Brazil: March 23, 2022

Reflections On The Adult Arnold Chiari I Syndrome

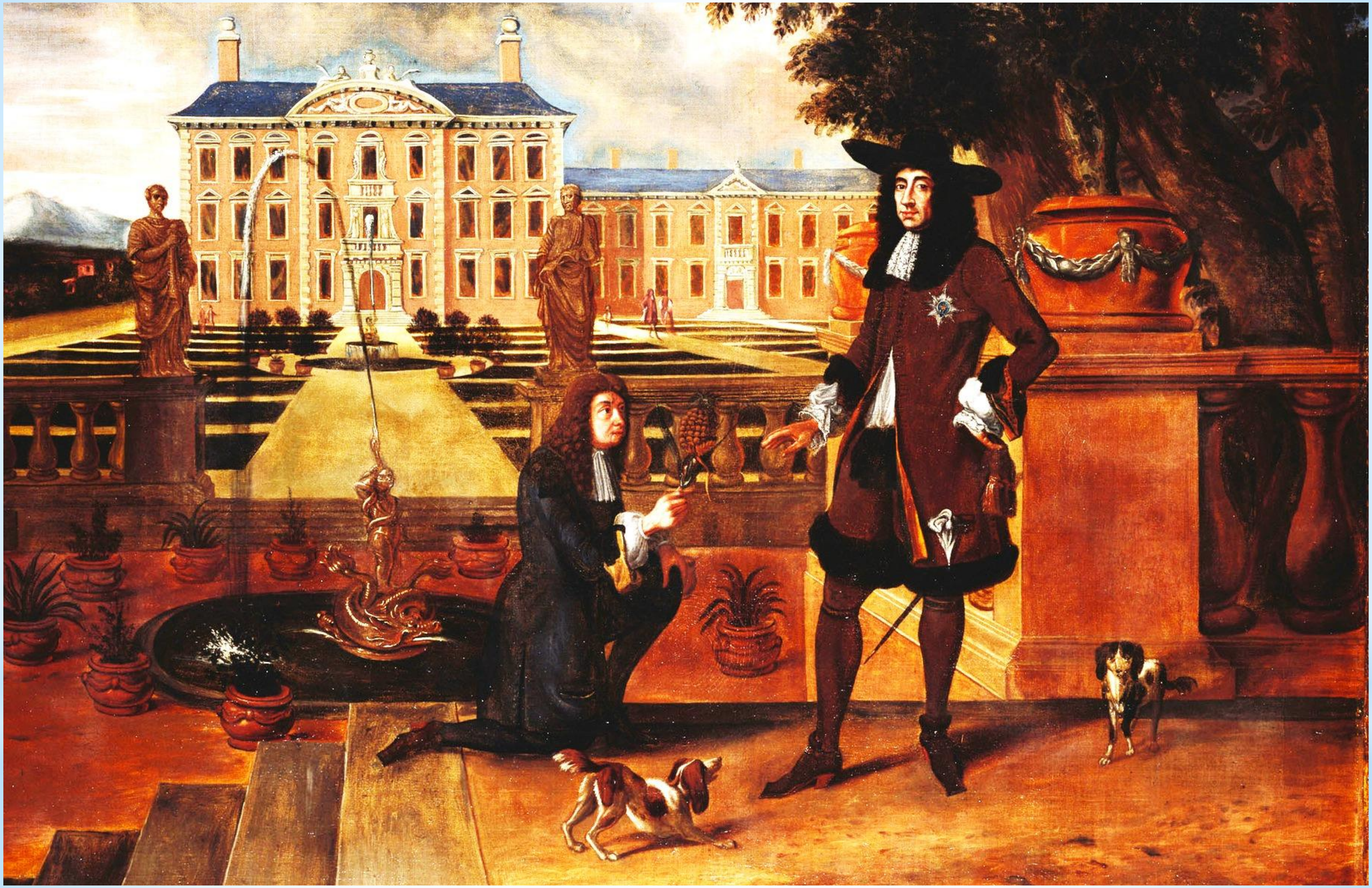
JOE SAM ROBINSON, Jr., MD

JIHYUN MOON

HARRY GROCE

MARCUS EVANS, MD

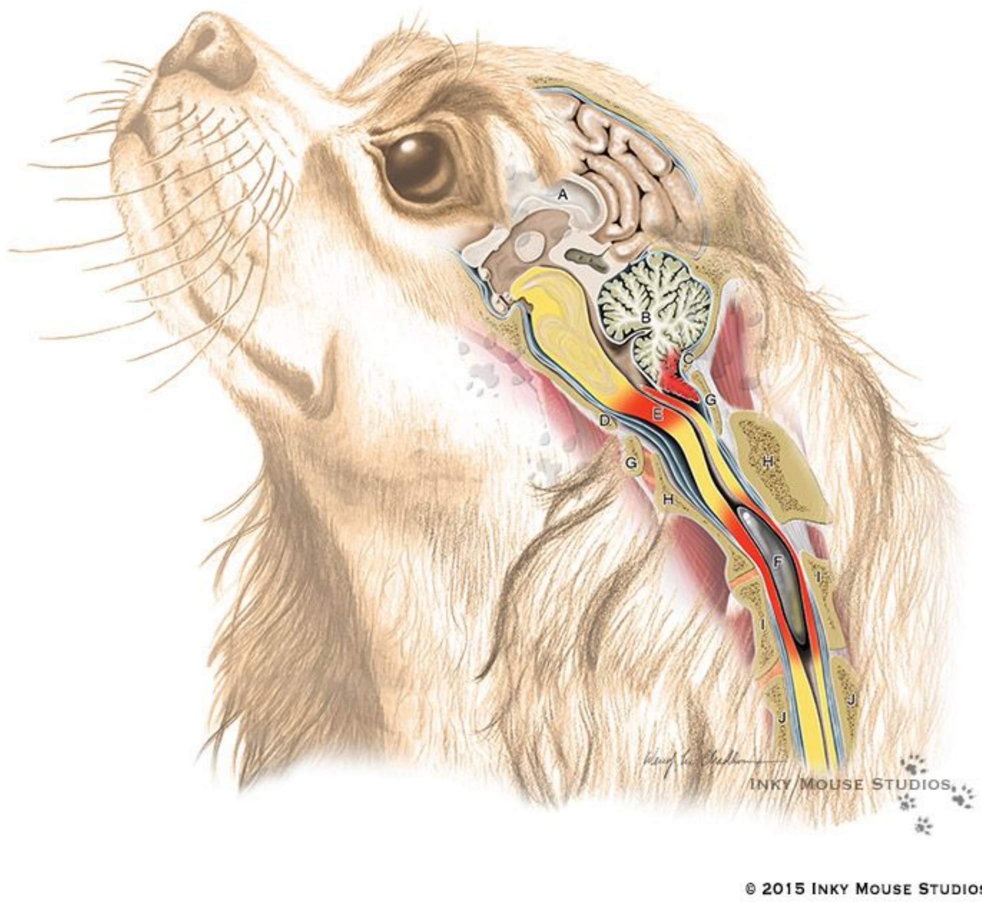
An Example From The Animal Kingdom



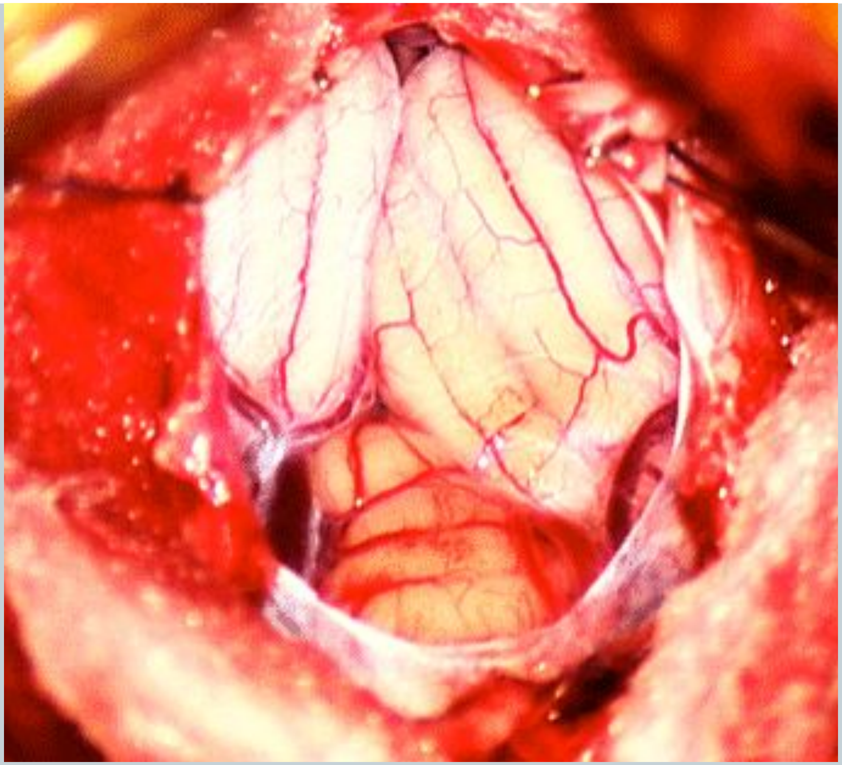
BRITISH SCHOOL, 17TH CENTURY
Charles II Presented with a Pineapple c.1675-80

Chiari in King Charles Cavalier Spaniels

- 90% of King Charles Cavalier Spaniels have Chiari malformation
- 30–70% have syringomyelia (Cerda-Gonzalez, 2015)



Definition



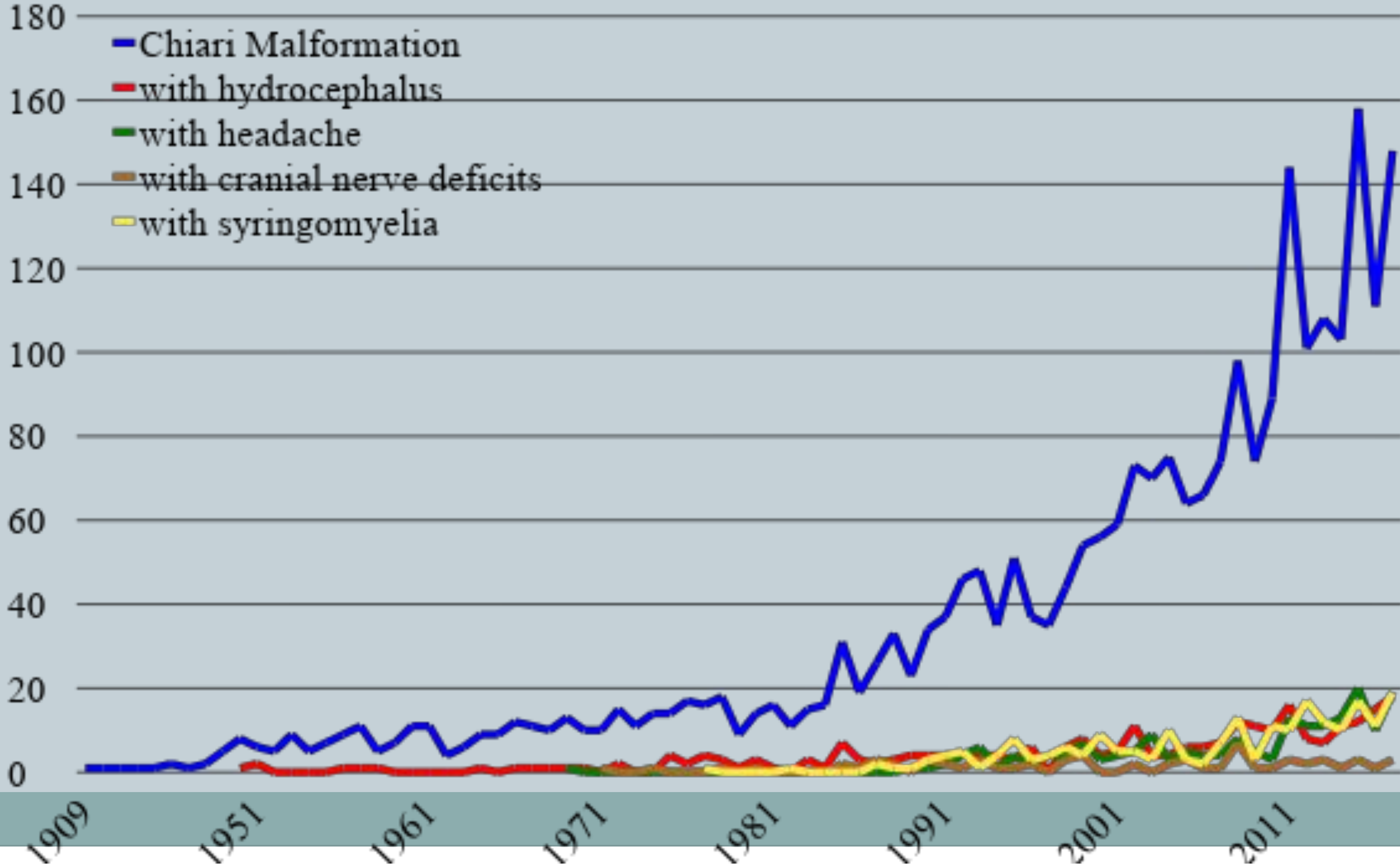
Chiari malformations, types I-IV, refer to a spectrum of congenital hindbrain abnormalities affecting the structural relationships between the cerebellum, brainstem, the upper cervical cord, and the bony cranial base.

OVERVIEW

Type	Age of presen-ta tion	Prevalence	Pathology	Clinical features	Treatment	Complications
Chiari I ✔	Usually adults	Adults – 0.77% Children – 3.6%	Descended cerebellar tonsils	headache	Decompression	⇧
			syringomyelia	Cape-like sensory loss (temperature)	Decompression +/- tonsilar resection or plugging obex	⇧⇧⇧⇧
			hydrocephalus	Papilledema, headache	shunting	⇧⇧
Chiari II	At childhood	1:1000	Caudal herniation of cerebellar vermis, brainstem and IV ventricle. Strongly associated with myelomeningocele	Abundant neurological deficits	FM decompression, myelomeningocel e repair	⇧⇧⇧
Chiari III	At birth	Rare	encephalocele	severe neurological defects, seizures	-	
Chiari IV	At birth	Extremely rare	extreme cerebellar hypoplasia	incompatible with life	-	

- An increasing worldwide inquiry and interest in Chiari related healthcare issues.

