CHIARI: SURGICAL MANAGEMENT

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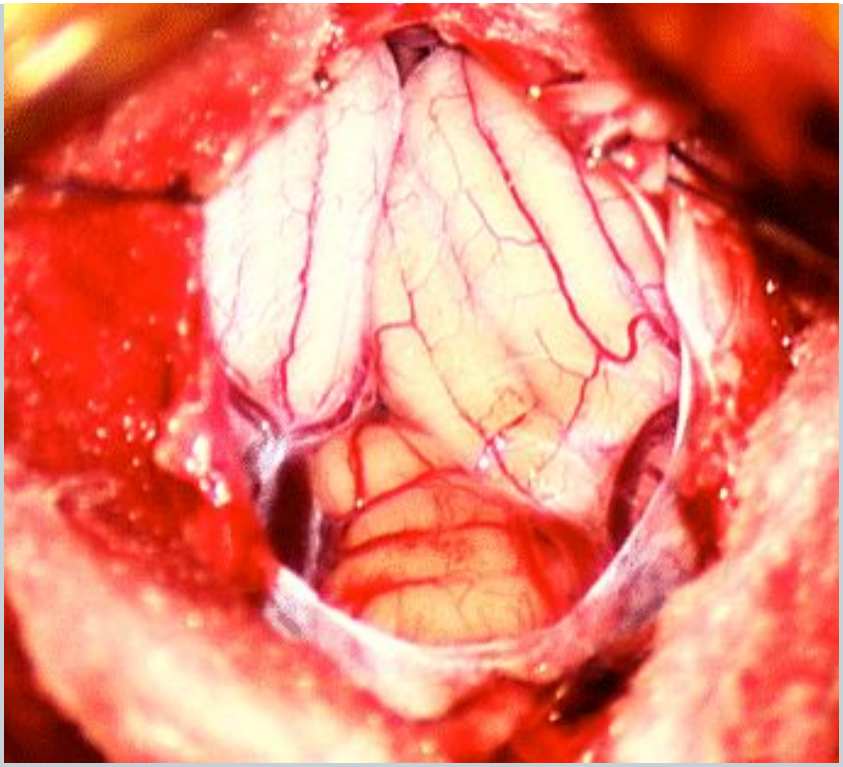
Definition



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.

Introduction – A work in progress

- A subject of great confusion referable to the absence of appropriate outcome analysis and classification issues.
- There is a great uncertainty and confusion about the treatment of people with the problem.
- For purposes of our discussion we will focus on Chiari I (tonsils descended ≈ 5mm or more) as this is where the most uncertainty is.

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 +/- tonsillar 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	-	

Outline

- Etiology of anatomical alteration
- Epidemiology
- History
- Confusion
- Why is there confusion?
- Suggestions/Explanation for confusion
- Treatment protocols

Etiology of anatomical alteration

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

Causal interconnections?

Epidemiology is uncertain

Epidemiology of Chiari

Prevalence

World population – 7.6 billion

- **Chiari prevalence by retrospective review of MRI films**
  - Adult population – 0.77% (review of 22,000 MRIs >18 years old) [Meadows J et al. 2000]

    43.9 million adult Chiari patients
  - Pediatric population – 3.6% (review of 14,116 MRIs <18 years old) [Strahle J et al. 2011]

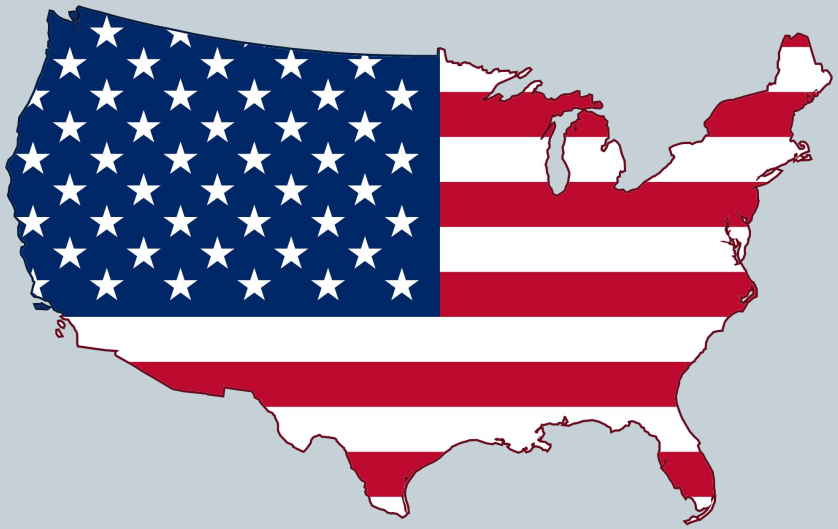
    68.4 million pediatric Chiari patients
- **Symptomatic Chiari in general population - 0.1% with a slight female predominance**

  ?

  7.6 million symptomatic Chiari patients

US Data

- There are over 300,000 diagnosed Chiari patients in US
- About 10,000 decompressive procedures per year for Chiari in US



Trends in Chiari I surgery in US

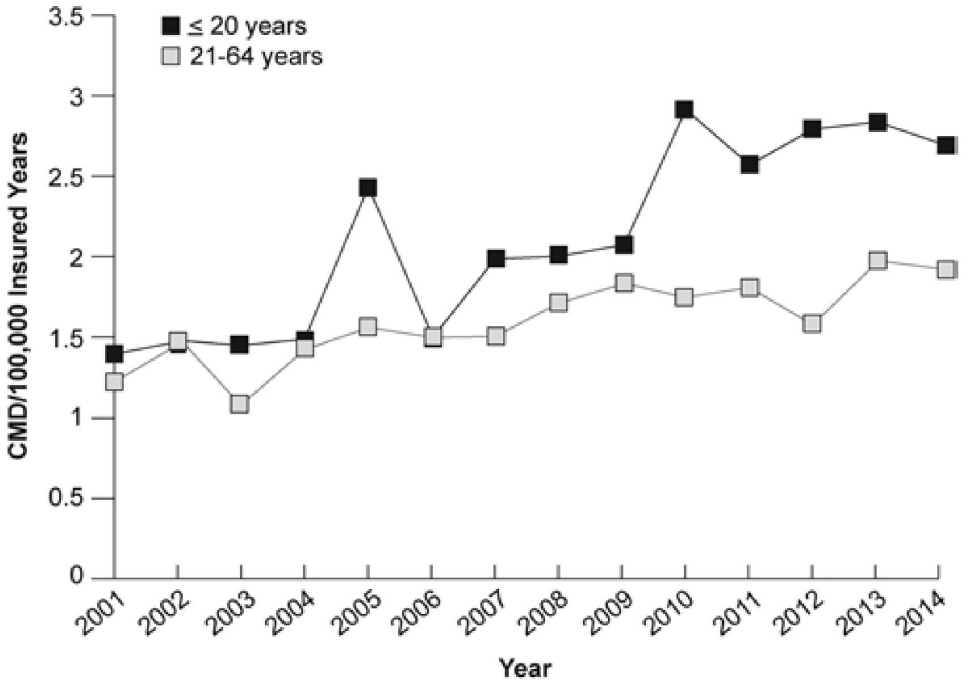


FIG. 2. Annual rate of CMD over a 14-year interval. Pediatric patients (age ≤ 20 years) had an increasing rate of CMD over the interval. Adult patients younger than 65 years also had a smaller, but still significant, increased rate of CMD.

Reference: Trends in surgical treatment of Chiari malformation Type I in the United States; D. Andrew Wilkinson, MD, Kyle Johnson, BS, Hugh J. L. Garton, MD, MSc, Karin M. Muraszko, MD, and Cormac O. Maher, MD; J Neurosurg Pediatr 19:208–216, 2017

History

Pathological description and resulting nomenclature dispute

How did treatment protocols arrive at current state?

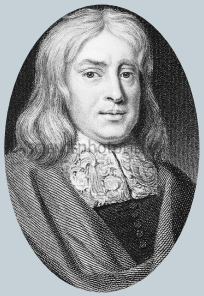
Color code

Clinical diagnosis

Surgical treatment

Pathological diagnosis

Radiographic diagnosis



Thomas Sydenham
first description of sign-symptom complexes,
1670

1670

Jean Pierre Flourens
first description of cerebral localizations
1820

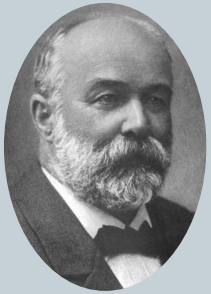
1820

Jean Cruveilhier
described Chiari as anatomical variant of development
1829

1829

Marshall Hall
first description of reflex arc
1837

1837



Theodor Langhans
first correlation between tonsillar ectopia and syringomyelia
1881

1881

Carl Wernicke
first sterile external ventricular drainage
1881

1881

Color code

Clinical diagnosis

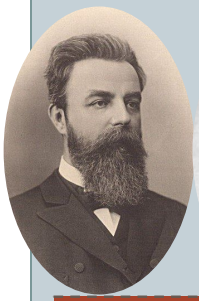
Surgical treatment

Pathological diagnosis

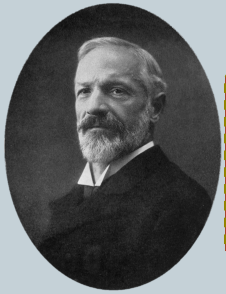
Radiographic diagnosis

William Gowers
first systematic neurological examination
1886

John Cleland
first Chiari case series
1883



Kahler and Schultze
first description of clinical syndrome associated with syringomyelia
1882



Hans Chiari
first classification,
1891

Henry Head
First description of spinal dermatomes
1893

Johann von Mikulicz-Radecki
First ventriculo-subgaleal shunt
1893

1882 1883 1886 1891 1893 1893

Color code

Clinical diagnosis

Surgical treatment

Pathological diagnosis

Radiographic diagnosis

Julius Arnold
description of a single case of Type 1 Chiari malformation
1894

Alfred Parkin
First posterior fossa decompression for hydrocephalus
1895

Aring CD
First publication on clinical presentation of Chiari (cerebellar syndrome)
1938

