2025 Intracranial Hypotension Conference

Amsterdam June 28-29

Superficial siderosis: Cerebral amyloid angiopathy vs SIH

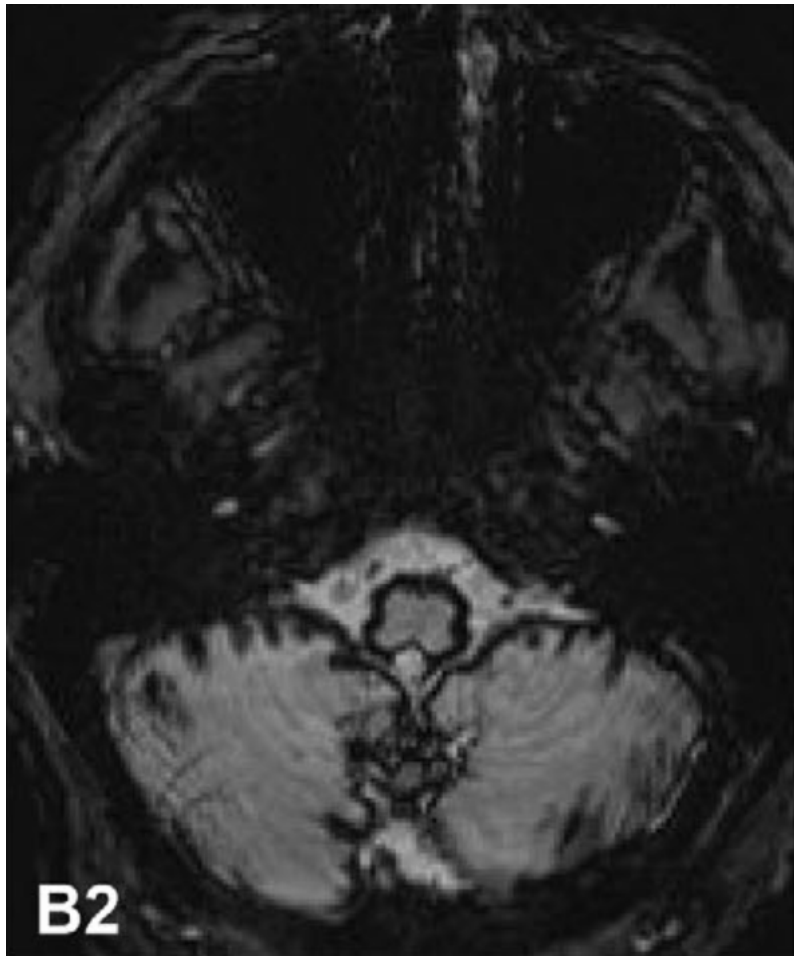
Jürgen Beck

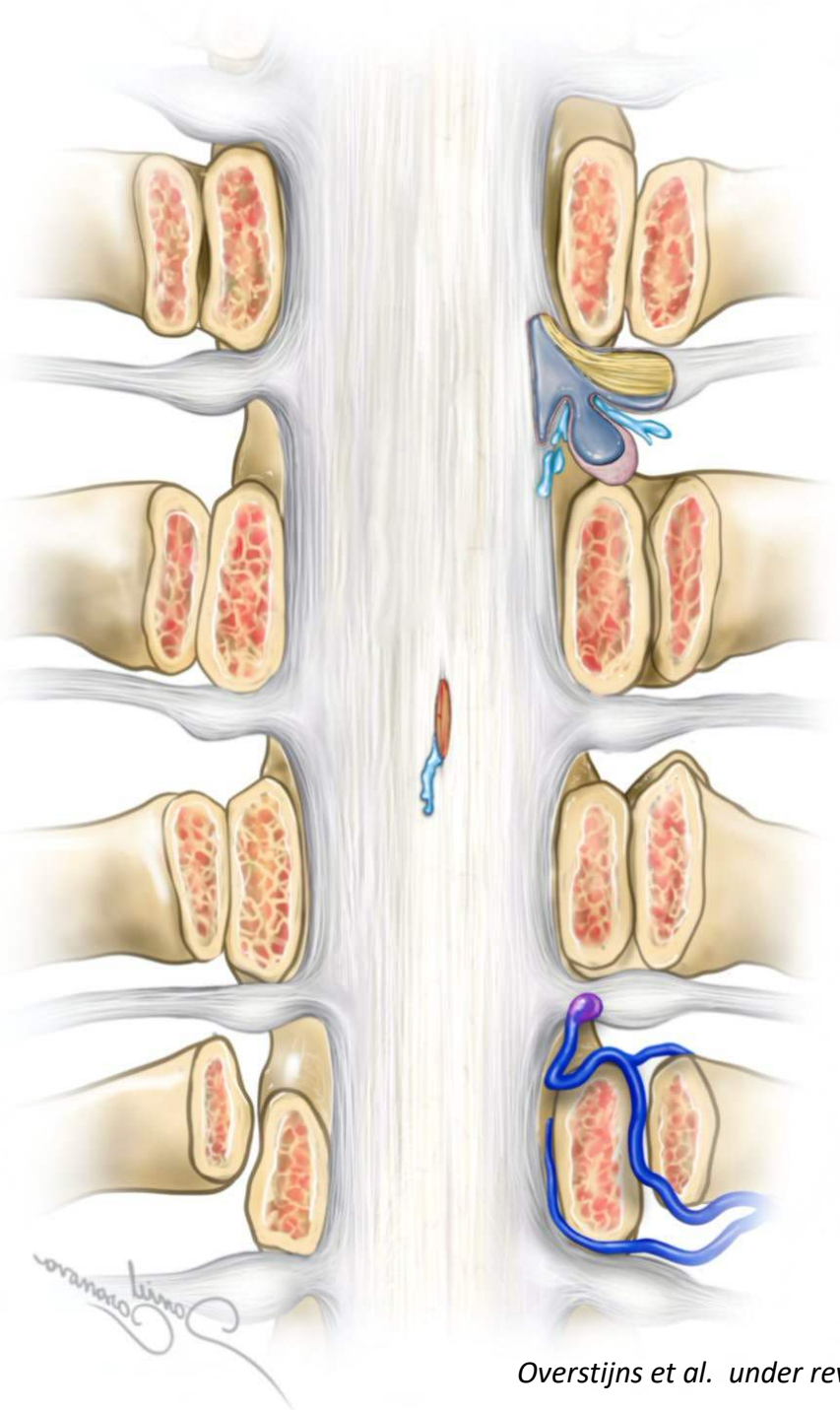
Director Dept. of Neurosurgery

Medical Center – University of Freiburg, Germany

Chair CSF-Section - European Association of Neurosurgical Societies

Superficial Siderosis





Overstijns et al. under review 2025

RUSSELL G. MACROBERT, M.B. (TOR.)
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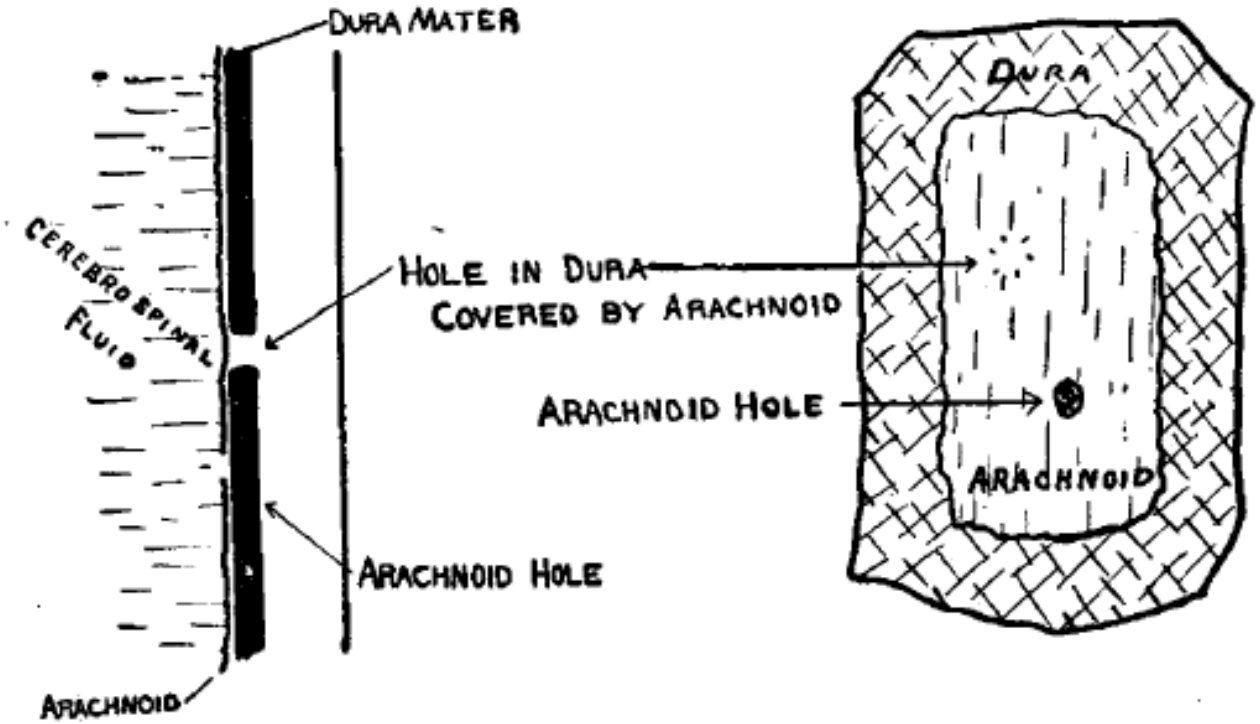