Cited Articles Related to Chiari Malformation – 2,613

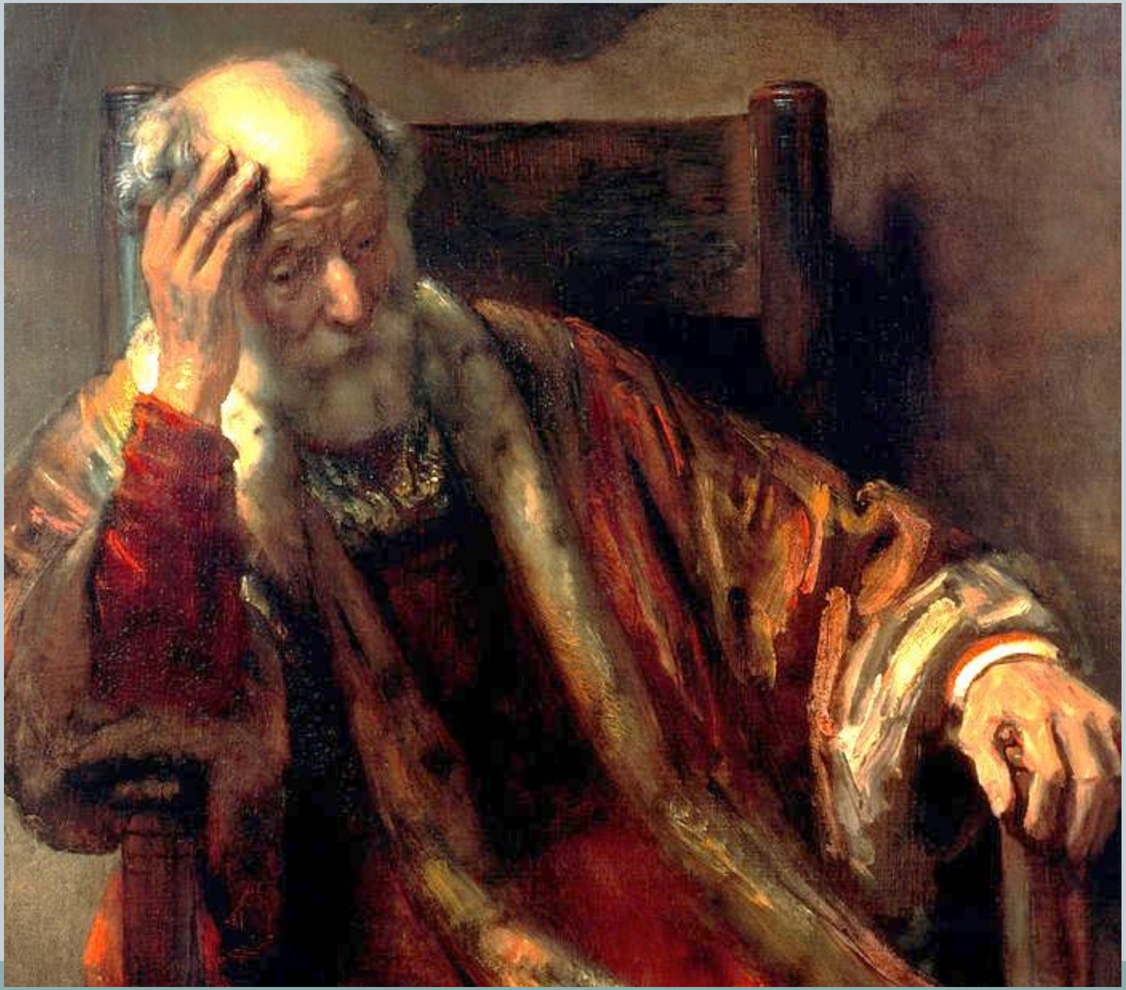


However…

Confusion

- Literature confusion
- Treatment confusion
- Outcome confusion

Why Is There Confusion?



Missing An Ideal Situation

- Have hard sign-symptom complex
- Radiographic studies that show the physiological difficulty
- The causal relationship is clearly established between physiological difficulties and the resulting clinical dysfunction
- Have maneuver to correct the problem
- Positive clinical change
- Measurable findings indicating that physiological difficulties are no longer present

- Unfortunately…

THE CLINICAL SITUATION IS COMPLICATED
(many disease processes)

A. Additional fixed anatomical associations
- small Posterior Fossa
- tethered cord
- congenital hydrocephalus

B. Progressive associations influencing CSF dynamics
- CSF production
- Cerebral atrophy
- Hormones
- Sleep apnea
- Cervical stenosis

(these associations can independently cause symptoms without Chiari and have reciprocal impact and can be transient)

C. TIME

CHIARI I

D. Impact
a. mechanism of impact is controversial
b. may not cause any symptoms

E. Compression
CSF disruption
+ headache
++ syringomyelia
+++ hydrocephalus

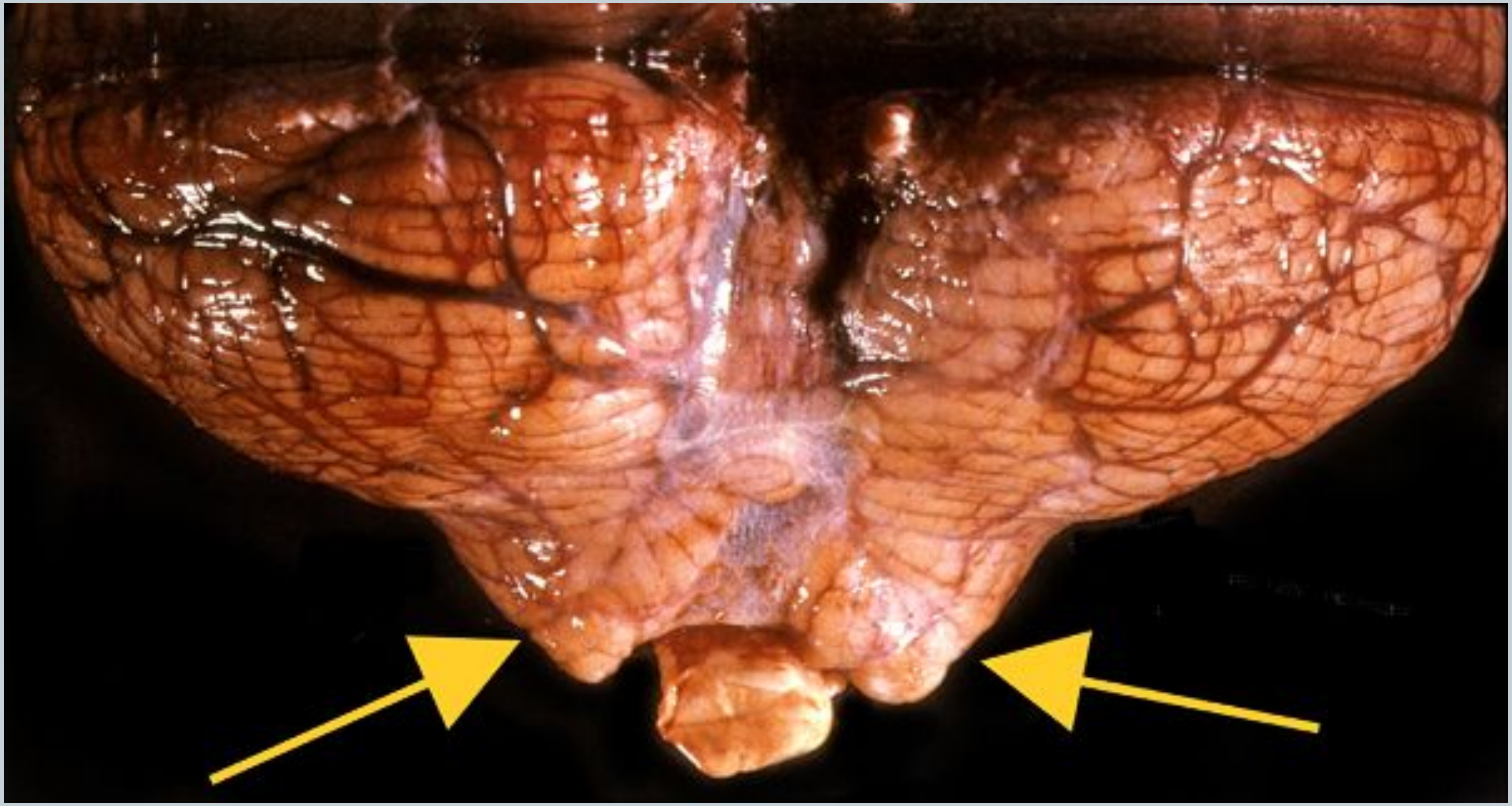
F. Symptoms
- a large host of subjective complaints
- poorly demarcated

G. Multiple causes of symptoms
- Migraine
- Cervical disc disease
- Occipital neuralgia
- Gout
- etc

Anatomical Alterations

- Progressive Change

Classification of the Degree of Hindbrain Abnormality



Anatomical Etiology – Non Exhaustive List

CONGENITAL Chiari

- **Abnormally small posterior fossa. (linkage to chromosomes 9 and 15)**
- **Other causes resulting in lack of development of posterior fossa:**
  - craniosynostosis (especially of the lambdoid suture)
  - hyperostosis (such as craniometaphyseal dysplasia, osteopetrosis, erythroid hyperplasia)
  - X-linked vitamin D-resistant rickets
  - neurofibromatosis type I

- **ACQUIRED Chiari**
  - space occupying lesions
  - head and spine trauma

Posterior Fossa Volume in Chiari I Patients vs. Healthy Controls

- Volumetric calculations for the posterior cranial fossa revealed:
  - a significant reduction of total volume (mean, 13.4 ml)
  - 40% reduction of cerebrospinal fluid volume (mean, 10.8 ml)
  - normal brain and cerebellum volume.

Variable	Control subjects	Chiari I	P
Volumetric measurements			
Total Posterior fossa	137.9±18.7	124.5±5.9	0.001
Brain	111.9±14.9	108.6±21.4	0.369
CSF	26.7±7.3	15.9±7.1	<0.001

Neurosurgery. 1999 May;44(5):1005-17.

Chiari I malformation redefined: clinical and radiographic findings for 364 symptomatic patients.

Milhorat TH[1], Chou MW, Trinidad EM, Kula RW, Mandell M, Wolpert C, Speer MC.

Tonsilar Position Relative to Foramen Magnum at Various Ages

Age (years)	Position
0-9	-1.5
10-19	-0.4
20-29	-1.1
30-39	0
40-49	0.1
50-59	0.2
60-69	0.2
70-79	0.6
80-89	1.3

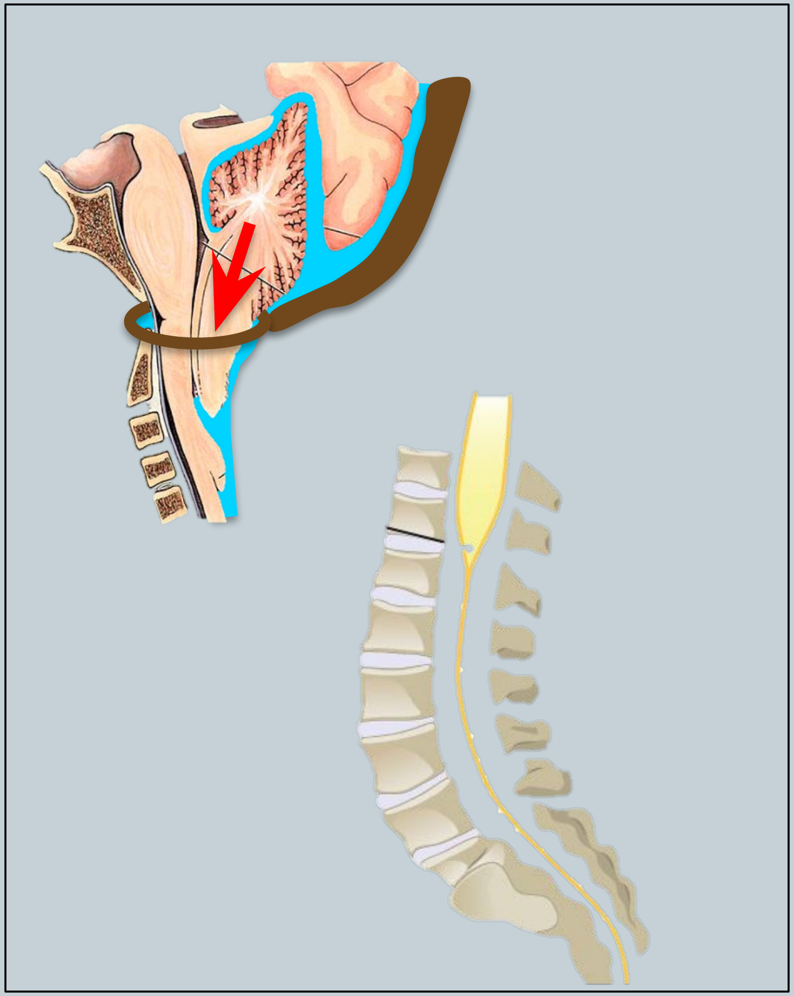
Reference: Mikulis, David & Diaz, O & Egglin, Thomas & Sanchez, R. (1992). Variance of the position of the cerebellar tonsils with age: Preliminary report. Radiology. 183. 725-8.

Tonsilar Descent in Chiari I

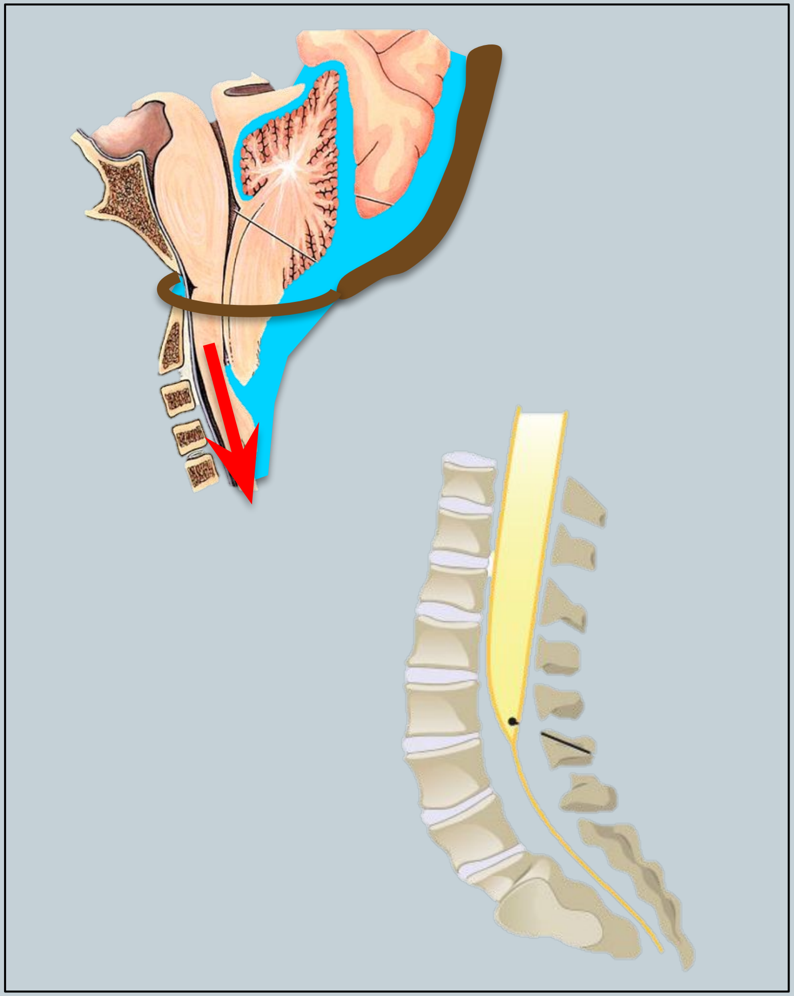
Tonsillar descent below Foramen Magnum	
C1	62%
C2	25%
C3	3%

Reference: S. Paul, Kamal & H. Lye, Richard & Alexander Strang, F & Dutton, John. (1983). Arnold-Chiari malformation. Review of 71 cases. Journal of neurosurgery. 58. 183-7.