Heppner F
First publication on Chiari causing hydrocephalus in children
1951

Nulsen Spitz
First valved shunts
1952

Gardner and Goodall
first Chiari decompressions in children
1957

1894 1895 1938 1951 1952 1957

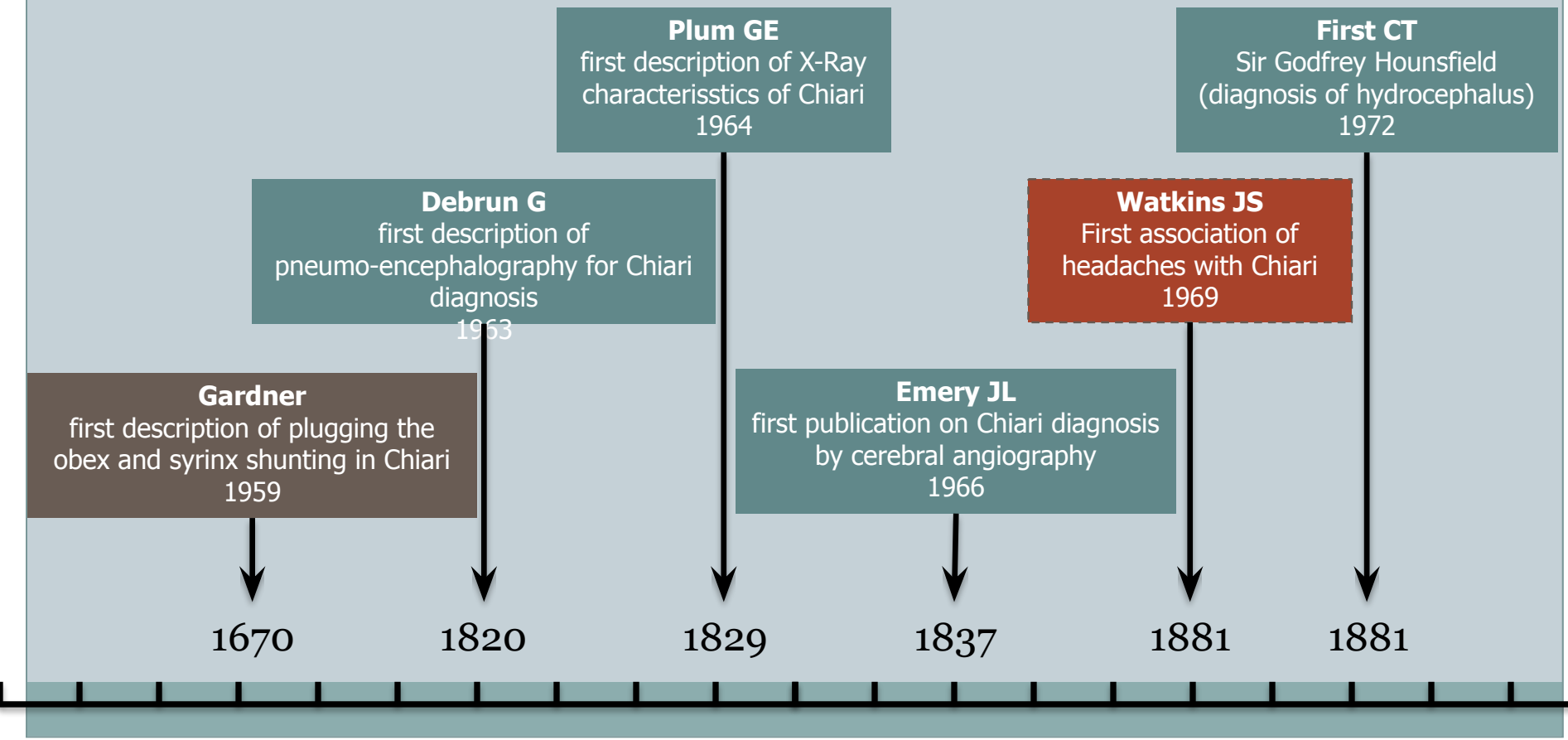
Color code

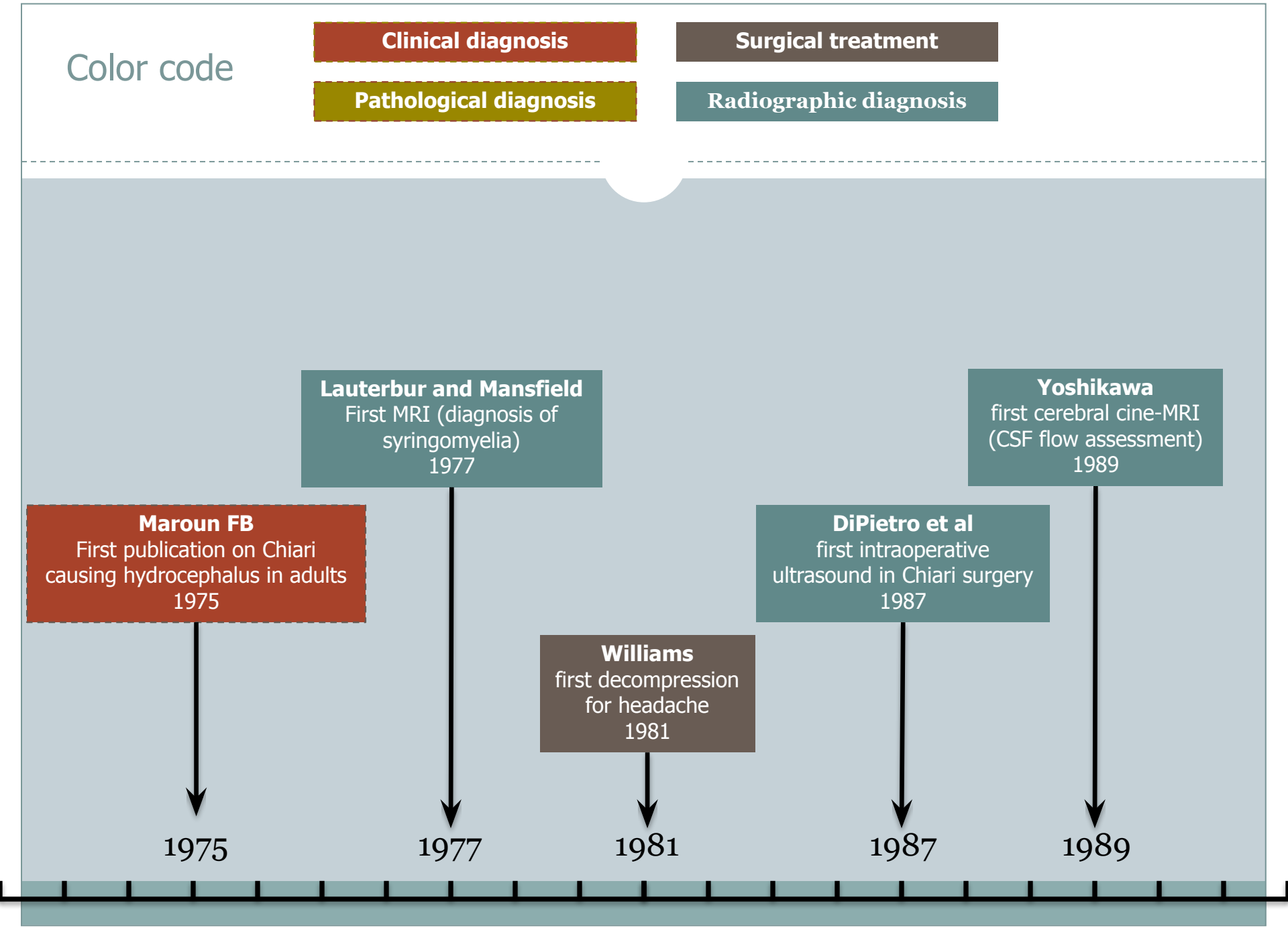
Clinical diagnosis

Surgical treatment

Pathological diagnosis

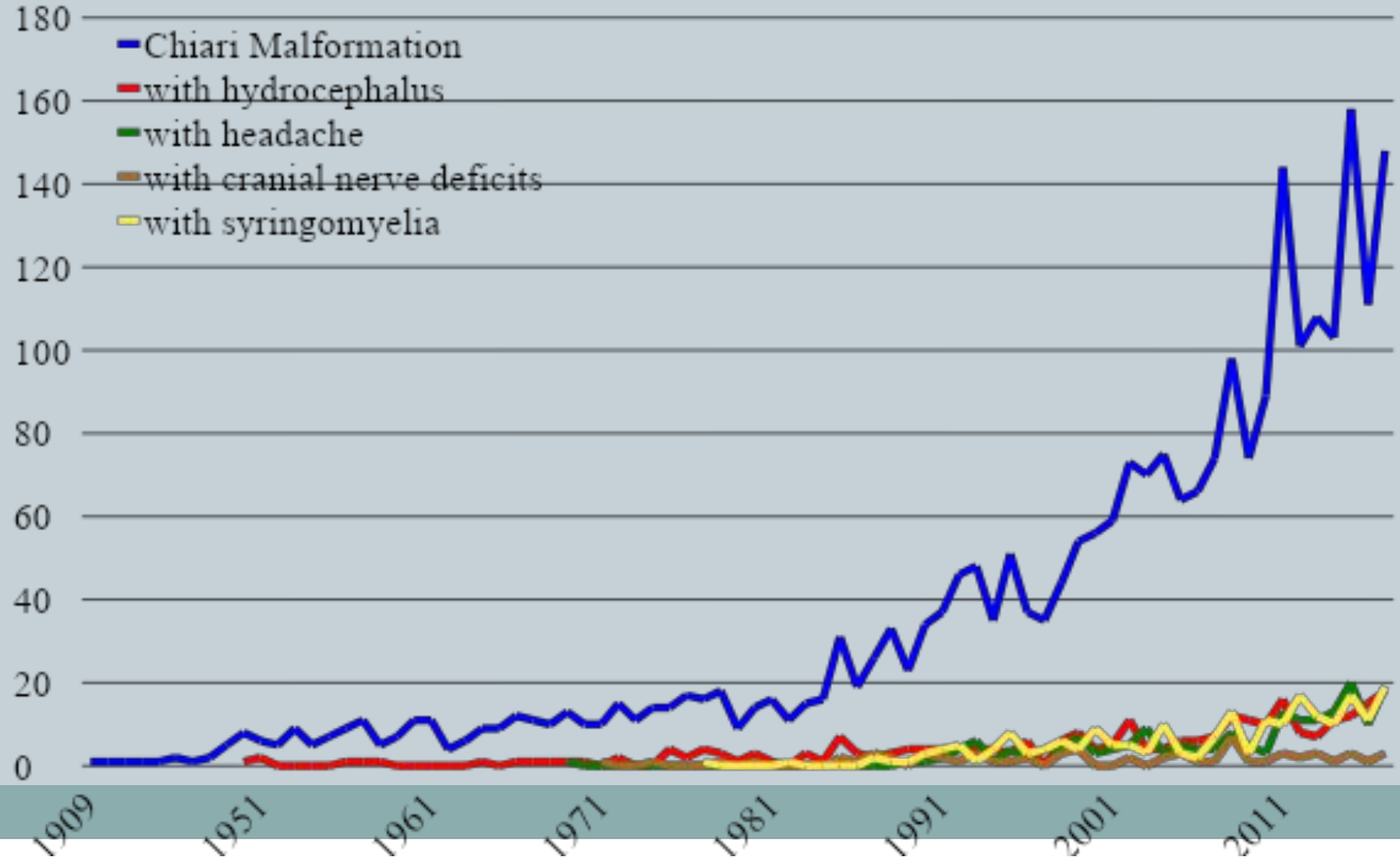
Radiographic diagnosis



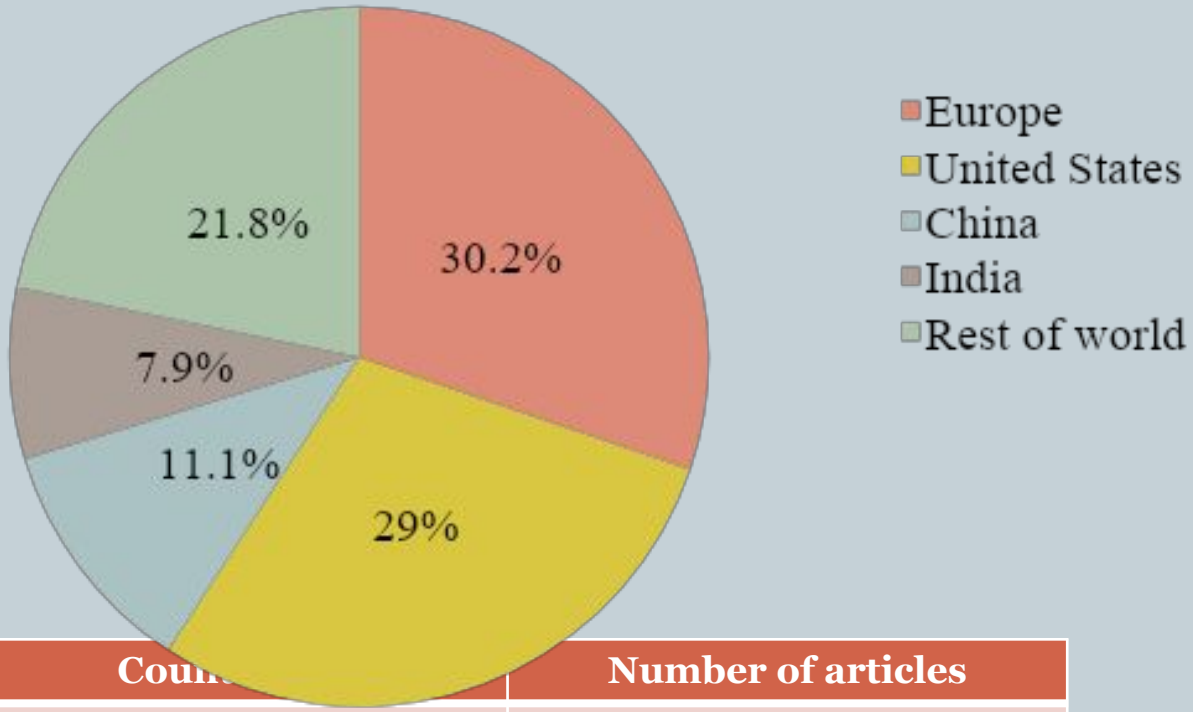

Color code
Clinical diagnosis
Surgical treatment
Pathological diagnosis
Radiographic diagnosis
Lauterbur and Mansfield
First MRI (diagnosis of syringomyelia)
1977
Yoshikawa
first cerebral cine-MRI (CSF flow assessment)
1989
Maroun FB
First publication on Chiari causing hydrocephalus in adults
1975
DiPietro et al
first intraoperative ultrasound in Chiari surgery
1987
Williams
first decompression for headache
1981
1975
1977
1981
1987
1989

