MACQUARIE University

Syringomyelia: Chiari vs SIH

Marcus Stoodley

MACQUARIE University

Disclosures: Nil

Chiari malformation

Syringomyelia

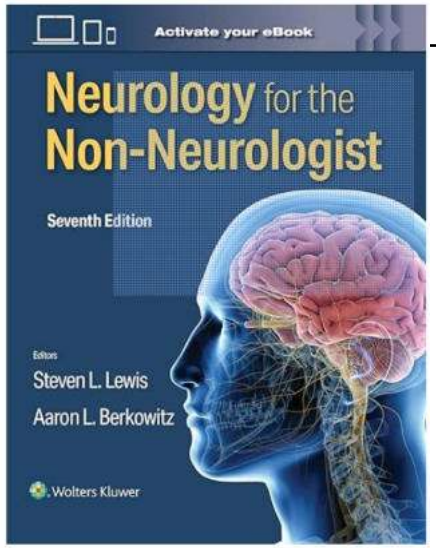
SIH + Chiari/Syrinx

Iatrogenic intracranial hypotension

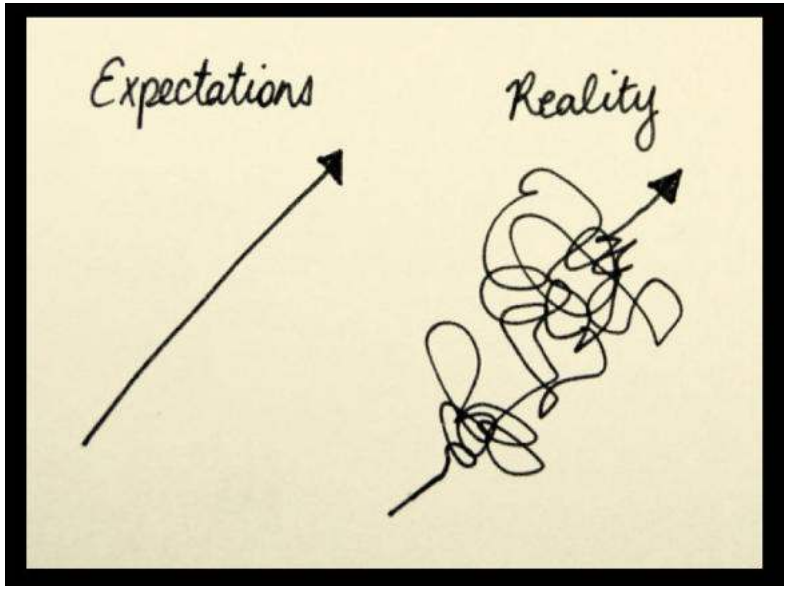
Chiari and Syringomyelia

MACQUARIE University

1. Textbook



2. The real world



https://www.timclue.com/blog/expectations-vs-reality

Chiari malformation

- Cerebellar tonsil herniation into spinal canal
  - ‘≥ 5 mm’

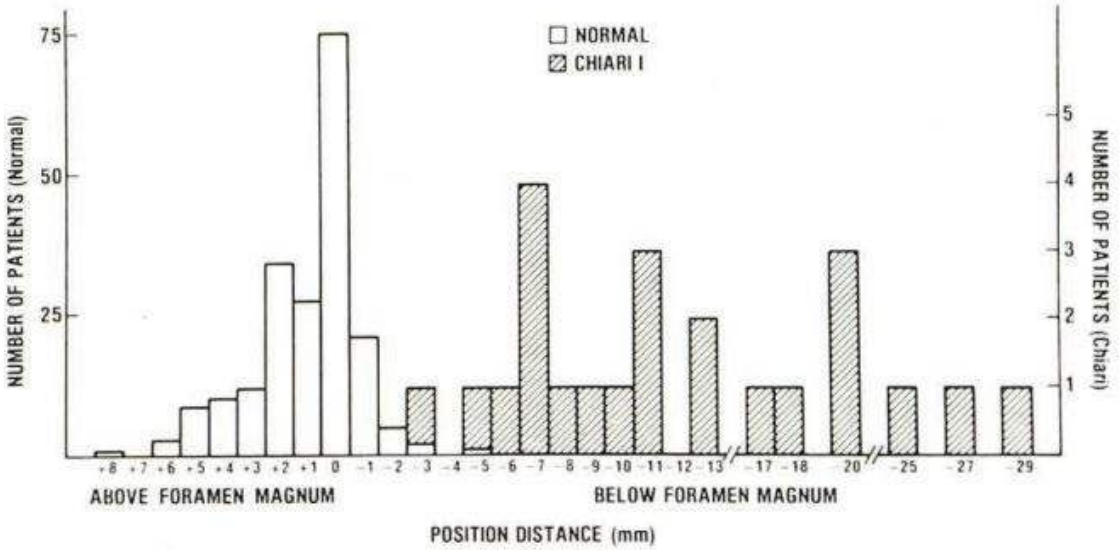


Fig. 3.—Histogram showing tonsillar positions in 200 normal and 25 Chiari I patients. Open boxes represent normal patients. Cross-hatched boxes represent patients with Chiari malformation.

Barkovich et al
AJNR 7:795–799, September/October 1986
0195–6108/86/0705–0795

Chiari malformation

- Cerebellar tonsil herniation into spinal canal
  - ‘≥ 5 mm’

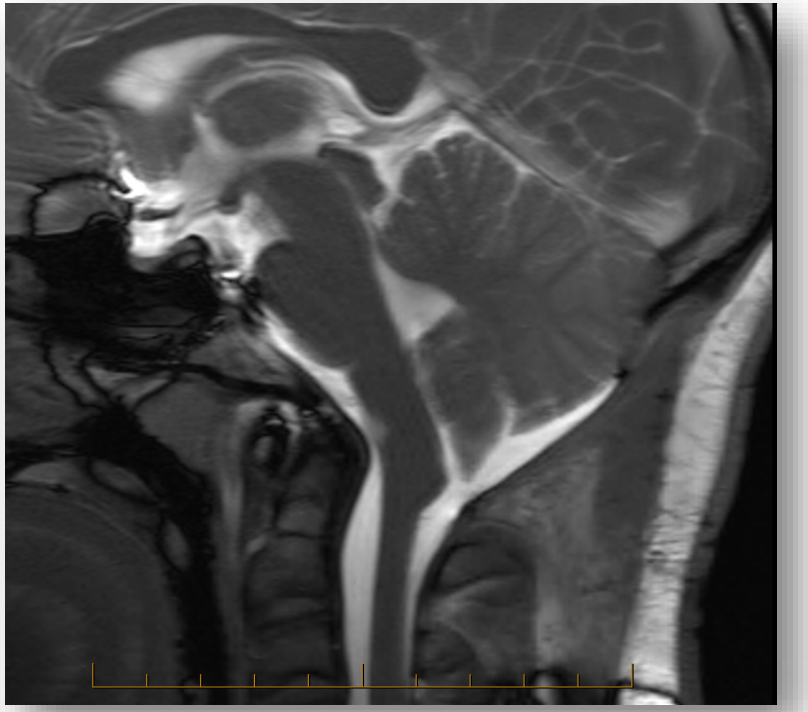
Pre-op

Post-op

Chiari malformation

- Cerebellar tonsil herniation into spinal canal
  - ‘≥ 5 mm’
  - Often associated skull base abnormalities
    - Platybasia, atlas assimilation, acute clival-axis angle

Pre-op



Post-op

Chiari malformation

- Cerebellar tonsil herniation into spinal canal
  - ‘≥ 5 mm’
  - Often associated skull base abnormalities
    - Platybasia, atlas assimilation, acute clival-axis angle
  - More common in EDS
    - Also have cranio-cervical instability → postural headache/neck pain!

Chiari malformation

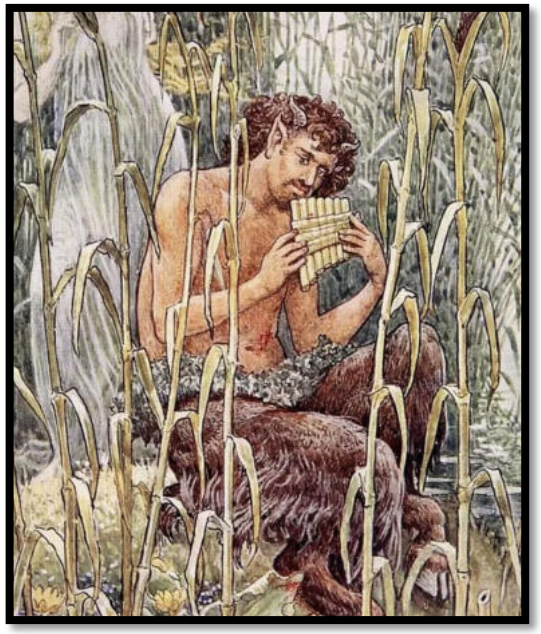
- Cerebellar tonsil herniation into spinal canal
  - ‘≥ 5 mm’

Pre-op

Post-op

Syringomyelia

Jan Brueghel the Younger
1601 – 1678



https://flutealmanac.com/syrinxs-serenade-the-mythical-origin-of-the-pan-flute/

Syringomyelia

Cesar Pietersz. (Bovetius) van EVERDINGEN
Alkmaar c. 1617-Alkmaar 1678

Dirck van der Lisse (fl.c.1635 - d.1669)

Pan Pursuing Syrinx c. 1589
Hendrick Goltzius

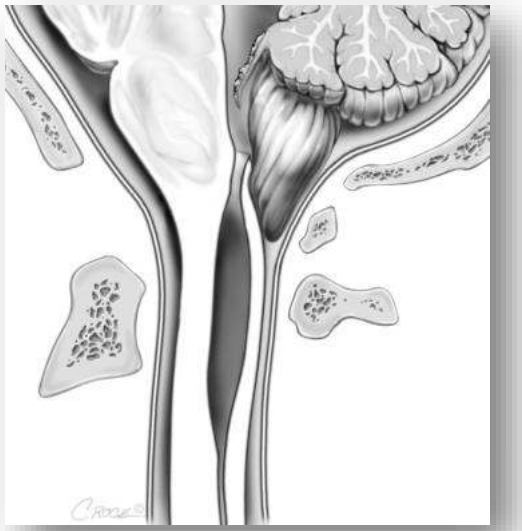
Gerard Hoet (I)
Pan and Syrinx, c. 1700

Willem de Heusch (1638–1692)