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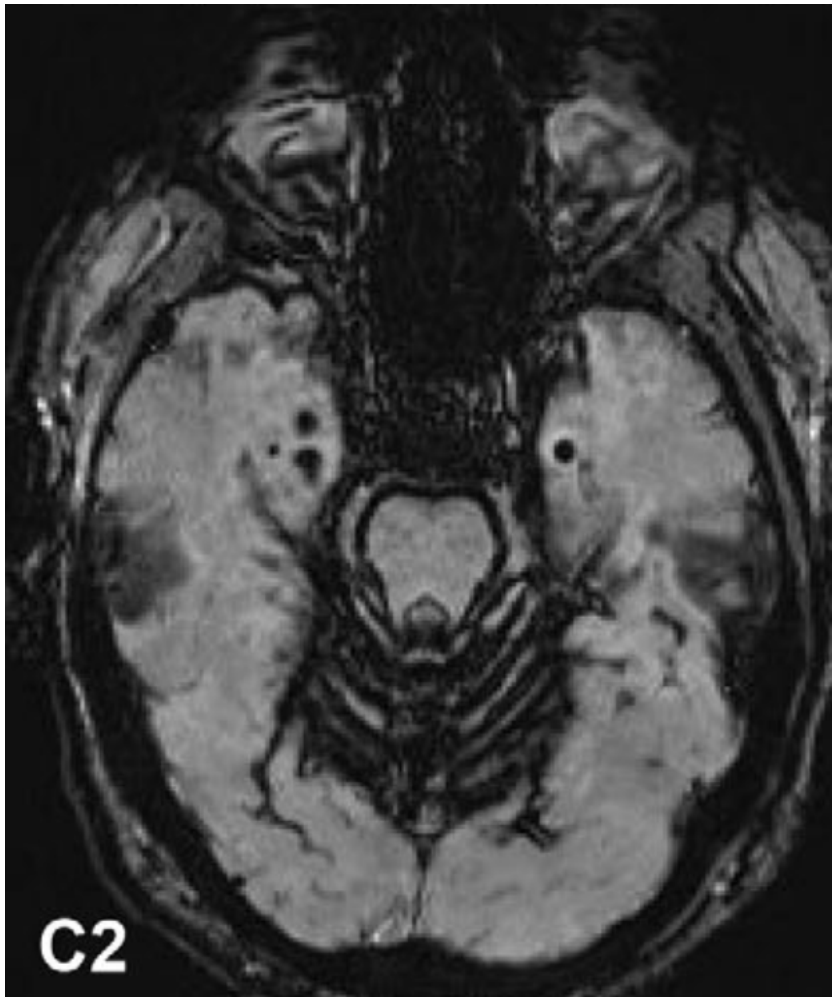
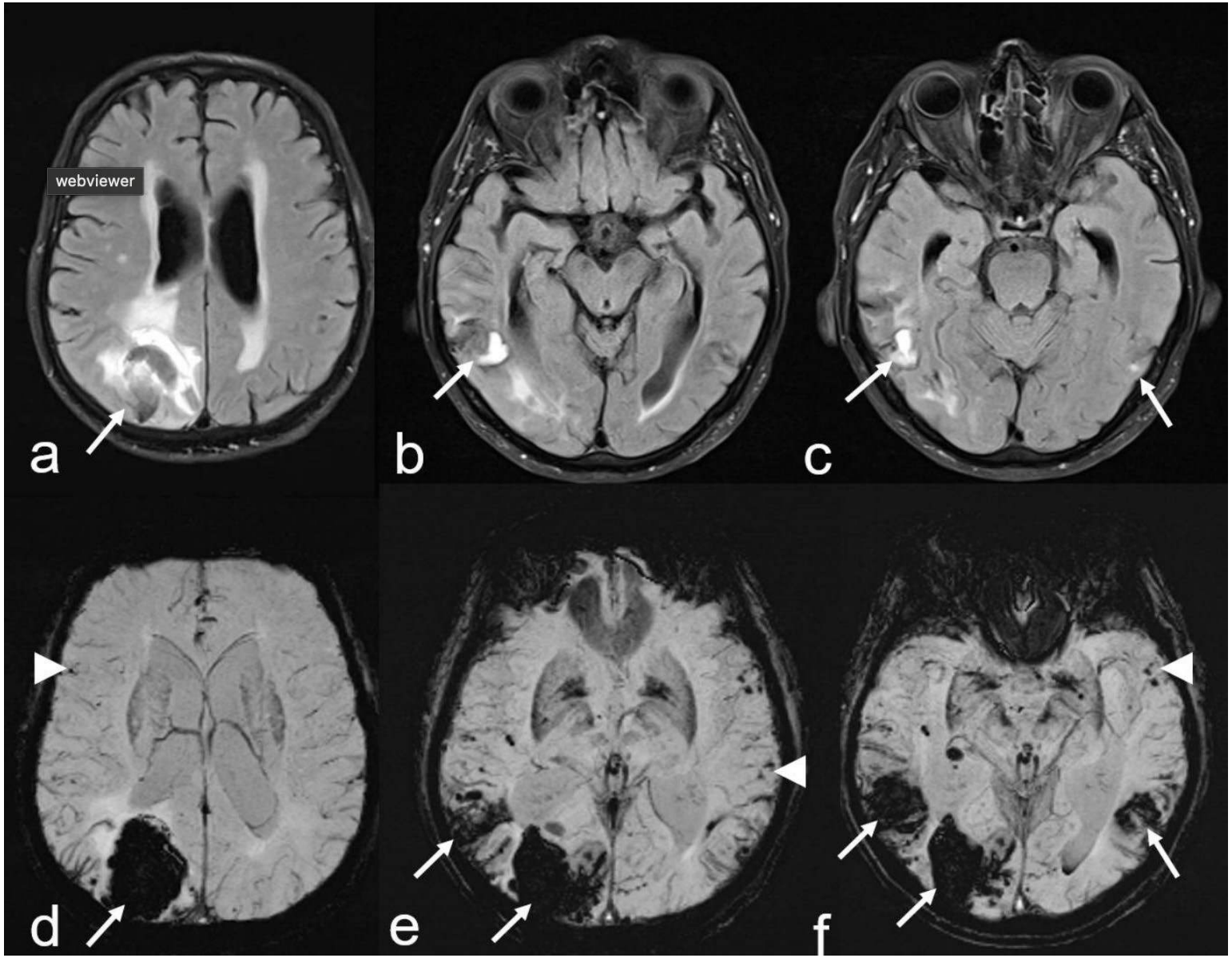
Superficial Siderosis