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

Cited articles related to Chiari malformation – 2,613



Publishing countries in 2017



Coun	Number of articles
Europe	51
United States	49
China	19
India	13
Rest of world	37

However…

Guidelines / Standard of Care

- No standard of care
- No Grade I evidence

Confusion

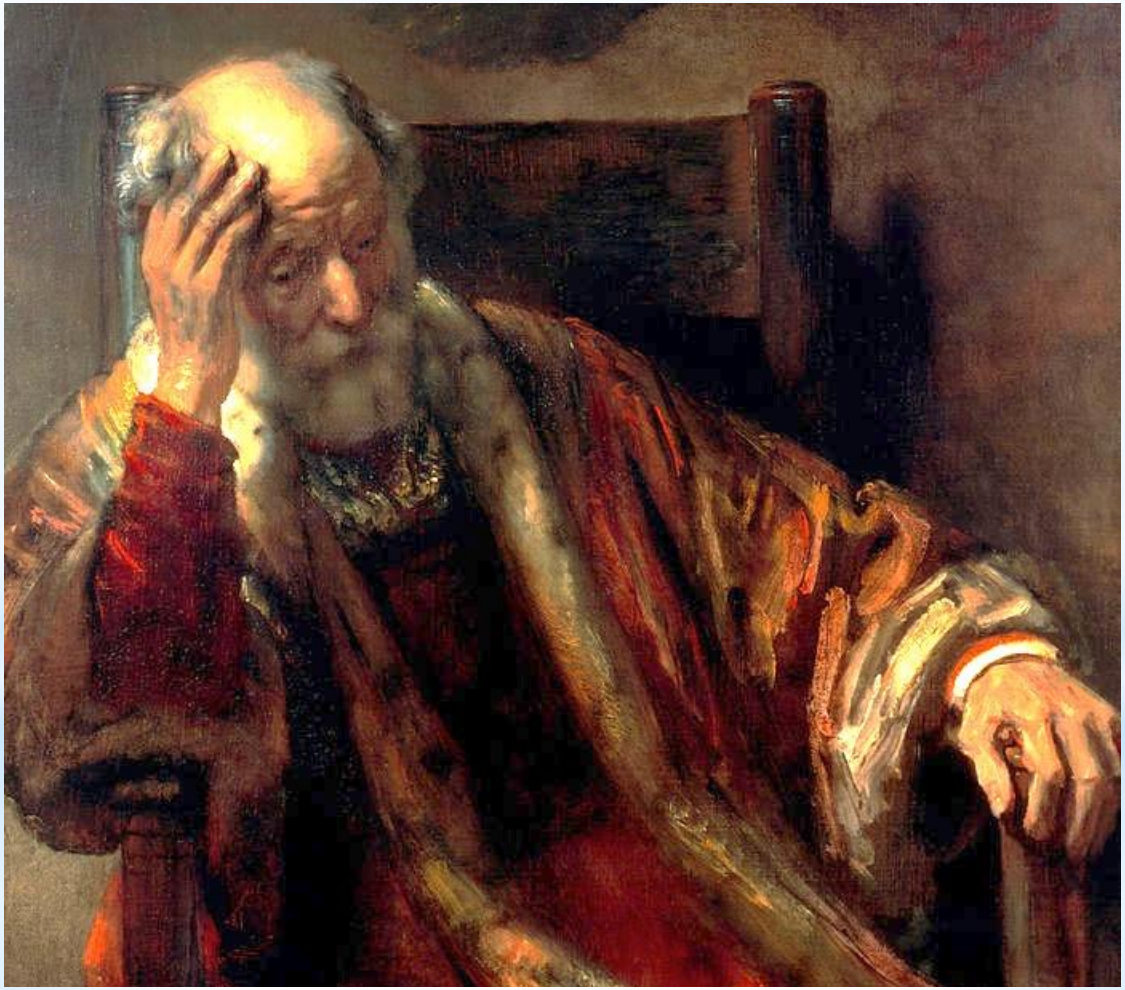
- Literature confusion
- Treatment confusion
- Outcome confusion

An example of confusion

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- no syrinx	8
1A	w/ 2mm wide syrinx	28
1B	w/ 8mm wide syrinx	75
2	9 yr old with headaches 10 mm tonsils- no syrinx	46
2A	w/ 2mm wide syrinx	64
2B	w/ 8mm wide syrinx	90
3	11 yr old w/progressive scoliosis 12 mm tonsils- no syrinx	58
3A	w/ small syrinx	85
3B	w/ 6mm wide syrinx	97

Why is there confusion?



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…

I. Causal connections are insecure as CSF abnormalities are often transient and may depend on secondary factors as well as being imperfectly demarcated.

II. Sign-symptom complexes are vague

- A. Direct Hindbrain Pressure
- B. Disruption of CSF pathways – Hydrocephalus / Syringomyelia / Headache
  1. Natural disease course is partially known
  2. Sign-symptom complexes can be caused by other pathological processes
  3. Chiari can exist without clinical findings and vice versa – correlation between the degree of Chiari malformation and presentation is not direct
  4. Ockam's razor - Multiple possible causes and influences to make better or worse
     - a. Age events
     - b. Hypertension
     - c. Sleep apnea
     - d. Medication related chronic headache
     - e. Estrogen toxicity
     - f. Cervical stenosis
     - g. Abnormally small posterior fossa
  5. In relatively small number Chiari may be the only cause

III. Radiographic evaluation is variable

IV. Therapeutic interventions are often imprecise

V. Bad outcome analysis

- A. Absence of control group
- B. Clinical changes difficult to demarcate
- C. Radiographic follow-up insecure
- D. Incorrect scaling – depending on anatomy and ignoring physiological issues
- E. Partially known long term follow-up

VI. Resulting imperfect guidance on operative protocols

THE CLINICAL SITUATION IS COMPLICATED
(many disease processes)

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

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

TIME

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

CHIARI I

Impact
- may not cause any symptoms
- mechanism of impact is controversial

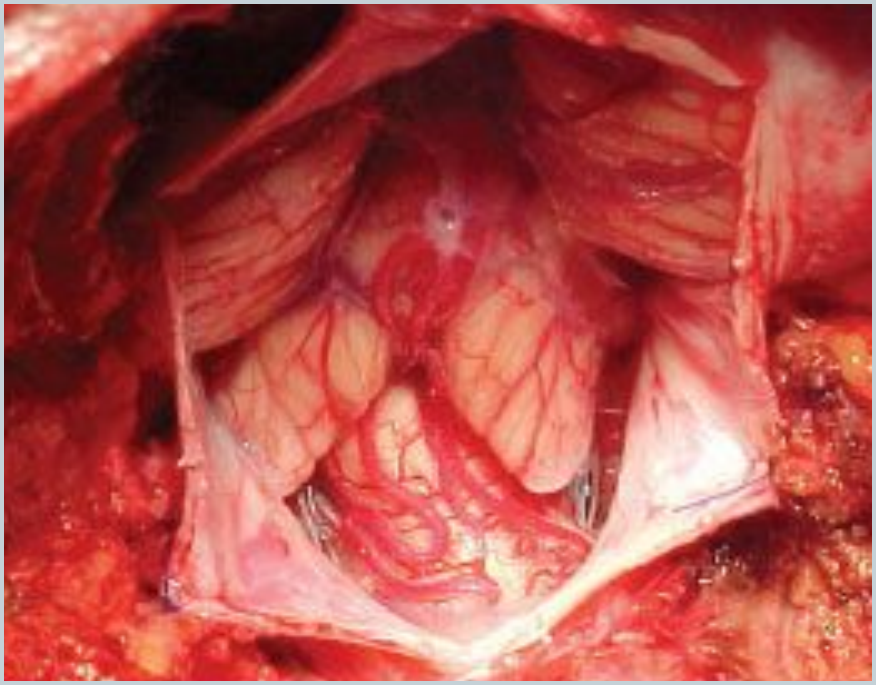
- Compression
- CSF disruption (can be transient)
  - \+ headache
  - ++ syringomyelia
  - +++ hydrocephalus

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

Symptoms
- a large host of subjective complaints
- poorly demarcated

A little light on the subject and ideas about improvement…

Surgery



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

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

A point of surgical dispute – posterior fossa and cervical decompressions

Extent of decompression

- 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)

