



# ACQUIRED CHIARI

Why is it so important?

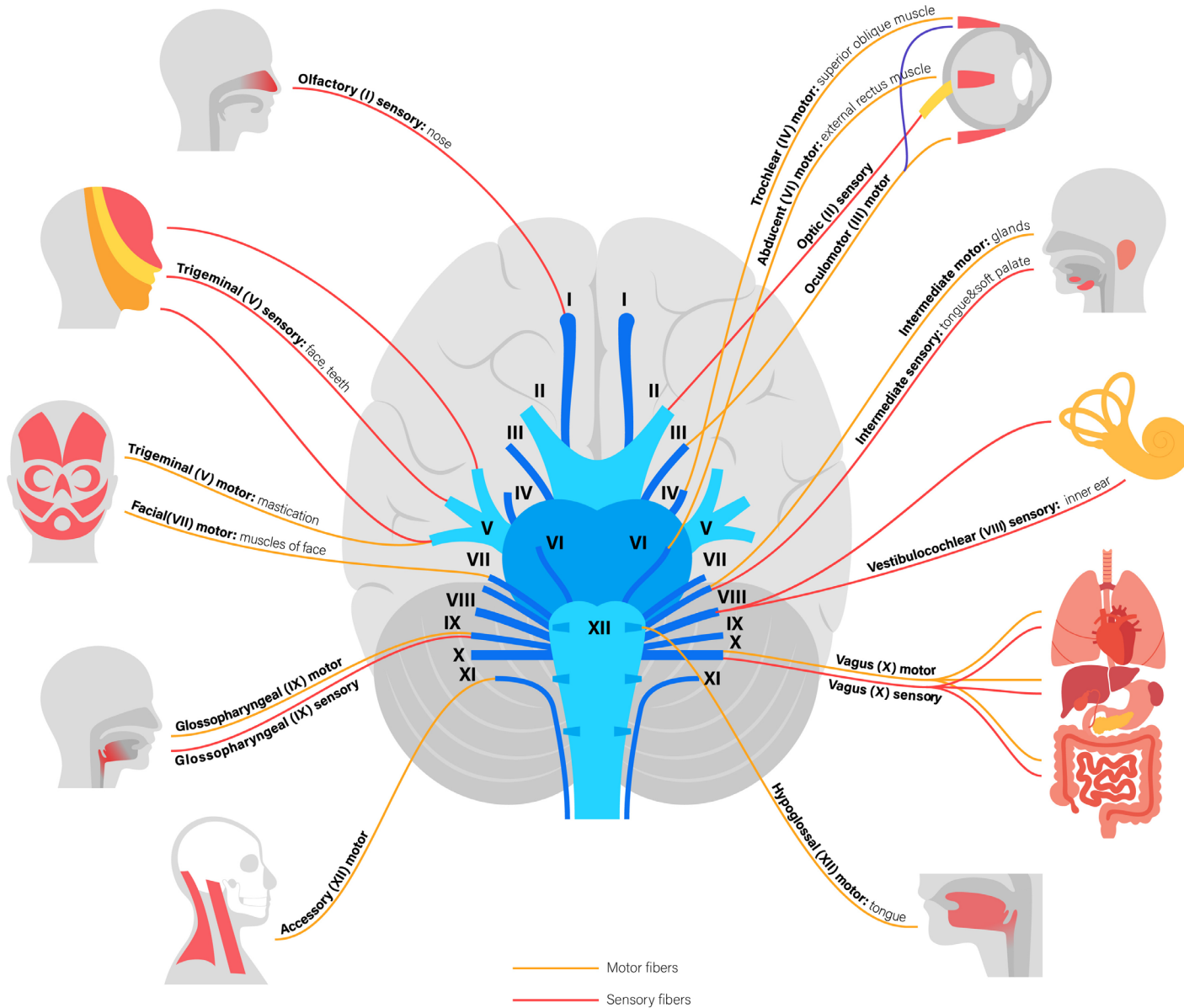
Courtesy of Chiari Bridges

## PRIMARY SYMPTOMS OF CHIARI

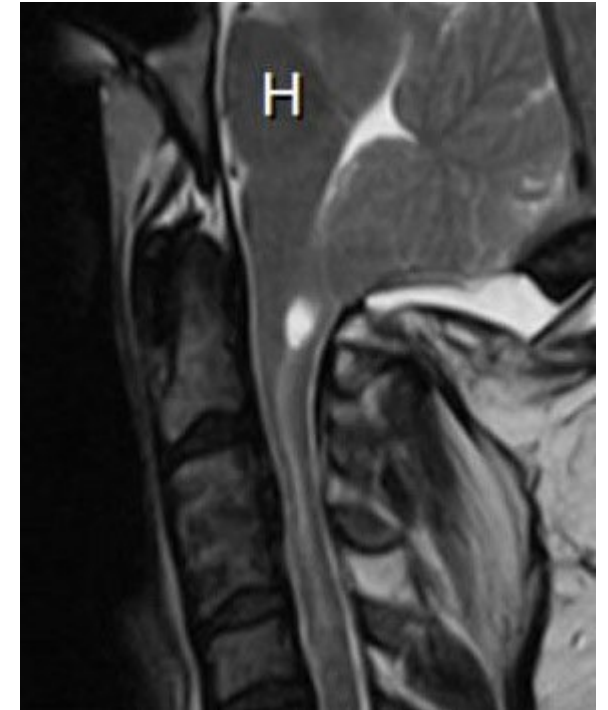


- **Occipital Pain:** pain at the occiput (base of the skull and upper neck), worsened by Valsalva maneuvers and being upright for a prolonged period.
- **Equilibrium Problems:** vertigo, dizziness, nausea/vomiting, balance, gait, and overall proprioception problems (heights, peripheral vision).
- **Cognitive Decline:** brain fog, memory issues often due to inflammation.
- **Throat and Esophageal Problems:** dysphagia, hoarseness, hiccups
- **Sleep Problems:** insomnia, Myalgic encephalomyelitis/chronic fatigue syndrome (ME/CFS) central/obstructive sleep apnea
- **Muscle Spasms:** eye twitching, painful neck spasms, restless leg/limbs
- **Numbness in the arms/hands**
- **Hormone Related Problems**
- **Autonomic Dysfunction (Dysautonomia):** POTS, tachycardia, neurocardiogenic syncope...