Cerebral amyloid angiopathy



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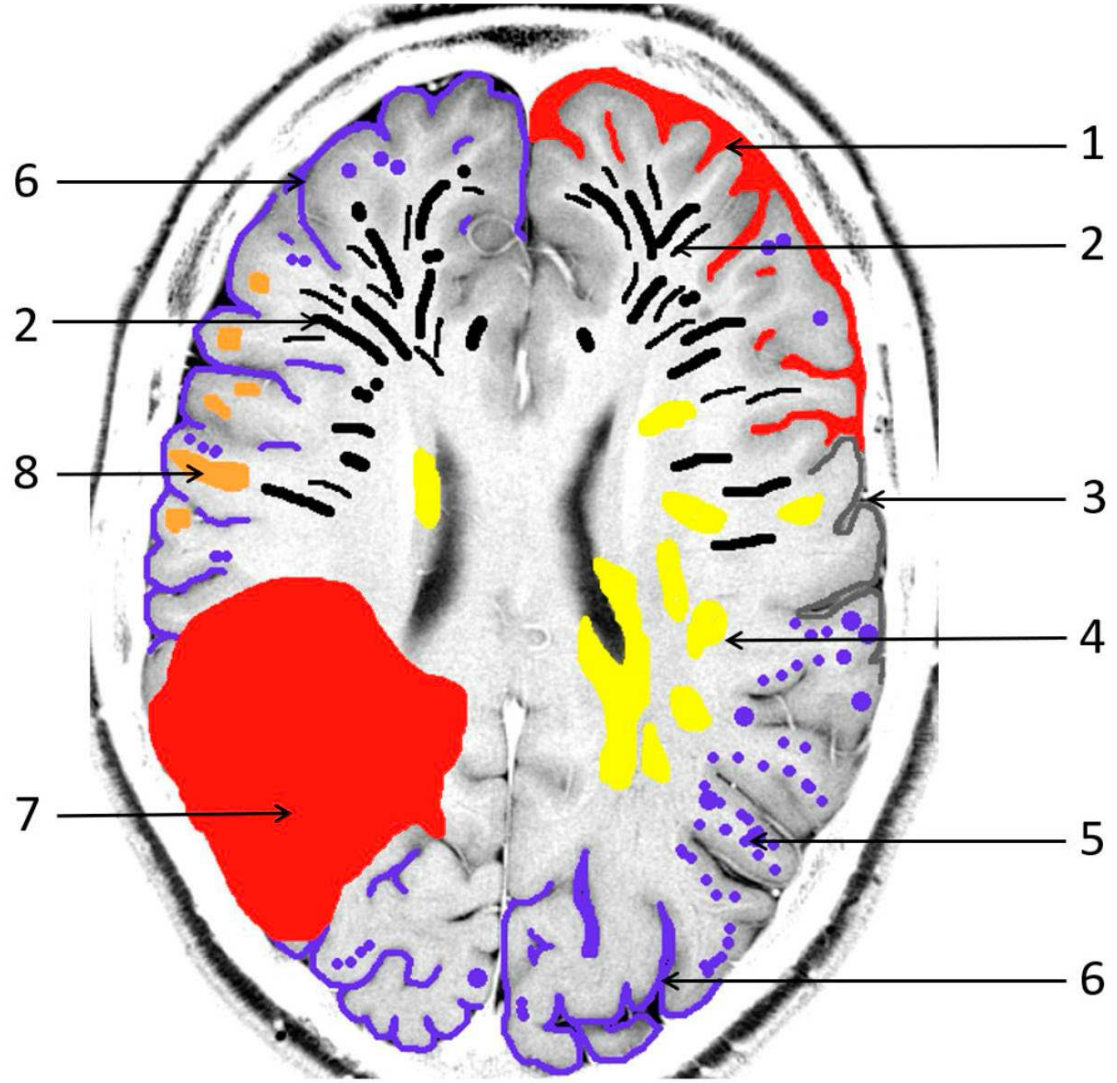
Cerebral amyloid angiopathy