Chiari and Tethered Cord



Chiari Without Tethered Cord



Chiari Due to Tethered Cord

Physiology

Causal Interconnections?

Increased Intracranial Pressure ⇄ Chiari Malformation

CSF Abnormalities May Be Transient and May Depend on Secondary Factors

24-hour erratic flow dynamics of CSF

Fluids Barriers CNS. 2014 May 1;11:10. doi: 10.1186/2045-8118-11-10. eCollection 2014.

A new look at cerebrospinal fluid circulation.

Brinker T[1], Stopa E[1], Morrison J[1], Klinge P[1].

Author information

1 Department of Neurosurgery, The Warren Alpert Medical School of Brown University, Rhode Island Hospital, 593 Eddy Street, Providence, RI 02903, USA.

Abstract

According to the traditional understanding of cerebrospinal fluid (CSF) physiology, the majority of CSF is produced by the choroid plexus, circulates through the ventricles, the cisterns, and the subarachnoid space to be absorbed into the blood by the arachnoid villi. This review surveys key developments leading to the traditional concept. Challenging this concept are novel insights utilizing molecular and cellular biology as well as neuroimaging, which indicate that CSF physiology may be much more complex than previously believed. The CSF circulation comprises not only a directed flow of CSF, but in addition a pulsatile to and fro movement throughout the entire brain with local fluid exchange between blood, interstitial fluid, and CSF. Astrocytes, aquaporins, and other membrane transporters are key elements in brain water and CSF homeostasis. A continuous bidirectional fluid exchange at the blood brain barrier produces flow rates, which exceed the choroidal CSF production rate by far. The CSF circulation around blood vessels penetrating from the subarachnoid space into the Virchow Robin spaces provides both a drainage pathway for the clearance of waste molecules from the brain and a site for the interaction of the systemic immune system with that of the brain. Important physiological functions, for example the regeneration of the brain during sleep, may depend on CSF circulation.

KEYWORDS: Aquaporin; Astrocyte; Blood brain barrier; Cerebrospinal fluid circulation; Virchow Robin space

PMID: 24817998 PMCID: PMC4016637 DOI: 10.1186/2045-8118-11-10

CSF Flow – Not a Constant Steady Stream

- CSF is not produced exclusively by choroid plexus
- CSF production changes by age
- CSF production changes throughout the day
- CSF flow alterations maybe transient and positional
- Intracranial pressure is variable

Fluids Barriers CNS. 2014 May 1;11:10. doi: 10.1186/2045-8118-11-10. eCollection 2014.

A new look at cerebrospinal fluid circulation.

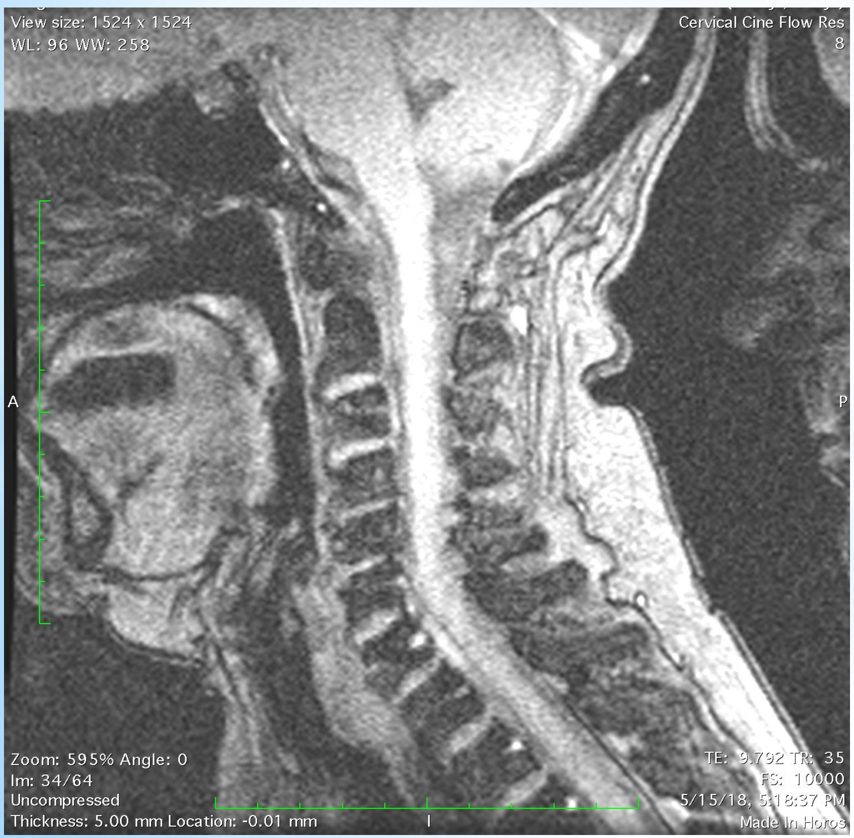
Brinker T[1], Stopa E[1], Morrison J[1], Klinge P[1].

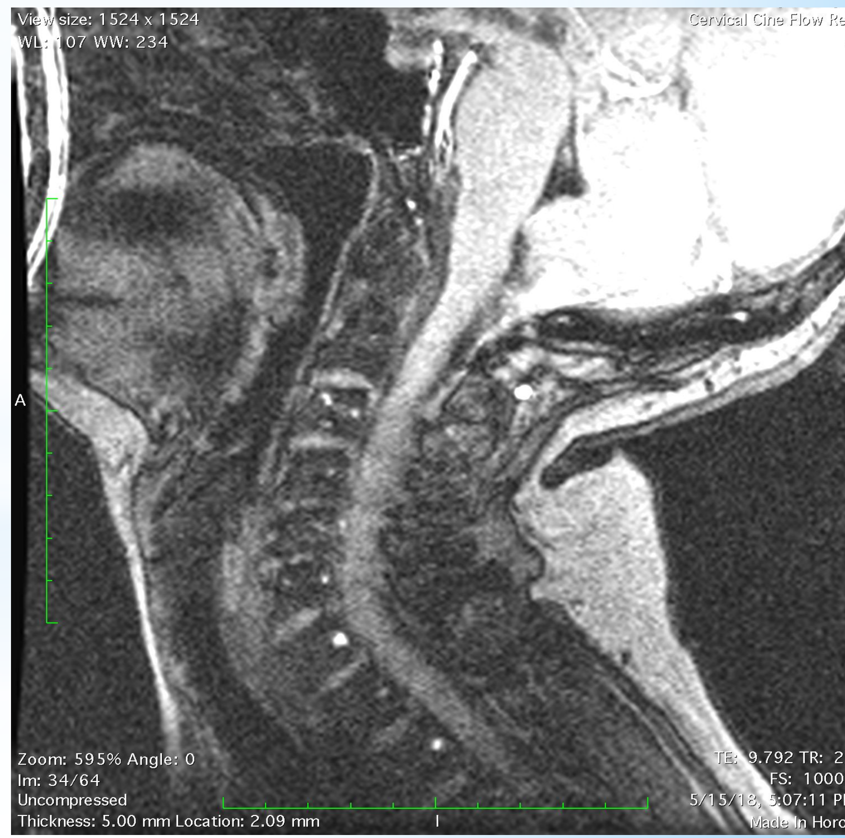
Author information

1 Department of Neurosurgery, The Warren Alpert Medical School of Brown University, Rhode Island Hospital, 593 Eddy Street, Providence, RI 02903, USA.

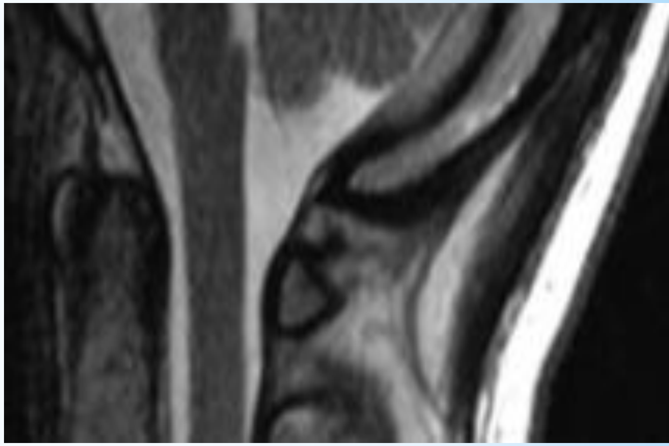
Moreover…

- Our observation is that flexion and extension of the neck is of substantial consequence
- Flexion of the cervical spine improves CSF flow significantly as compared to extension
- Improvement of CSF flow in prone position could be due to flexion of the neck.





In Our Recent Limited Study:

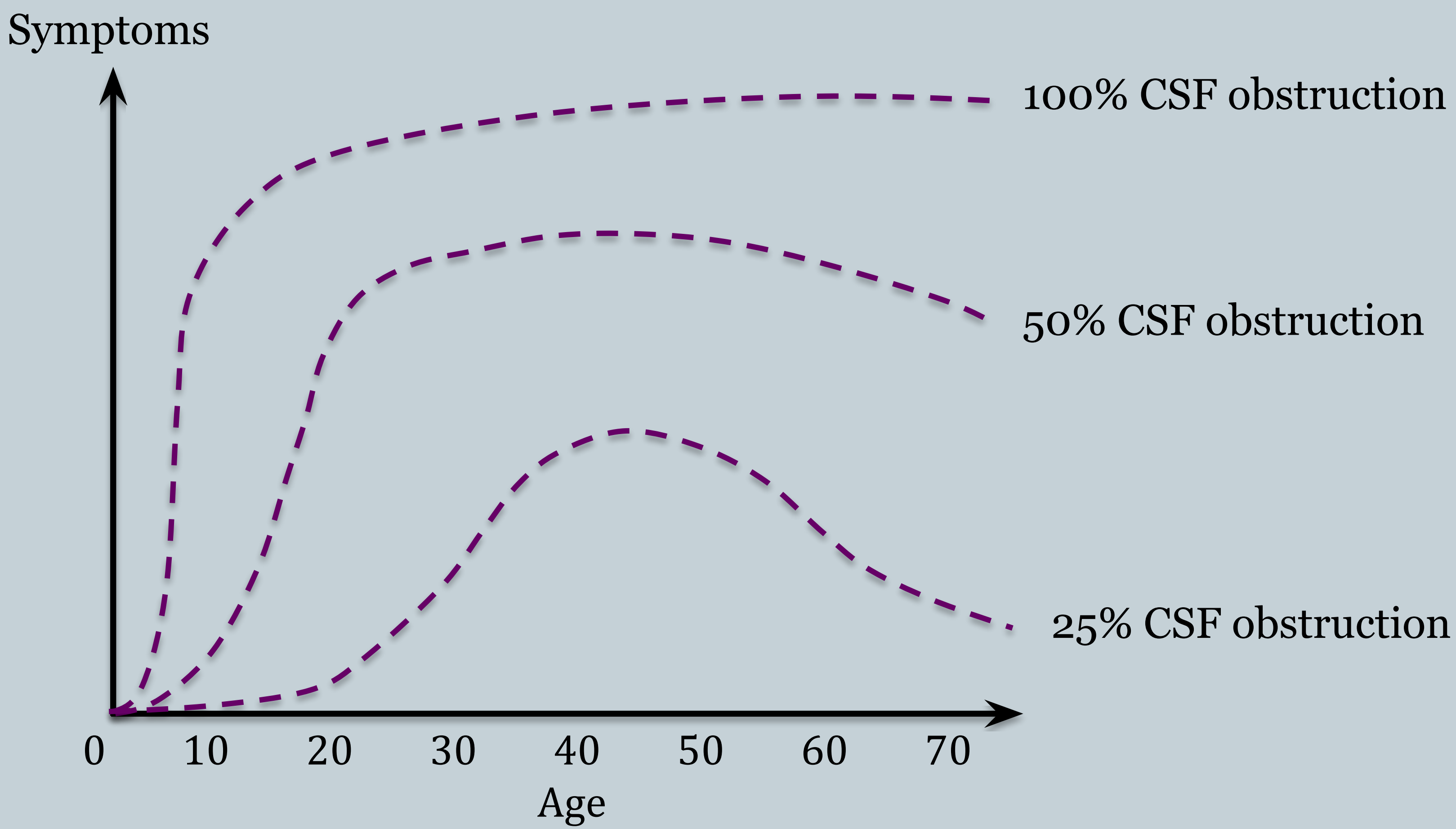


* 36 patients, 648 MRI measurements of CSF flow restriction (flexion, extension, and neutral position)
  * Foramen Magnum, Odontoid Base, and Base of C2
* Demonstrated episodic, substantial flow alteration in differing geographical areas, both in extension and flexion, highlighting the advantages of a **neutral** neck position