# CRANIAL NERVES



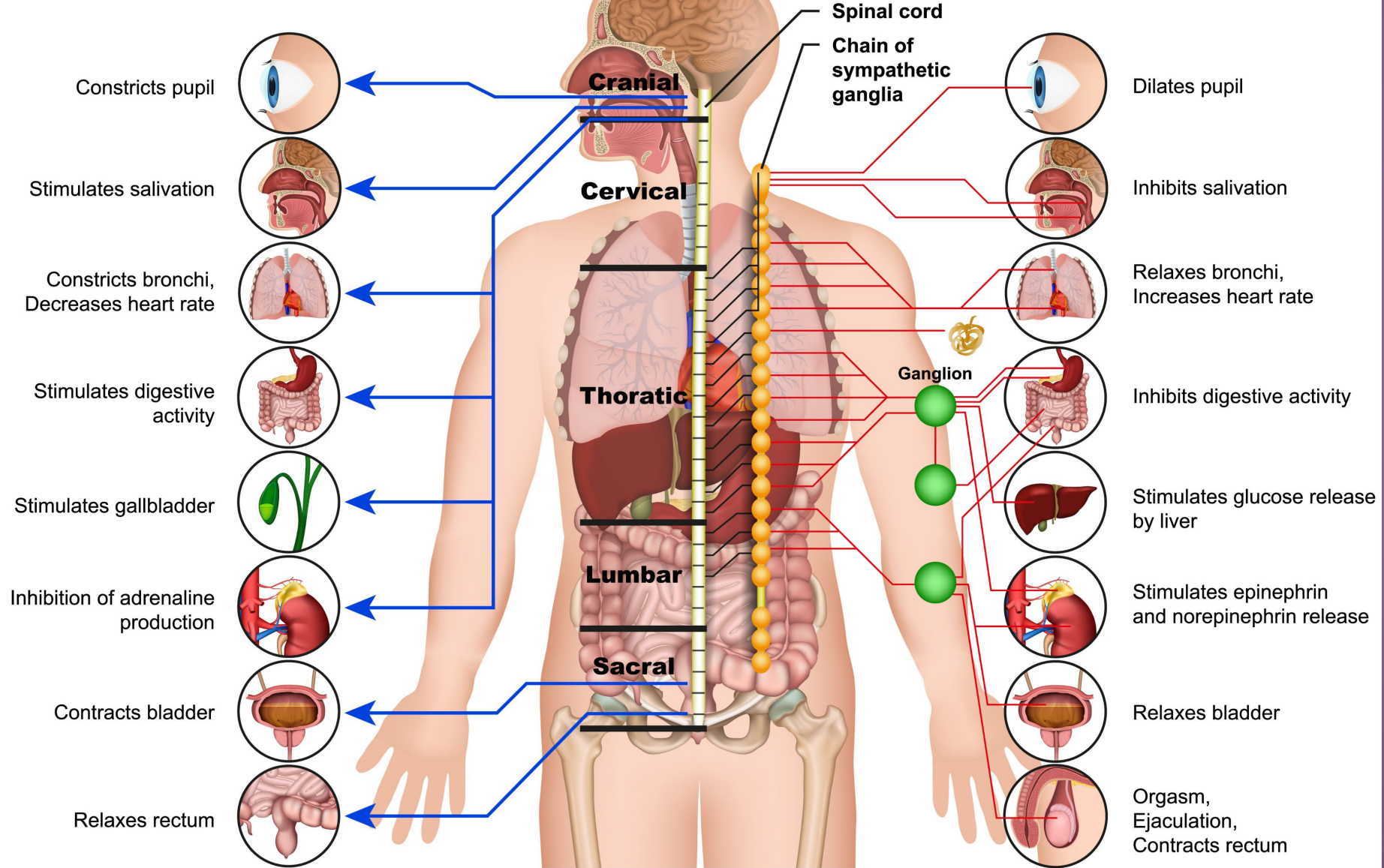
# CRANIAL NERVES



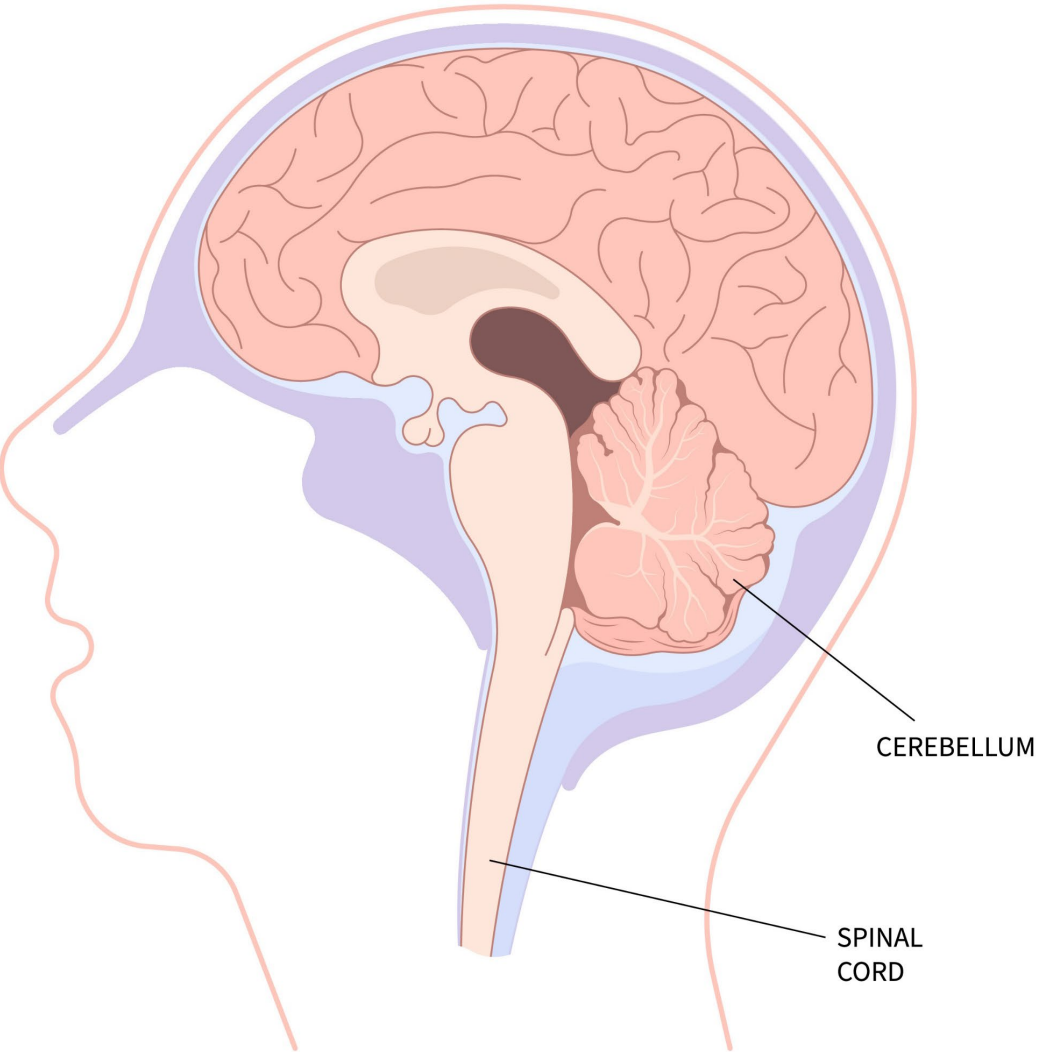


# Parasympathetic nervous system

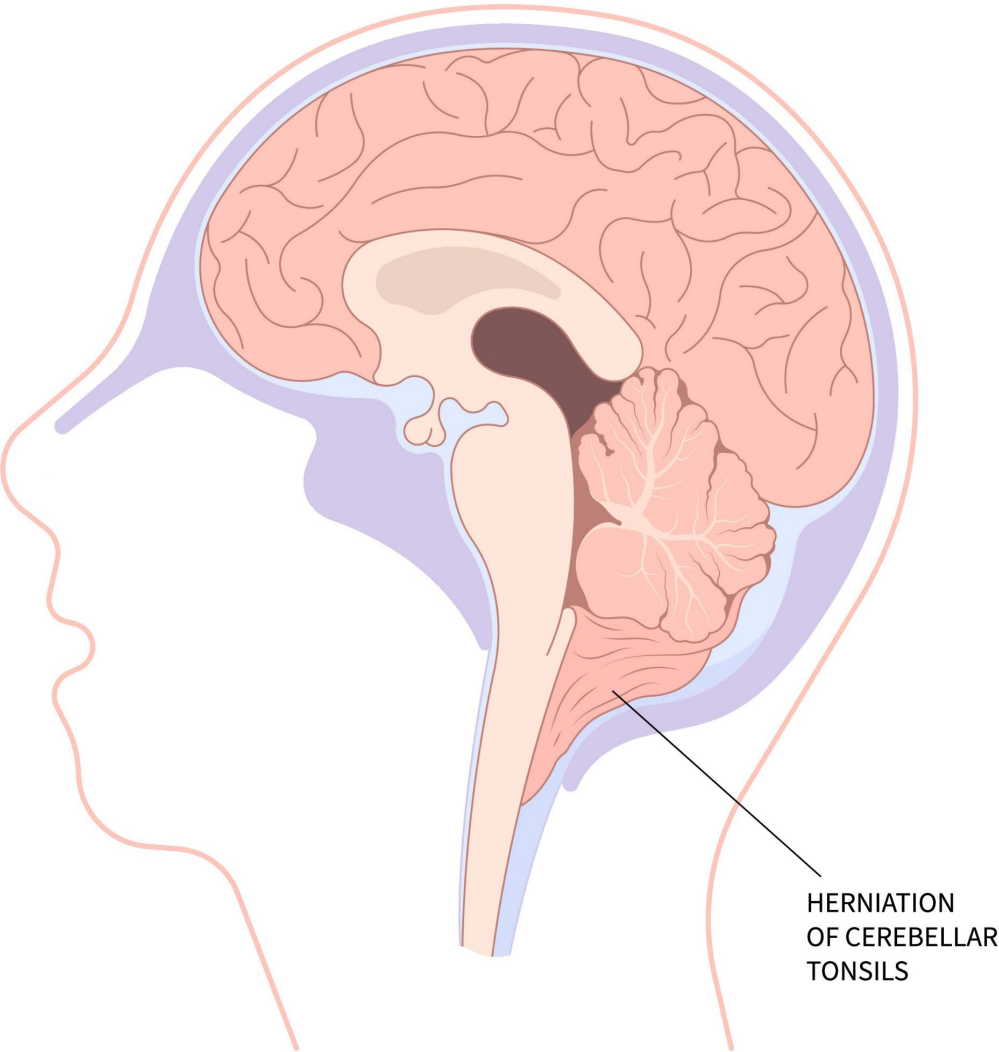
# Sympathetic nervous system



# NORMAL



# CHIARI MALFORMATION





# CONGENITAL defined

Structural or functional anomalies (for example, metabolic disorders) that occur during intrauterine life.





---

## ACQUIRED defined

---

Conditions that are NOT INHERITED or present at birth (congenital) but developed after birth.