superficial siderosis



superficial siderosis

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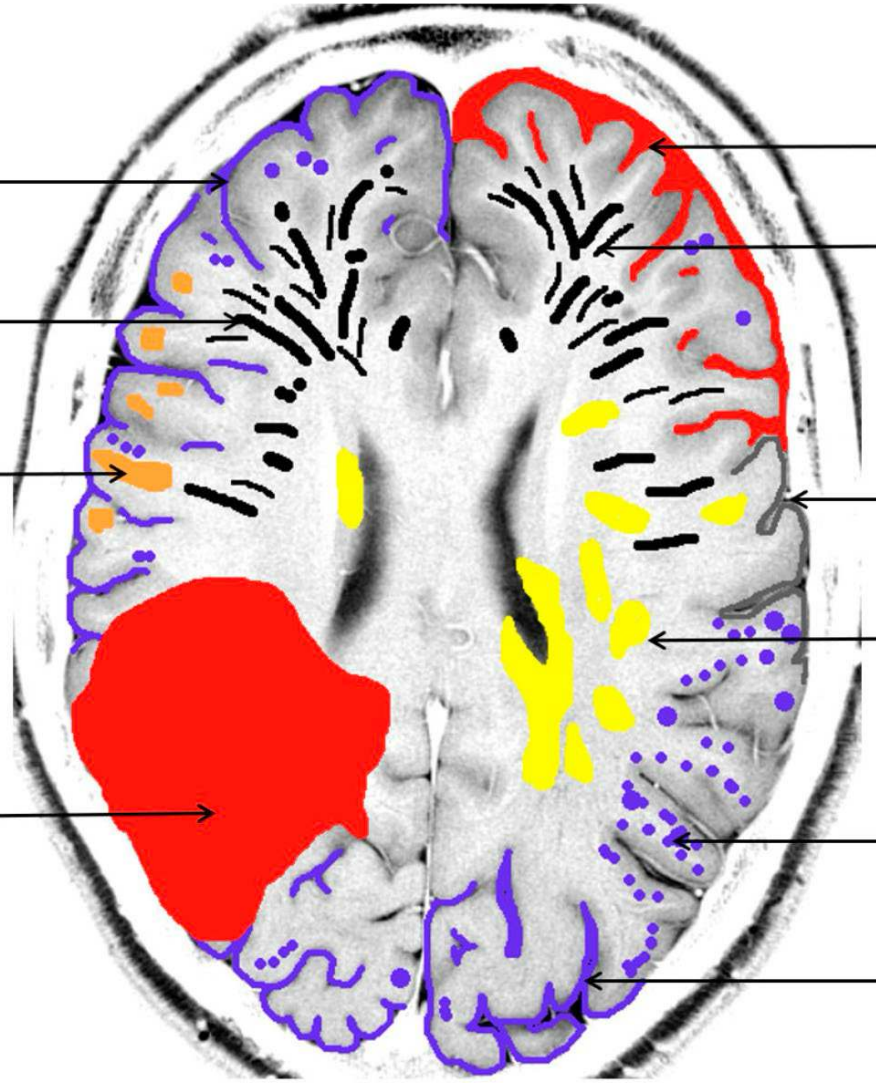
Cerebral amyloid angiopathy



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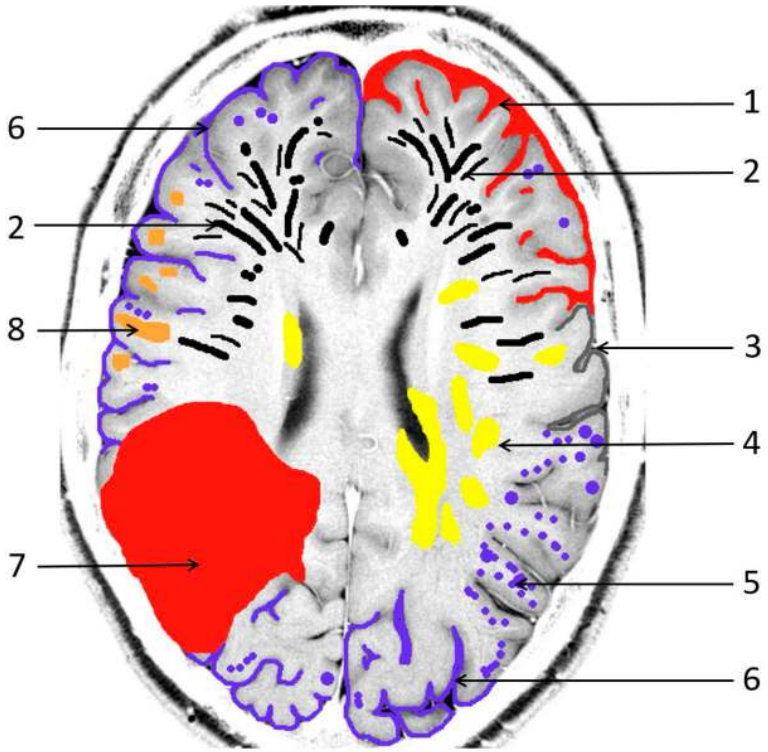
Cerebral amyloid angiopathy

superficial siderosis



Amyloid beta

- ↓ transport from ISF
- ↓ intramural periarterial drainage (IPAD)
- ↓ glymphatic transport