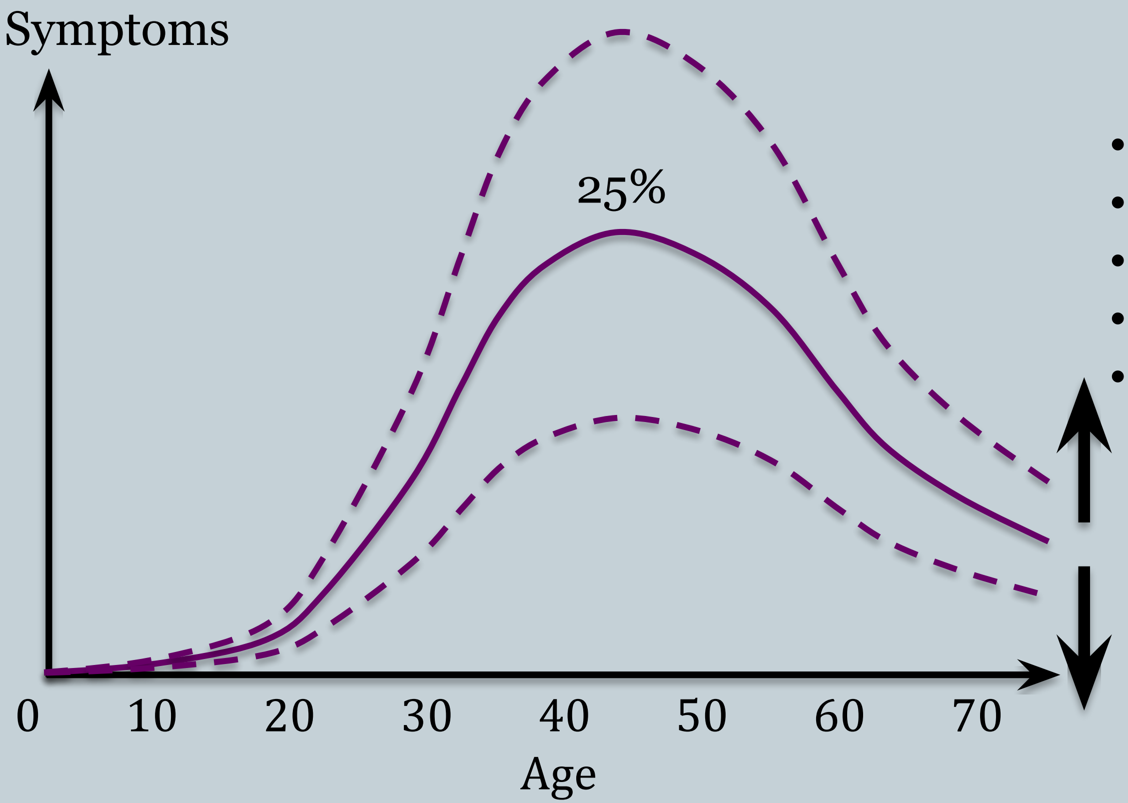
Average / S.D	Neutral		Flexion		Extension		Flexion Change		Extension Change	
	Ant	Post	Ant	Post	Ant	Post	Ant	Post	Ant	Post
Foramen Magnum	5.27 (Sd. 1.51)	14.54 (Sd. 1.92)	5.1 (Sd. 1.54)	14.38 (Sd. 2.29)	5.04 (Sd. 1.49)	14.62 (Sd. 2.57)	Decrease 0.16mm	Decreased 0.15mm	Decreased 0.23mm	Increased 0.16mm
Odontoid Base	2.93 (Sd. 1.12)	3.62 (Sd. 1.37)	3.03 (Sd. 1.13)	3.39 (Sd. 1.38)	2.14 (Sd. 1.06)	3.64 (Sd. 1.41)	Increased 0.1mm	Decreased 0.22mm	Decreased 0.78mm	Increased 0.03mm
Base of C2	2.76 (Sd. 1.04)	1.94 (Sd. 0.88)	3.38 (Sd. 1.06)	1.55 (Sd. 0.90)	1.88 (Sd. 0.97)	2.09 (Sd. 0.94)	Increased 0.62mm	Decreased 0.38mm	Decreased 0.38mm	Increased 0.17mm

Thus…

Compromised CSF circulation

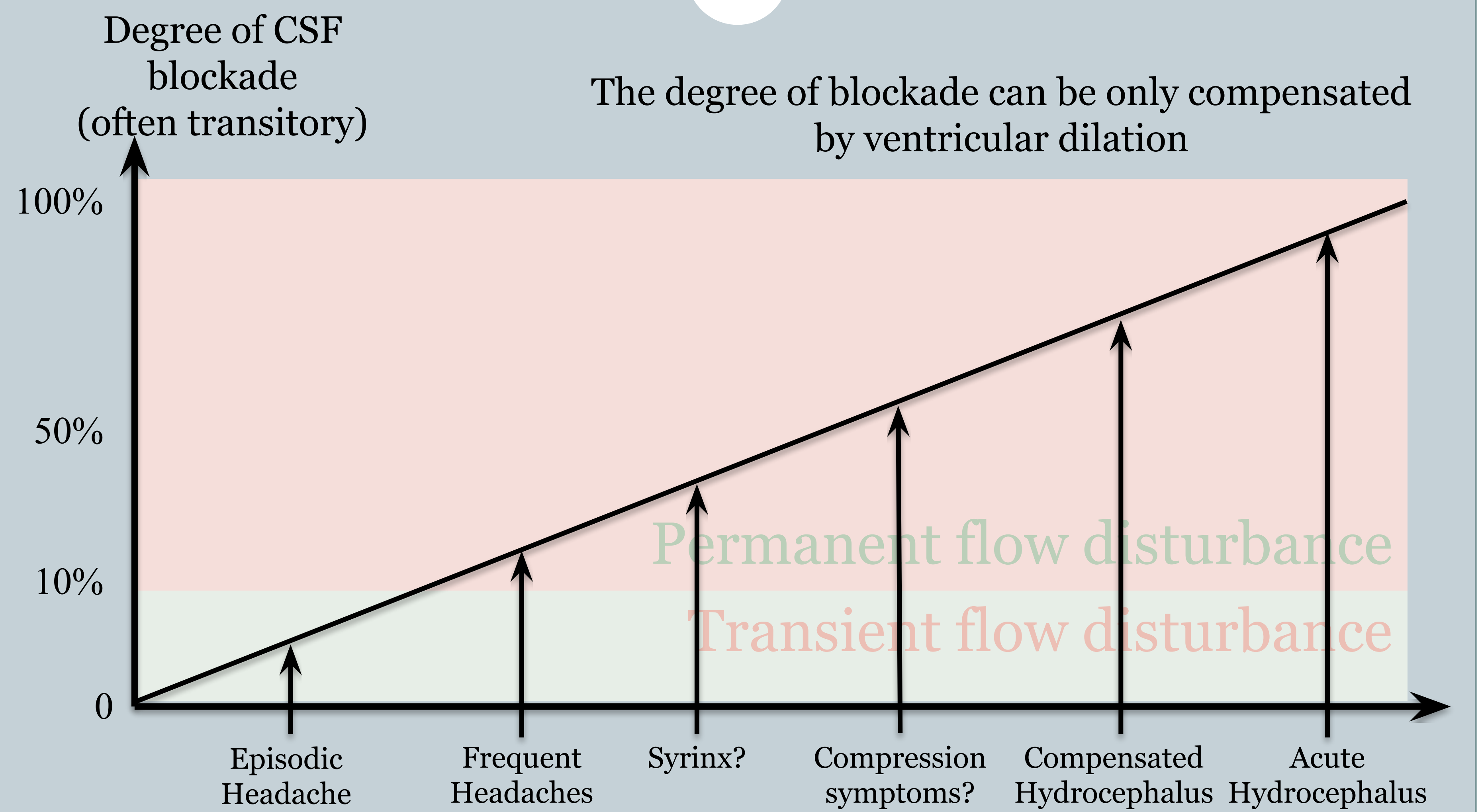


Multiple Causal Factors



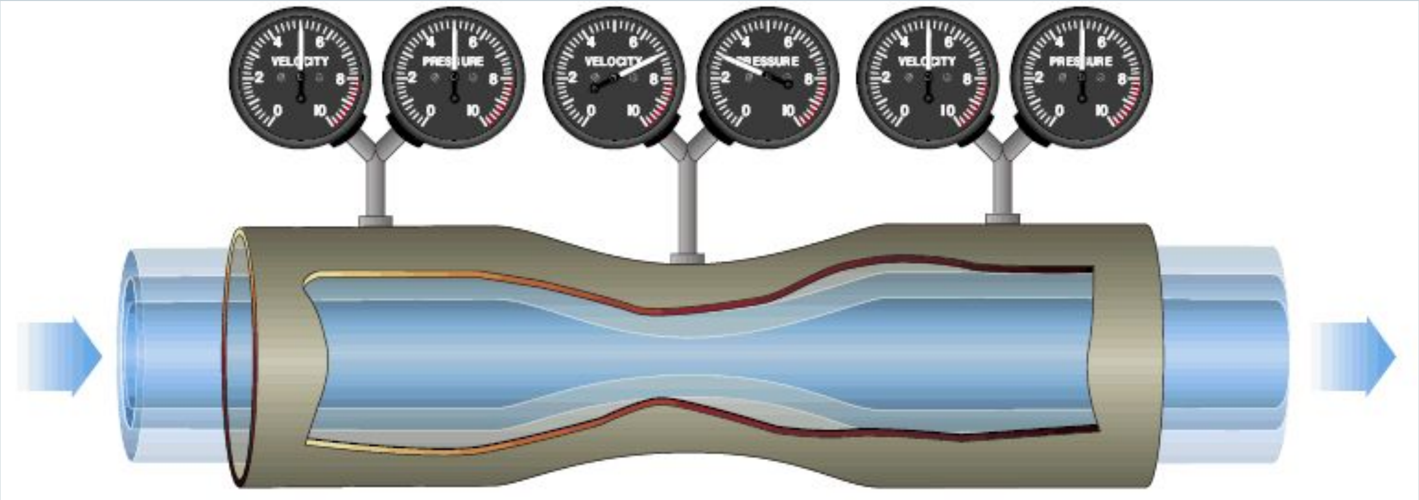
- Spine changes (stenosis)
- Sleep apnea
- Elevated blood pressure
- Improper head position
- Neuronal swelling (hormonal impact)

- Ageing
- Decreased CSF production
- Cerebellar atrophy
- Hormonal effects (Menopause)
- Proper head position
- Blood pressure correction



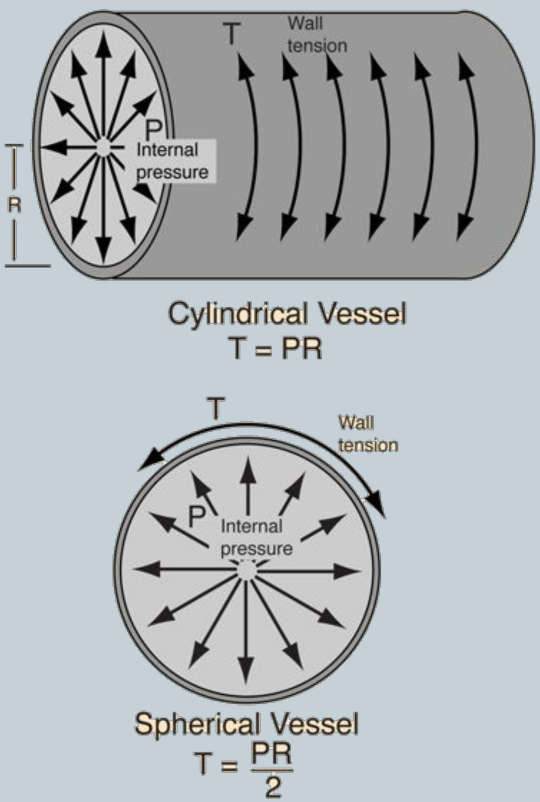
Degree of CSF blockade (often transitory)
The degree of blockade can be only compensated by ventricular dilation
100%
50%
10%
0
Permanent flow disturbance
Transient flow disturbance
Episodic Headache
Frequent Headaches
Syrinx?
Compression symptoms?
Compensated Hydrocephalus
Acute Hydrocephalus

Fluid Mechanics



Fluid Mechanics

LaPlace's Law

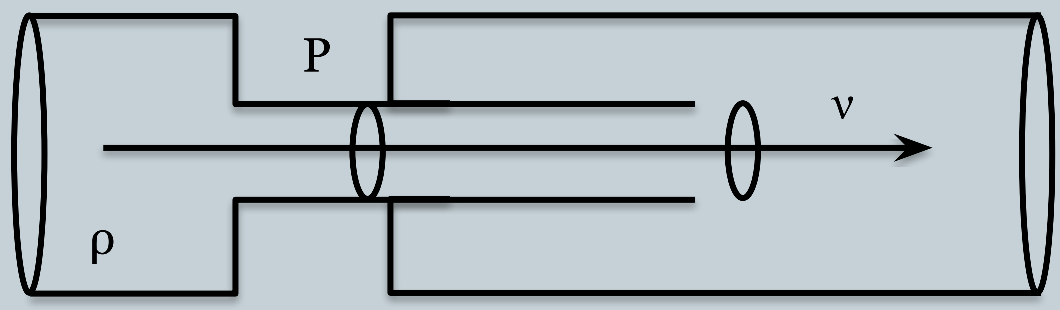


Bernoulli's principle

$$\frac{v^2}{2} + gz + \frac{p}{\rho} = \text{constant} \quad (A)$$

where:

v is the fluid flow speed at a point on a streamline,
g is the acceleration due to gravity,
z is the elevation of the point above a reference plane, with the positive z-direction pointing upward – so in the direction opposite to the gravitational acceleration,
p is the pressure at the chosen point, and
ρ is the density of the fluid at all points in the fluid.