# Congenital Assumption & Failed Decompressions

Congenital  
Chiari



Small  
Posterior  
Fossa

Acquired  
Chiari



Pathological  
Conditions

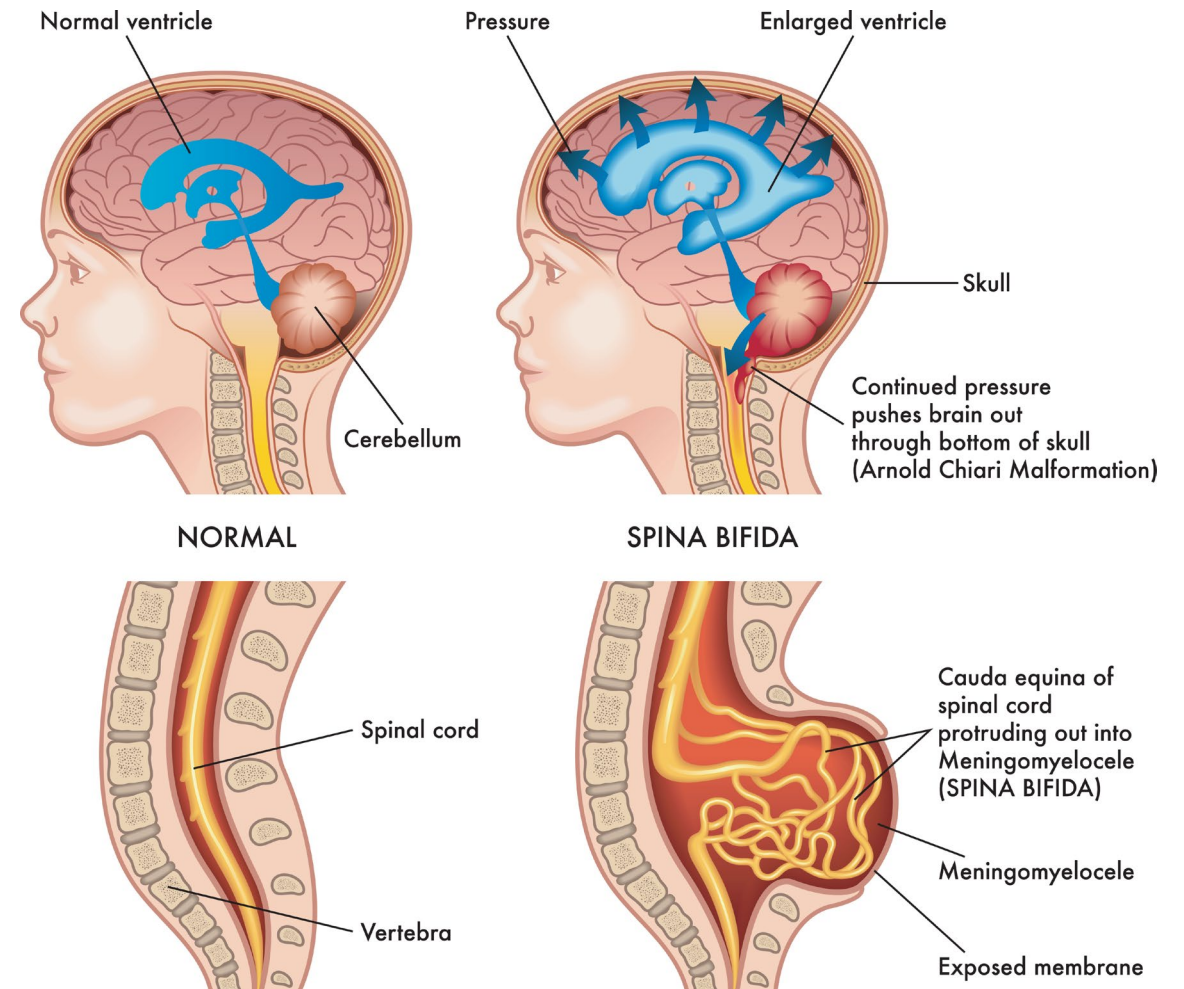




# Arnold Chiari

## Chiari 2 Malformations

- Spina Bifida (myelomeningocele)
- Tethered Cord Syndrome
- Hydrocephalus induced IH