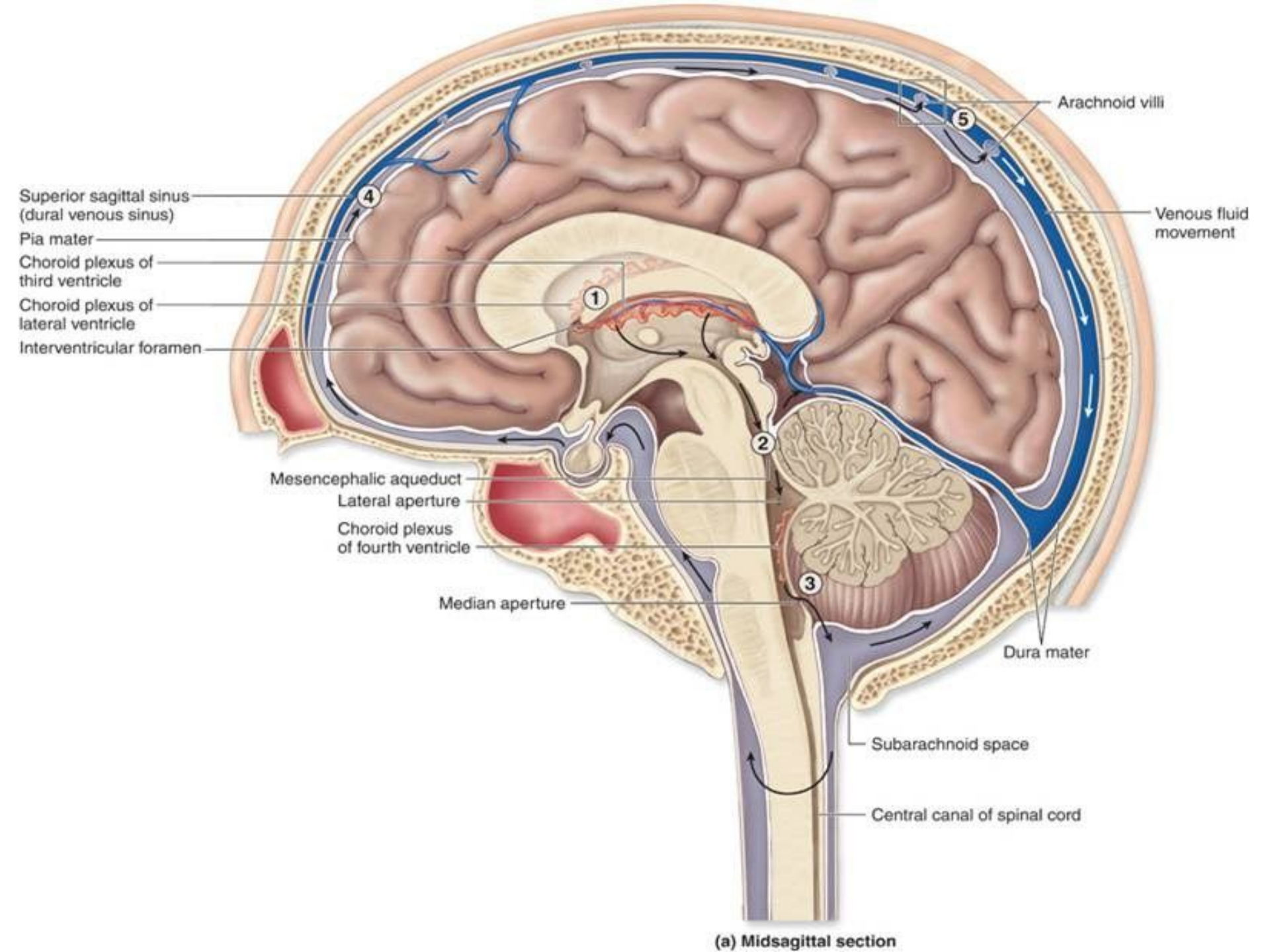
Tonsillar descent in Chiari I

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

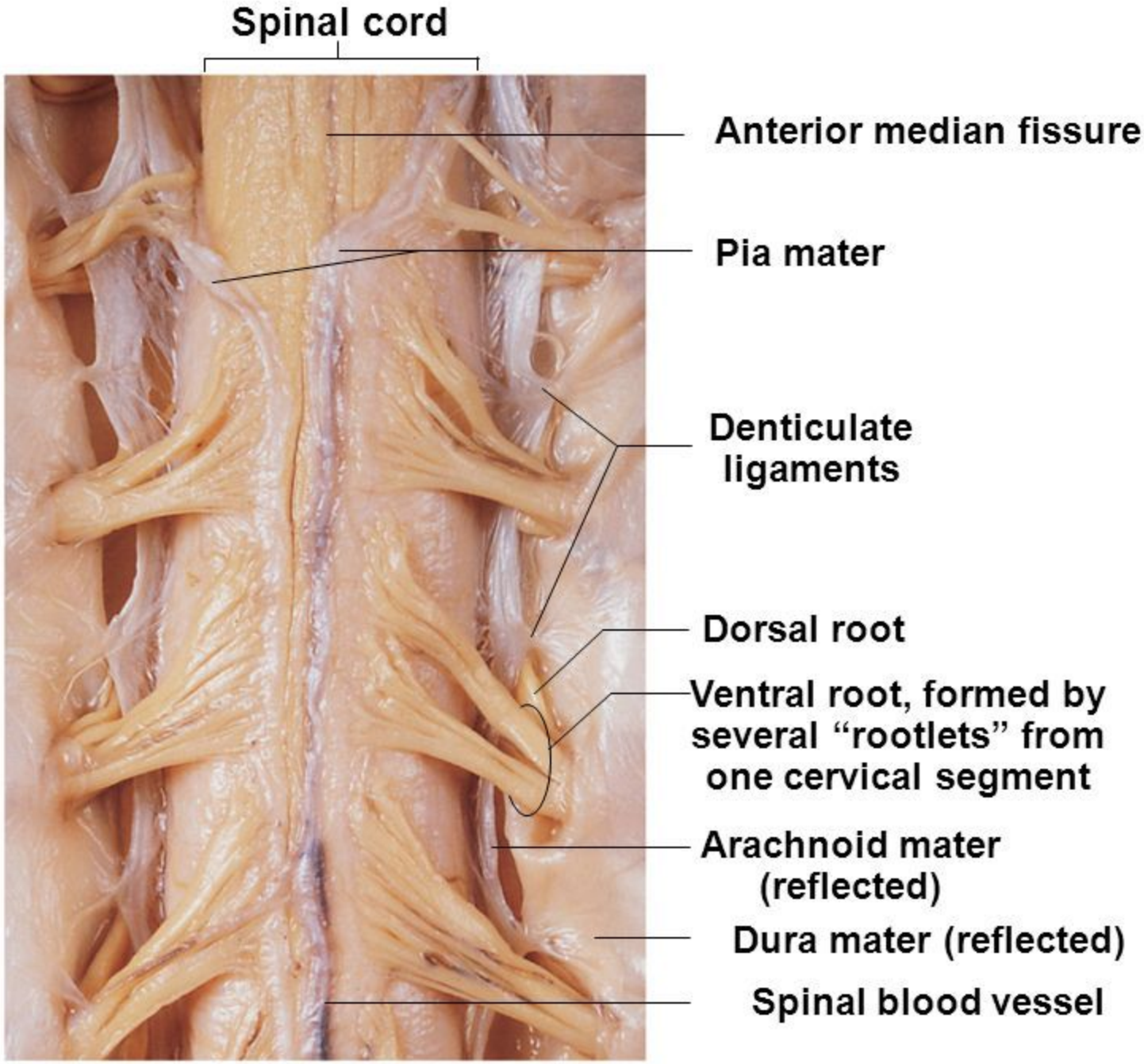
Reference: S. Paul, Kamal & H. Lye, Richard & Alexander Strang, F & Dutton, John. (1983). Arnold-Chiari malformation. Review of 71 cases. Journal of neurosurgery. 58. 183-7.

To open, or not to open… the dura

Anatomy

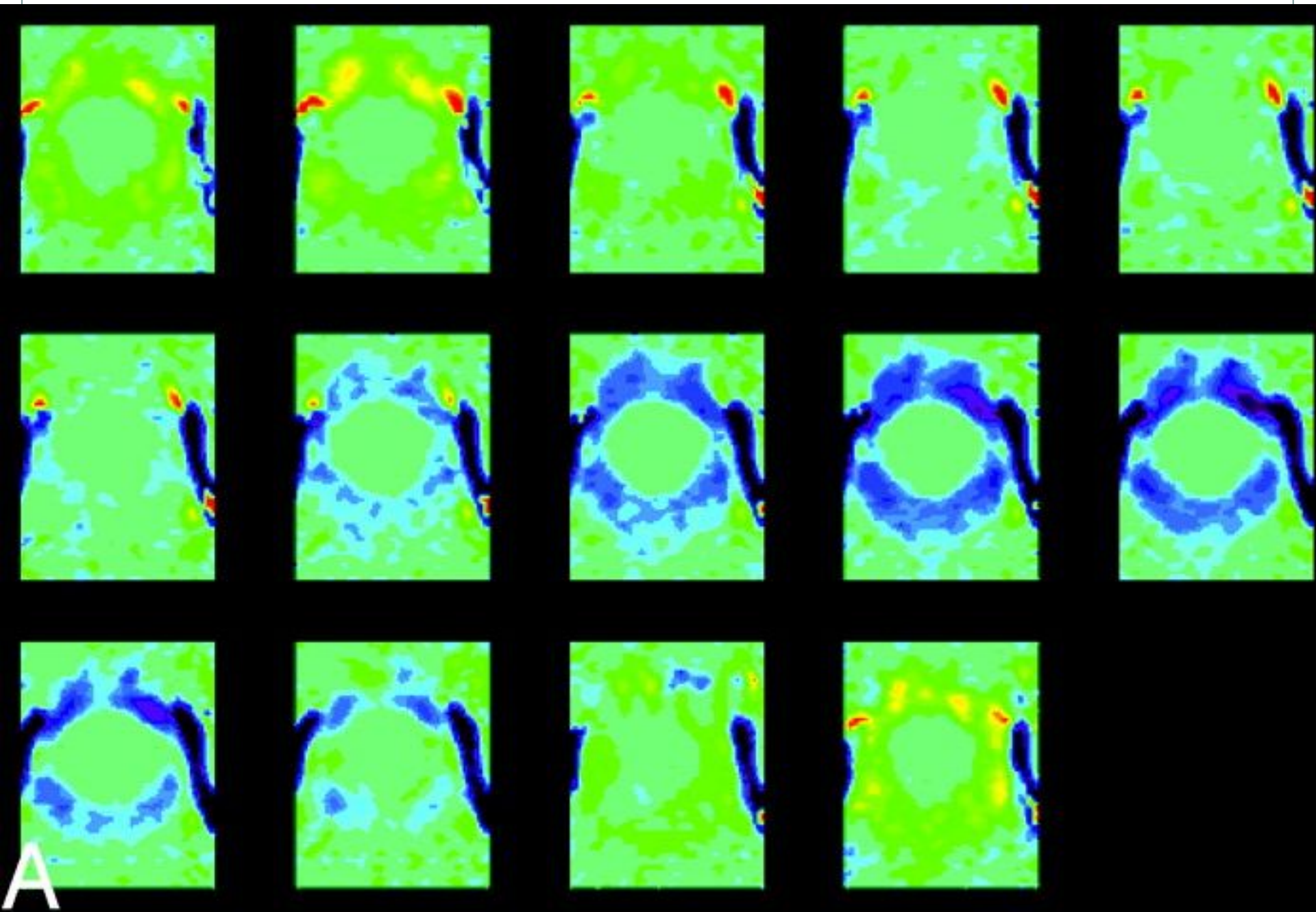


(a) Midsagittal section

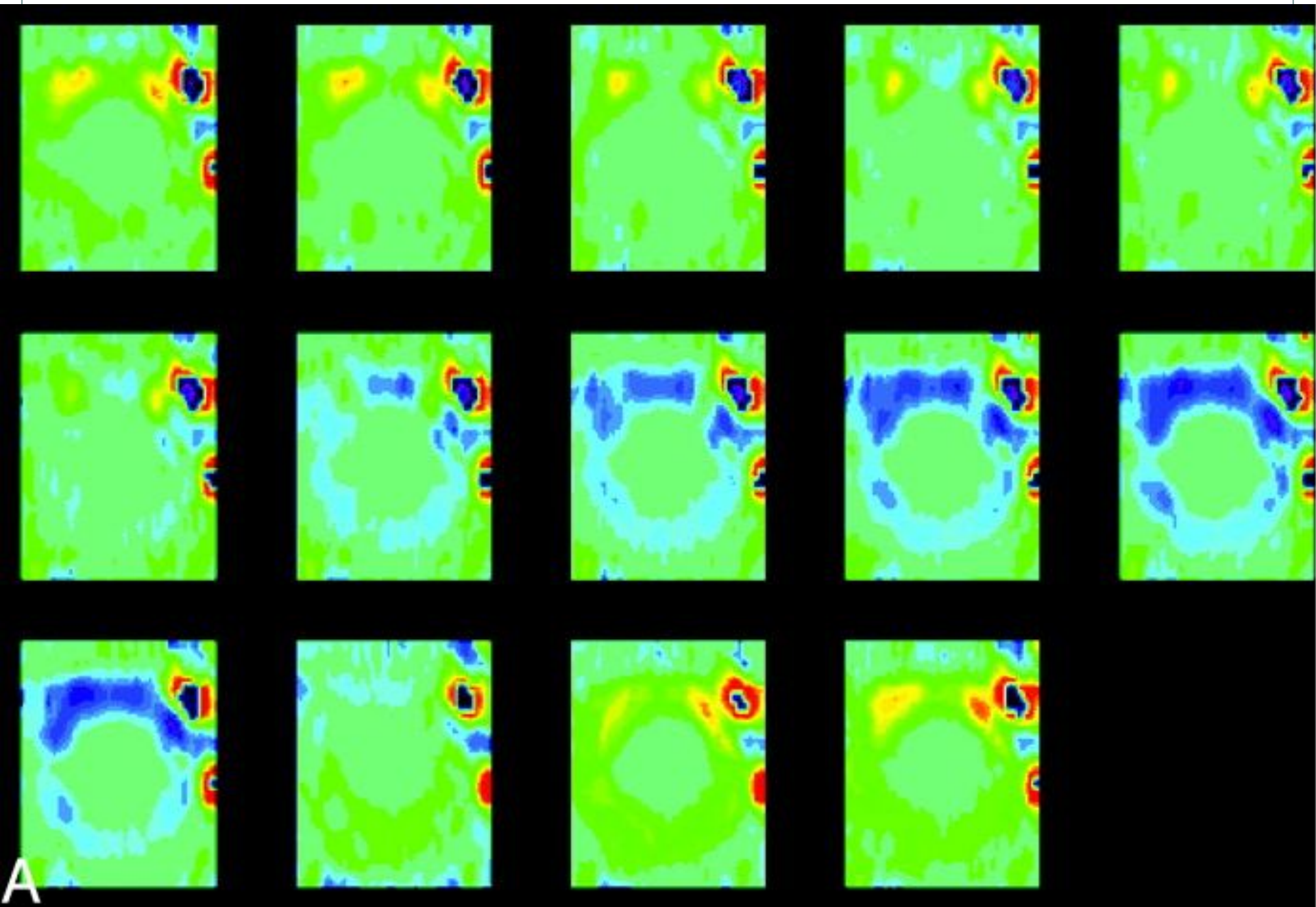

Spinal cord
Anterior median fissure
Pia mater
Denticulate ligaments
Dorsal root
Ventral root, formed by several "rootlets" from one cervical segment
Arachnoid mater (reflected)
Dura mater (reflected)
Spinal blood vessel