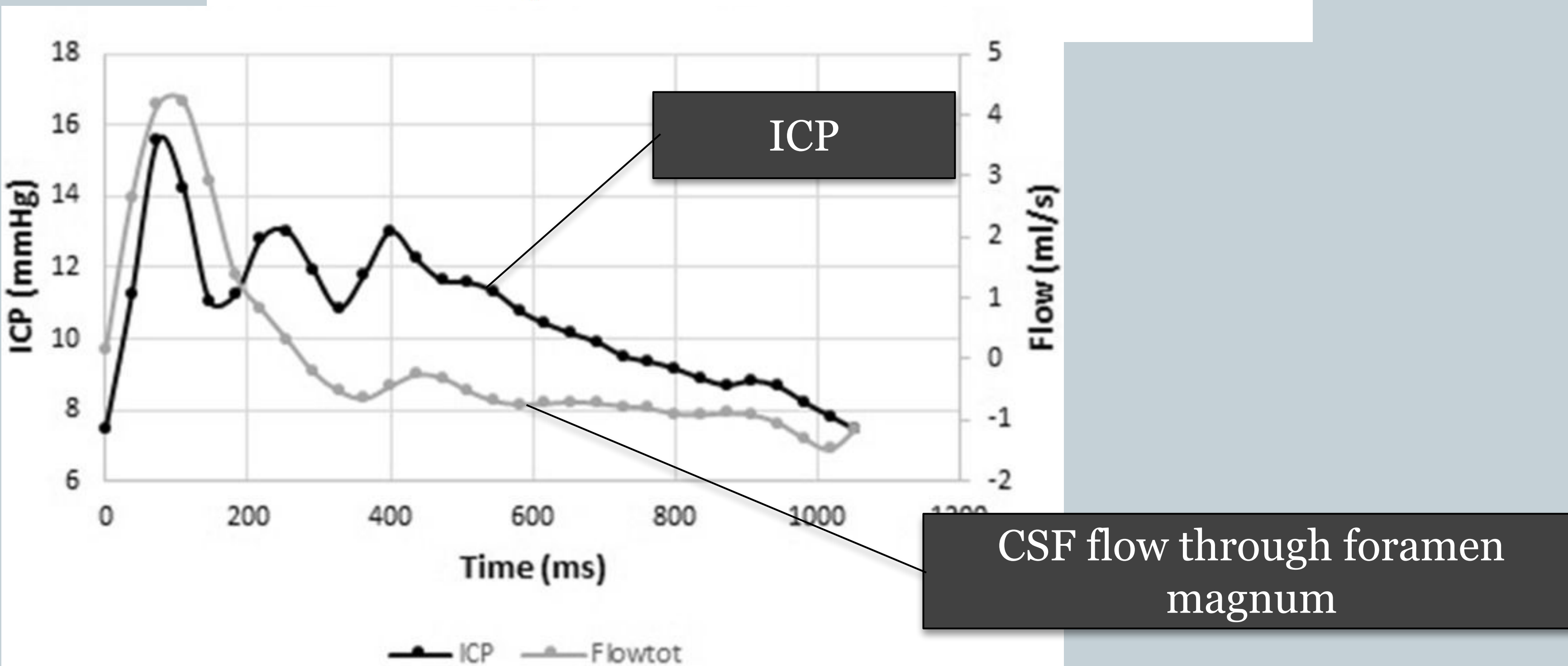


$\nu^2/2+P=const$ $\quad$ $T=Pr$ $\quad$ $\nu^2/2+T/r=const$

$$Q_{(flow\ rate)} = \nu_{(velocity)} \cdot S_{(surface)}$$

Cine MRI Significance

$$\Delta\, ICV(\tau) = \int_{\tau_0}^{\tau} (CBFa\,(t) - CBF\nu\,(t) - CSFF\,(t))\, dt$$



Acta Neurochir (Wien). 2018 Feb;160(2):219-224. doi: 10.1007/s00701-017-3435-2. Epub 2017 Dec 22.

ICP curve morphology and intracranial flow-volume changes: a simultaneous ICP and cine phase contrast MRI study in humans.

Unnerbäck M[1], Ottesen JT[2], Reinstrup P[3].

CSF Flow Velocity

Peak CSF velocity in FM

- Peak velocity of CSF is higher in Chiari patients than in healthy individuals (p=0.01)
  - Systolic - 1.8 to 4.8 cm/s vs. 1.2 to 3.3 cm/s
  - Diastolic - 2.5 to 5.3 cm/s vs. 1.6 to 4.5 cm/s vs.
  - Phase-Contrast MRI studies used to calculate
- Craniocervical decompression in patients with Chiari I malformations decreases peak CSF velocities in the foramen magnum.

AJNR Am J Neuroradiol. 2003 Feb;24(2):169-76.

Peak systolic and diastolic CSF velocity in the foramen magnum in adult patients with Chiari I malformations and in normal control participants.

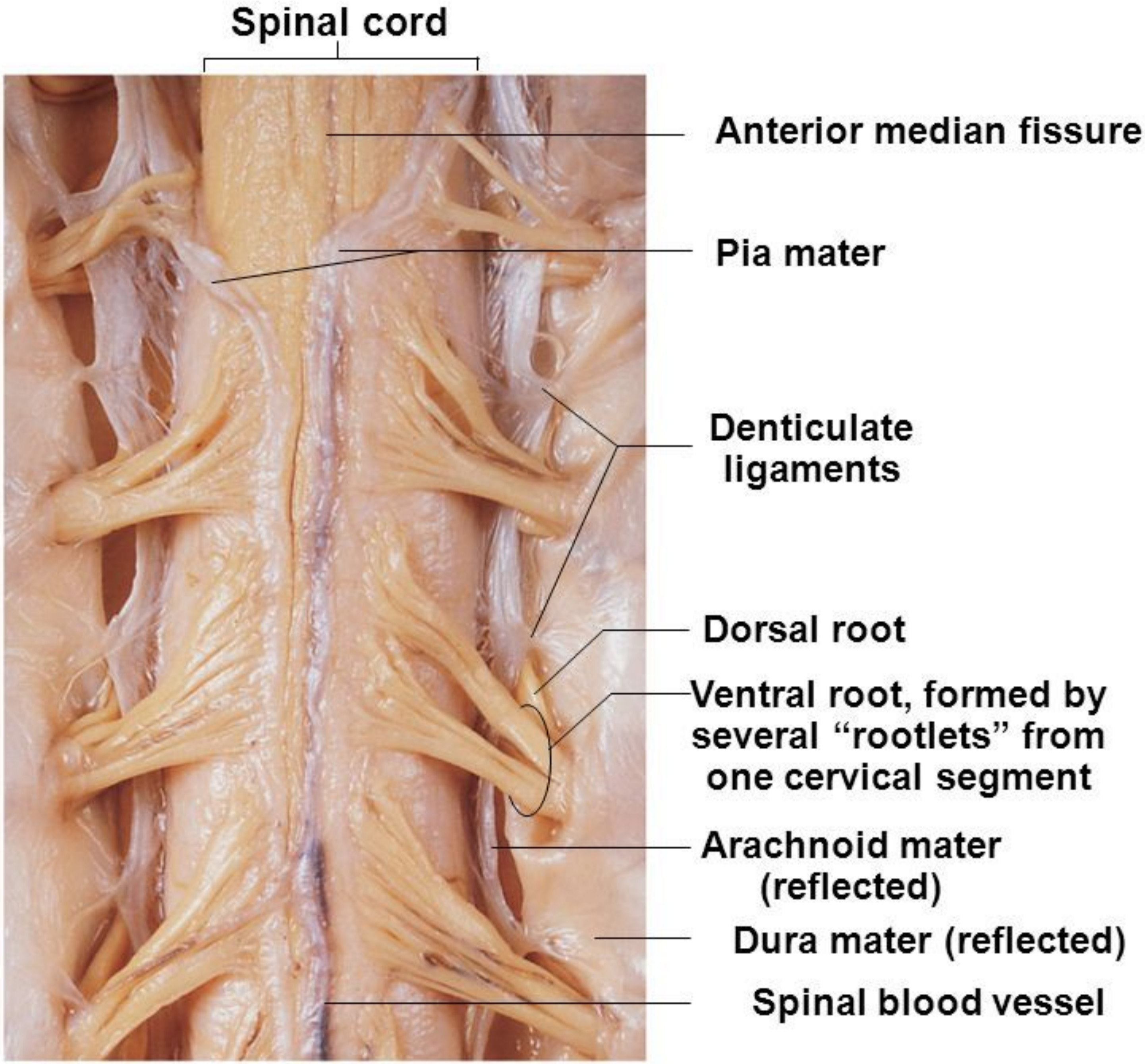
Haughton VM[1], Korosec FR, Medow JE, Dolar MT, Iskandar BJ.

Poiseuille's Law

- The flow (Q) of fluid through a tube is related to a number of factors:
  - the viscosity (η) of the fluid,
  - the pressure gradient across the tubing (P),
  - and the length (L) and diameter (r) of the tubing.
- Doubling the diameter of a tube increases the flow rate by 16 fold (r4).
- Flow is inversely proportional to the viscosity of the fluid. Increasing viscosity decreases flow through a pipe.

Q	Flow rate
P	Pressure
r	Radius
η	Fluid viscosity
l	Length of tubing

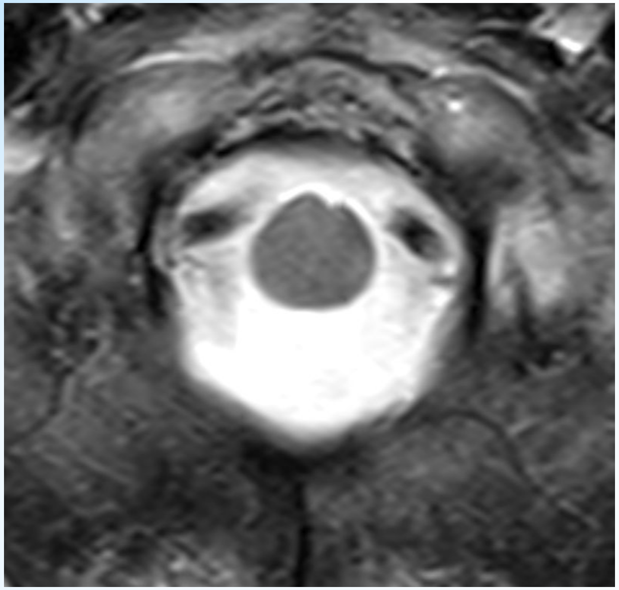
$$Q = \frac{\pi P r^4}{8 \eta l}$$

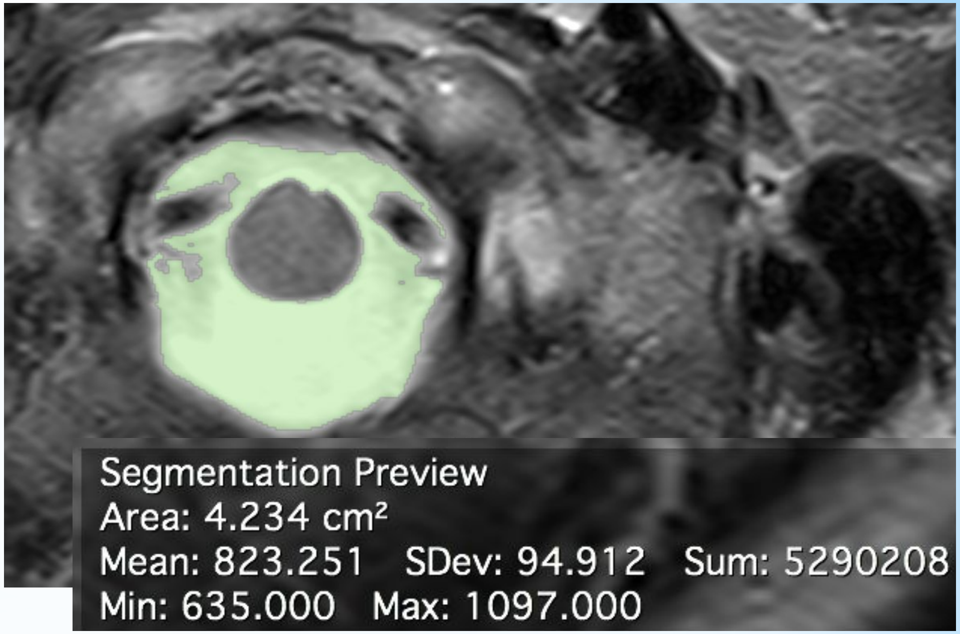


Spinal cord
Anterior median fissure
Pia mater
Denticulate ligaments
Dorsal root
Ventral root, formed by several “rootlets” from one cervical segment
Arachnoid mater (reflected)
Dura mater (reflected)
Spinal blood vessel

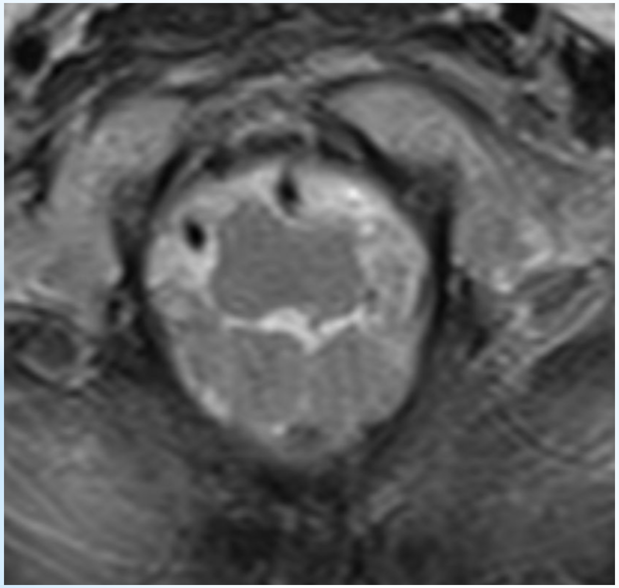
Two-Way Highway

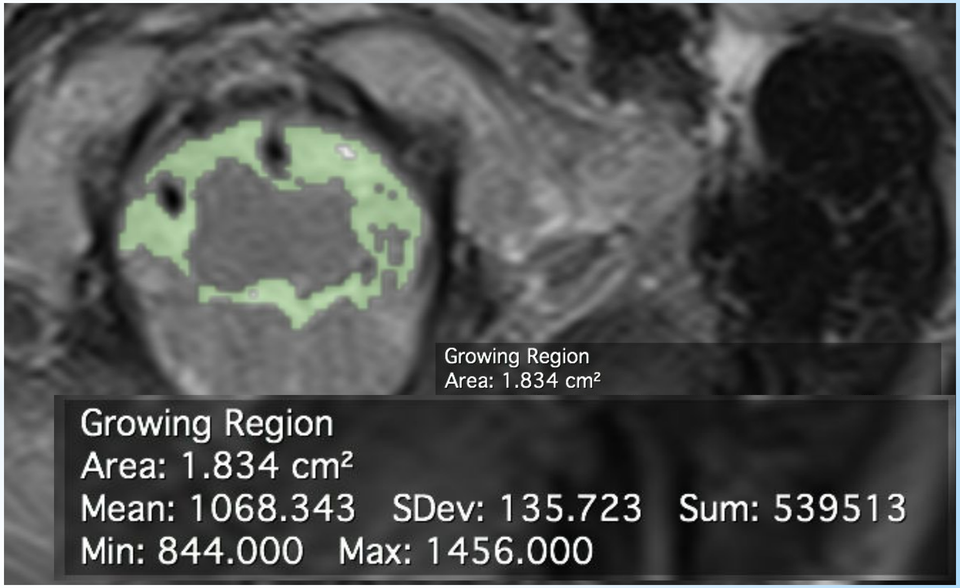
Normal





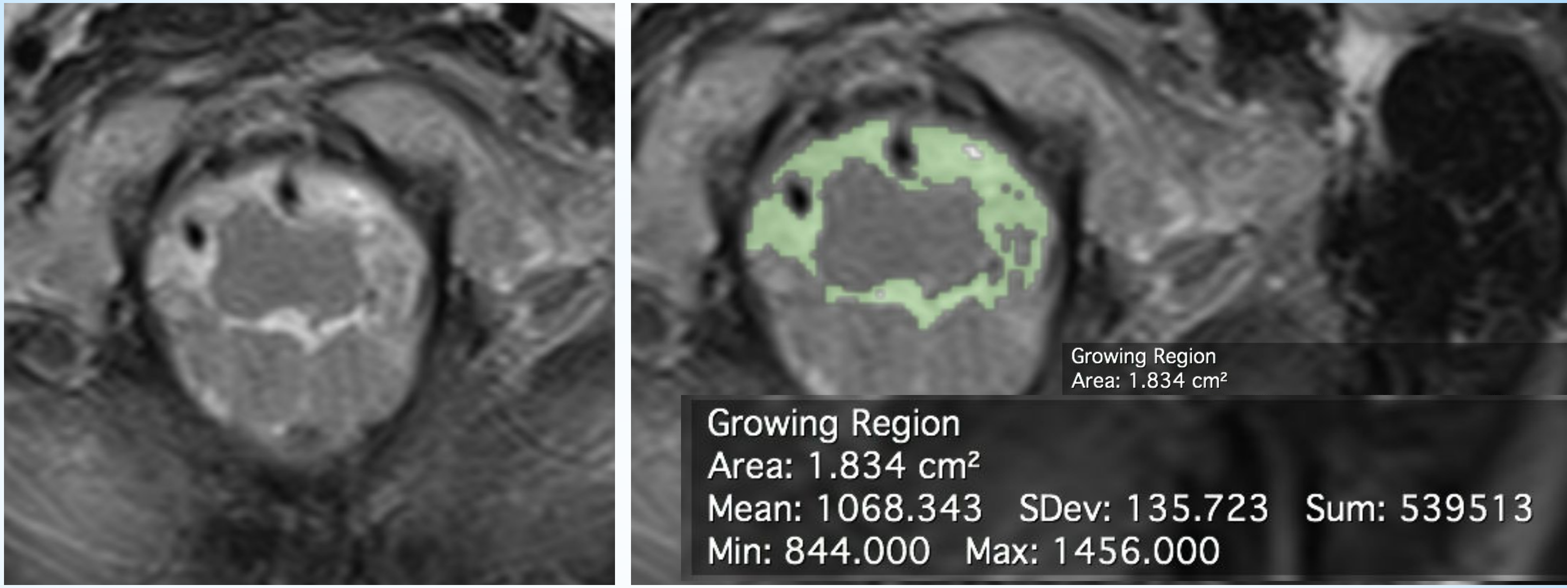
Chiari I (episodic headache)