# The Occipital Bone

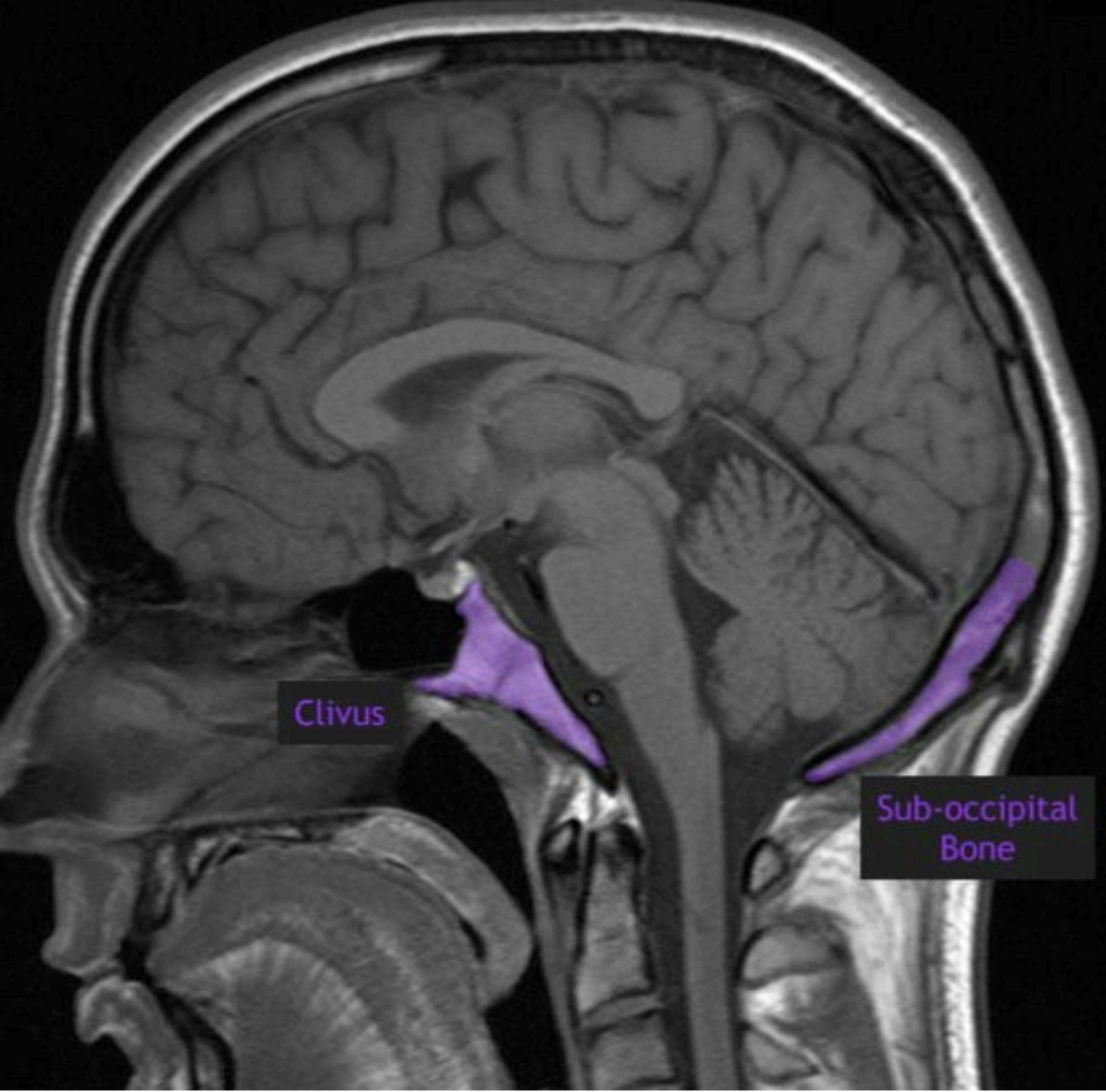




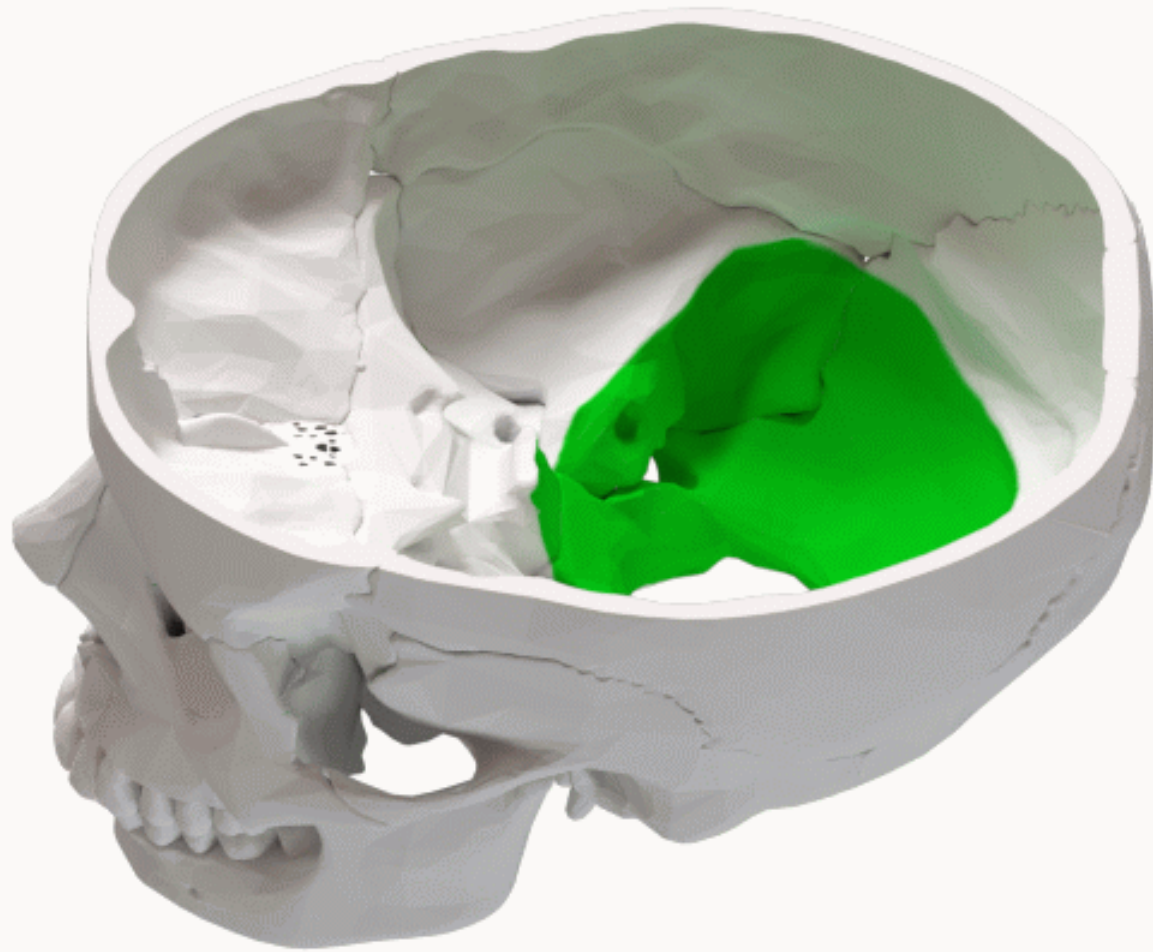


# The Foramen Magnum





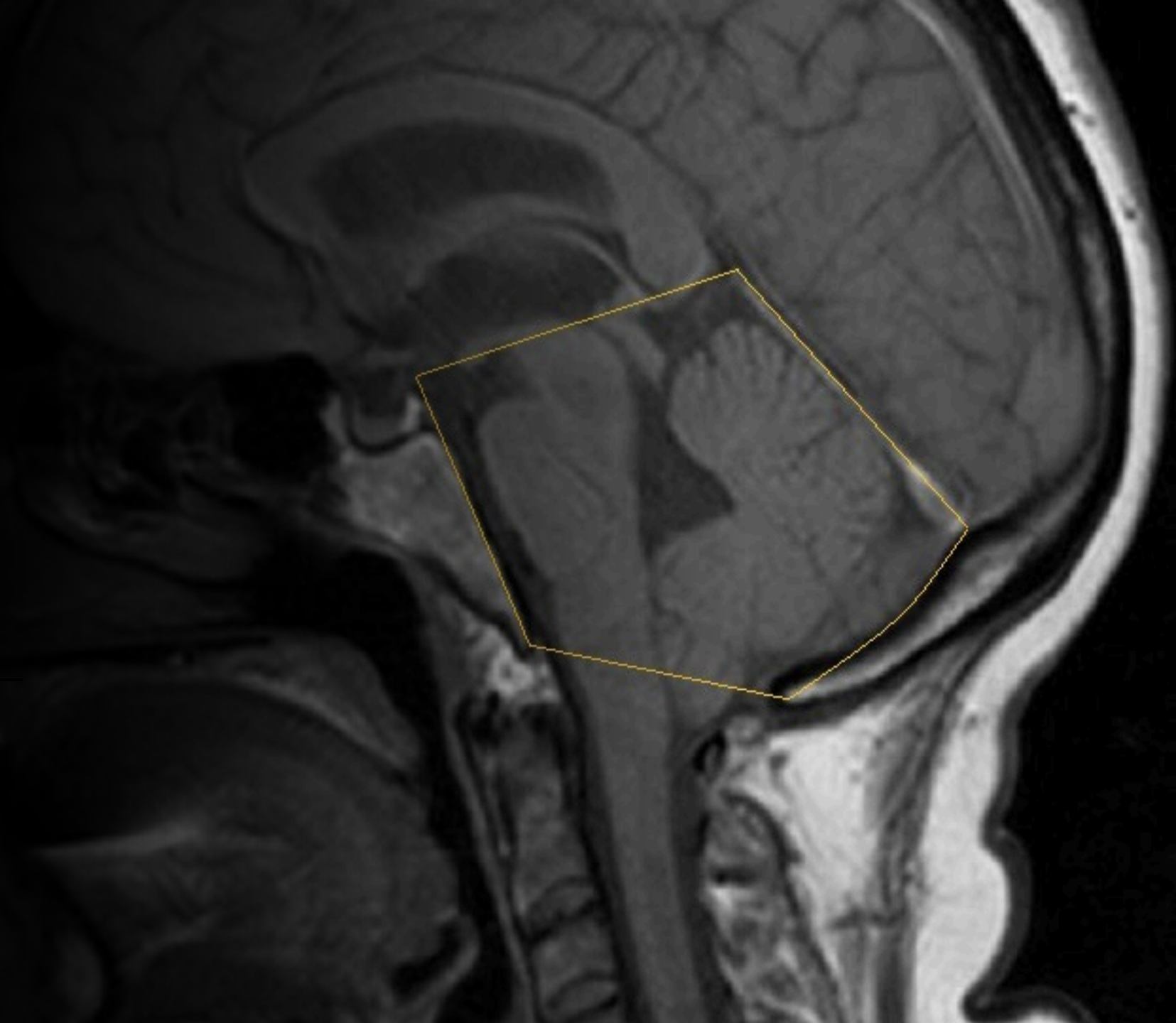
# The Foramen Magnum



# Posterior Fossa



# Posterior Fossa Hypoplasia





**Does Size  
Matter?**



How many millimeters does it take to block the flow of cerebrospinal fluid?