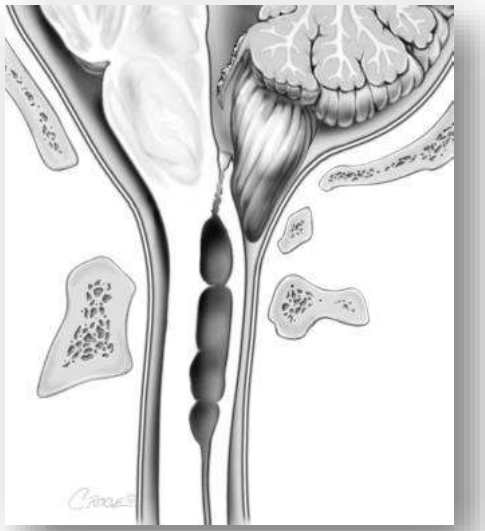
Anthonie Waterloo, 1609 - 1690

Franciscus Xaverius Xavery, 1770

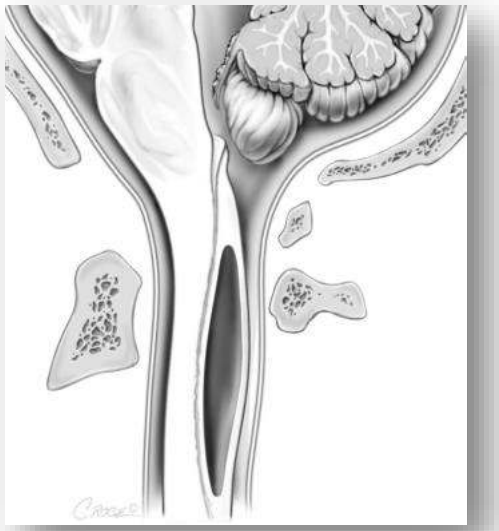
Syringomyelia



Communicating canalicular



Non-communicating canalicular

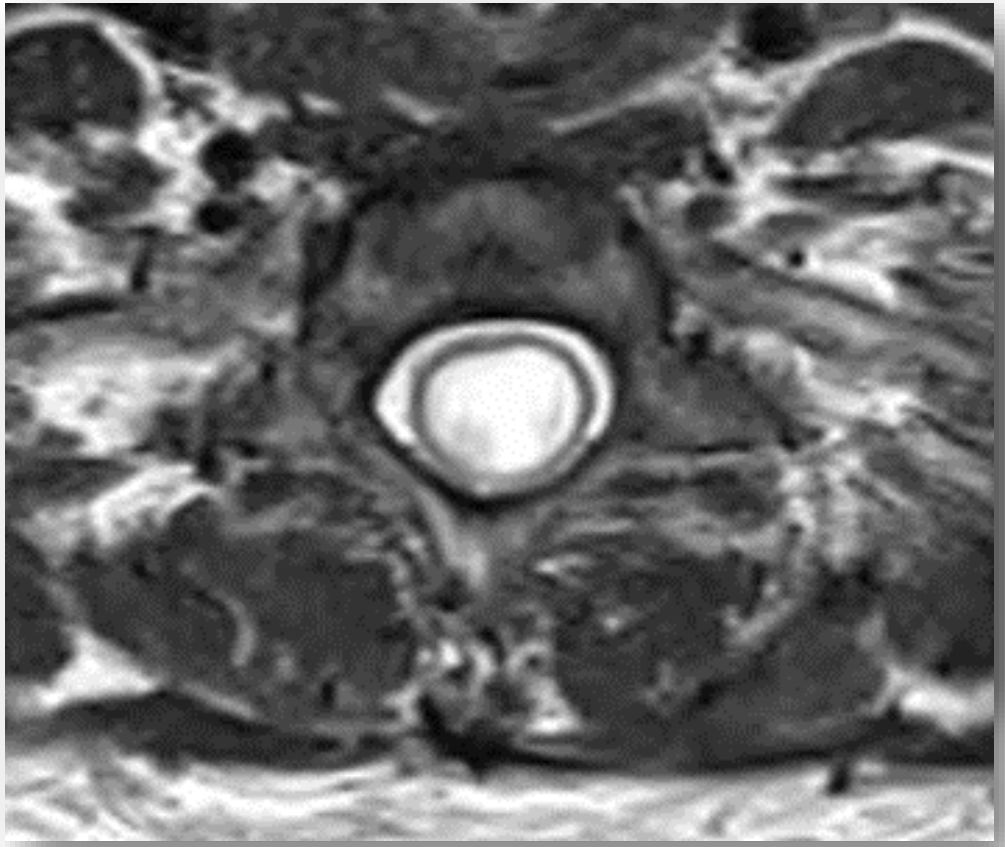


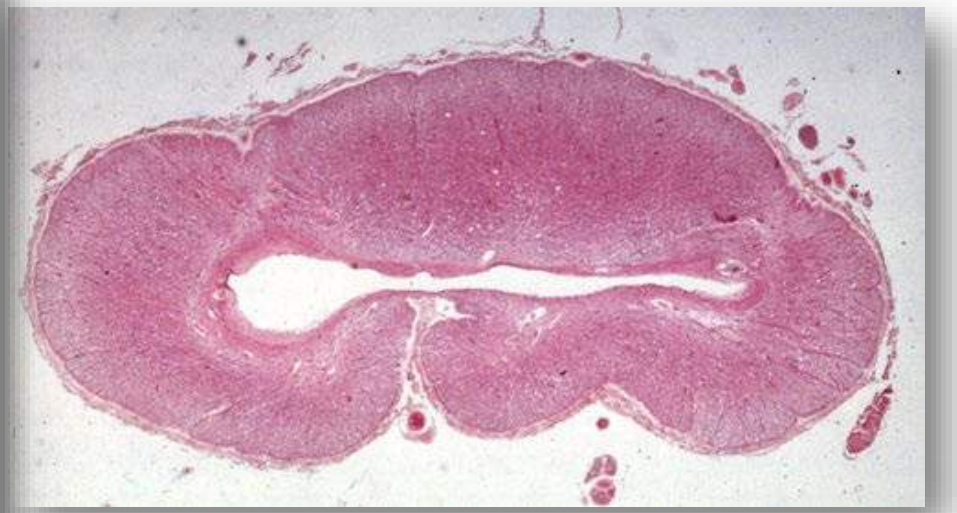
Extracanalicular

Syringomyelia

MACQUARIE University

"Destruction or degeneration … leads to cavitation and accumulation of fluid"

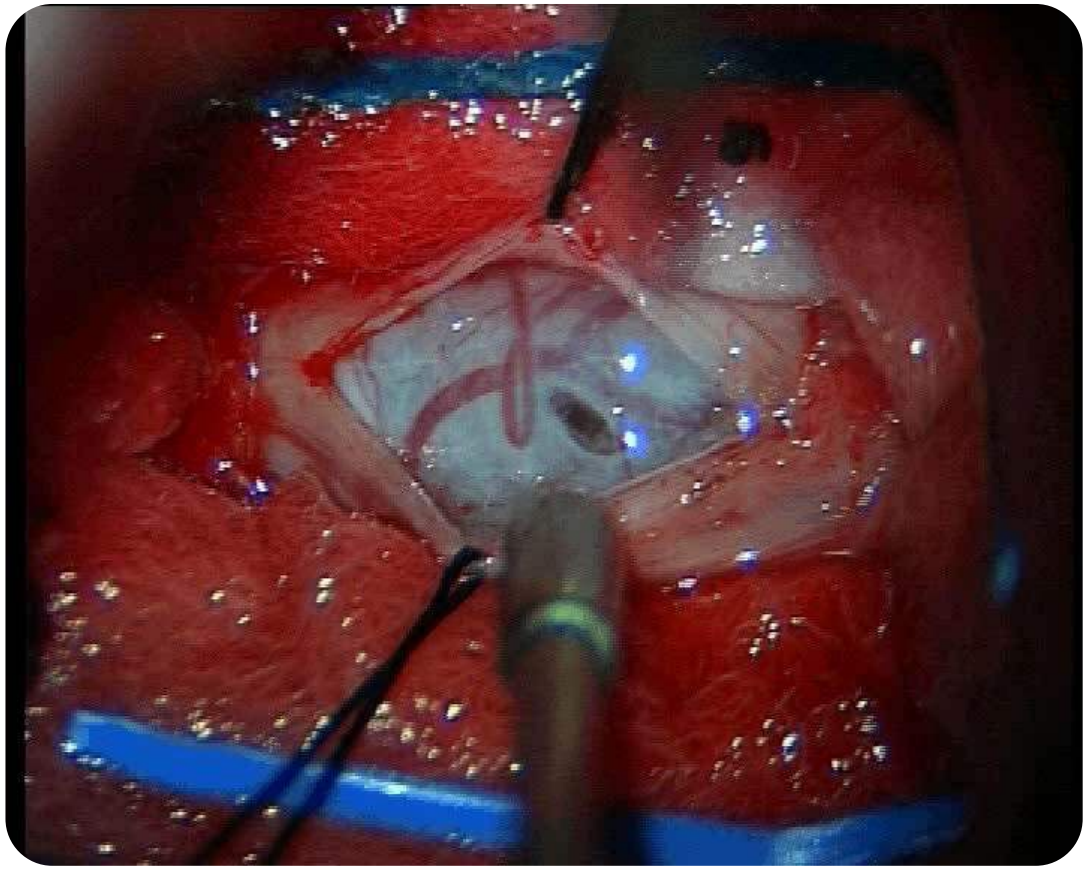




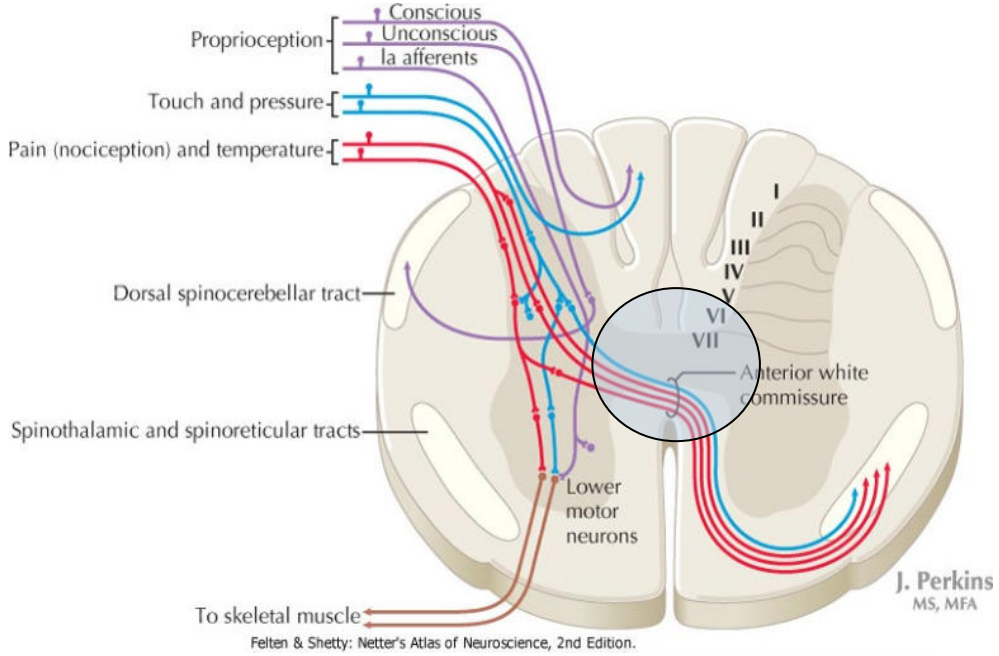
Neuropathology-web.org

Syringomyelia

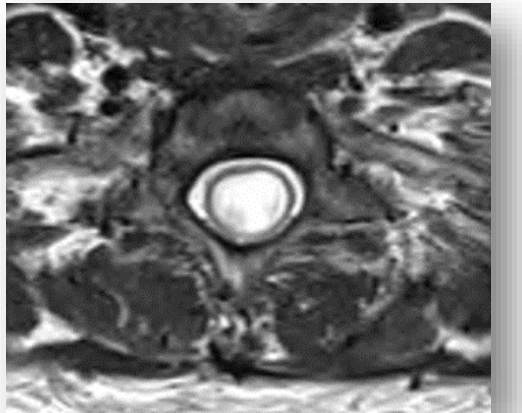
MACQUARIE University

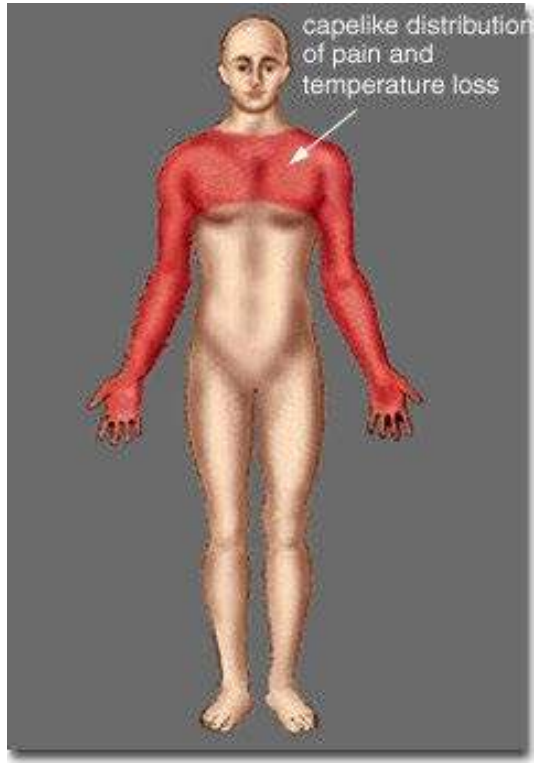


Syringomyelia



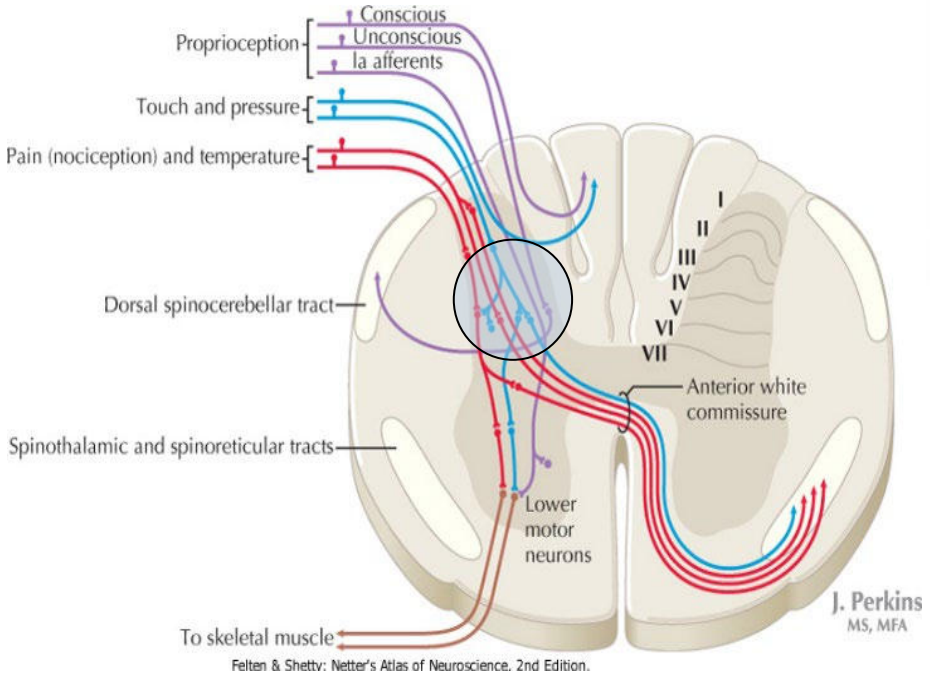
Felten & Shetty: Netter's Atlas of Neuroscience, 2nd Edition.