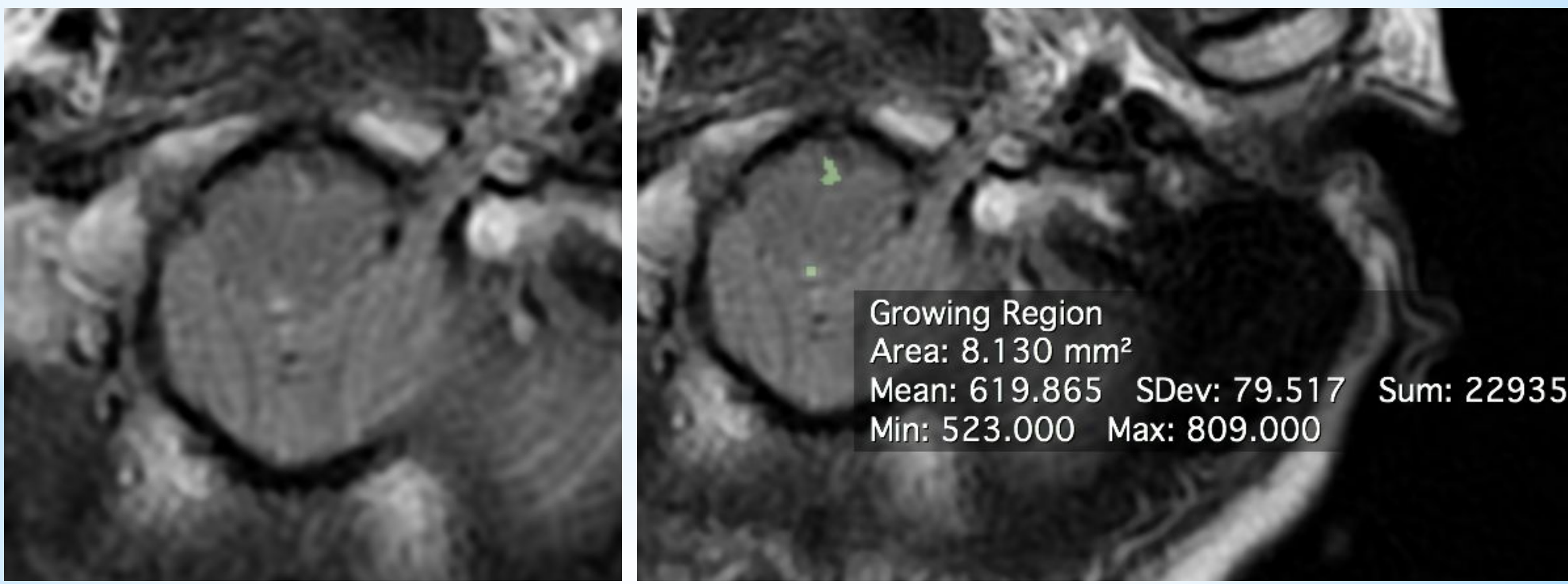


$$Q_{(\text{flow rate})} = \nu_{(\text{velocity})} \cdot S_{(\text{surface})}$$

Chiari I (episodic headache)



Chiari I (progressive, unremitting headaches)



$$Q_{(\text{flow rate})} = \nu_{(\text{velocity})} \cdot S_{(\text{surface})}$$

Sign-Symptom Complex

Sign-Symptom Complexes Associated With Chiari Referable to Pressure

Direct Hindbrain Pressure

- ✓ Paralysis due to pressure at the cervico-medullary junction: may progress in a so-called "clockwise" fashion, affecting the right arm, then the right leg, then the left leg, and finally the left arm; or the opposite way around. facial pain, tinnitus, Lhermitte's sign, bulbar symptoms (impaired gag reflex, dysphagia, bulbar palsy)

Disruption of CSF pathways (indirect pressure)

- ✓ Headache
  - Associated with cough and sneezing, exacerbated by Valsalva, occipital location, aggravated by Valsalva maneuvers
- ✓ Chronic increase of intracranial pressure
  - Nausea, vomiting, papilledema on fundoscopic exam, pupillary dilation, dysautonomia: tachycardia , syncope, polydipsia, chronic fatigue, Vertigo, Nausea, Nystagmus
- ✓ Syringomyelia
  - Muscle weakness, restless leg syndrome, impaired coordination, sensory loss in cape-like distribution (especially temperature), tingling and numbness

Natural History of Sign-Symptom Complexes in Chiari 1 Without Treatment – No Cine MRIs Done

Direct Hindbrain Pressure

- ✓ Paresthesia (2 years follow-up) (Chavez et al. 2014)
  - improved or remained unchanged in 75.4%
- ✓ Ataxia
  - improved or remained unchanged in 82.1%

Disruption of CSF pathways

- ✓ Nausea
  - (2 years follow-up) - 88.9% improved and 11.1% remained unchanged (Chavez et al. 2014)
- ✓ Headache (5 years follow-up)
  - 37% improvement, 51.9% no change, 11.1% worsening of headaches (Killeen et al. 2015)
- ✓ Syringomyelia
  - 47% decrease, 41% unchanged, 12% progress (Singhal et al., 2011)

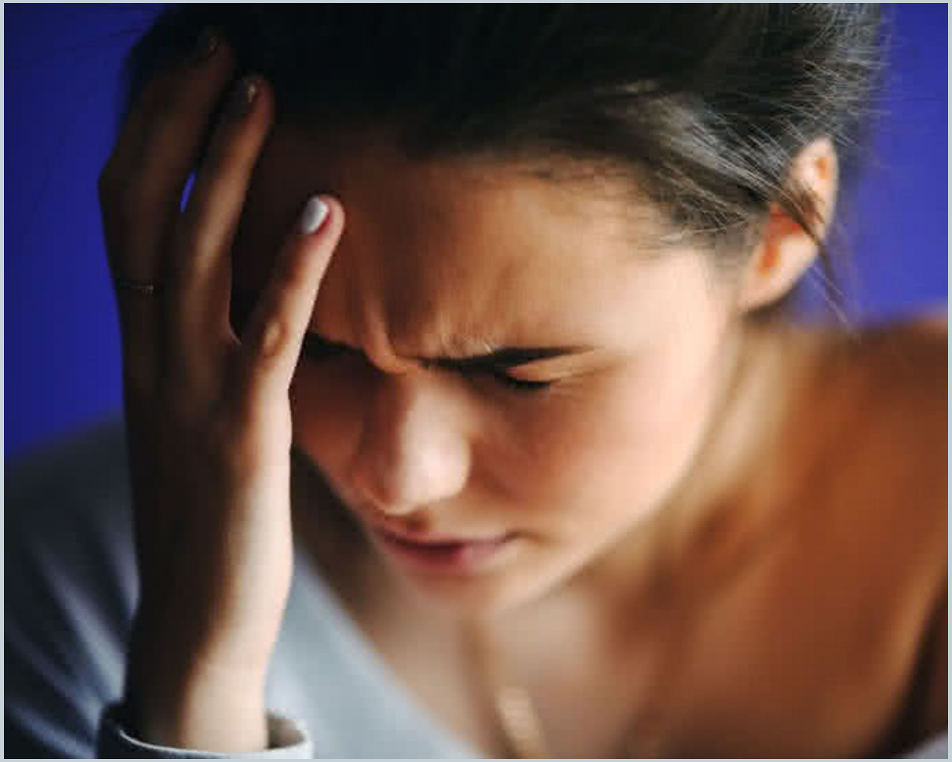
Stated Presenting Symptoms in Chiari I – Vague and Scattered

Pain (headache 34%)	**69%**
Weakness (1 or more limbs)	56%
Numbness (1 or more limbs)	52%
Loss of temperature sensation	40%
Painless burns	15%
Unsteadiness	40%
Diplopia	13%
Dysphasia	8%
Tinnitus	7%
Vomiting	5%
Dysarthria	4%
Other	14%

Reference: S. Paul, Kamal & H. Lye, Richard & Alexander Strang, F & Dutton, John. (1983). Arnold-Chiari malformation. Review of 71 cases. Journal of neurosurgery. 58. 183-7.

Multiple Possible Causes of Occipital Headaches

- Cervical disc disease
- Occipital neuralgia
- Trauma
- Muscle spasm or sprain
- Osteoarthritis
- Tumors
- Chiari malformation
- Infection
- Gout
- Diabetes
- Vasculitis



Headache and Chiari I

- Headache is the most common presenting symptom of Chiari I malformation
- However - lifetime prevalence of headache (with any form)
  - Men - 93%
  - Women - 99%
- The point prevalence of headache
  - Men - 11%
  - Women – 22%

J Clin Epidemiol. 1991;44(11):1147-57.

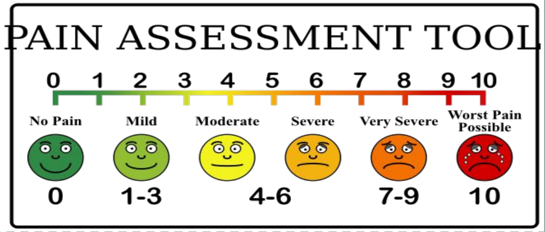
Epidemiology of headache in a general population--a prevalence study.

Rasmussen BK[1], Jensen R, Schroll M, Olesen J.

Data from International Headache Society

- Over 300 different headache types and etiologies are described
- 75% of migraine headaches are accompanied by neck pain
- In patients suffering from migraine and coincidental Chiari malformation treating the Chiari may lessen the incidence and/or severity of the migraines.
- Unfortunately, rarely does Chiari treatment eliminate the Migraine attacks. Proper Migraine management is still usually necessary, even after surgery.

Pain Scales



- Dozens of scales used to measure pain
  - Pain is subjective, so scales are difficult to standardize
- Most rely on some linear scale, usually 1-10
  - Patient marks a number, a face, or some other device to illustrate their pain level
  - e.g. Wong-Baker, VAS, NRS
- Some use more comprehensive questionnaires to get as objective a result as possible
  - e.g. McGill Pain Index, Brief Pain Inventory
- Some pain scales are designed for specific disease circumstances, such as dementia, insect bites/stings, and osteoarthritis

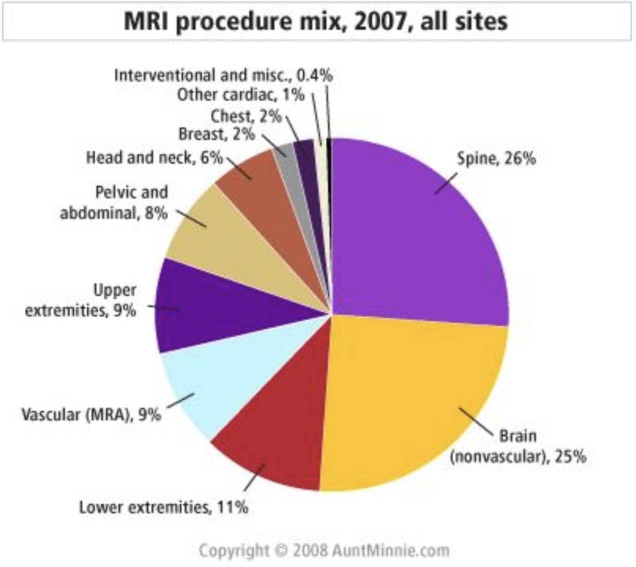
Radiological Diagnosis

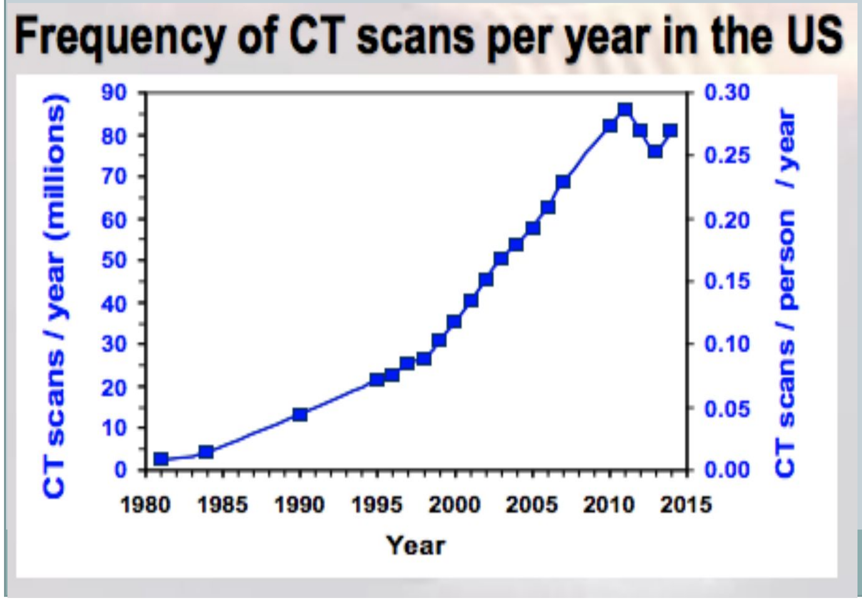
Incidental Diagnosis

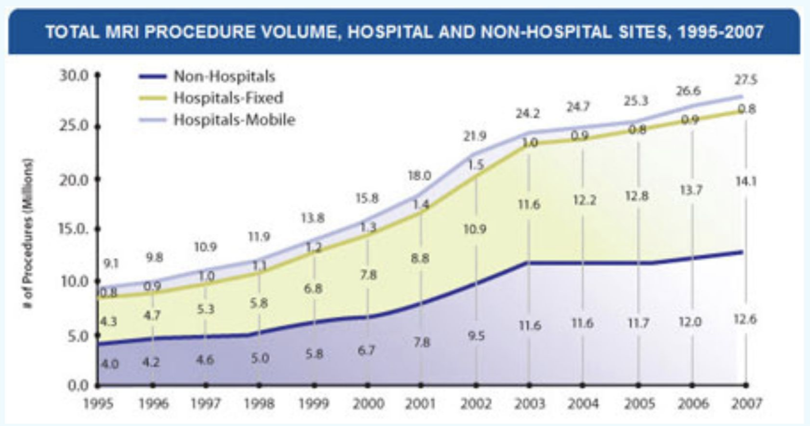
Number of head and neck diagnostic procedures in US - 2007