Two-way highway

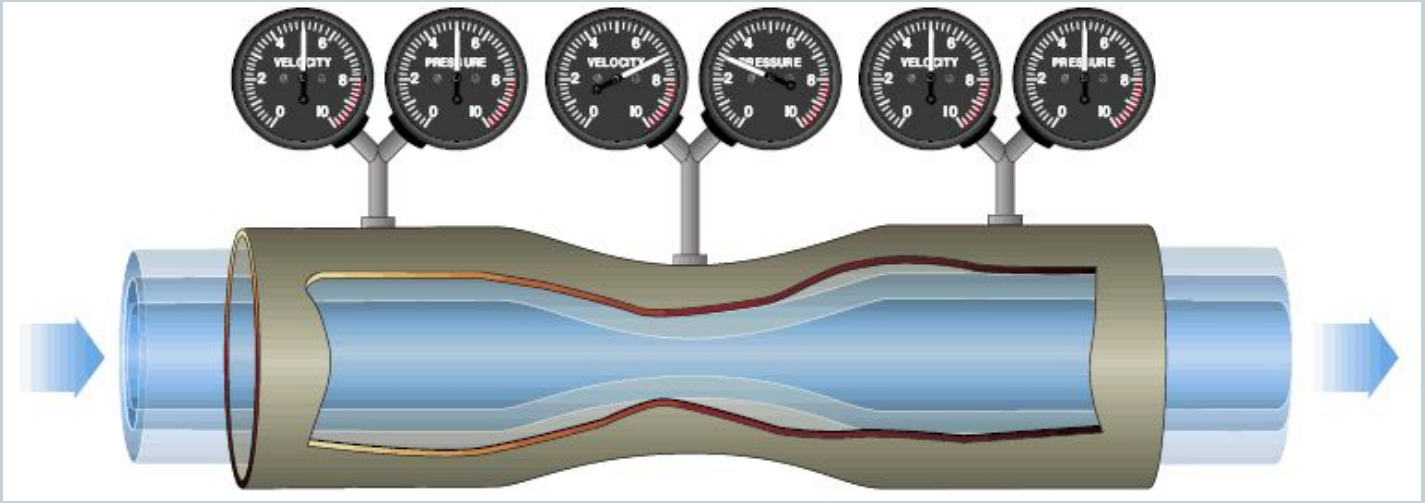
Normal



Chiari I

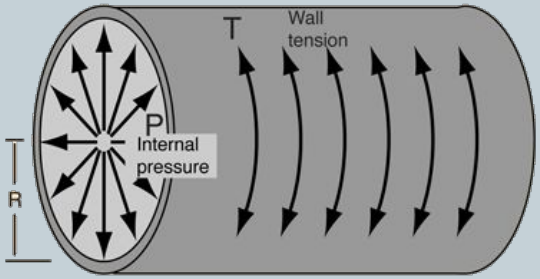


Fluid Mechanics



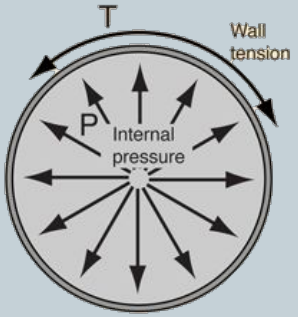
Fluid Mechanics

LaPlace's Law



Cylindrical Vessel

$T = PR$



Spherical Vessel

$T = \frac{PR}{2}$

Bernoulli's principle

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

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.

$\nu^2/2+P=\text{const}$ $T=Pr$ $\nu^2/2+T/r=\text{const}$

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

Poiseuille's law

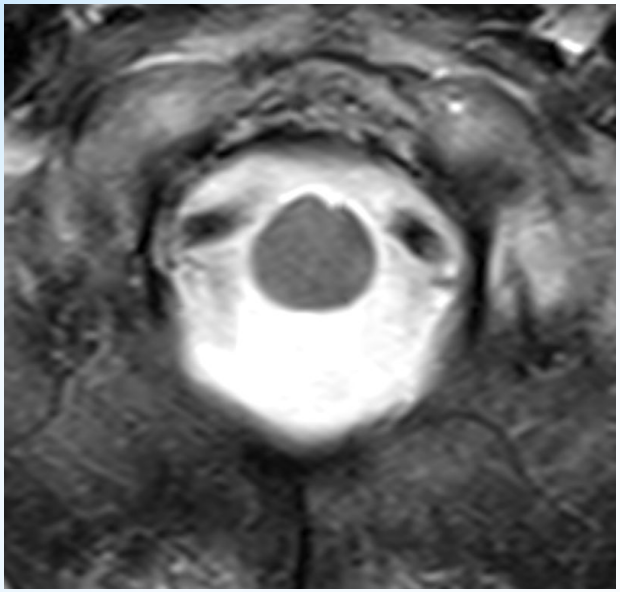
Q	Flow rate
P	Pressure
r	Radius
η	Fluid viscosity
l	Length of tubing

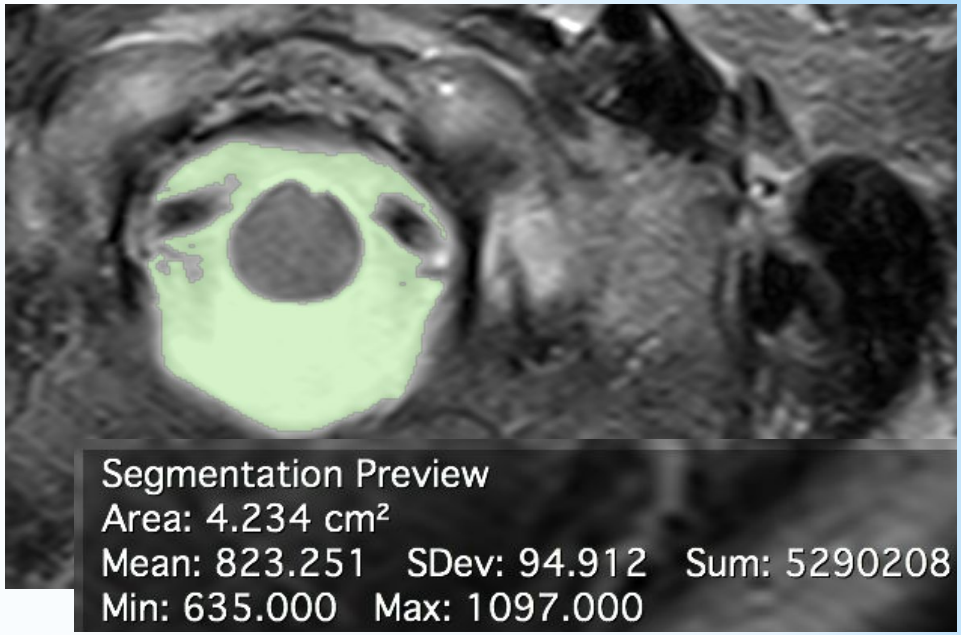
$$Q = \frac{\pi \mathrm{P} r^4}{8 \eta l}$$

- 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.

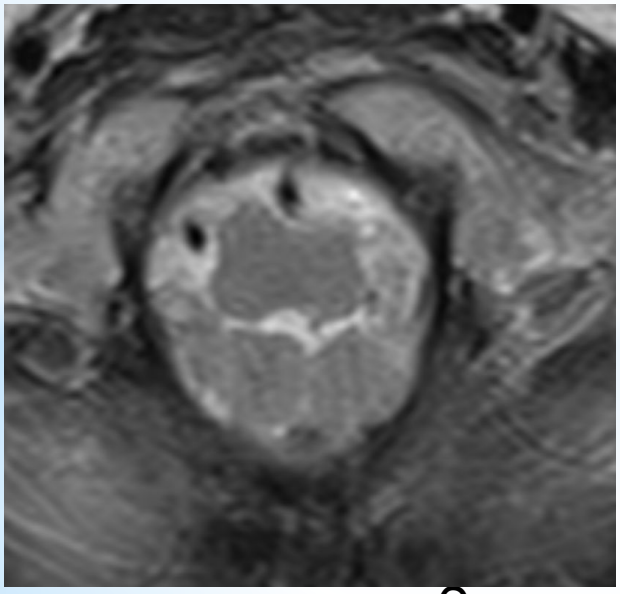
T2 MRI

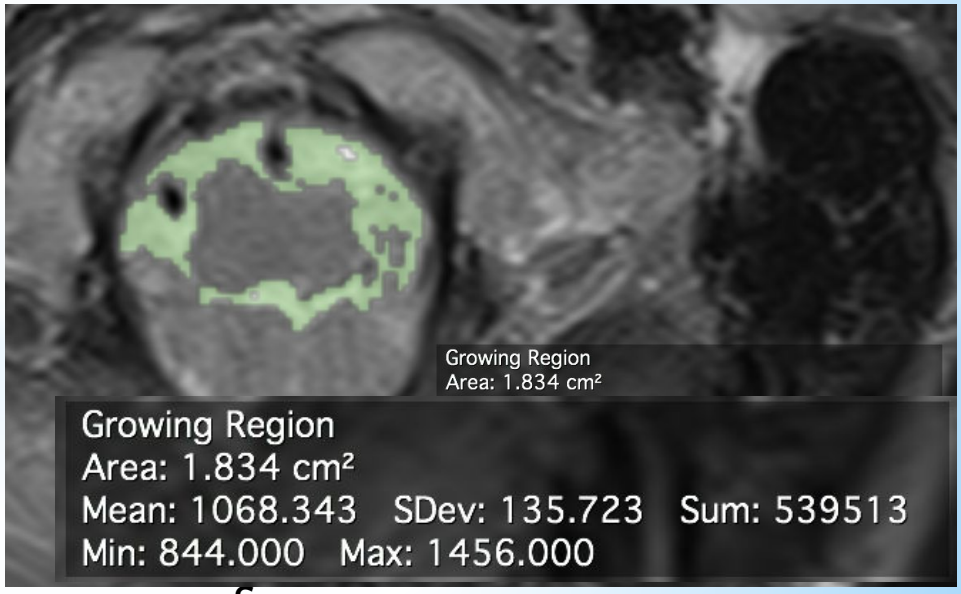
Normal





Chiari I (episodic headache)