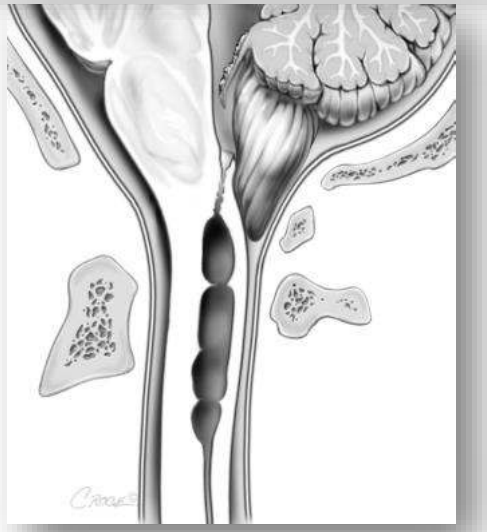




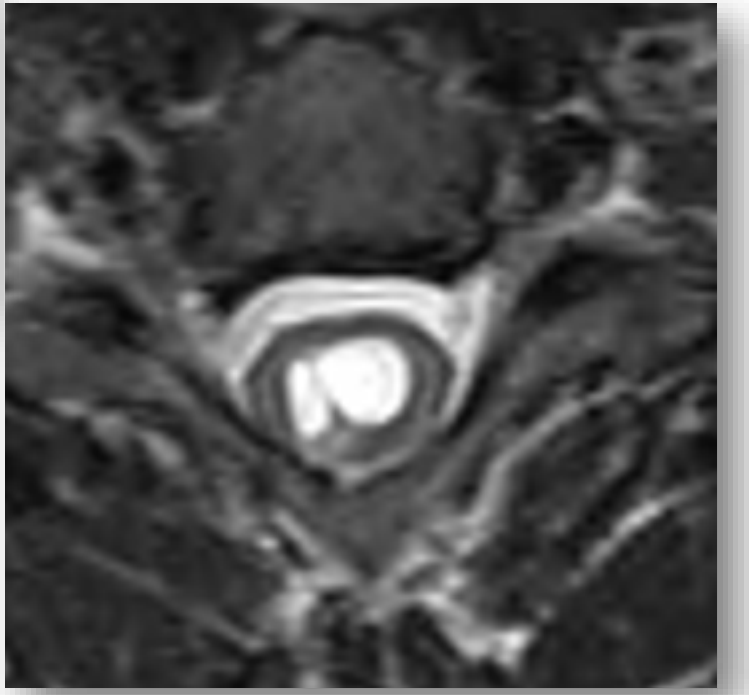
www.intermed.med.uottawa.ca

Syringomyelia



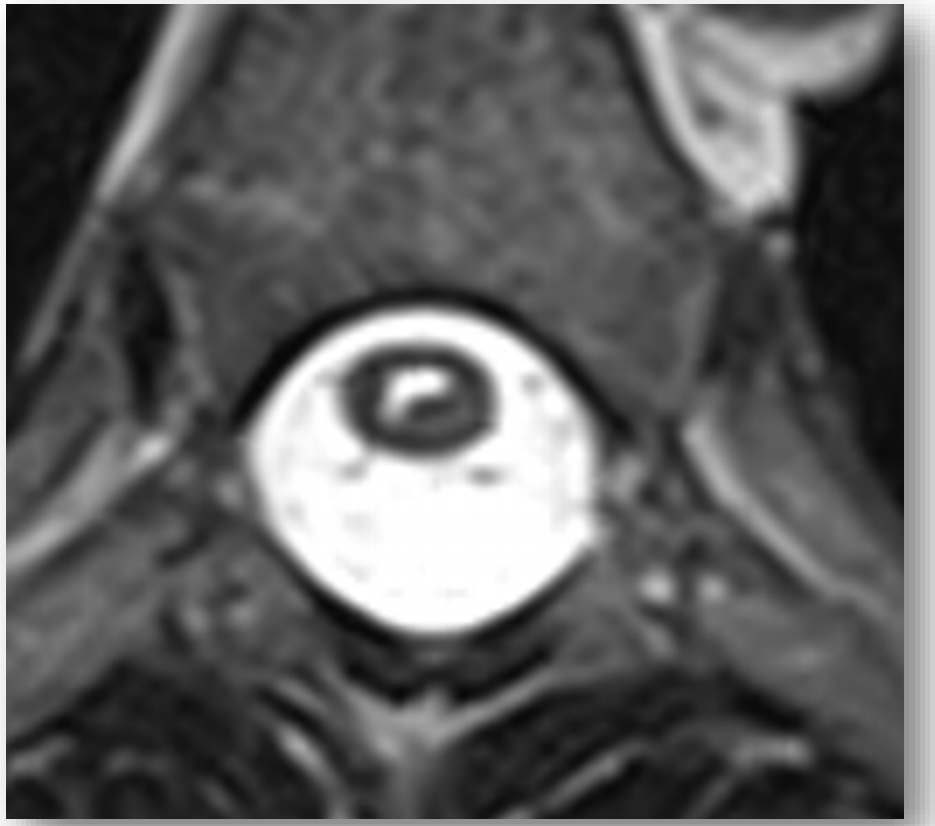


Non-communicating canalicular



Syringomyelia

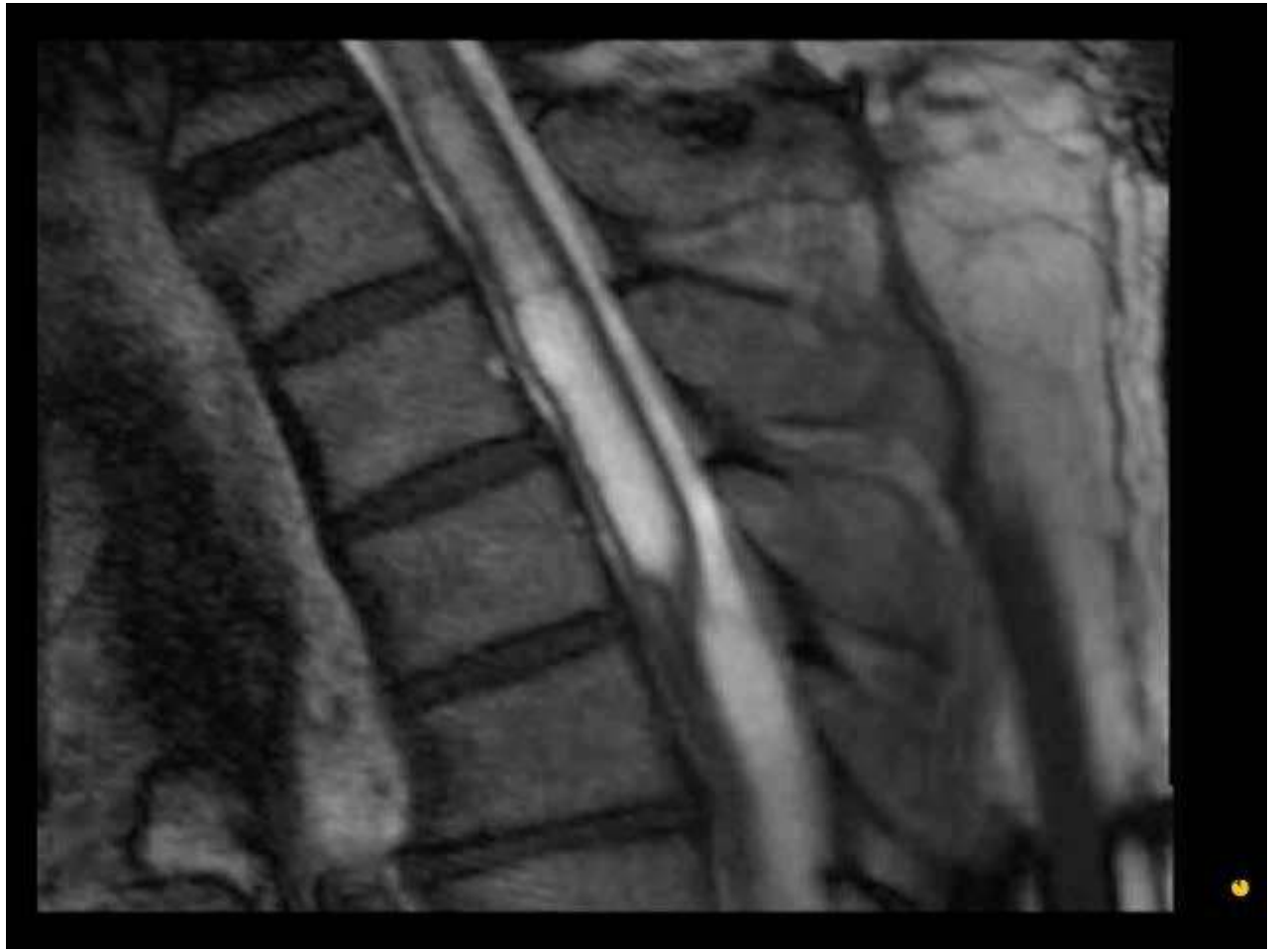
MACQUARIE University



Syringomyelia

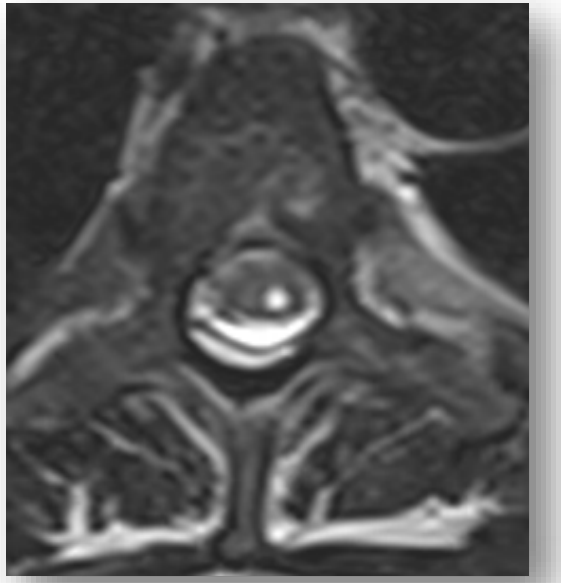
MACQUARIE University

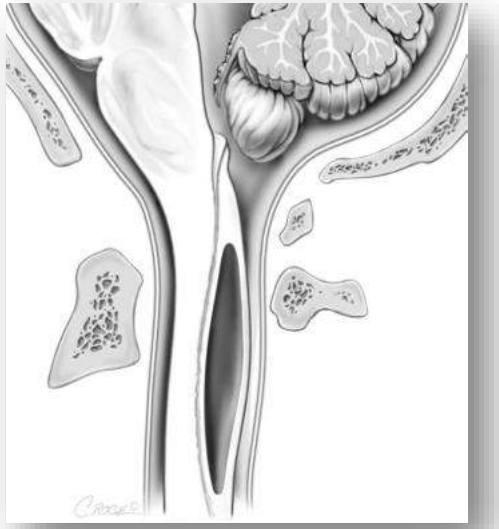
- Spinal subarachnoid obstruction



Syringomyelia

- Spinal arachnoiditis

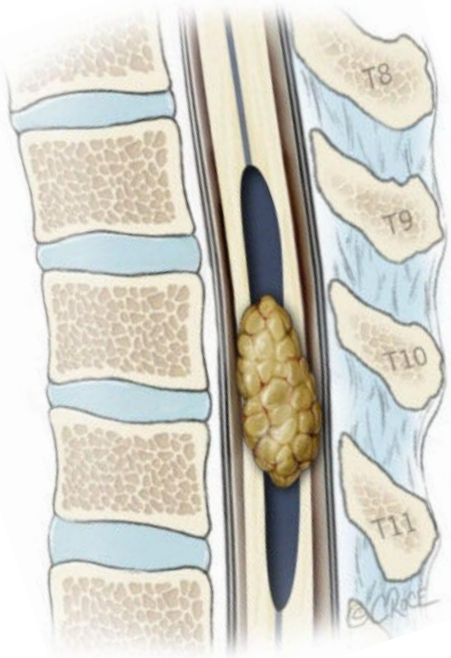




Extracanalicular

Syringomyelia

- Tumour



- Tethered cord

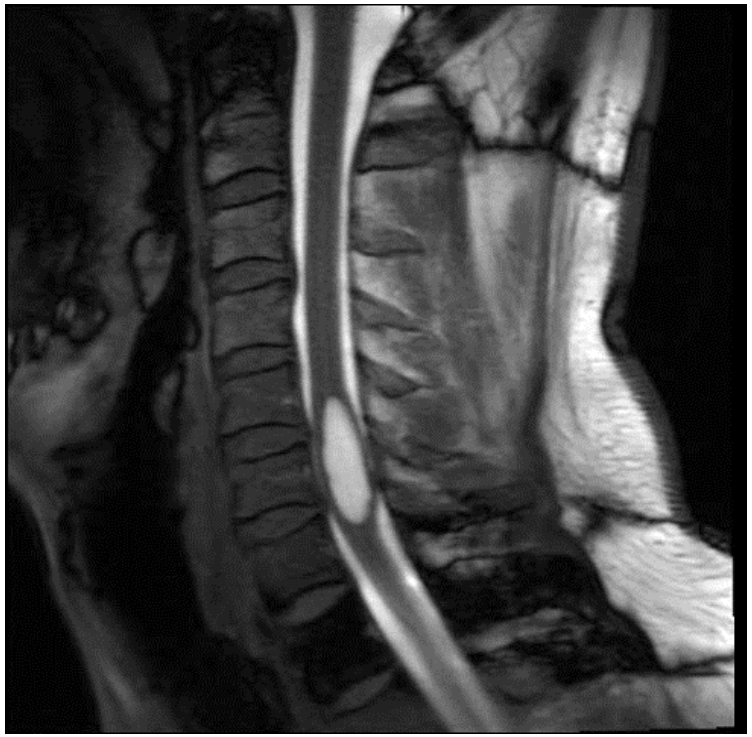
Syringomyelia

MACQUARIE University

- Uncertain cause?

Syringomyelia

- ‘Idiopathic’



Syringomyelia

MACQUARIE University

- Cranio-cervical junction pathology
  - Chiari malformation
  - Arachnoiditis: haemorrhage, trauma, surgery