Number of head and neck MRI procedures in US – 8,525,000

Number of head and neck CT procedures in US – 27,900,000







MRI Procedures: Total U.S. MRI procedure volume, hospital and non-hospital sites, 1995-2007.*

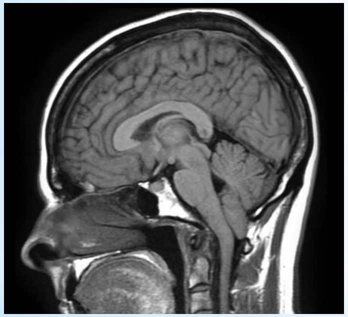
Disconnection Between Radiographic Diagnosis and Symptoms Asymptomatic Chiari

- The majority of patients with Chiari I malformation are asymptomatic. If symptomatic, patients usually present in adulthood
- In normal individuals cerebellar tonsils can extend up to 5 mm below the foramen magnum. However, in Chiari I patients it can be anywhere from 3 to 29 mm [Barkovich et al. 1986]
- 14% of patients with tonsillar ectopia greater than 5mm are asymptomatic. [Meadows J et al. 2000]
- Therefore, cerebellar tonsillar ectopia should not serve as the sole indicator for diagnosis and surgical treatment in the absence of clinical or other radiographic evidence.

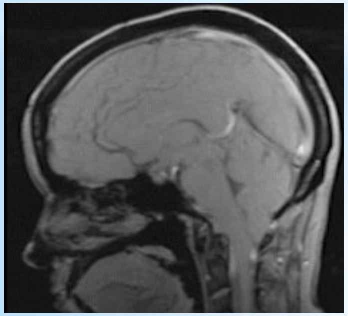
Now…

Problems With Radiographic Diagnosis

CT- poor visualization of soft tissues



MRI – anatomical picture without functional assessment



Cine MRI – absence of a range of normal

Surgery - Bad Outcome Analysis

- No appropriate subcategorization of outcome analysis
- Short follow-up
- Absence of randomized trials
- Scaling is not well done

Surgical Treatment Options for Chiari

- **Observation (Asymptomatic Chiari)**
- **Surgery for symptomatic patients**
  - Standalone extradural decompression ± C1 laminectomy
  - Dural opening without duroplasty ± arachnoid opening
  - Dural opening with duroplasty ± arachnoid opening
  - Cerebellar tonsillectomy ± duroplasty
  - Electrocauterization of tonsils
- **Surgery for hydrocephalus**
  - decompressive procedures
  - shunting

Surgical Treatment Options for Syringomyelia

Surgery for symptomatic syringomyelia

- Suboccipital and cervical decompression and duraplasty with or without plugging of the obex.
  - Rate of recurrent/residual syrinx after decompression only is about 6.7% (Schuster J.M. et al, 2013)
- Laminectomy and syringotomy (dorsolateral myelotomy)
- Shunts
  - Ventriculoperitoneal shunt
  - Syringosubarachnoid dorsal root entry zone shunt
  - Syringoperitoneal shunt
- Percutaneous needling
- Terminal ventriculostomy (in syrinx without Chiari malformation)
- Neuroendoscopic surgery
- Surgical untethering in select cases with tethering associated with syringomyelia

Surgery

- Many surgical options – we are only going to discuss posterior decompressions
- Extent of craniectomy and cervical decompression
- Open the dura or not
  - CSF anatomy
  - CSF Physiology
  - CSF flow compromise
  - Cine MRI
  - Fluid mechanics
  - Clinical data
- Benefits of intraoperative ultrasound

A Point of Surgical Dispute – Posterior Fossa and Cervical Decompressions

Extent of Decompression

- Decompression at least to the bottom of C2
- Craniectomy size (Jamie Baisden, 2012)

  Better outcome with small craniectomy vs extensive posterior fossa decompression for syringomyelia (Klekamp *et al*.)

  Larger posterior fossa decompression is more effective in the short-term postoperative period (1–4 weeks). However, smaller PF craniectomy showed clearly improved long-term efficacy (Zhang *et al*.).

  Fewer postoperative complications in smaller PFD *versus* those undergoing extended PFD (Zhang *et al*.).

- Other recommendations – historical procedures like plugging the obex, 4th ventricular shunting, terminal ventriculostomy and opening of foramen Magendie are not warranted. (Greenberg, 2016)

To Open, or Not to Open… the Dura

Higher Complication Rate With Duroplasty

Decompression With vs. Without Dural Opening

Children

- Duroplasty is associated with lower rate of reoperation (2.1% vs 12.6%)
- Duroplasty is associated with higher CSF related complications (18.5% vs. 1.8%)
- No significant difference between 2 techniques in clinical improvement or syringomyelia decrease.

Reference: (Kennedy BC et al., 2015, Lin W et al, 2017)

Adults

- Duroplasty is associated with:
  - slightly lower rate of reoperation (0.7% vs 2.1%)
  - significantly higher rate of aseptic meningitis (27.1% vs. 6.1%)
  - more procedure-related complications (2.3% vs. 0.8%)
  - longer length of hospital stay (4.4% vs. 3.8%)
  - higher hospital charges (USD 35,321 vs. 31,483)
- Similar short- and long-term clinical outcomes.

Reference: (Junchen C et al., 2017)

Complications of Chiari I Surgery

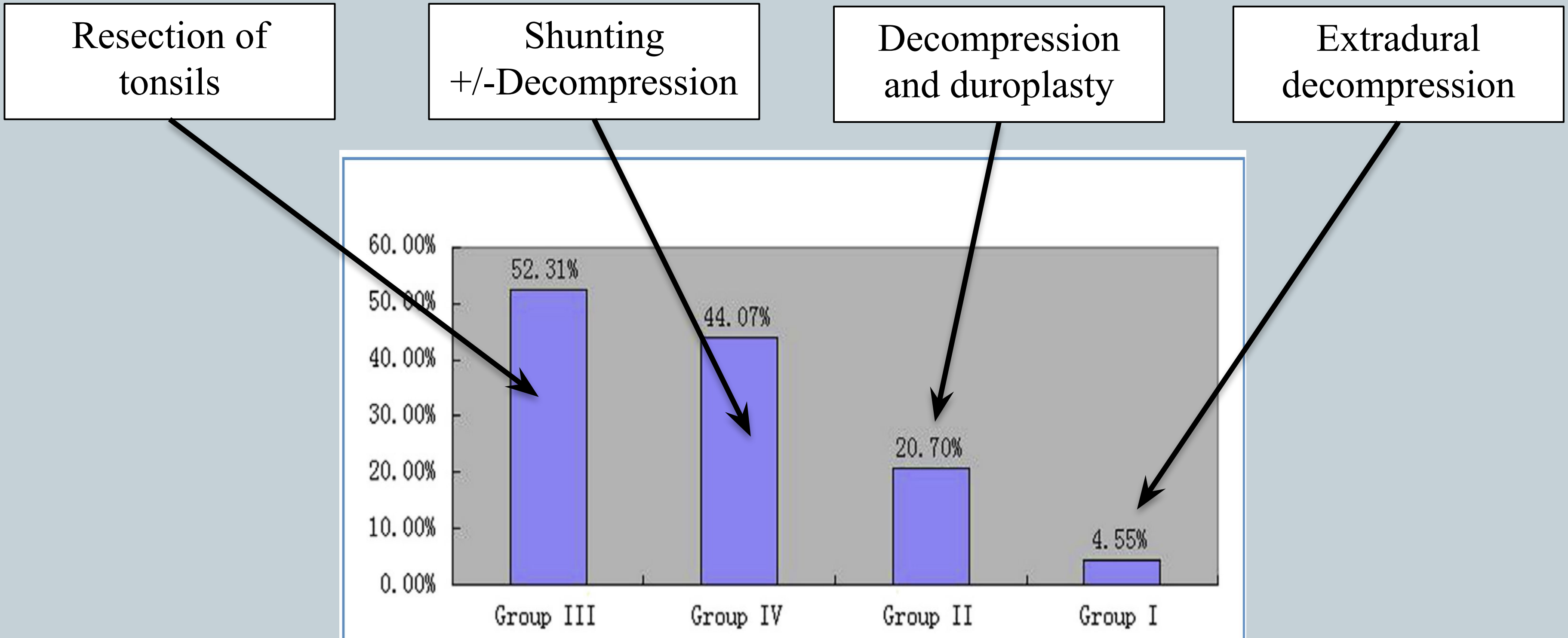


Figure 4. The distribution of adverse events according to the surgical method.

World Neurosurg. 2016 Apr;88:7-14. doi: 10.1016/j.wneu.2015.11.087. Epub 2015 Dec 28.

A Systematic Review of Chiari I Malformation: Techniques and Outcomes.

Zhao JL[1], Li MH[2], Wang CL[1], Meng W[1].

Results of Chiari I Surgery

Type of surgery	Improvement	Operative Complications	Postoperative deterioration	Reoperation	Benefit/risk ratio
Extradural decompression	73.6%	4.55%	7.8%	10.56%	16.2
Duroplasty	82.2%	20.7%	3.7%	7.72%	3.9
tonsillectomy	86%	52.3%	2.2%	9.04%	1.6

World Neurosurg. 2016 Apr;88:7-14. doi: 10.1016/j.wneu.2015.11.087. Epub 2015 Dec 28.

A Systematic Review of Chiari I Malformation: Techniques and Outcomes.

Zhao JL[1], Li MH[2], Wang CL[1], Meng W[1].

- Thus, we generally would advocate conservatism in regards to opening the dura.
- A relatively small expansion of Foramen Magnum CSF space should be adequate.

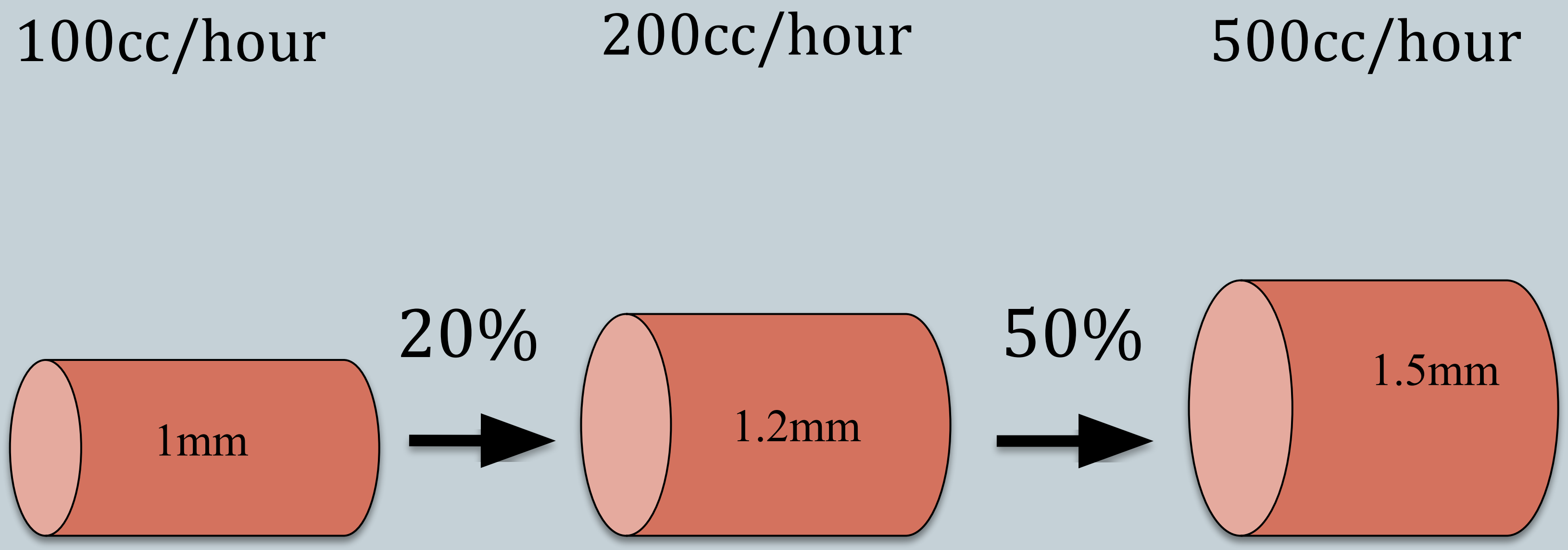
Q	Flow rate
P	Pressure
r	Radius
η	Fluid viscosity
l	Length of tubing

$$Q = \frac{\pi P r^4}{8\eta l}$$

radius

viscosity

How Much Bigger Do We Need?



However… Dural Elasticity is an Issue

Biotechnol Lett. 2013 May;35(5):825-30. doi: 10.1007/s10529-012-1127-9. Epub 2013 Feb 2.

An optofluidic mechanical system for elasticity measurement of thin biological tissues.

Cha C[1], Oh J.

J Neurotrauma. 2008 Jan;25(1):38-51. doi: 10.1089/neu.2007.0348.

Mechanical properties of dura mater from the rat brain and spinal cord.

Maikos JT[1], Elias RA, Shreiber DI.

J Neurosurg. 1977 Sep;47(3):391-6.

Elasticity of the spinal cord dura in the dog.

Tunituri AR.

Additionally… Ultrasound for Intraoperative Evaluation of CSF Flow

Given the higher rate of complications after duroplasty, and higher rate of reoperation without duroplasty, intraoperative ultrasonography was advocated for selection of candidates for duroplasty VS. bony decompression alone.