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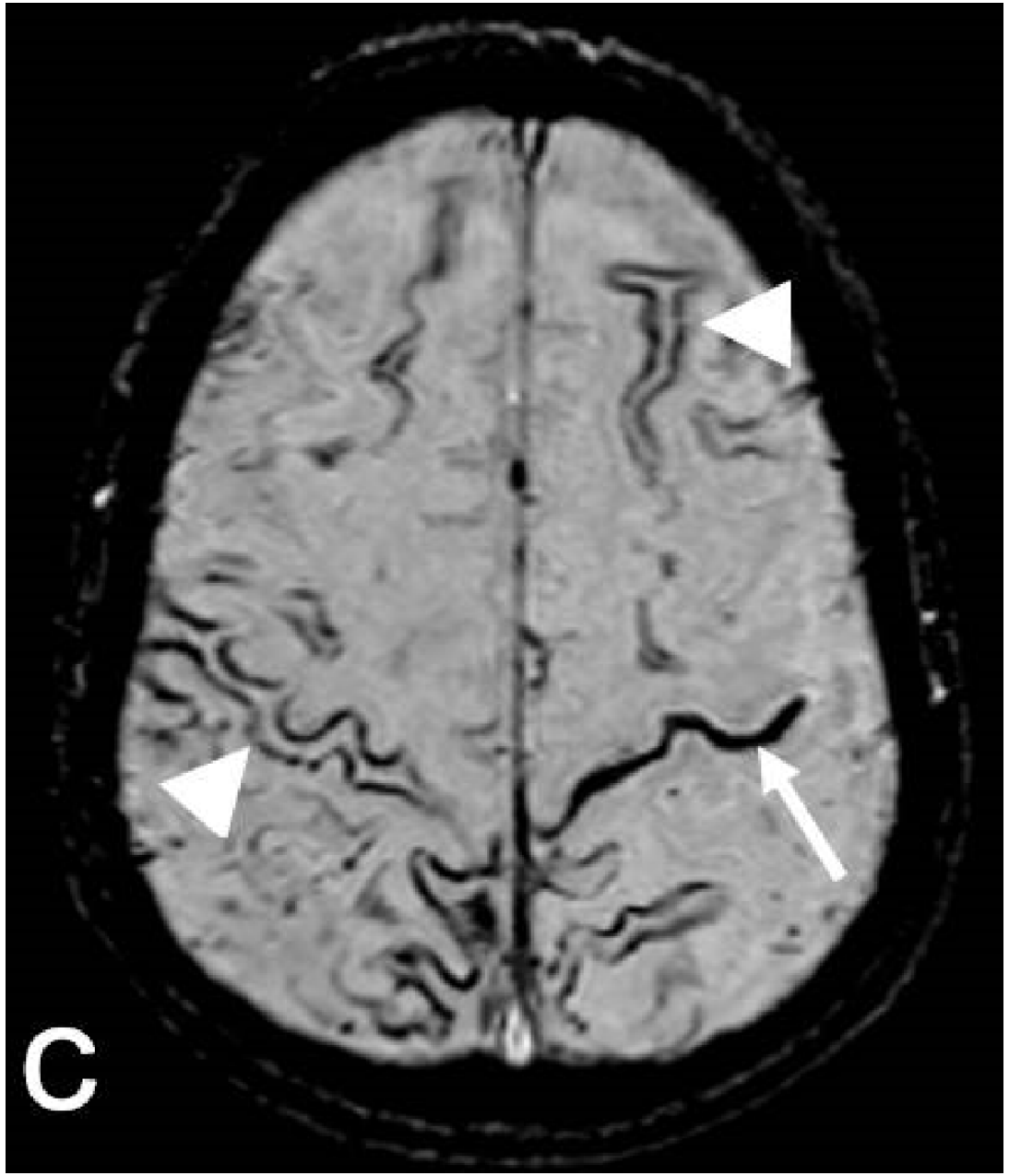

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Validation of the Boston Criteria Version 2.0 for Cerebral Amyloid Angiopathy in Patients Presenting With Intracerebral Hemorrhage