  - Tumours
  - Arachnoid cysts

Chiari + syringomyelia

MACQUARIE University

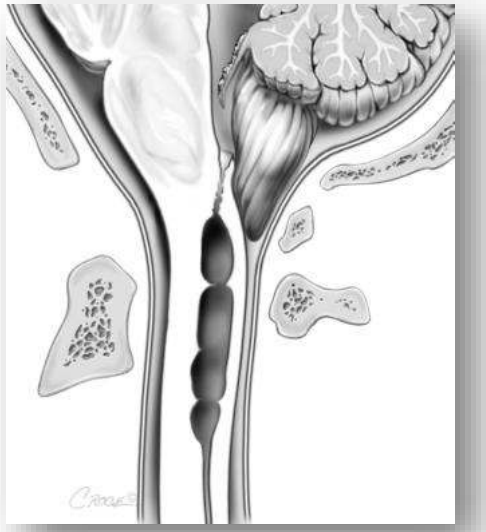
- ~ 50% of Chiari patients

Chiari + syringomyelia

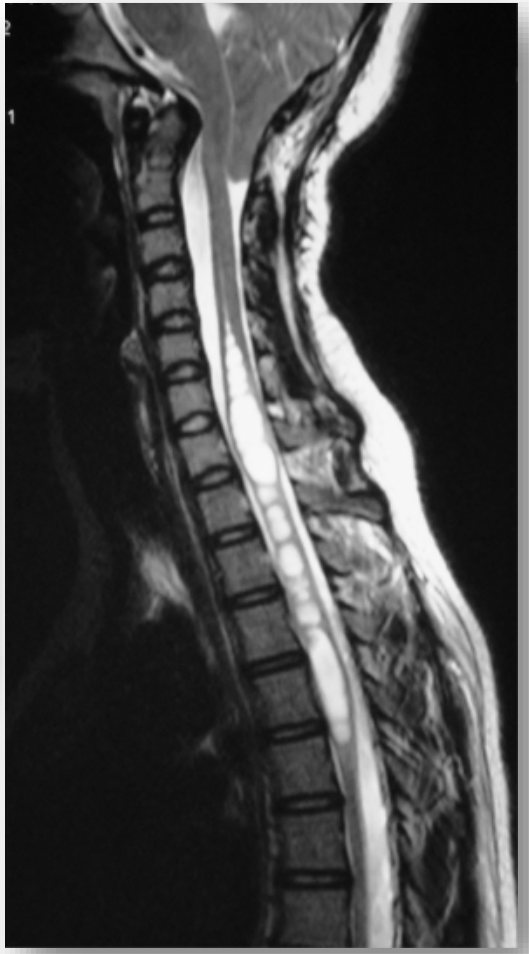
- ~ 50% of Chiari patients
- Pathogenesis unknown
  - CSF from SAS? (not from 4th ventricle)

Gardner

Williams



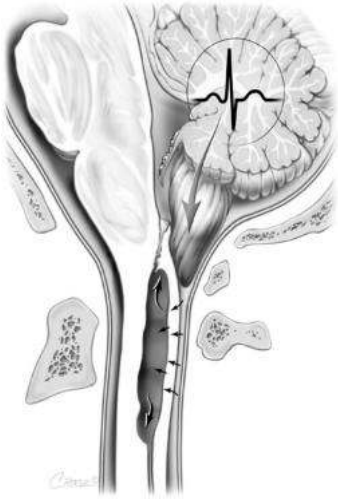
Non-communicating canalicular



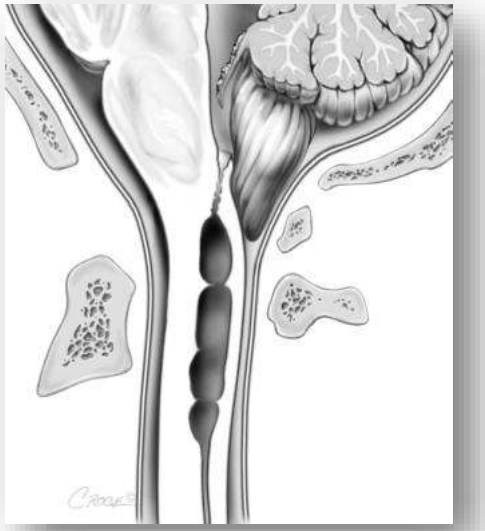
Chiari + syringomyelia

- ~ 50% of Chiari patients
- Pathogenesis unknown
  - CSF from SAS? (not from 4th ventricle)

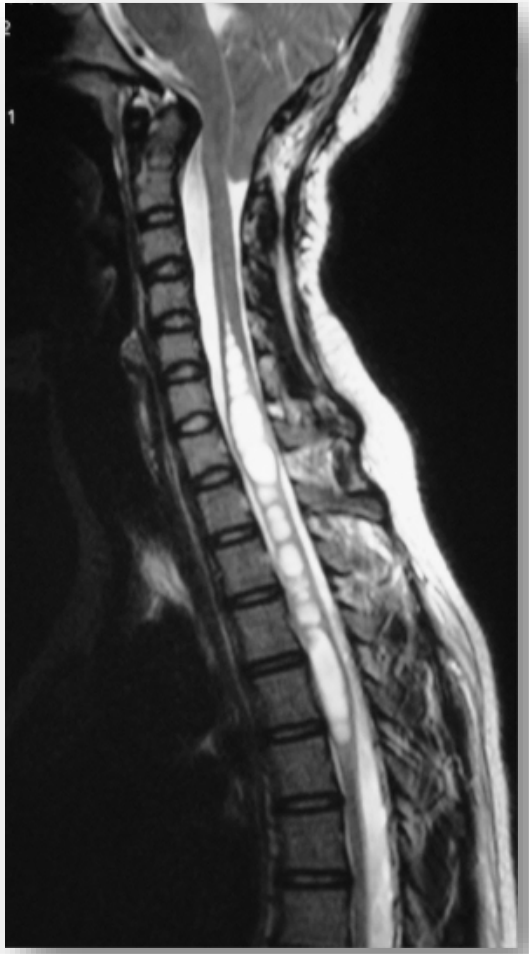
Ball & Dayan



Oldfield



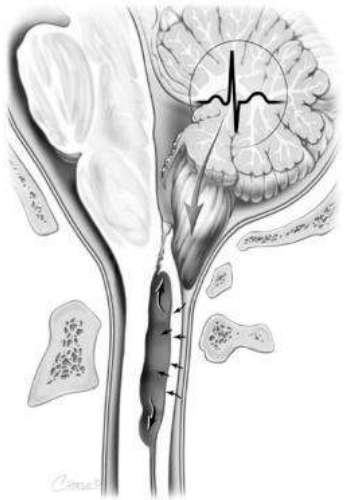
Non-communicating canalicular



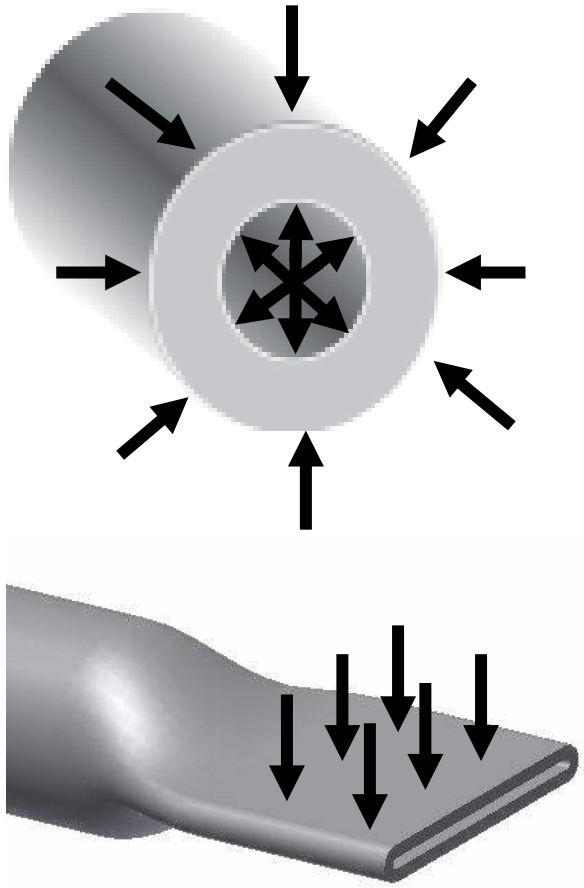
Chiari + syringomyelia

- ~ 50% of Chiari patients
- Pathogenesis unknown
  - CSF from SAS? (not from 4th ventricle)