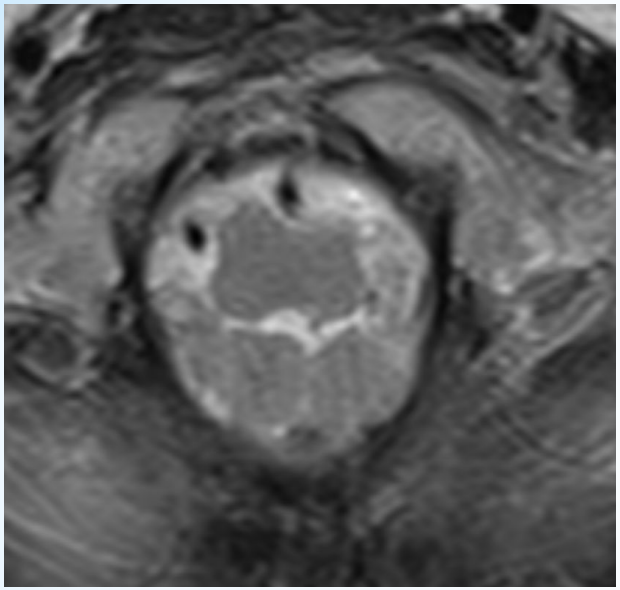


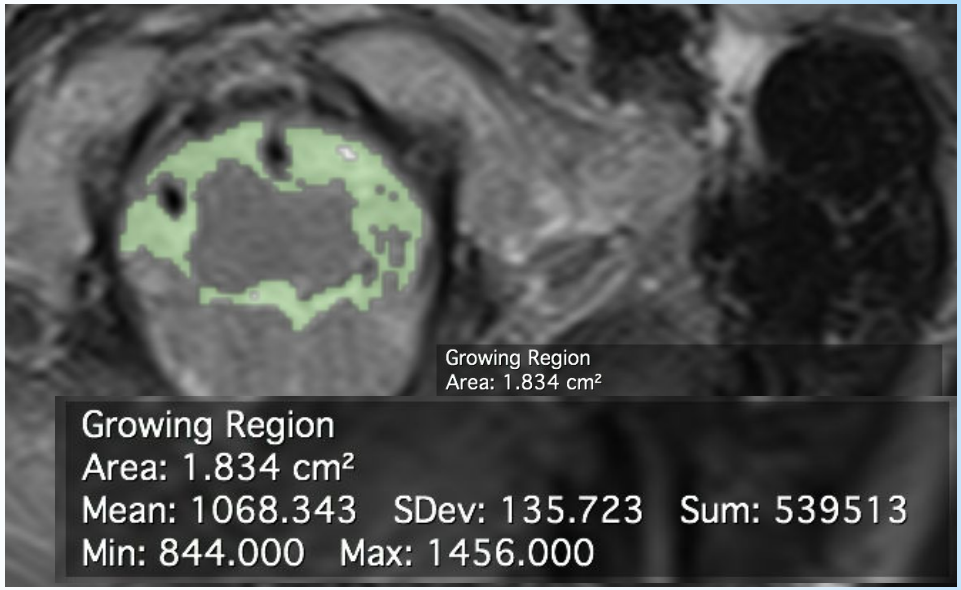


$$Q_{(\text{flow rate})} = \nu_{(\text{velocity})} \cdot S_{(\text{surface})}$$

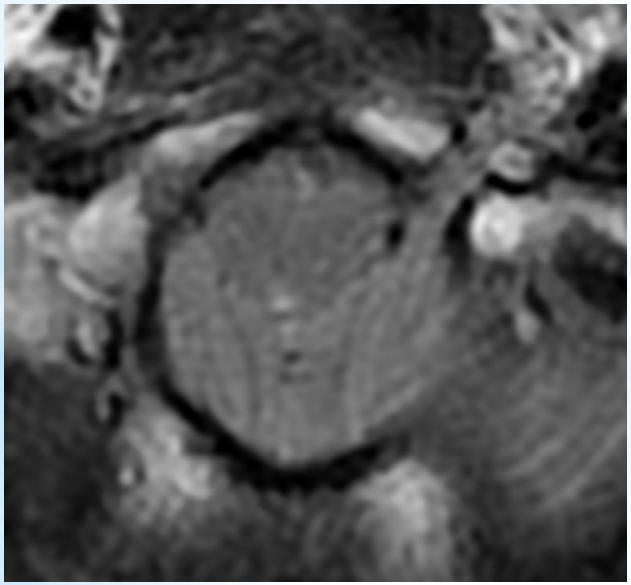
T2 MRI

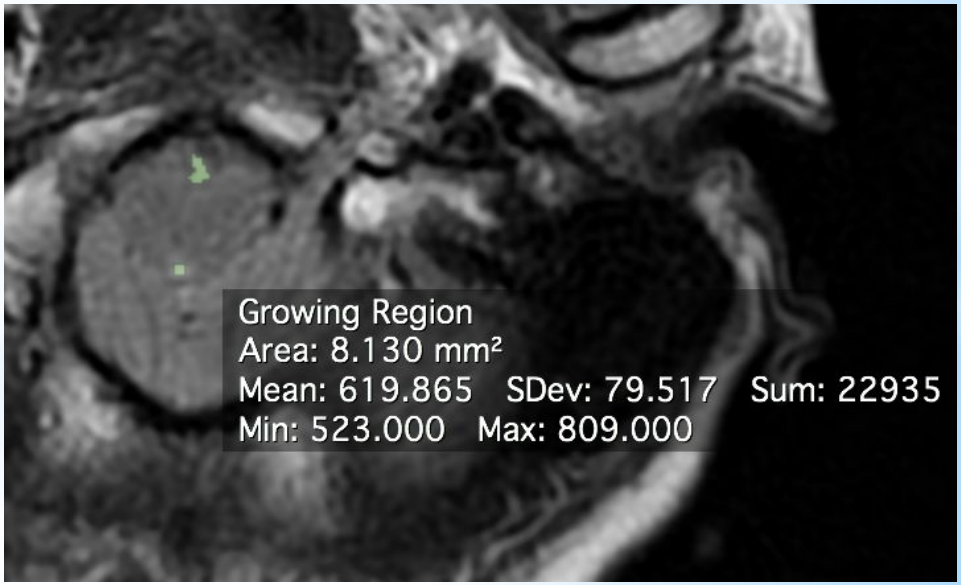
Chiari I (episodic headache)





Chiari I (progressive unremitting headache)





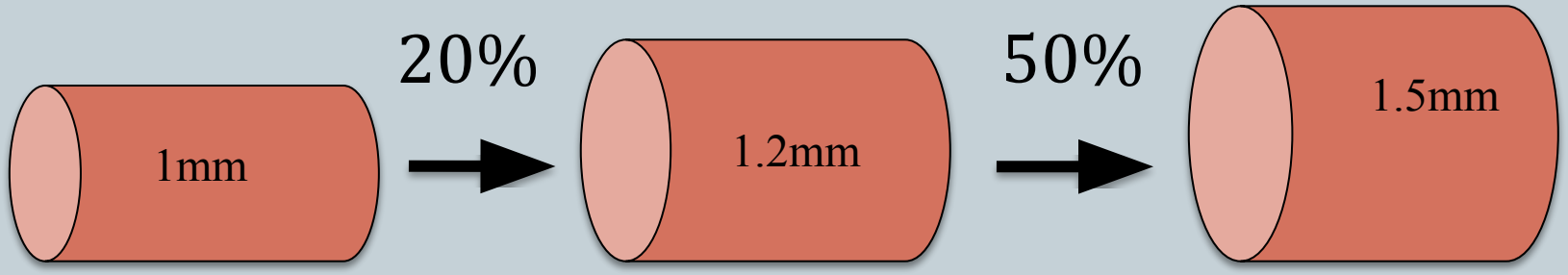
$$Q_{(\text{flow rate})} = \nu_{(\text{velocity})} \cdot S_{(\text{surface})}$$

How much bigger do we need?

100cc/hour

200cc/hour

500cc/hour

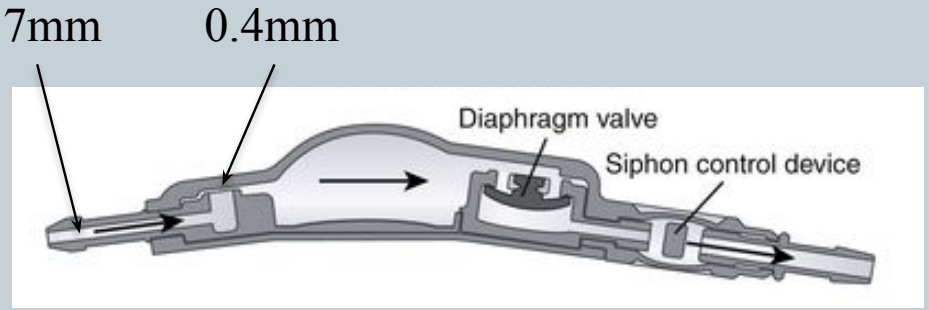


Q	Flow rate
P	Pressure
r	Radius
η	Fluid viscosity
l	Length of tubing

$$Q = \frac{\pi \mathrm{Pr}^4}{8\eta l}$$

Shunt valve

This is an example how a small shut conduit can successfully decrease pressure.



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.

Clinical data

- 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.

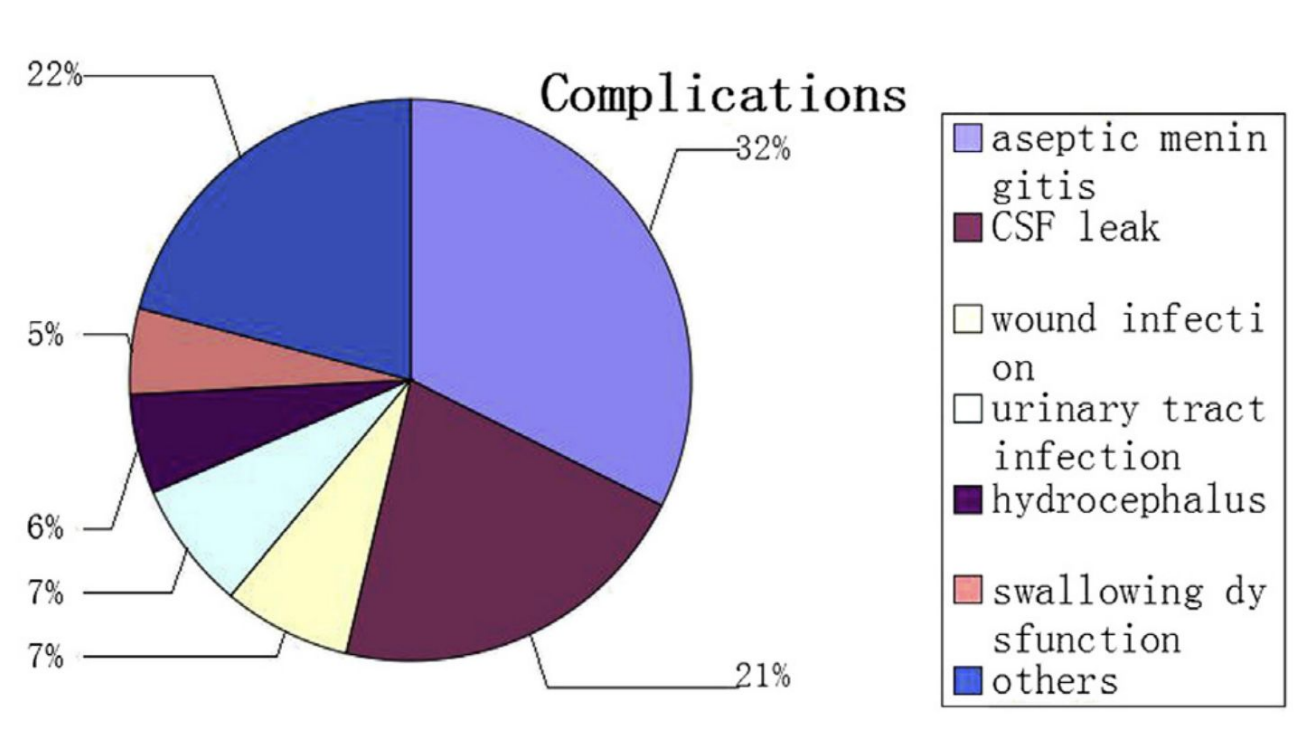
Reference: (Kennedy BC et al., 2015, Lin W et al, 2017)

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



All types of surgical interventions

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].

Complications of Chiari I Surgery

Resection of tonsils

Shunting +/-Decompression

Decompression and duroplasty

Extradural decompression

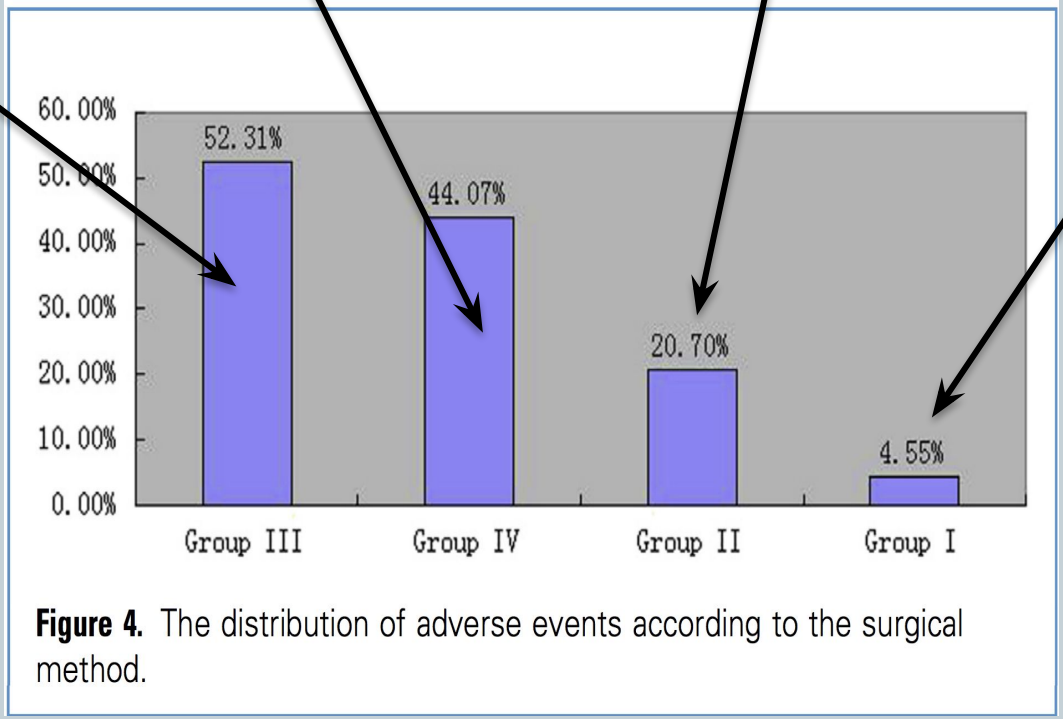


Figure 4. The distribution of adverse events according to the surgical method.