# HYPERMOBILE EHLERS-DANLOS CONNECTS THE DOTS

## INTRACRANIAL PRESSURE PROBLEMS

- Idiopathic Intracranial HYPERTension\*
- Spontaneous Intracranial HYPOtension

## CRANIOVERTEBRAL ABNORMALITIES

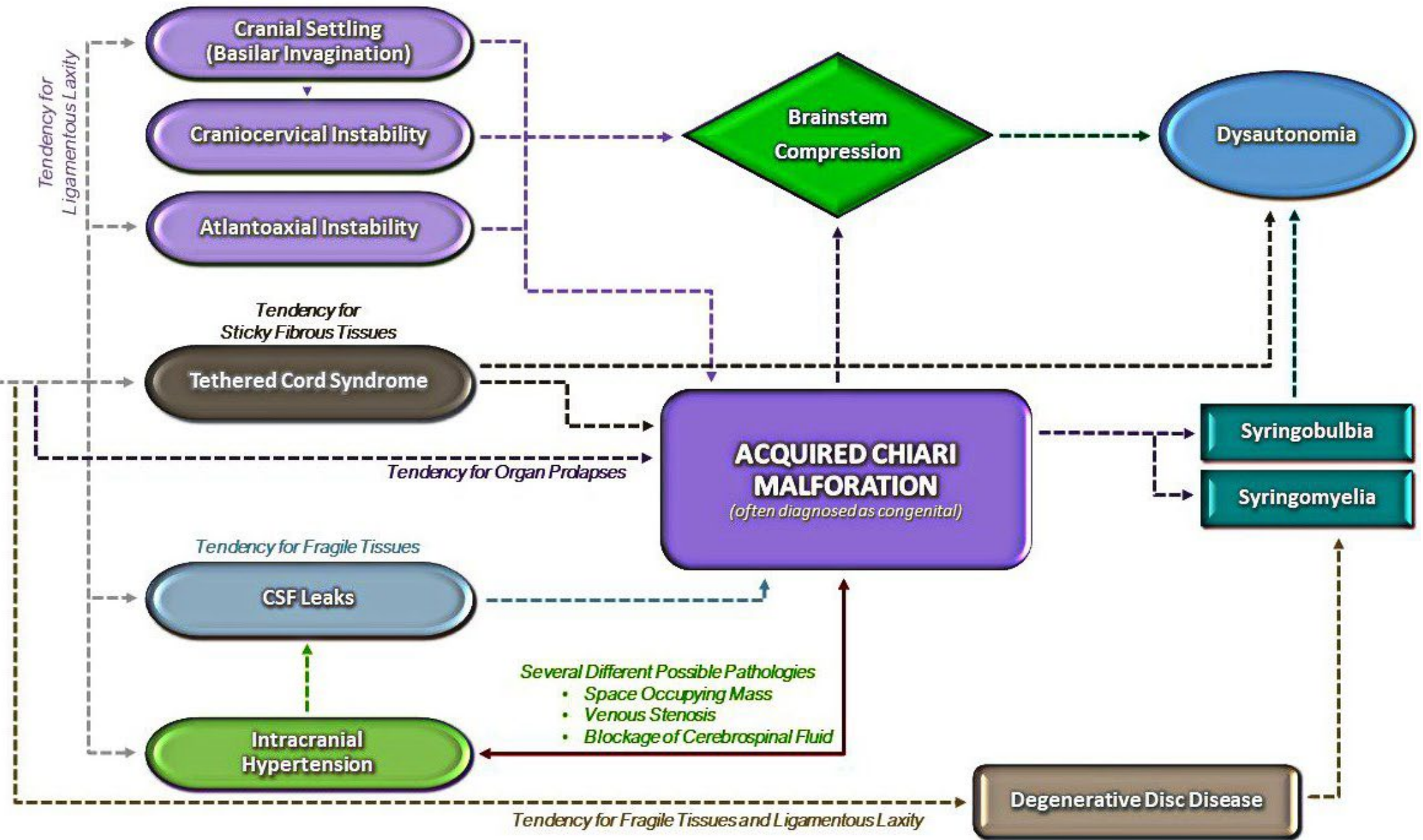
- Craniocervical Instability
- Atlantoaxial Instability
- Basilar Invagination & Cranial Settling