Ball & Dayan



Oldfield

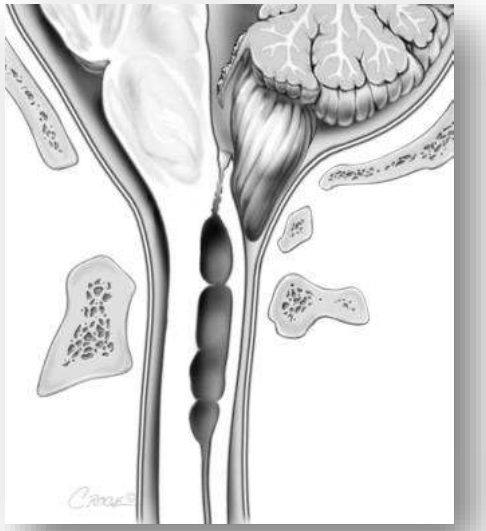


Chiari + syringomyelia

- ~ 50% of Chiari patients
- Pathogenesis unknown
  - CSF from SAS? (not from 4th ventricle)

Pre-op

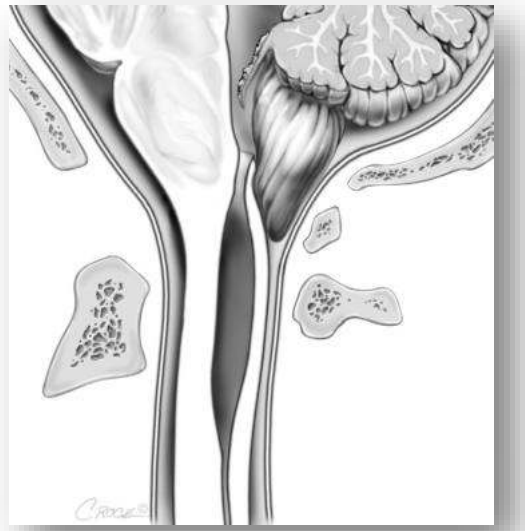
Post-op



Non-communicating canalicular

Chiari + syringomyelia

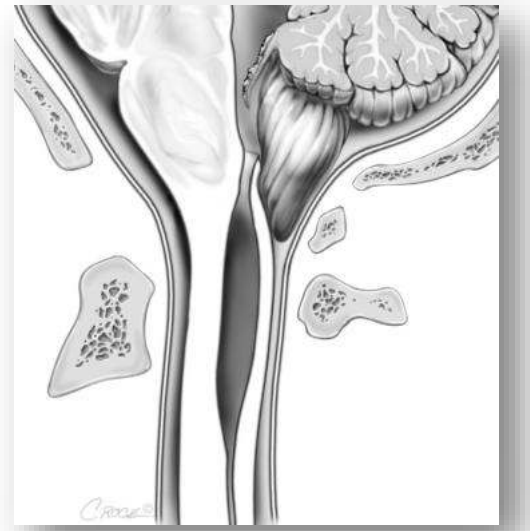
- ~ 50% of Chiari patients
- Pathogenesis unknown
  - Sometimes is communicating



Communicating canalicular

Chiari + syringomyelia

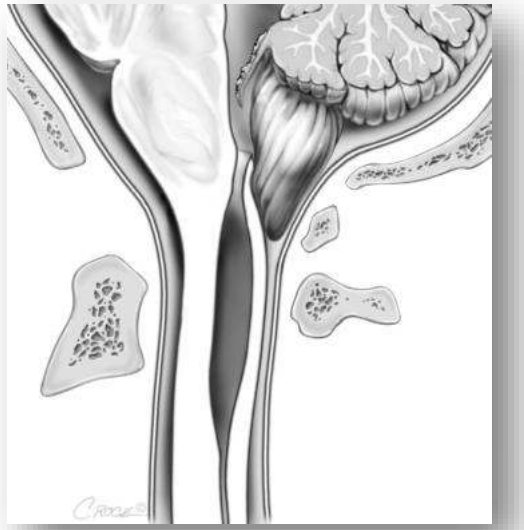
- ~ 50% of Chiari patients
- Pathogenesis unknown
  - Sometimes is communicating



Communicating canalicular

Chiari + syringomyelia

- ~ 50% of Chiari patients
- Pathogenesis unknown
  - Sometimes is communicating

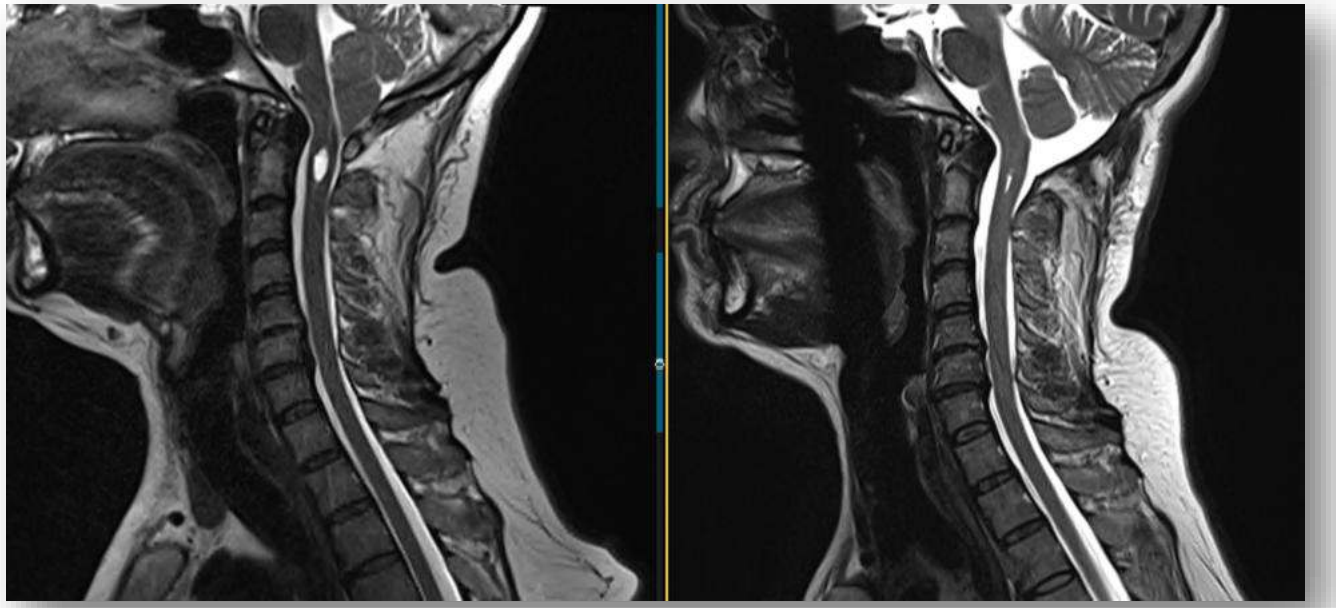


Communicating canalicular

Chiari + syringomyelia

MACQUARIE University

- ~ 50% of Chiari patients
- Pathogenesis unknown
  - Sometimes is communicating



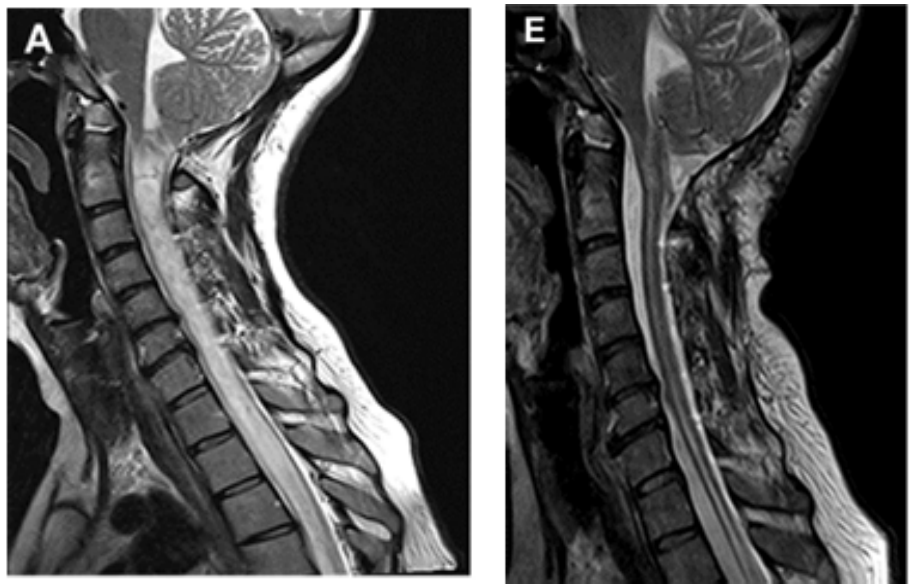
Syringomyelia + other cranio-cervical

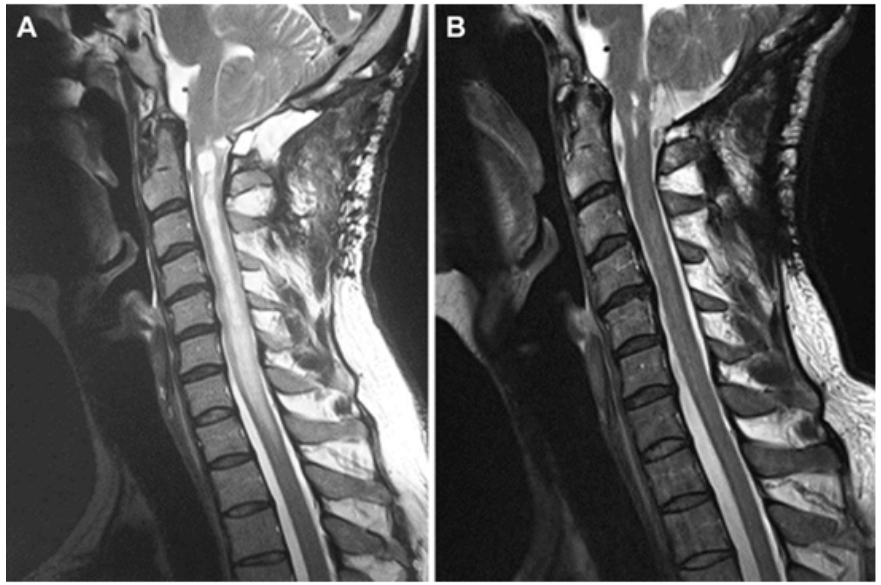
MACQUARIE University

- Arachnoiditis
  - Trauma, infection, surgery
- Arachnoid cysts
- Tumours

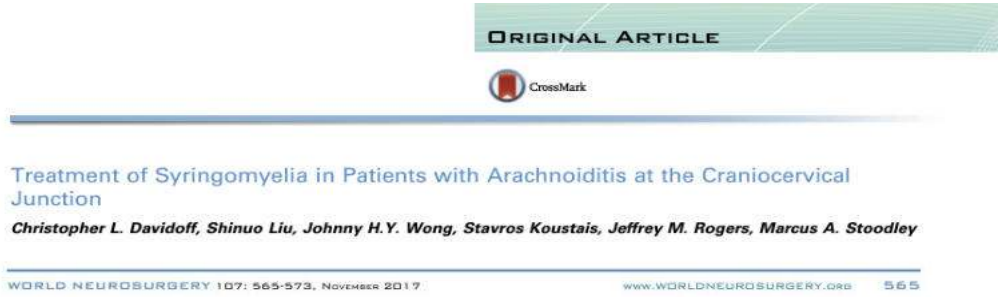
Syringomyelia + other cranio-cervical

Cranio-cervical junction arachnoiditis





Trauma



ORIGINAL ARTICLE

CrossMark

Treatment of Syringomyelia in Patients with Arachnoiditis at the Craniocervical Junction