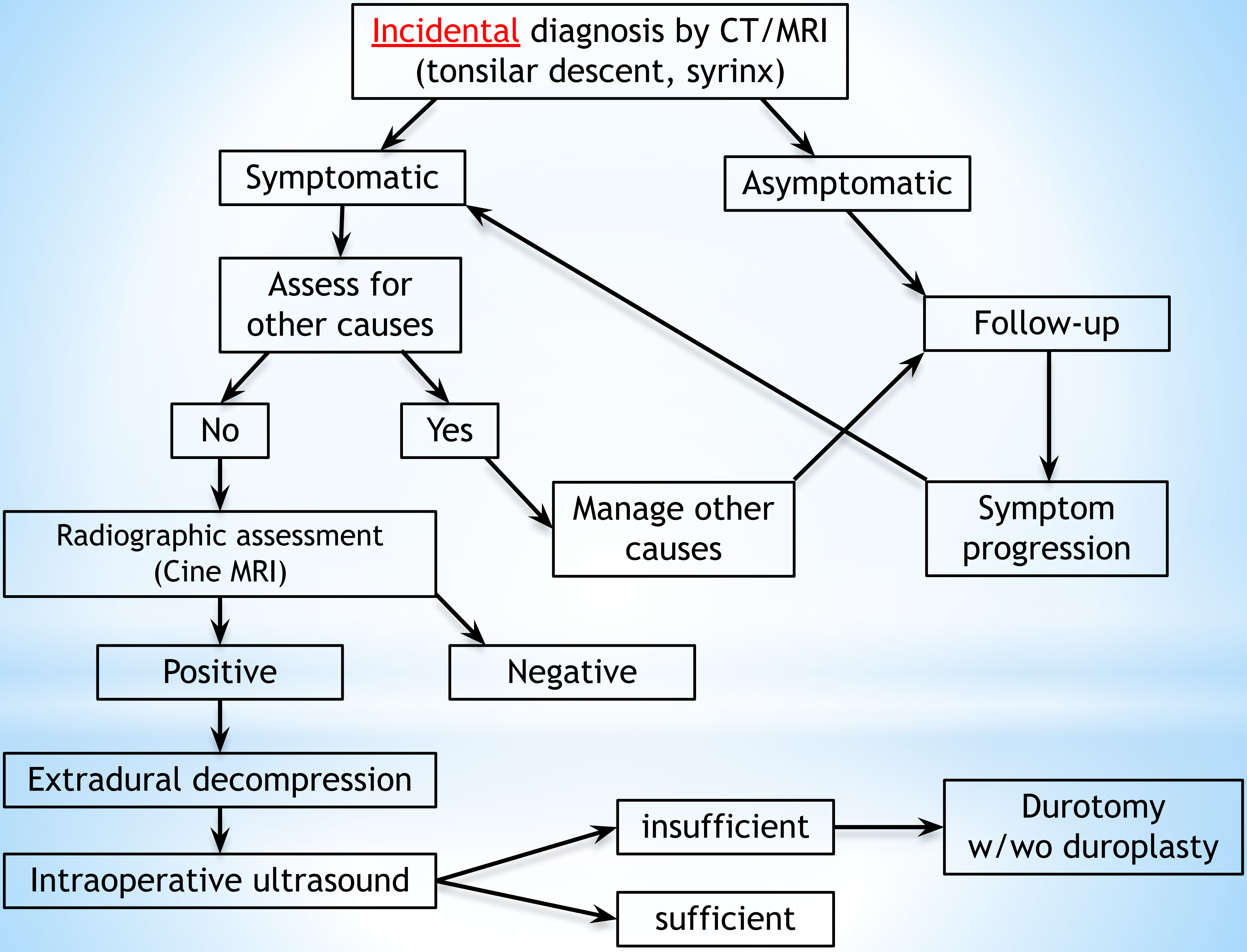
References:

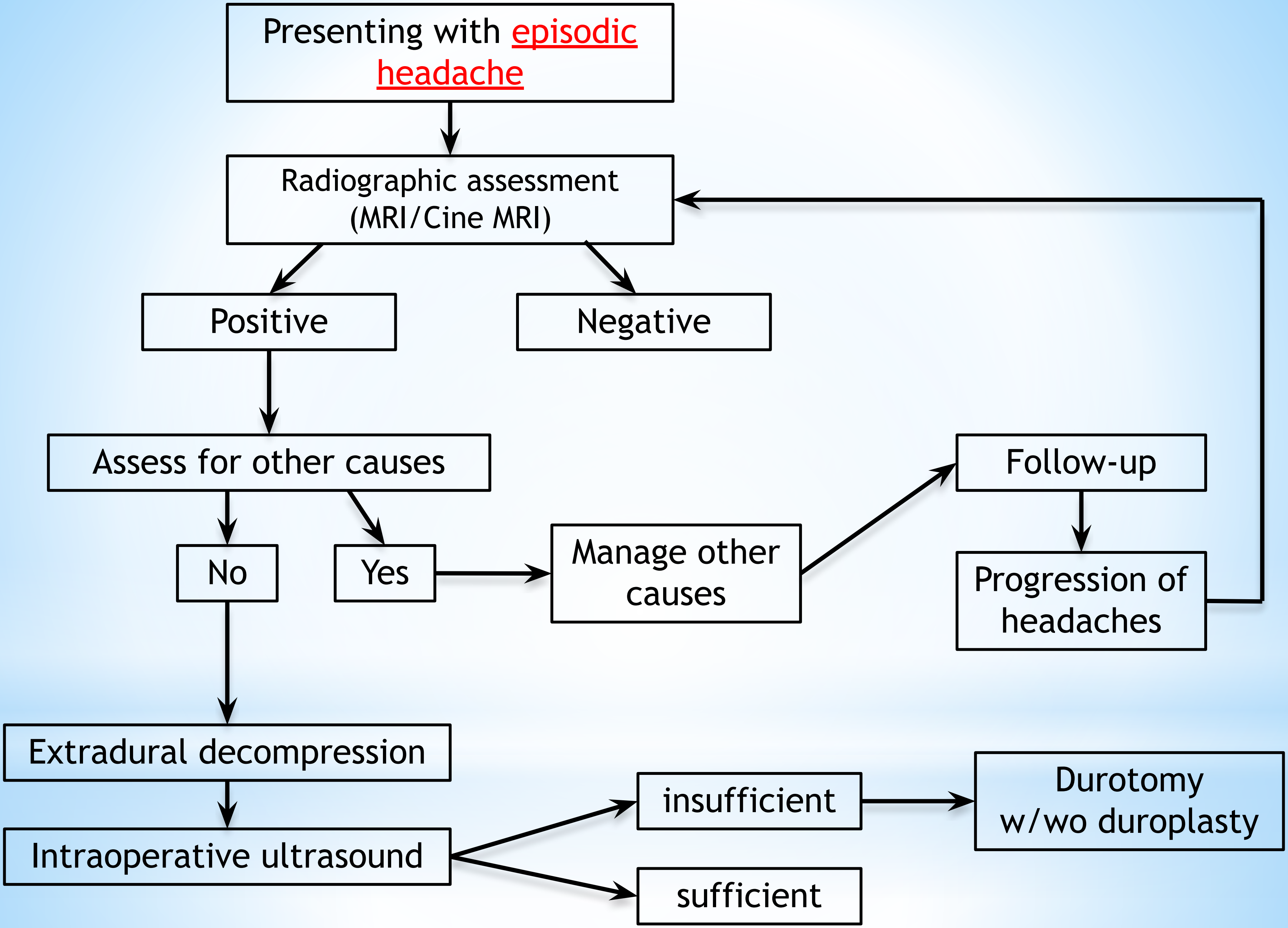
1. Intraoperative Ultrasonography for Definition of Less Invasive Surgical Technique in Patients with Chiari Type I Malformation. Roger SB, Mario AT, Matheus FO, Marcelode LO, Manoel JT, Edson BS, World Neurosurgery Volume 101, May 2017, Pages 466-475
2. Current opinions for treatment of symptomatic hindbrain herniation or Chiari type I malformation. Menezes AH. World Neurosurg. 2011 Feb;75(2):226-8
3. Intraoperative ultrasonography as a guide to patient selection for duraplasty after suboccipital decompression in children with Chiari malformation Type I. McGirt MJ, Attenello FJ, Datoo G, Gathinji M, Atiba A, Weingart JD, Carson B, Jallo GI. J Neurosurg Pediatr. 2008 Jul;2(1):52-7

Summary of Surgery Suggestions

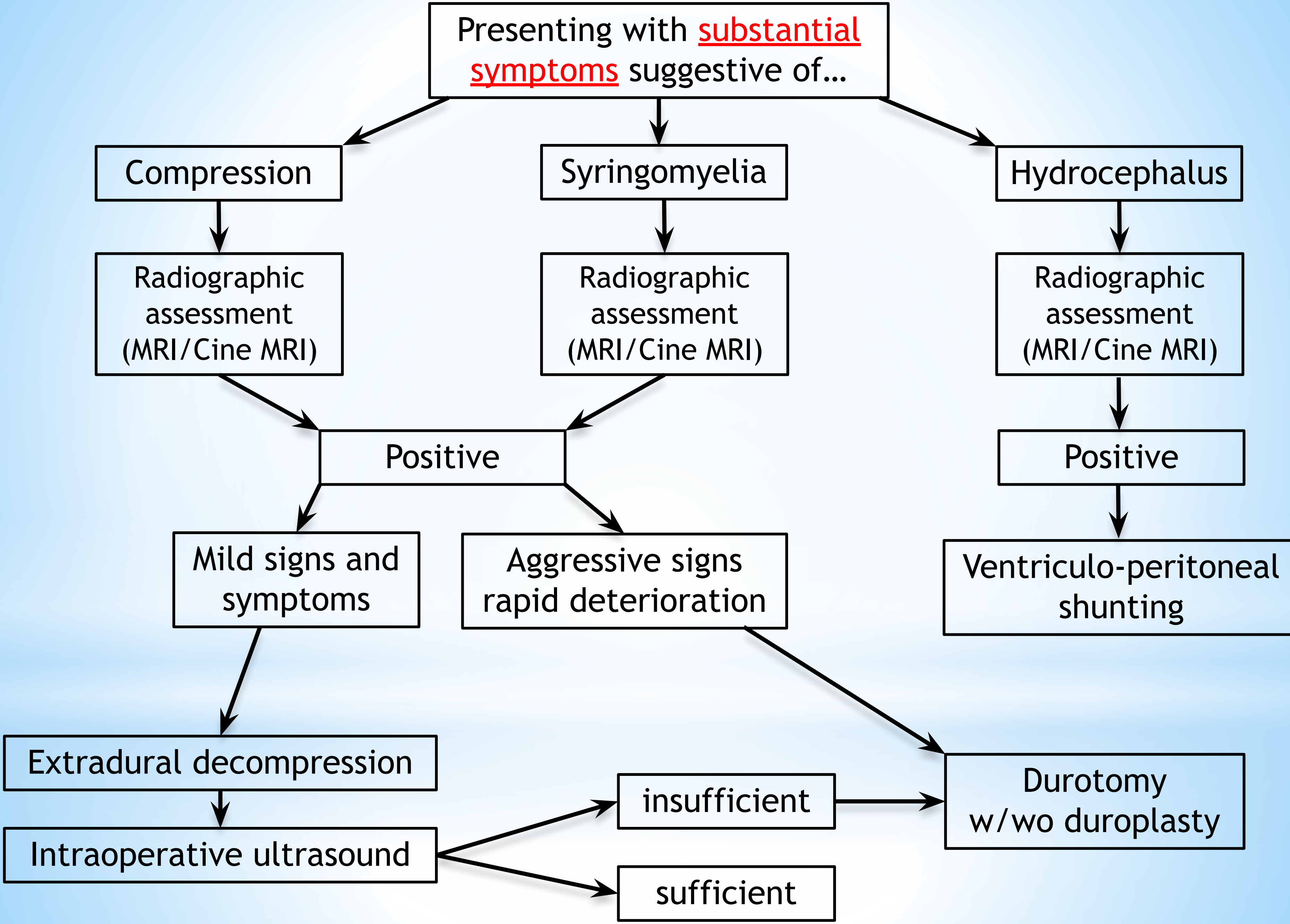
- Surgical indications/treatment options
- Level and extent of Posterior Fossa and cervical decompression
  - Decompress below the tonsils (at least to the bottom of C2)
  - Adequate but not excessive decompression
  - Get guidance from preoperative cine MRI
- Less is more – minimal required extent of decompression, don’t open the dura routinely
  - no good comparative papers
  - no evidence that opening dura is better
  - substantial complications with durotomy
  - aseptic meningitis from durotomy can cause alterations in flow dynamics
- In case of duroplasty exercise great care to avoid blood in intradural space.
- Intraoperative assessment
  - Intraoperative ultrasound for assessment of the degree of decompression
  - Intraoperative doppler/duplex for assessment of CSF flow

Treatment Algorithms


Incidental diagnosis by CT/MRI
(tonsilar descent, syrinx)
Symptomatic
Asymptomatic
Assess for
other causes
Follow-up
No
Yes
Manage other
causes
Symptom
progression
Radiographic assessment
(Cine MRI)
Positive
Negative
Extradural decompression
Intraoperative ultrasound
insufficient
sufficient
Durotomy
w/wo duroplasty



Presenting with episodic headache
Radiographic assessment (MRI/Cine MRI)
Positive
Negative
Assess for other causes
No
Yes
Manage other causes
Follow-up
Progression of headaches
Extradural decompression
Intraoperative ultrasound
insufficient
sufficient
Durotomy w/wo duroplasty


Presenting with substantial symptoms suggestive of…
Compression
Syringomyelia
Hydrocephalus
Radiographic assessment (MRI/Cine MRI)
Radiographic assessment (MRI/Cine MRI)
Radiographic assessment (MRI/Cine MRI)
Positive
Positive
Mild signs and symptoms
Aggressive signs rapid deterioration
Ventriculo-peritoneal shunting
Extradural decompression
Intraoperative ultrasound
insufficient
sufficient
Durotomy w/wo duroplasty

Questions to Ask

- Can the cascade of CSF flow abnormalities develop into a secondary pathology, e.g. NPH? Can temporary flow obstructions account for episodic posterior fossa neuronal dysfunction.
- Should headache sufferers sleep in prone position?
  - Should a soft collar be employed more commonly?
- Can CSF flow abnormalities regress over time due to age related changes, e.g. atrophy, menopause?
- Is Chiari malformation always associated with CSF flow abnormalities?
- Should scaled cine MRI studies be more widely employed in evaluation of chronic headaches and neck pain?

THANK YOU