## SPINAL CORD PROBLEM

- Tethered Cord Syndrome
- Occult Tethered Cord
  - Scoliosis



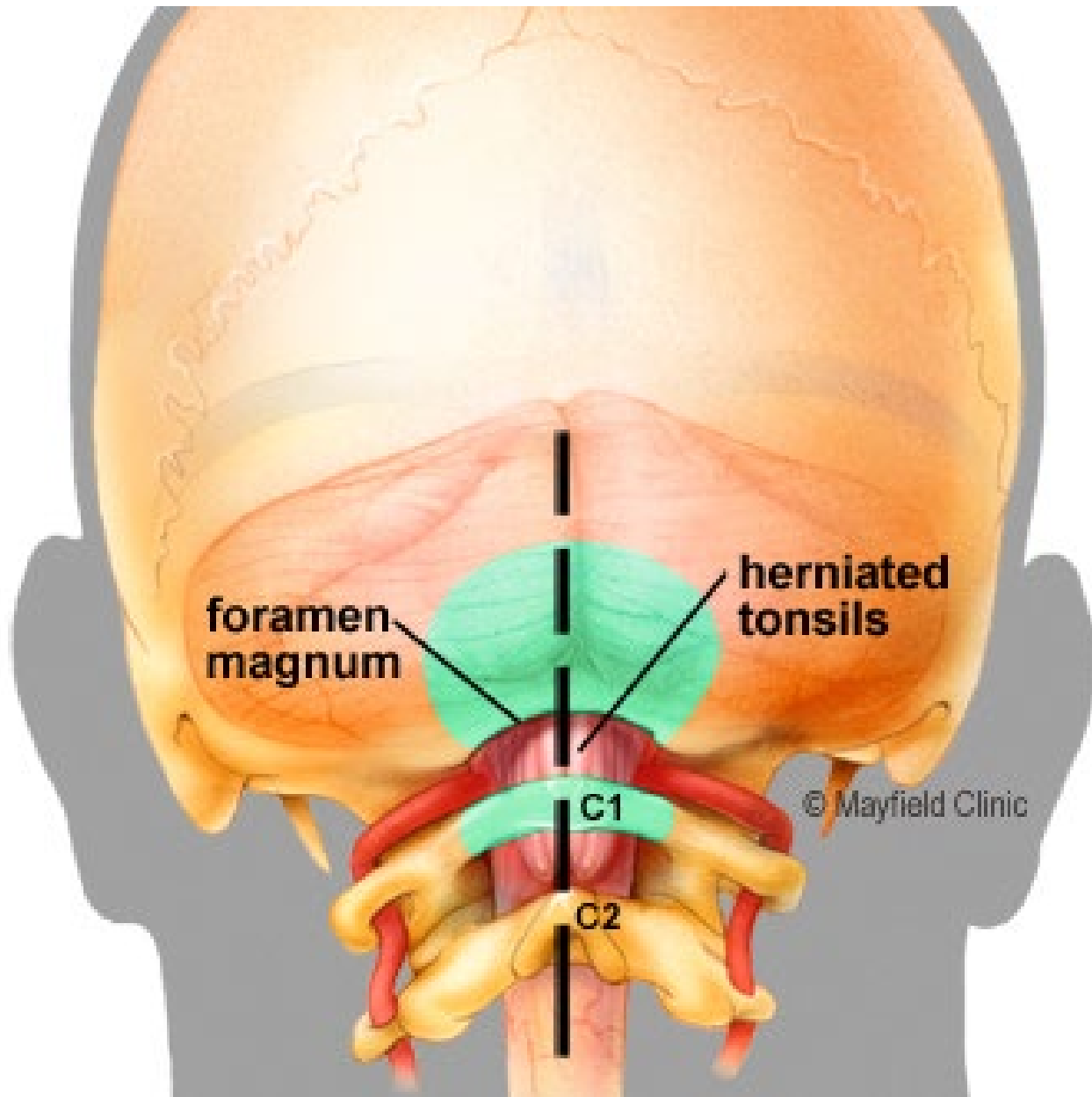
# EHLERS-DANLOS SYNDROME



# EDS Related Pathologies:

- Craniocervical Instability (CCI)
- Basilar Invagination (BI) & Impression
- Tethered Cord (TCS)
- CSF Leak (SIH)
- Intracranial Hypertension (IH/IIH)





## POSTERIOR FOSSA DECOMPRESSION

### DURAPLASTY

- Controversial
- Pseudomeningocele

### CAUTERIZATION VS. RESECTION

- What's the difference?

### LAMINECTOMY

- Posterior Arch of C1
- C1/C2 combination
- Craniocervical Instability

### CRANIECTOMY VS. CRANIOTOMY

- Removal of the only bone opposing the prolapse
- Titanium mesh plates



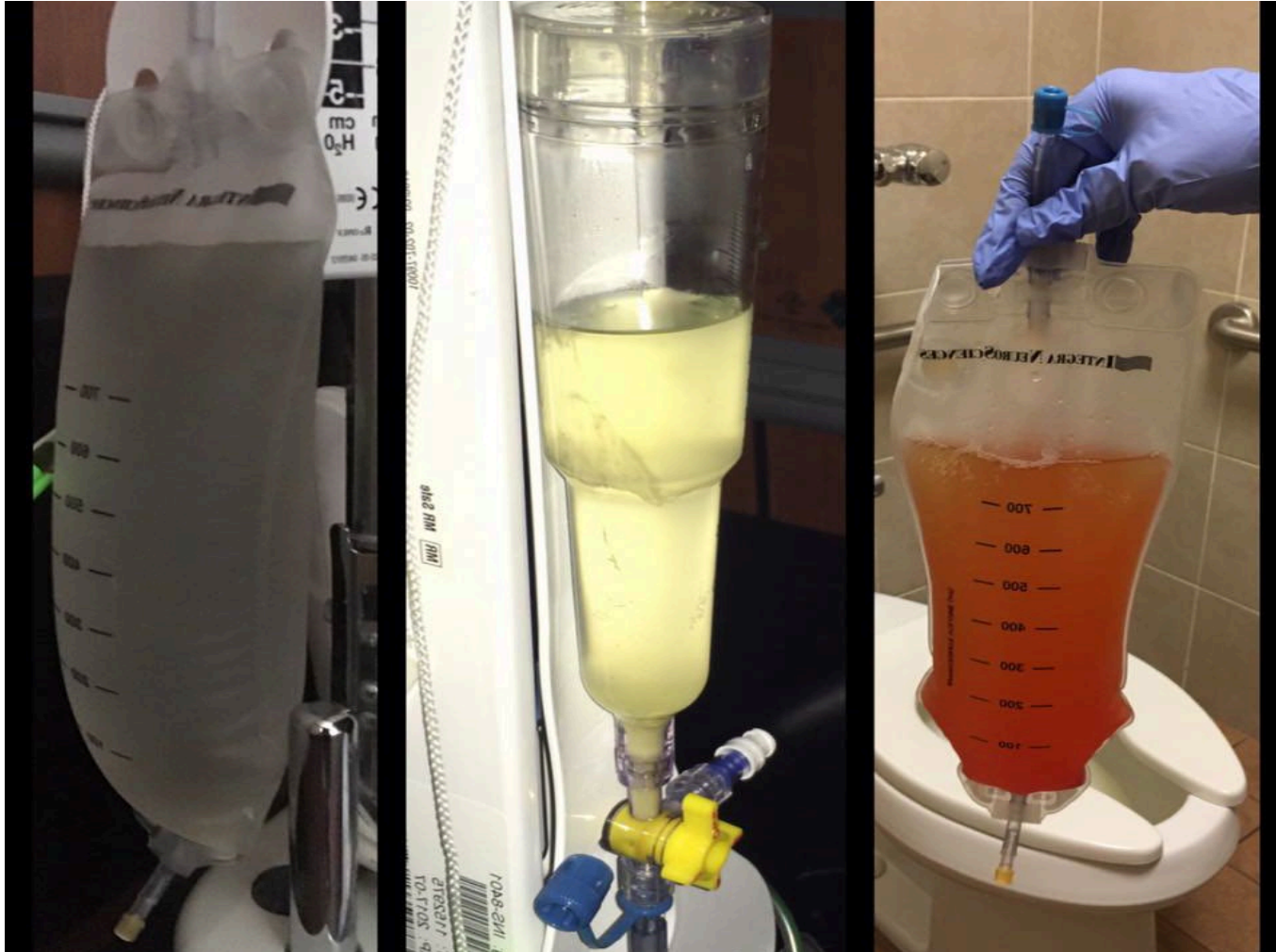
# Chicago Chiari Outcome Scale

Details of the CCOS, as reported in Aliaga et al.