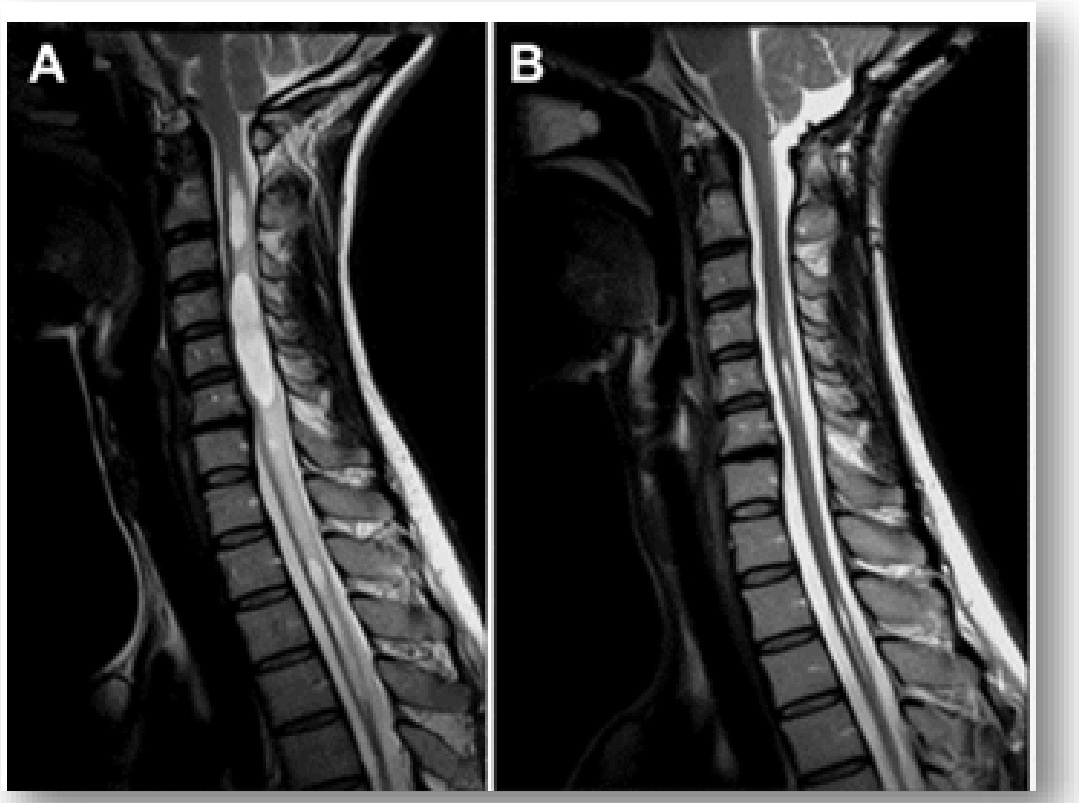
Christopher L. Davidoff, Shinuo Liu, Johnny H.Y. Wong, Stavros Koustais, Jeffrey M. Rogers, Marcus A. Stoodley

WORLD NEUROSURGERY 107: 565-573, NOVEMBER 2017 www.WORLDNEUROSURGERY.org 565

Syringomyelia + other cranio-cervical

MACQUARIE University

Cranio-cervical junction arachnoiditis



Meningitis

Syringomyelia + other cranio-cervical

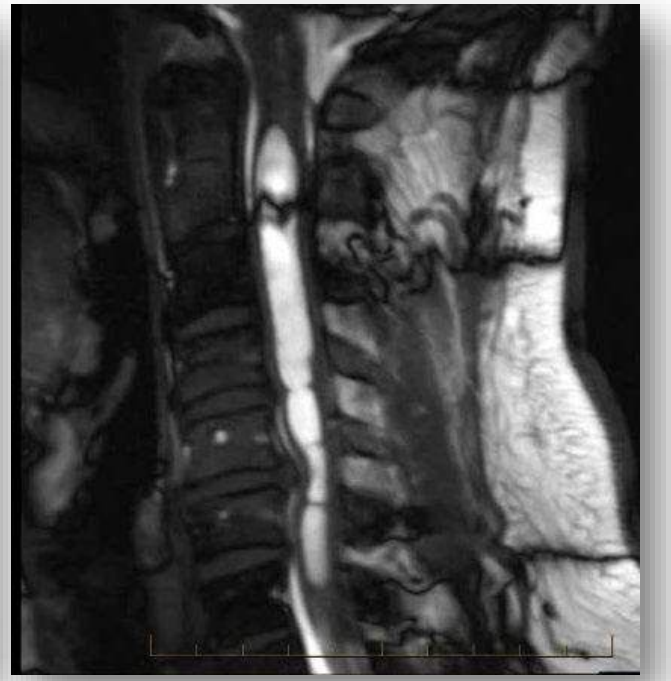
MACQUARIE University

Cranio-cervical junction arachnoiditis

Birth trauma

Syringomyelia + other cranio-cervical

Cranio-cervical junction arachnoiditis

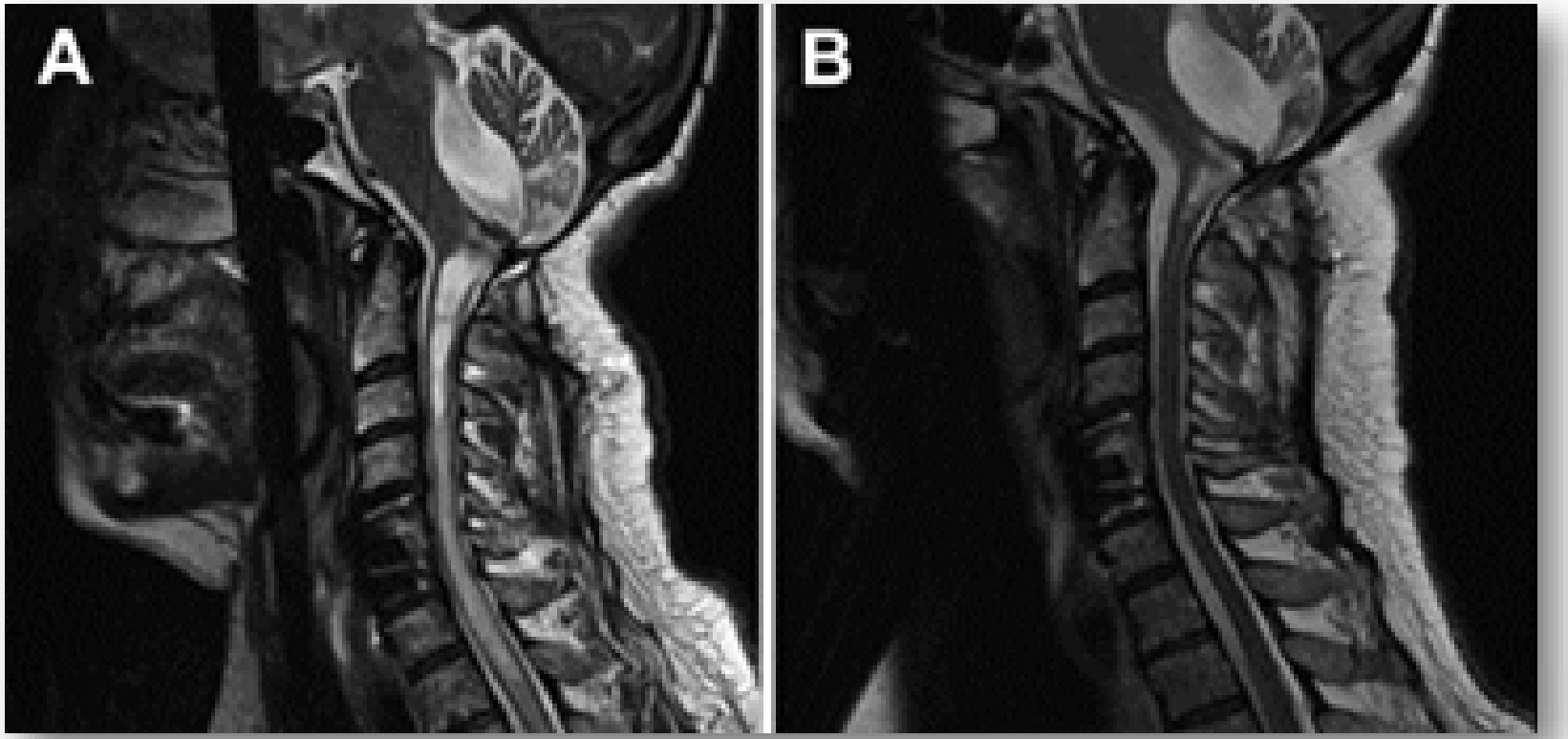


Birth trauma

Syringomyelia + other cranio-cervical

MACQUARIE University

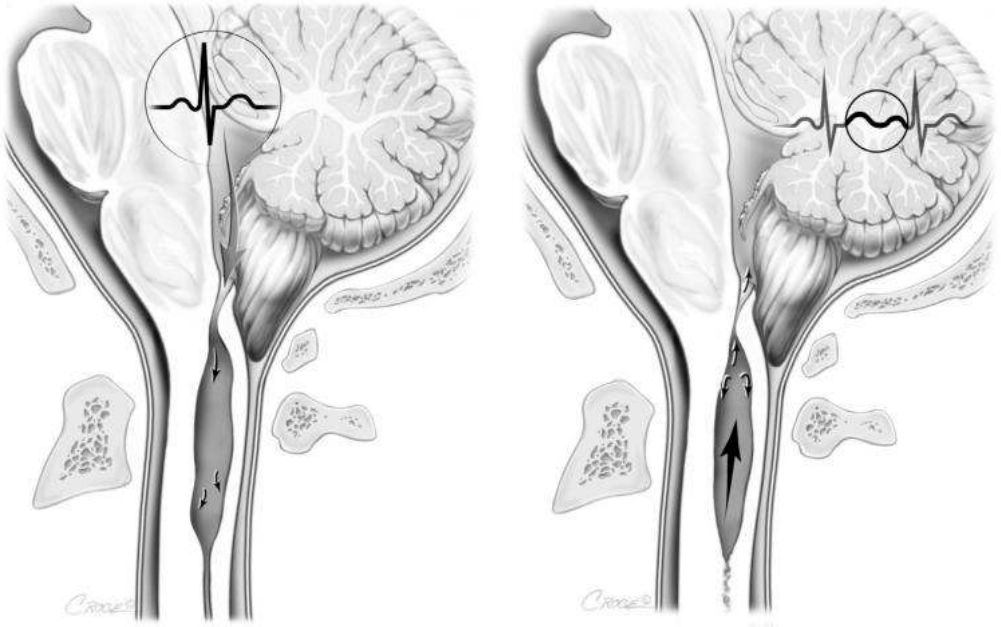
Cranio-cervical junction arachnoiditis



SAH

Syringomyelia + other cranio-cervical

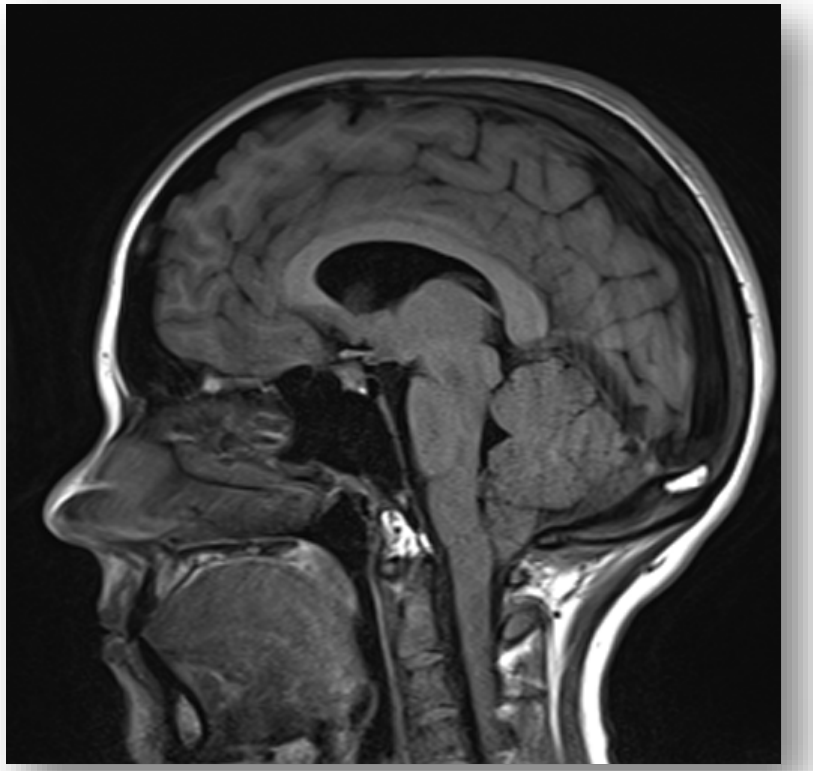
Cranio-cervical junction arachnoiditis



Gardner

SIH + Chiari/syrinx

Tonsillar herniation in SIH



JAMA Neurology | Original Investigation

Assessing Spinal Cerebrospinal Fluid Leaks in Spontaneous Intracranial Hypotension With a Scoring System Based on Brain Magnetic Resonance Imaging Findings

Tomas Dobrocky, MD; Lorenz Grunder, MD; Philipe S. Breiding, MD; Mattia Branca, MSc; Andreas Limacher, PhD; Pascal J. Mosimann, MD; Pasquale Mordasini, MSc; Felix Zibold, MD; Levin Haeni, MD; Christopher M. Jesse, MD; Christian Fung, MD; Andreas Raabe, MD; Christian T. Ulrich, MD; Jan Gralla, MSc; Jürgen Beck, MD; Elke I. Piechowiak, MD

IMPORTANCE Various signs may be observed on brain magnetic resonance imaging (MRI) in patients with spontaneous intracranial hypotension (SIH). However, the lack of a classification system integrating these findings limits decision making in clinical practice.