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

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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.

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- 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

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

radius

viscosity

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:

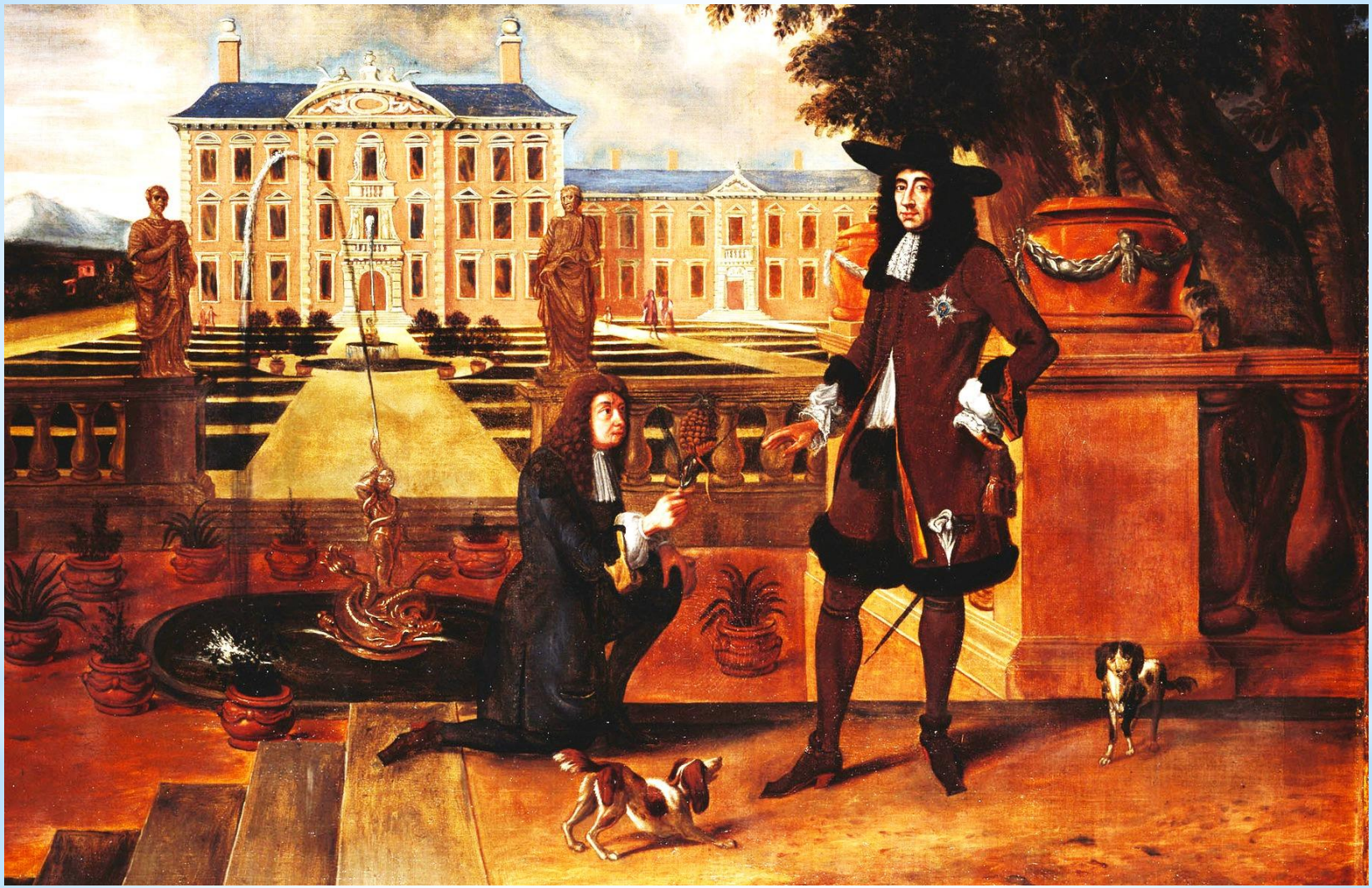
1. Intraoperative Ultrasonography for Definition of Less Invasive Surgical Technique in Patients with Chiari Type I Malformation. Roger SB, Mario AT, Matheus FO, Marcelode LO, Manoel JT, Edson BS, World Neurosurgery Volume 101, May 2017, Pages 466-475
2. Current opinions for treatment of symptomatic 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
  - 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