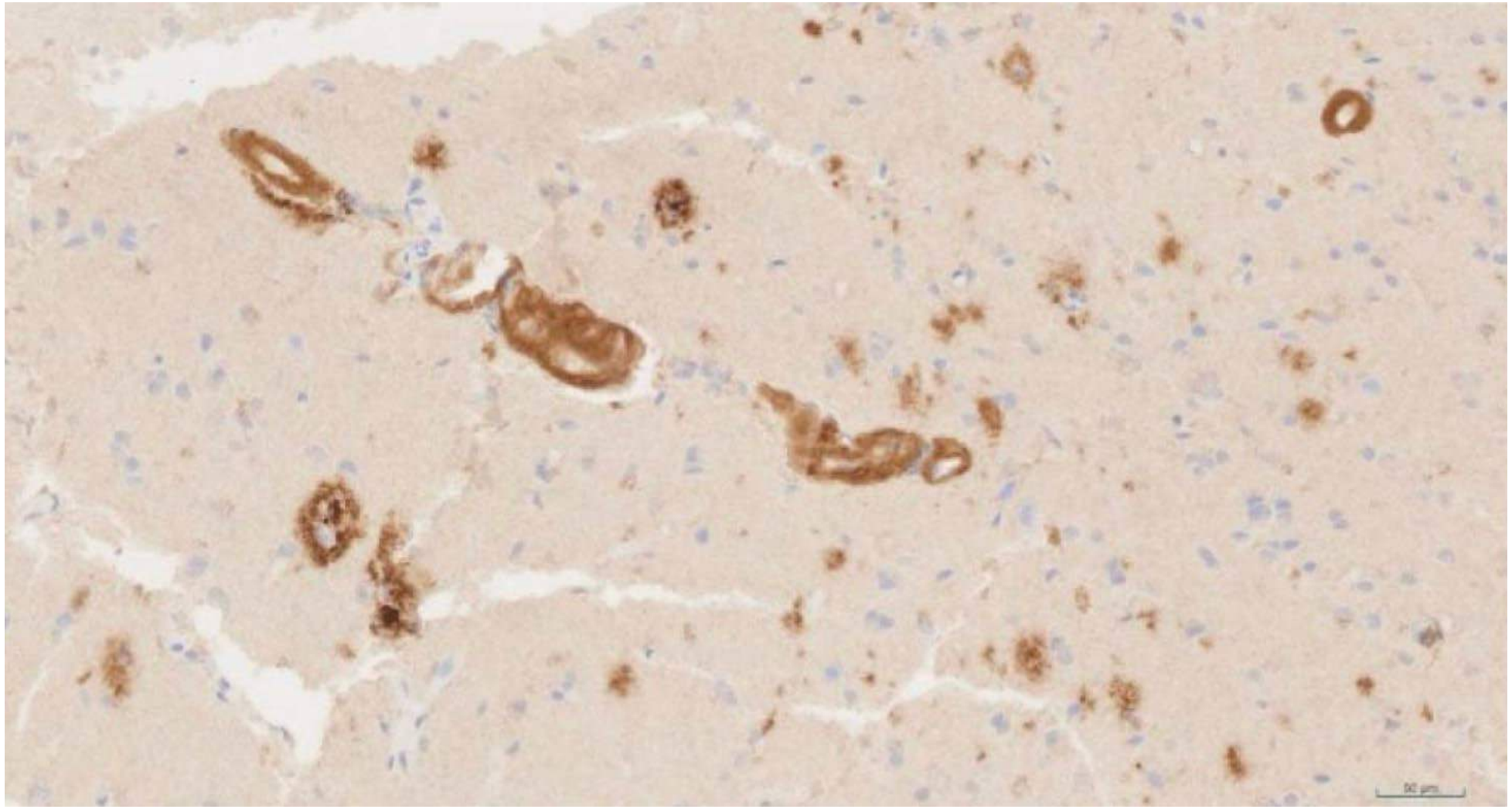
Margaret H. Downes,[1] Roshini Kalagara,[1] Christina P. Rossitto,[1] Vikram Vasan,[1] Devarshi Vasa,[1] Susmita Chennareddy,[1] Daniel R. Lefton,[2] Sema Yildiz,[2] Melissa Umphlett,[3] Carolyn Brockington,[4] and Christopher P. Kellner[1]

Neurology® 2025;104:e213460. doi:10.1212/WNL.0000000000213460

ORIGINAL CONTRIBUTION

Diagnostic Accuracy of Finger-Like Projections and Subarachnoid Hemorrhage for Cerebral Amyloid Angiopathy: Pathological Validation From Lobar Hematoma Evacuation or Brain Biopsy

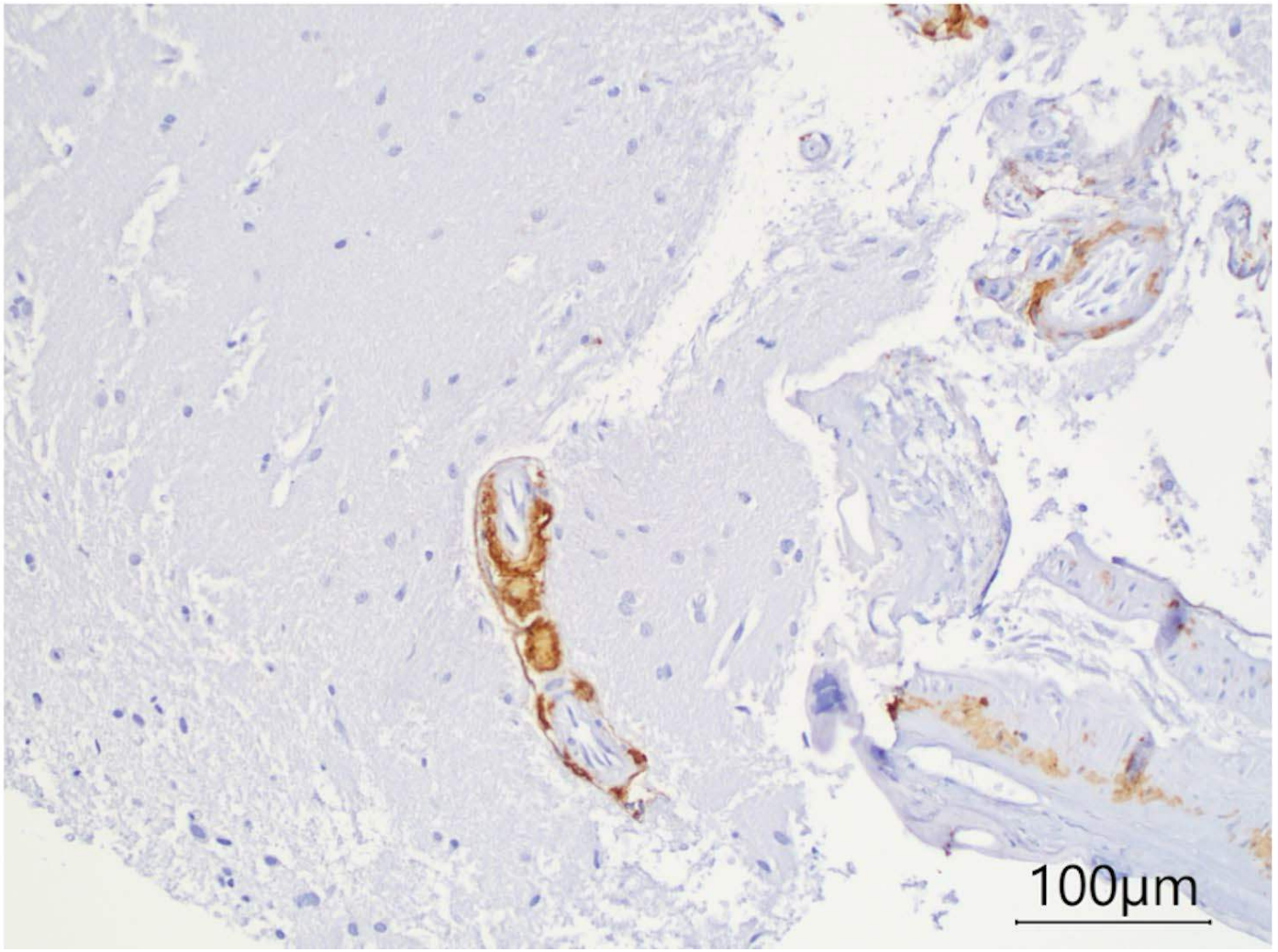
Alexandra Maury, MD; Joseph Benzakoun, MD, PhD; Alexandre Bani-Sadr, MD, PhD; Adrien ter Schiphorst, MD; Peggy Reiner, MD; Hassan Hosseini, MD, PhD; Pierre Seners, MD, PhD; Johan Pallud, MD, PhD; Catherine Oppenheim, MD, PhD; David Calvet, MD, PhD; Andreas Charidimou, MD, PhD; Pascale Varlet, MD, PhD*; Guillaume Turc, MD, PhD*; Jean-Claude Baron, MD, ScD*; on behalf of the HEMALOB Study Group†



Cerebral amyloid angiopathy in spontaneous intracerebral hemorrhage – radiology and immunohistochemistry in a consecutive surgical series

A.