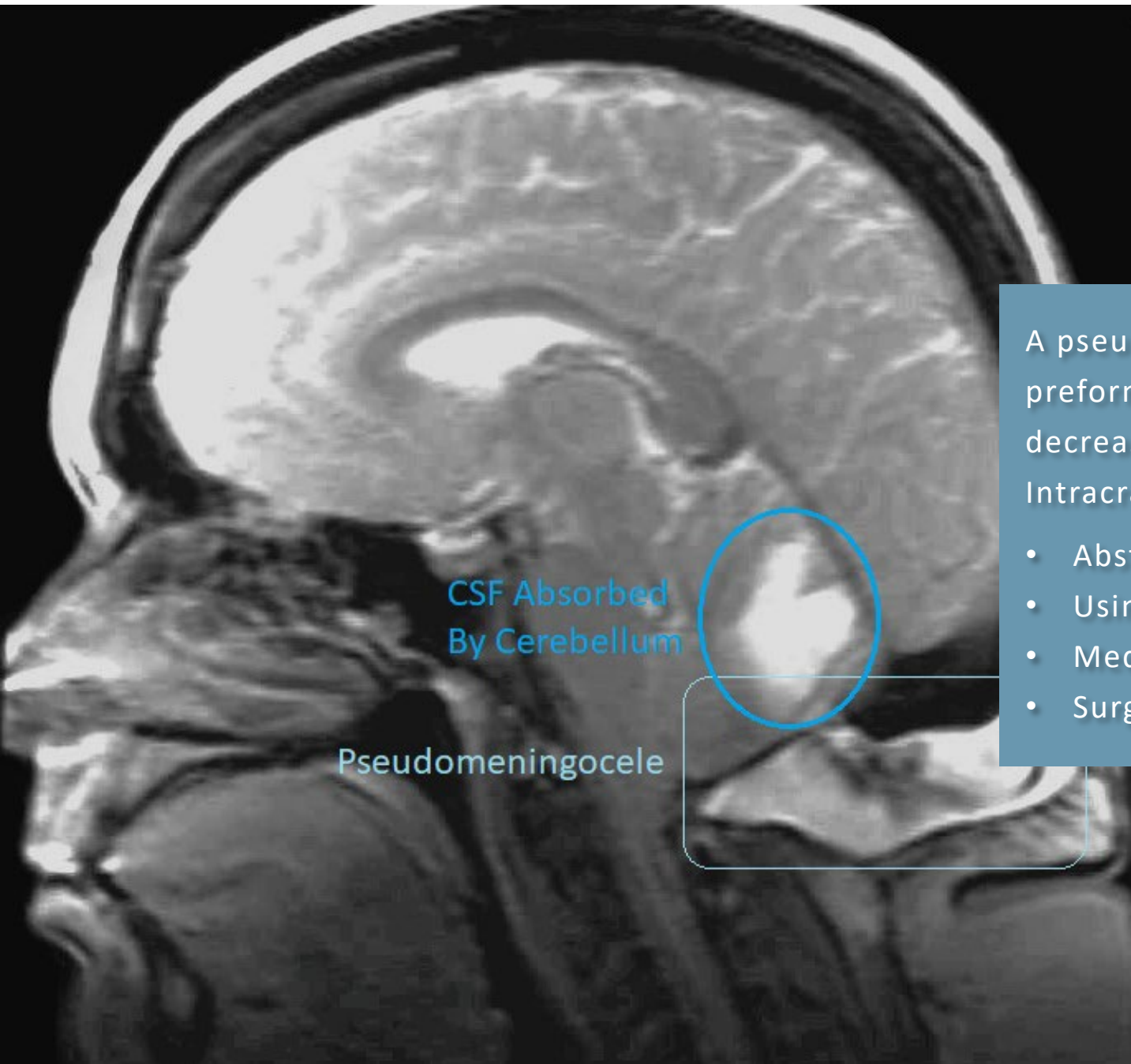
Score	1	2	3	4
<b>Pain</b>	Worse	Unchanged and refractory to medication	Improved or controlled with medication	Resolved
<b>Non-pain</b>	Worse	Unchanged or improved but impaired	Improved and unimpaired	Resolved
<b>Functionality</b>	Unable to attend	Moderate impairment (<50% attendance)	Mild impairment (>50% attendance)	Fully functional
<b>Complication</b>	Persistent complication, poorly controlled	Persistent complication, well-controlled	Transient complication	Uncomplicated course
<b>Location of the cerebellum</b>	Completely slumped and dysfunctional	Slumping and mostly dysfunctional	Slightly slumping and somewhat functional	Buoyant brain and fully functional

*Purple Chiari Bridges addition.*

# COMPLICATIONS OF DECOMPRESSION



- INFECTIONS
  - Surgical site infections
  - Aseptic meningitis
  - Bacterial meningitis
- BLEEDING & ANESTHESIA-RELATED COMPLICATIONS
  - Excessive blood loss
  - Anesthesia risks
- CSF LEAKS
  - Pseudomeningocele
  - Spinal leaks
- BONY REGROWTH
- SCAR TISSUE & ADHESIONS
- OCCIPITAL NEURALGIA
- CRANIOCERVICAL INSTABILITY
- CEREBELLAR SLUMPING
- FAILED DECOMPRESSION



## DECREASE YOUR RISK OF COMPLICATIONS

A pseudomeningocele is caused by DURAPLASTY being performed while your cranial pressure is high. You can decrease the risk of a pseudomeningocele by bringing your Intracranial Pressure (ICP) down BEFORE DECOMPRESSION.

- Abstaining from caffeine
- Using non-hormonal forms of birth control
- Medicinal Options: Topamax, Diamox
- Surgical Options: Shunt, Stent



**INSIST ON  
EXPLORING AND TREATING  
ALL PATHOLOGICAL COMORBIDITIES  
BEFORE DECOMPRESSION!**

**YOU HAVE A RIGHT TO BE A PART OF  
THE DECISIONS REGARDING YOUR  
TREATMENT OPTIONS, AND  
YOU'RE WORTH FIGHTING FOR!**