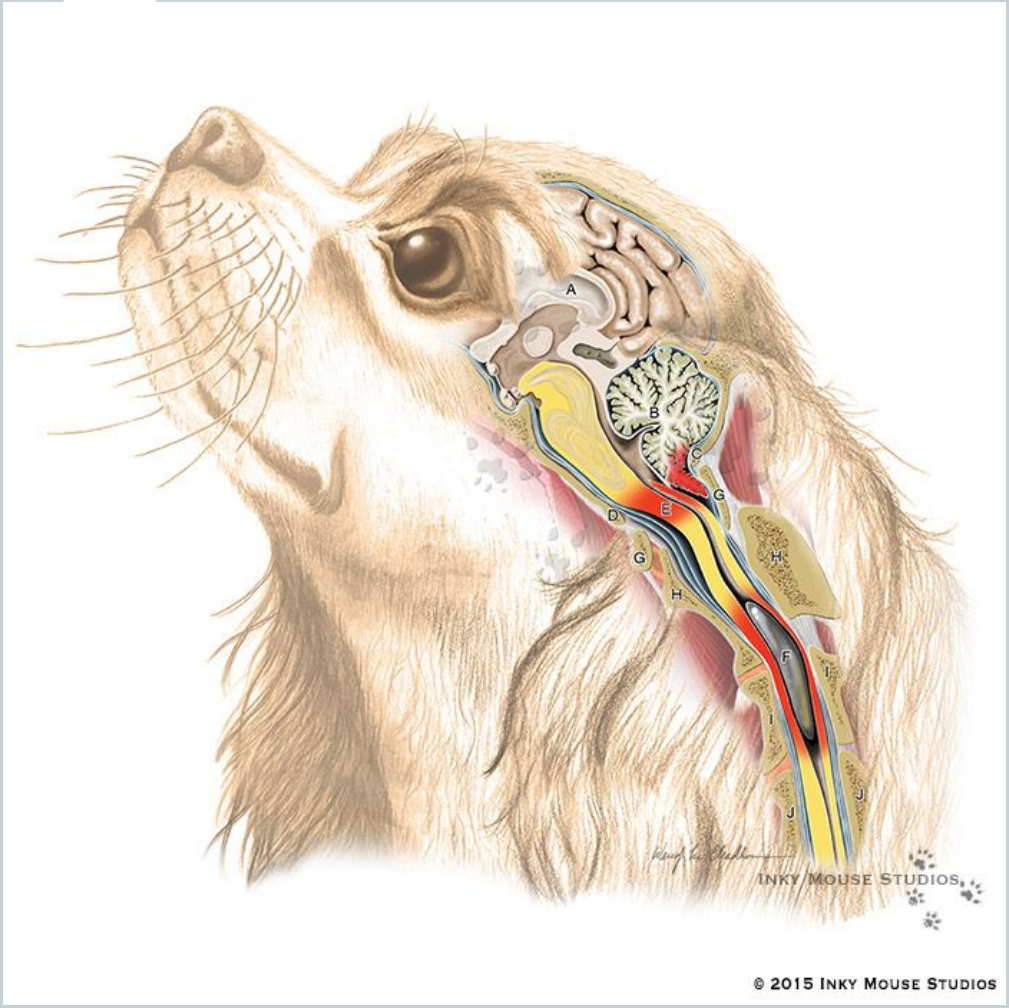
An example from the animal kingdom

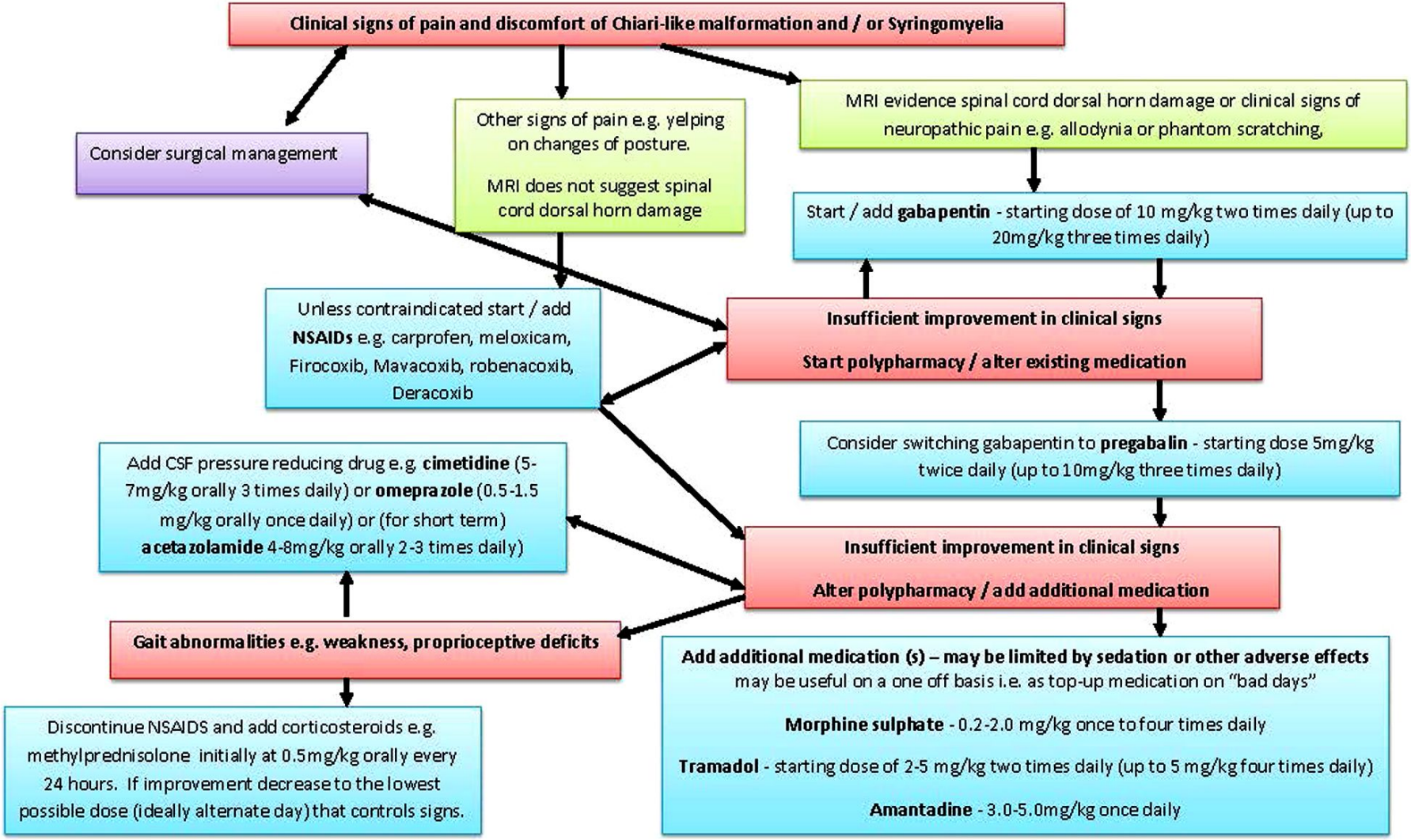


BRITISH SCHOOL, 17TH CENTURY
Charles II Presented with a Pineapple c.1675-80

Chiari in King Charles Cavalier Spaniels

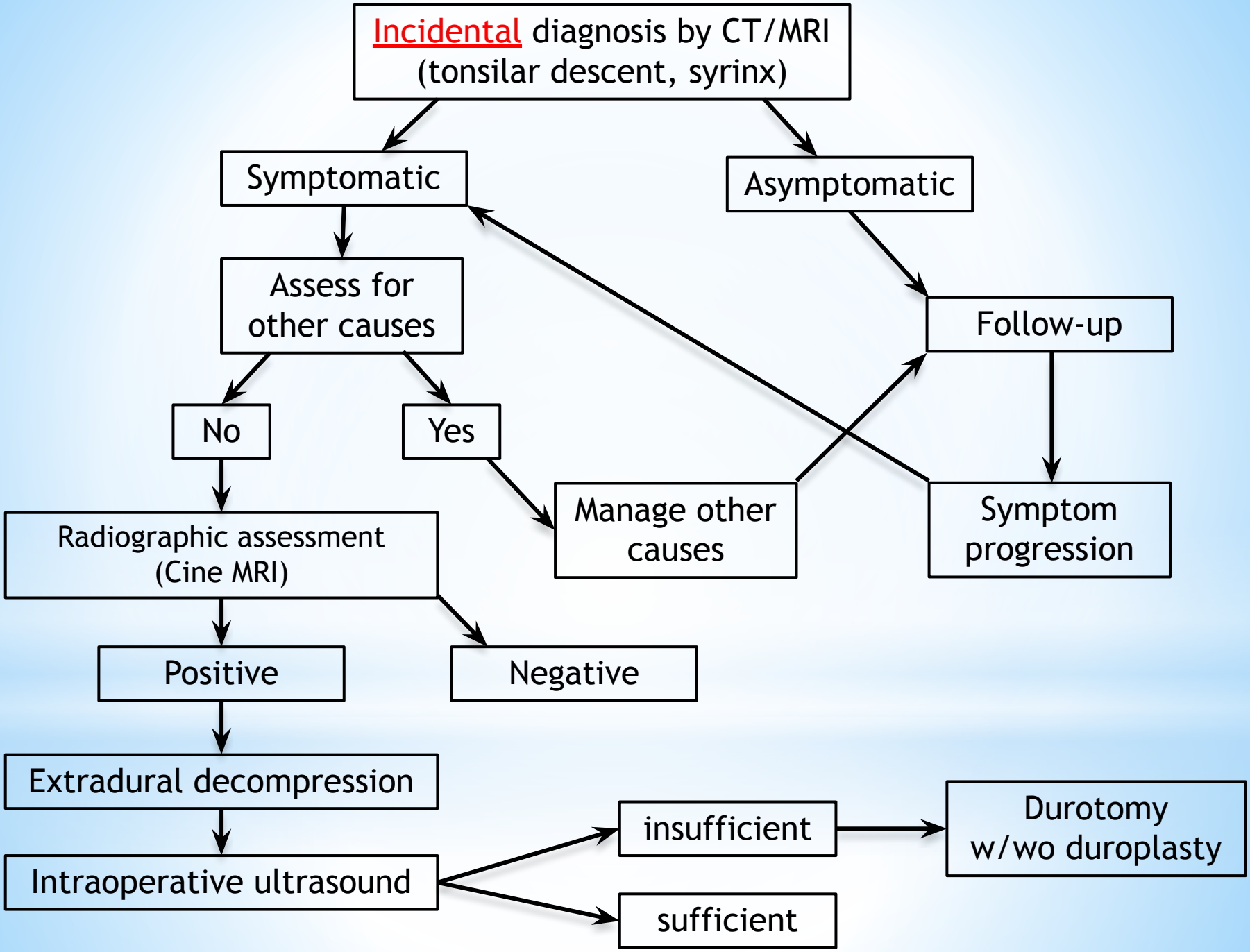
- 90% of King Charles Cavalier Spaniels have Chiari malformation
- 30–70% have syringomyelia (Cerda-Gonzalez, 2015)

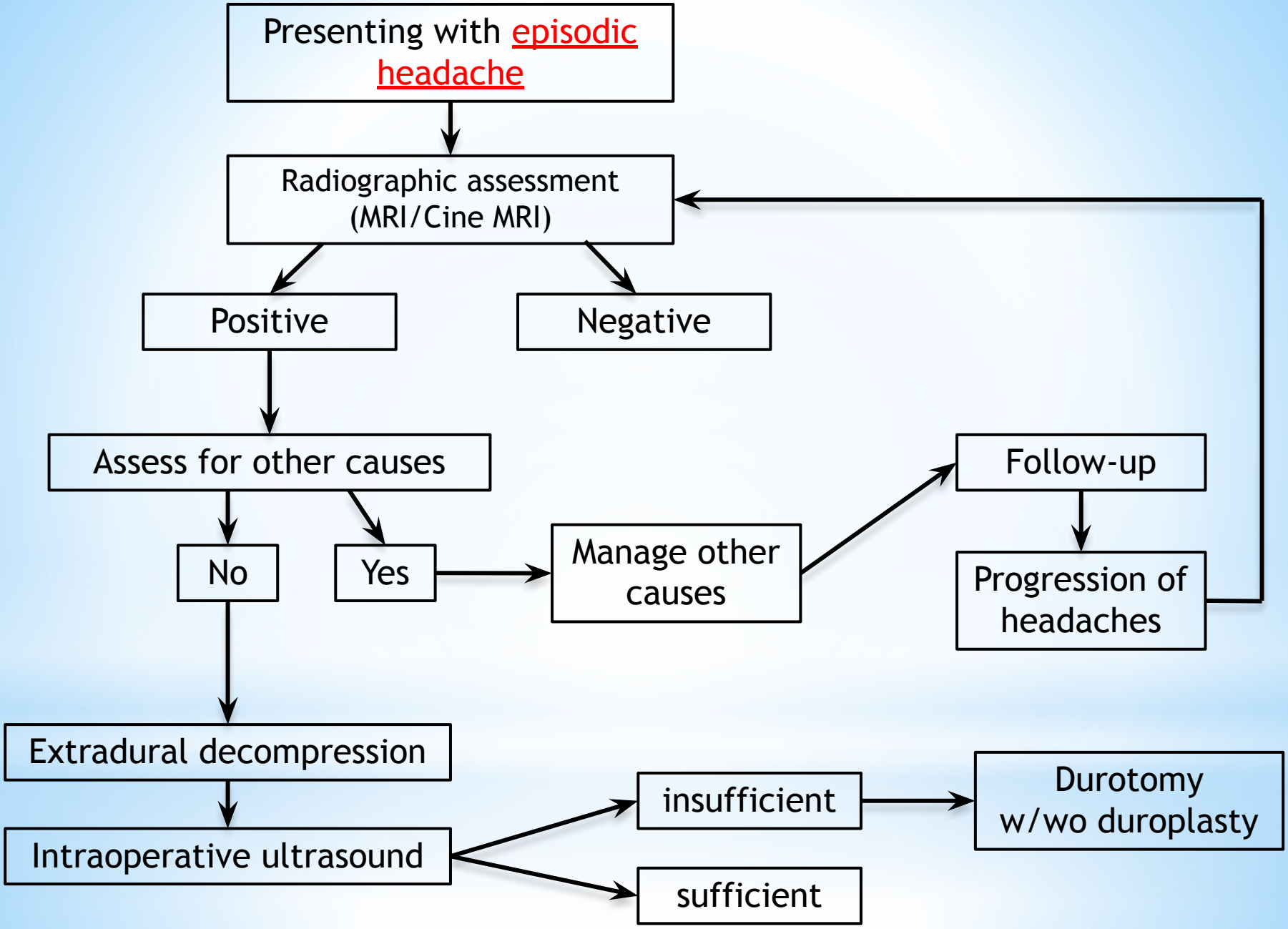


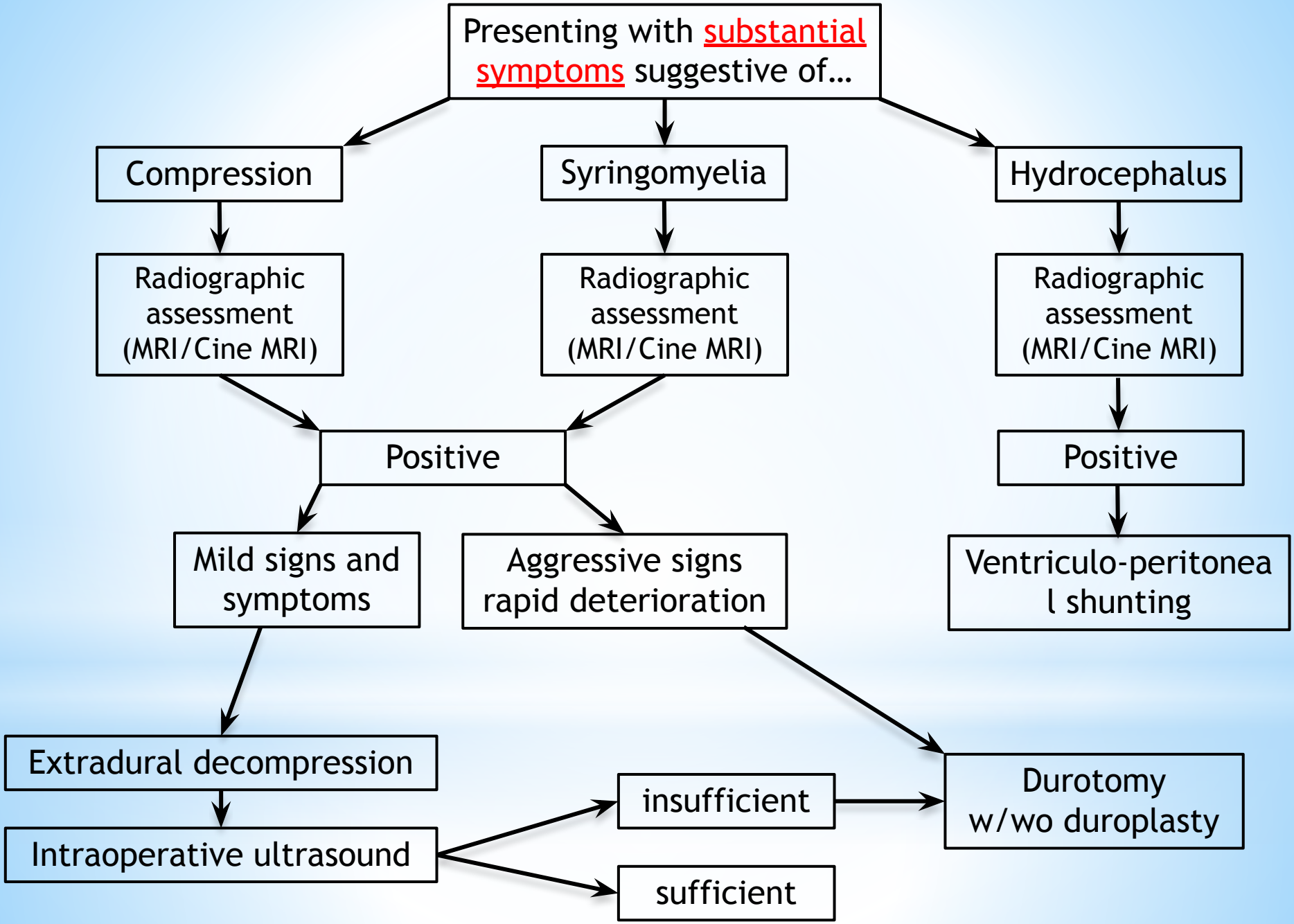


These treatment guidelines are for veterinary surgeons. With the exception of NSAIDS and cimetidine, none of these drugs are licensed for veterinary medicine and no drug is licensed with a specific indication for syringomyelia
Hematology and biochemistry should be monitored on at least a 12 monthly basis for any dog receiving long term medication.

© Clare Rusbridge BVMS PhD DECVN MRCVS 2013


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?
- 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 cine MRI studies be more widely

employed in evaluation of chronic headaches

and neck pain?

THANK YOU