B.



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label free molecular imaging

Neuro- , Oral- and Maxillofacial Surgery

Pathology and Neuropathology, Neuroanatomy

Stimulated Raman Imaging

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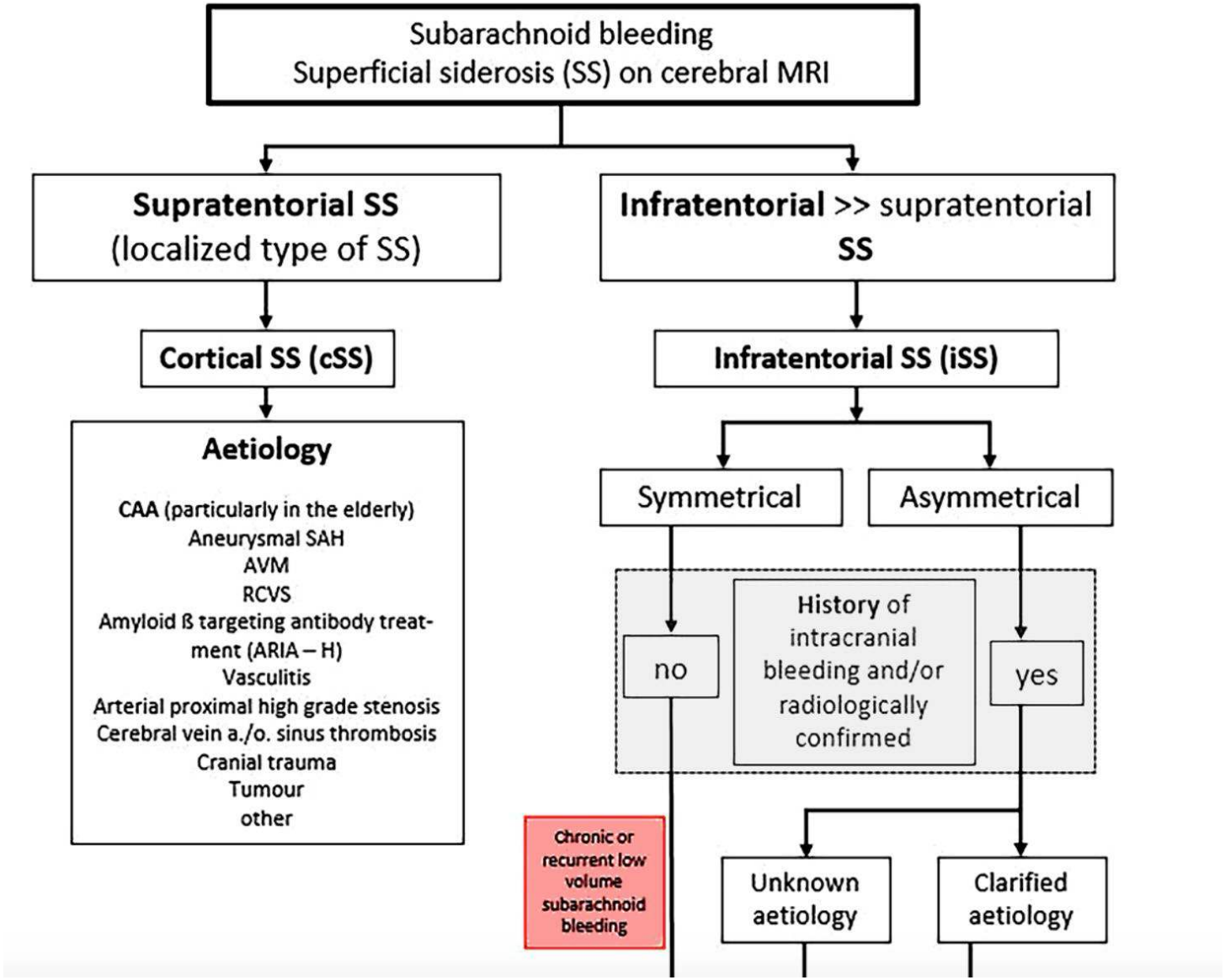
Superficial Siderosis