OBJECTIVE To develop a probability score based on the most relevant brain MRI findings to assess the likelihood of an underlying spinal cerebrospinal fluid (CSF) leak in patients with SIH.

DESIGN, SETTING, AND PARTICIPANTS This case-control study in consecutive patients investigated for SIH was conducted at a single hospital department from February 2013 to October 2017. Patients with missing brain MRI data were excluded. Three blinded readers retrospectively reviewed the brain MRI scans of patients with SIH and a spinal CSF leak, patients with orthostatic headache without a CSF leak, and healthy control participants, evaluating 9 quantitative and 7 qualitative signs. A predictive diagnostic score based on multivariable backward logistic regression analysis was then derived. Its performance was validated internally in a prospective cohort of patients who had clinical suspicion for SIH.

MAIN OUTCOMES AND MEASURES Likelihood of a spinal CSF leak based on the proposed diagnostic score.

RESULTS A total of 152 participants (101 female [66.4%]; mean [SD] age, 46.1 [14.3] years) were studied. These included 56 with SIH and a spinal CSF leak, 16 with orthostatic headache without a CSF leak, 60 control participants, and 20 patients in the validation cohort. Six imaging findings were included in the final scoring system. Three were weighted as major (2 points each): pachymeningeal enhancement, engorgement of venous sinus, and effacement of the suprasellar cistern of 4.0 mm or less. Three were considered minor (1 point each): subdural fluid collection, effacement of the prepontine cistern of 5.0 mm or less, and mamillopontine distance of 6.5 mm or less. Patients were classified into groups at low, intermediate, or high probability of having a spinal CSF leak, with total scores of 2 points or fewer, 3 to 4 points, and 5 points or more, respectively, on a scale of 9 points. The discriminatory ability of the proposed score could be demonstrated in the validation cohort.

CONCLUSIONS AND RELEVANCE This 3-tier predictive scoring system is based on the 6 most relevant brain MRI findings and allows assessment of the likelihood (low, intermediate, or high) of a positive spinal imaging result in patients with SIH. It may be useful in identifying patients with SIH who are leak positive and in whom further invasive myelographic examinations are warranted before considering targeted therapy.

Supplemental content

Author Affiliations: University Institute of Diagnostic and Interventional Neuroradiology, University of Bern, Inselspital, Bern, Switzerland (Dobrocky, Grunder, Breiding, Mosimann, Mordasini, Zibold, Gralla, Piechowiak); Clinical Trials Unit Bern, Institute of Social and Preventive Medicine, University of Bern, Bern, Switzerland (Branca, Limacher); Department of Neurosurgery, University of Bern, Inselspital, Bern, Switzerland (Haeni, Jesse, Fung, Raabe, Ulrich, Beck); Department of Neurosurgery, Medical Center–University of Freiburg, Freiburg, Germany (Fung, Beck).

Corresponding Author: Tomas Dobrocky, MD, Institute of Diagnostic and Interventional Neuroradiology, University of Bern, Inselspital Bern, Freiburgstrasse 8, Bern CH-3010, Switzerland (tomas.dobrocky@insel.ch).

JAMA Neurol. 2019;76(5):580-587. doi:10.1001/jamaneurol.2018.4921
Published online February 18, 2019.

jamaneurology.com

Table 2. Six Imaging Signs With Good Discriminative Power and Interrater Agreement That Were Included in the Final Diagnostic Score and Assigned Score Points

Characteristic	Coefficient (95% CI)	Odds Ratio (95% CI)	*P* Value	Score Points
Engorgement venous sinus	2.95 (1.18-4.72)	19.12 (3.26-112.30)	.001	2
Pachymeningeal enhancement	4.04 (2.50-5.59)	57.01 (12.18-266.78)	<.001	2
Subdural fluid collection	1.54 (−0.10 to 3.17)	4.65 (0.90-23.92)	.07	1
Suprasellar cistern[a]	3.48 (2.36-4.60)	32.32 (10.55-99.02)	<.001	2
Prepontine cistern[b]	1.47 (0.41-2.52)	4.34 (1.51-12.47)	.007	1
Mamillopontine distance[c]	1.13 (0.07-2.19)	3.08 (1.07-8.90)	.04	1

[a] ≤4 mm.
[b] ≤5 mm.
[c] ≤6.5 mm.

SIH + Chiari/syrinx

MACQUARIE University

Syrinx in SIH

Syrinx Secondary to Chiari-like Tonsillar Herniation in Spontaneous Intracranial Hypotension

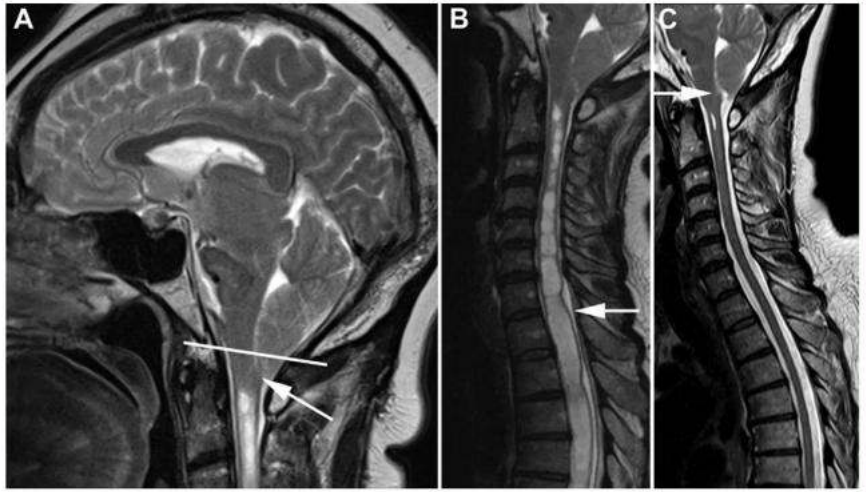
Erik H. Middlebrooks[1], Lela Okromelidze[1], George K. Vilanilam[1], Neethu Gopal[1], Patrick H. Luetmer[2], Vivek Gupta[1]

World Neurosurg. (2020) 143:e268-e274.

Table 1. Clinical and Radiographic Characteristics of Patients with Spontaneous Intracranial Hypotension Stratified by the Presence of Syrinx

Characteristic	SIH with Syrinx ($n = 6$)	SIH without Syrinx ($n = 8$)	*P* Value
Clinical			
Suboccipital headache	2 (40)	1 (12.5)	0.54
Postural headache	5 (83.3)	6 (75)	0.7
Somnolence	4 (66.7)	0 (0)	0.02*
Paresthesia	4 (66.7)	1 (12.5)	0.09
Upper extremity weakness	3 (50)	0 (0)	0.04*
MRI			
Pachymeningeal enhancement	4 (66.7)	6 (75)	0.63
Protrusion of pituitary into suprasellar cistern	2 (33.3)	3 (37.5)	0.4
Collapsed optic nerve sheath	5 (83.3)	2 (25)	0.1
Obex displacement below foramen magnum	5 (83.3)	1 (12.5)	0.03*
Flattening of pons	6 (100)	2 (25)	0.01*
Sagging of brain	6 (100)	7 (87.5)	1.00
Foramen magnum crowding	6 (100)	8 (100)	1.00

SIH, spontaneous intracranial hypotension; MRI, magnetic resonance imaging.
*Statistically significant.



SIH + Chiari/syrinx

MACQUARIE University

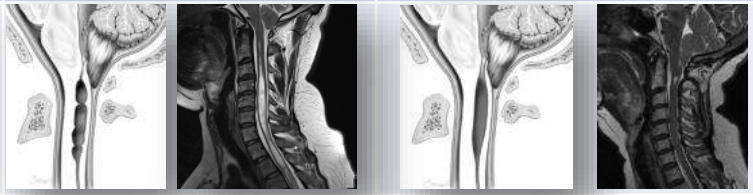
Tonsillar herniation in SIH: clinical features

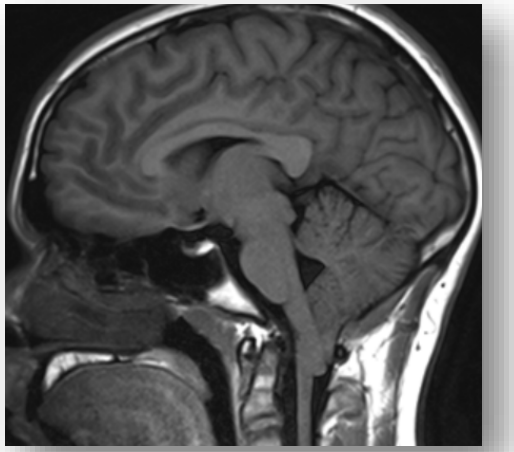
	Chiari	SIH
Orthostatic headache	✗	✓
Headache location	Occipital	Frontal, global, +/- occipital
Duration	Long	Shorter
Onset	Gradual	May be sudden onset
Cognitive effects	✓	✓
Fronto-temporal dementia	✗	✓
Hearing changes	✓	✓
Tremor	✗	✓
Brain stem signs	✓	✓

