for failure before undergoing additional treatment*

# State of Research



## ***Known***

Affects >300,000 in the US  
Surgery helps most people  
Can cause syringomyelia  
Affects everyone differently

## ***Suspected***

Several distinct sub-groups/causes  
CSF dynamics play a role  
Children have better outcomes than adults

## ***Questions***

How to identify good candidates for surgery?  
Is there an ideal surgical technique?  
What role do genetics play, and what genes?  
Why do people become symptomatic?

## ***Controversies***

The relationship between tethered cord and Chiari  
Whether children with Chiari should be restricted from physical activity before and/or after surgery  
Whether brain tissue should be removed during surgery

*Conquer Chiari funds research grants and organizes research conferences to advance our understanding of Chiari and improve experiences and outcomes*

# 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*

# Conquer Chiari – Learn More



*In order to Conquer Chiari, it is important to educate yourself. There is a wealth of information available on the Conquer Chiari website:*

**[Research Articles](#)**

**[Personal Stories \(Good & Bad\)](#)**

**[Research Videos](#)**

**[Handouts](#)**

**[Conquer Chiari: A Patient's Guide](#)**

**[Contents Under Pressure](#)**

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