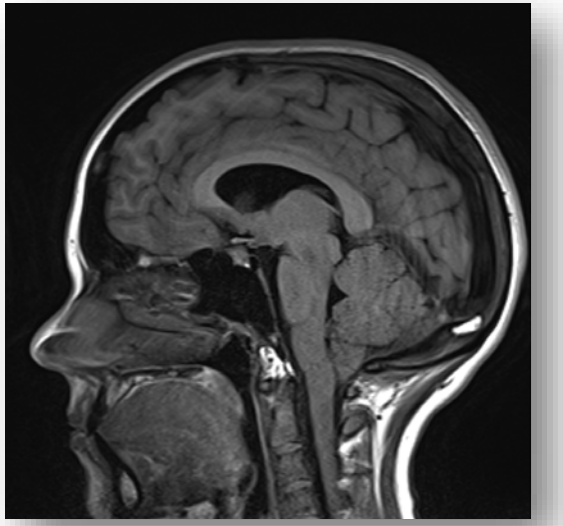
SIH + Chiari/syrinx

MACQUARIE University

Tonsillar herniation in SIH: imaging features

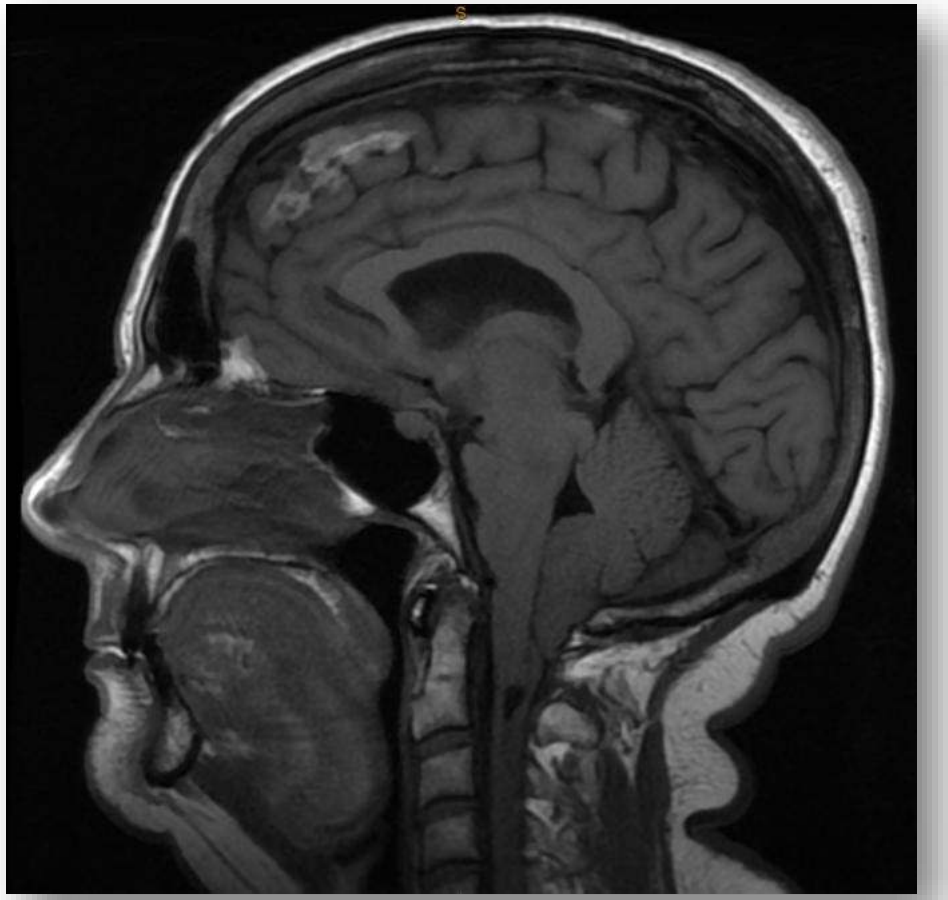
	Chiari	SIH
Tonsillar herniation	✓	✓
'Bern signs'	✗	✓
IIH signs	✓	✓
Buckling of medulla	✓	✗
Syrinx	Common	Uncommon
Syrinx type		
Syrinx symptoms	✓	✗
Cerebellar slump post-op	✓ ✗	✓

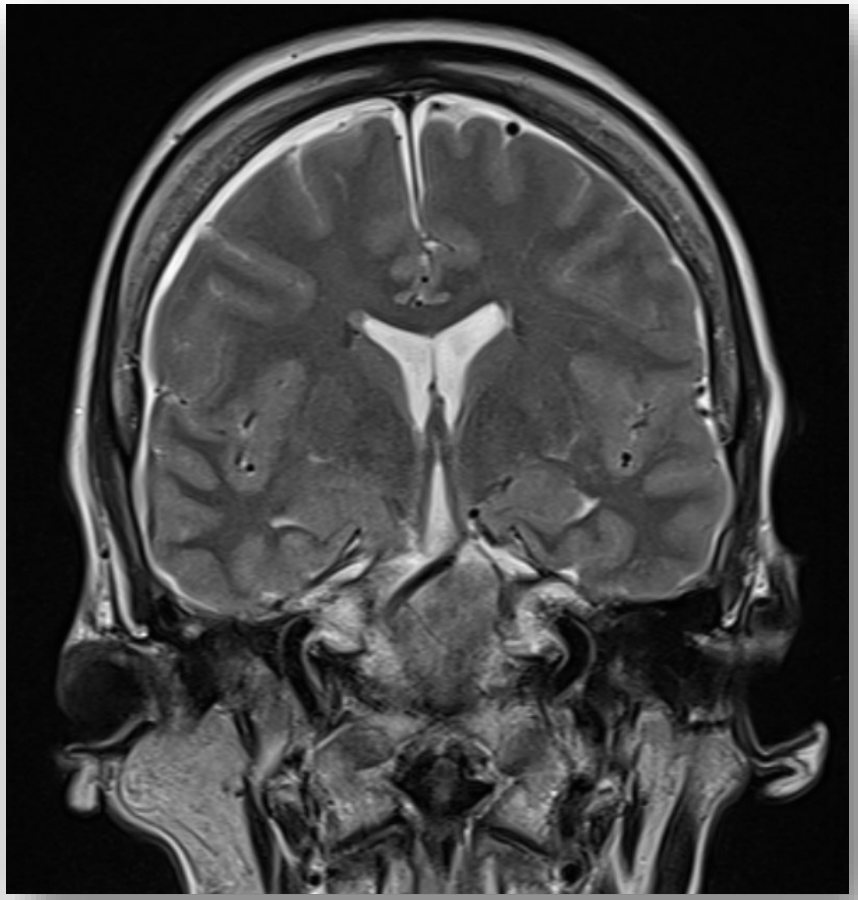


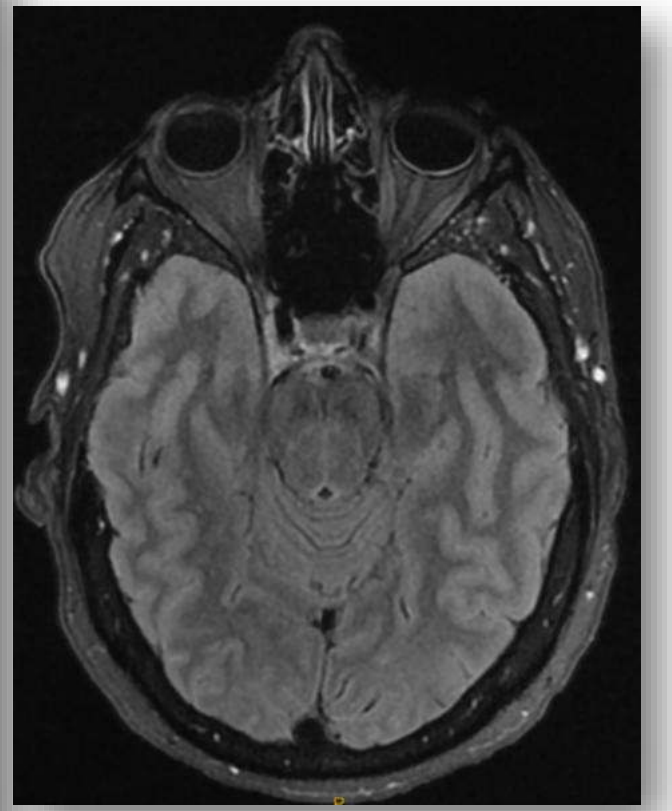


SIH + Chiari/syrinx

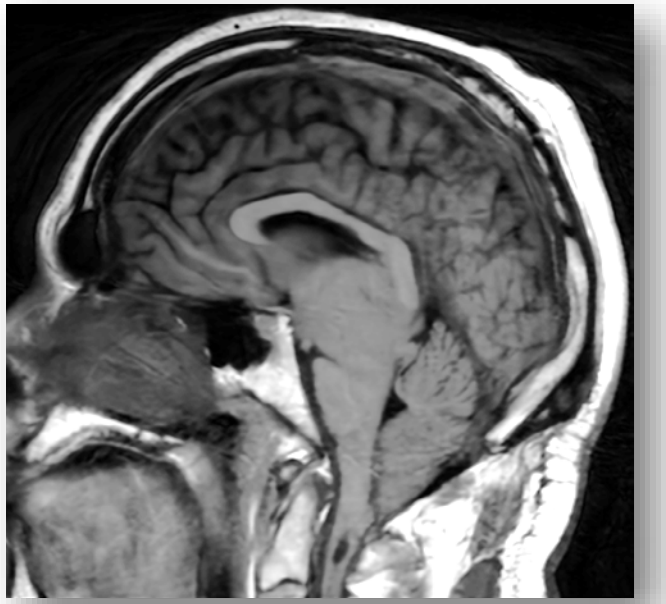
MACQUARIE University







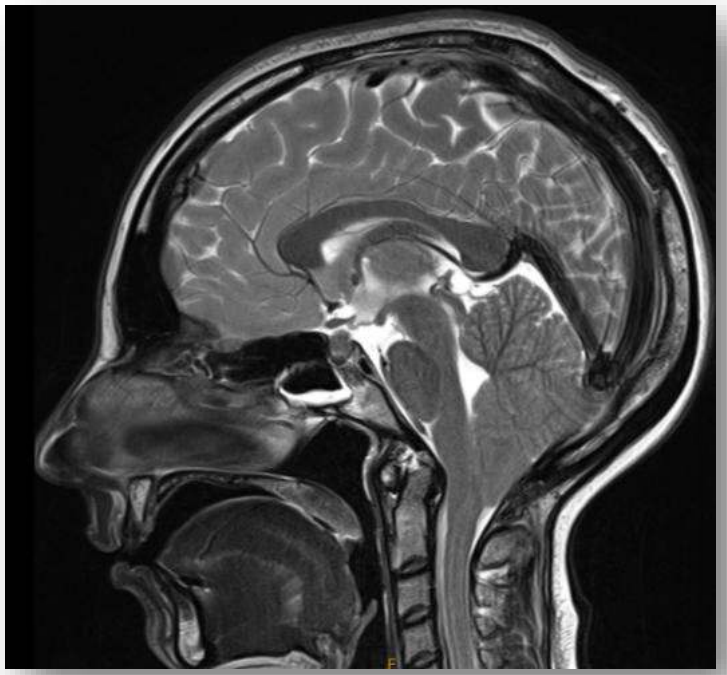
SIH + Chiari/syrinx



** Shown with patient permission*

SIH + Chiari/syrinx

- SIH Imaging can mimic Chiari malformation

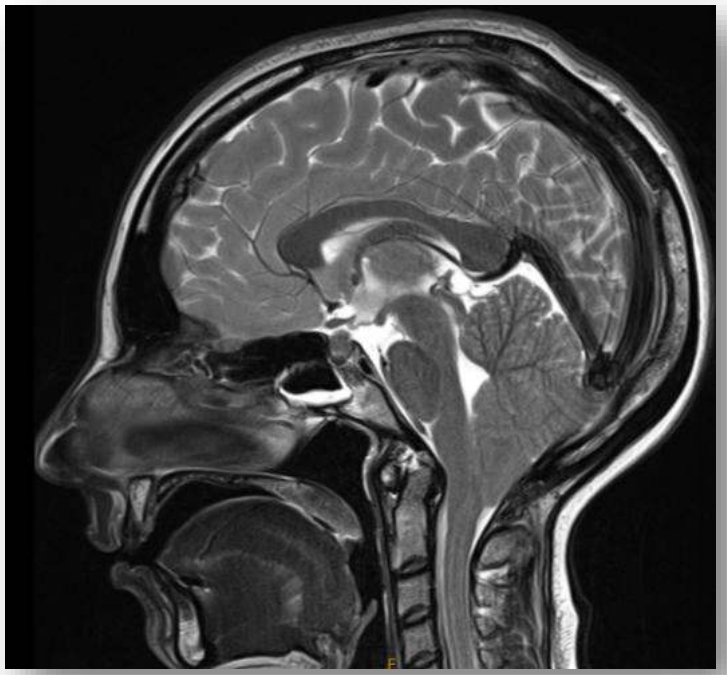


Pre-op

Post-op

SIH + Chiari/syrinx

- SIH Imaging can mimic Chiari malformation



Pre-op

Post-op

SIH + Chiari/syrinx

- SIH Imaging can mimic Chiari malformation

Pre-op

Post-op

Iatrogenic SIH + Chiari/syrinx

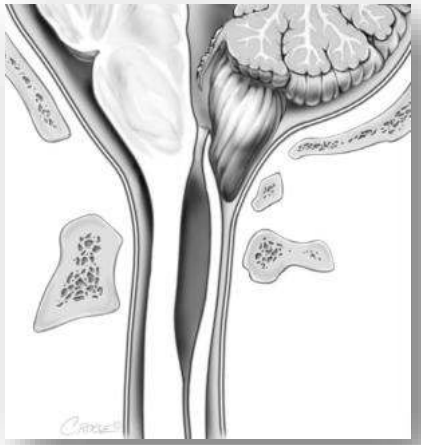
SIH + Chiari/syrinx

MACQUARIE University

Conclusions

- Tonsillar descent is common in SIH
  - Can mimic Chiari malformation on imaging
  - Clinical features usually different

- Syringomyelia is rare in SIH
  - Communicating
  - Asymptomatic
  - Improves with SIH treatment



CSF MDT

MACQUARIE University