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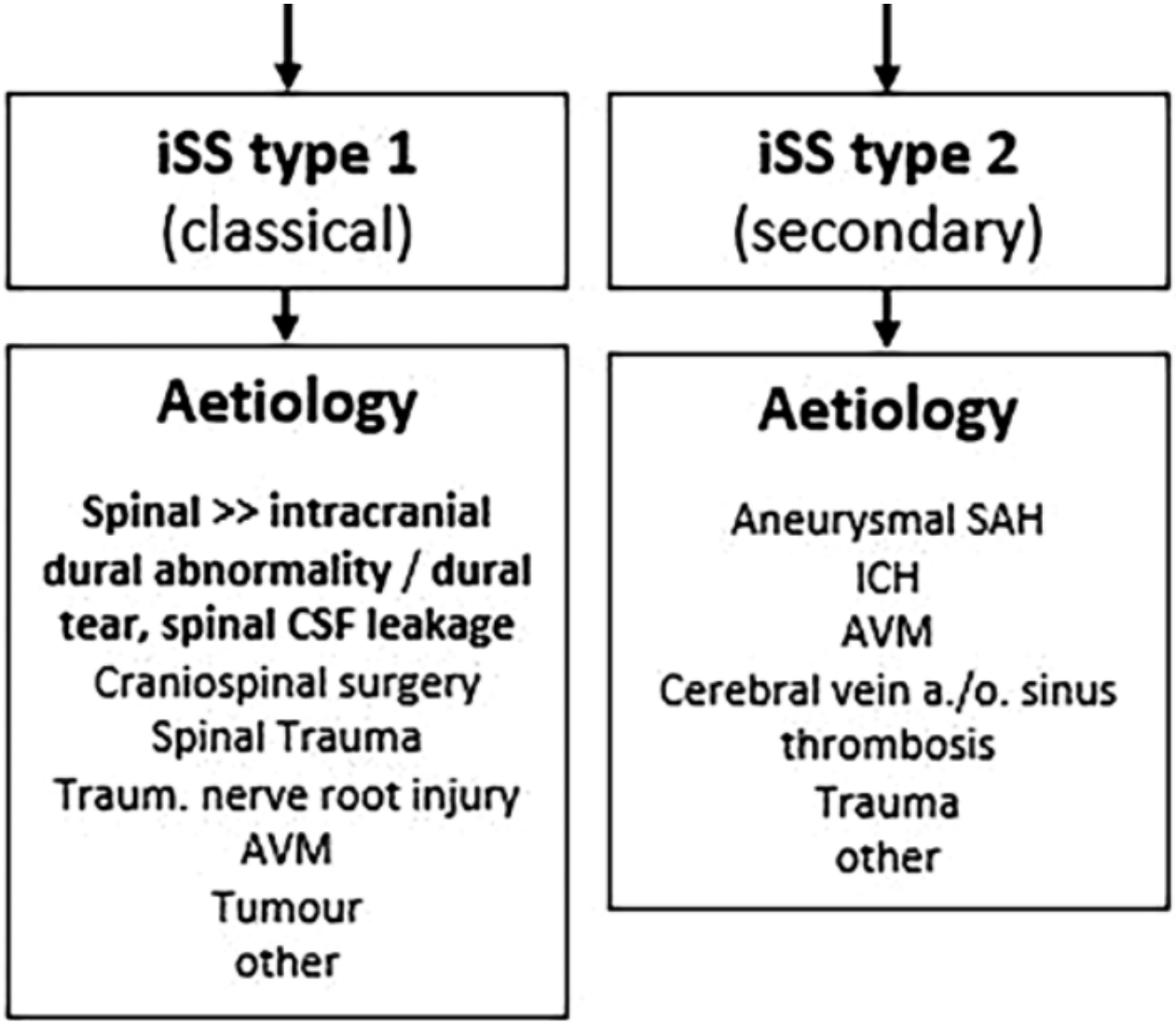
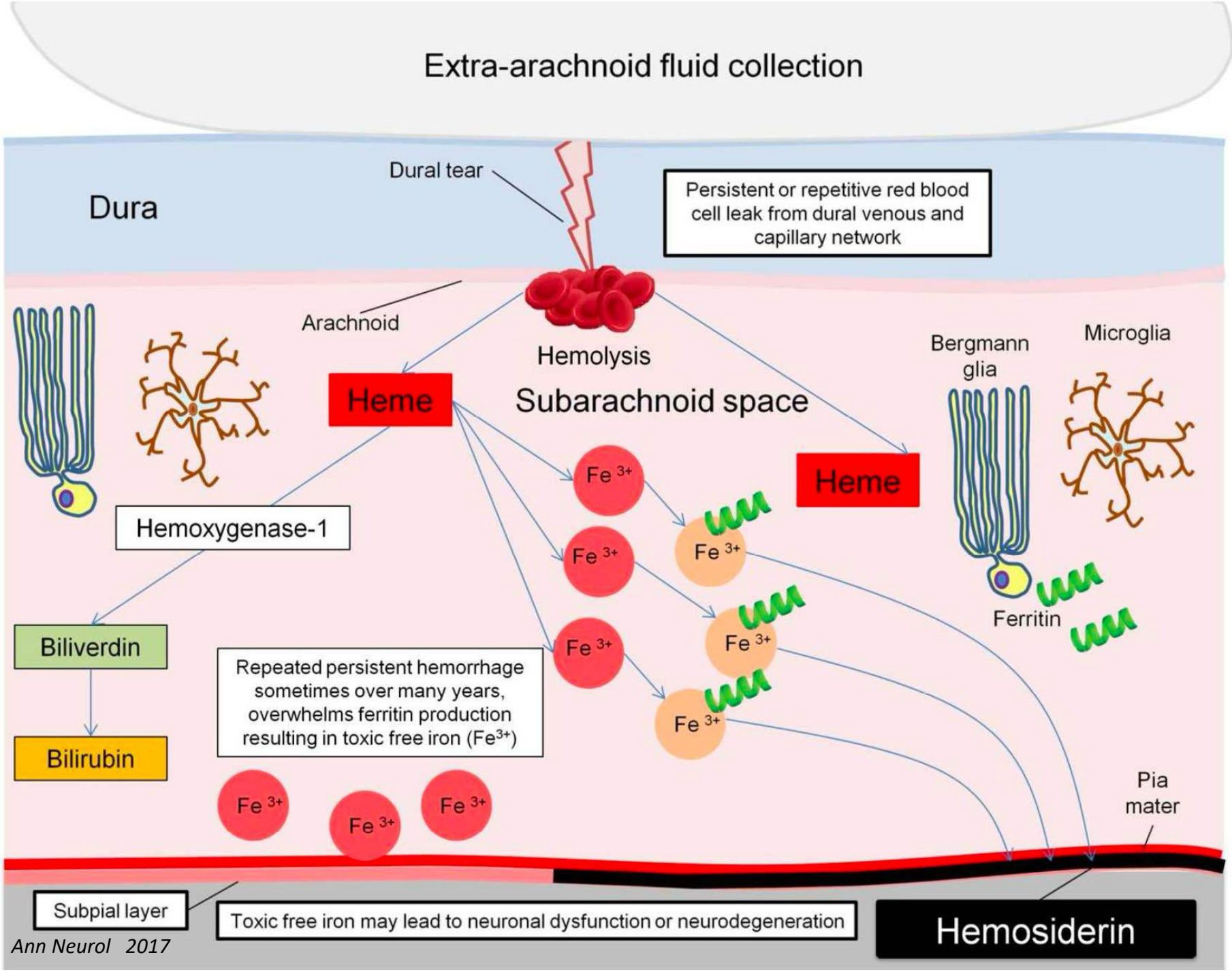
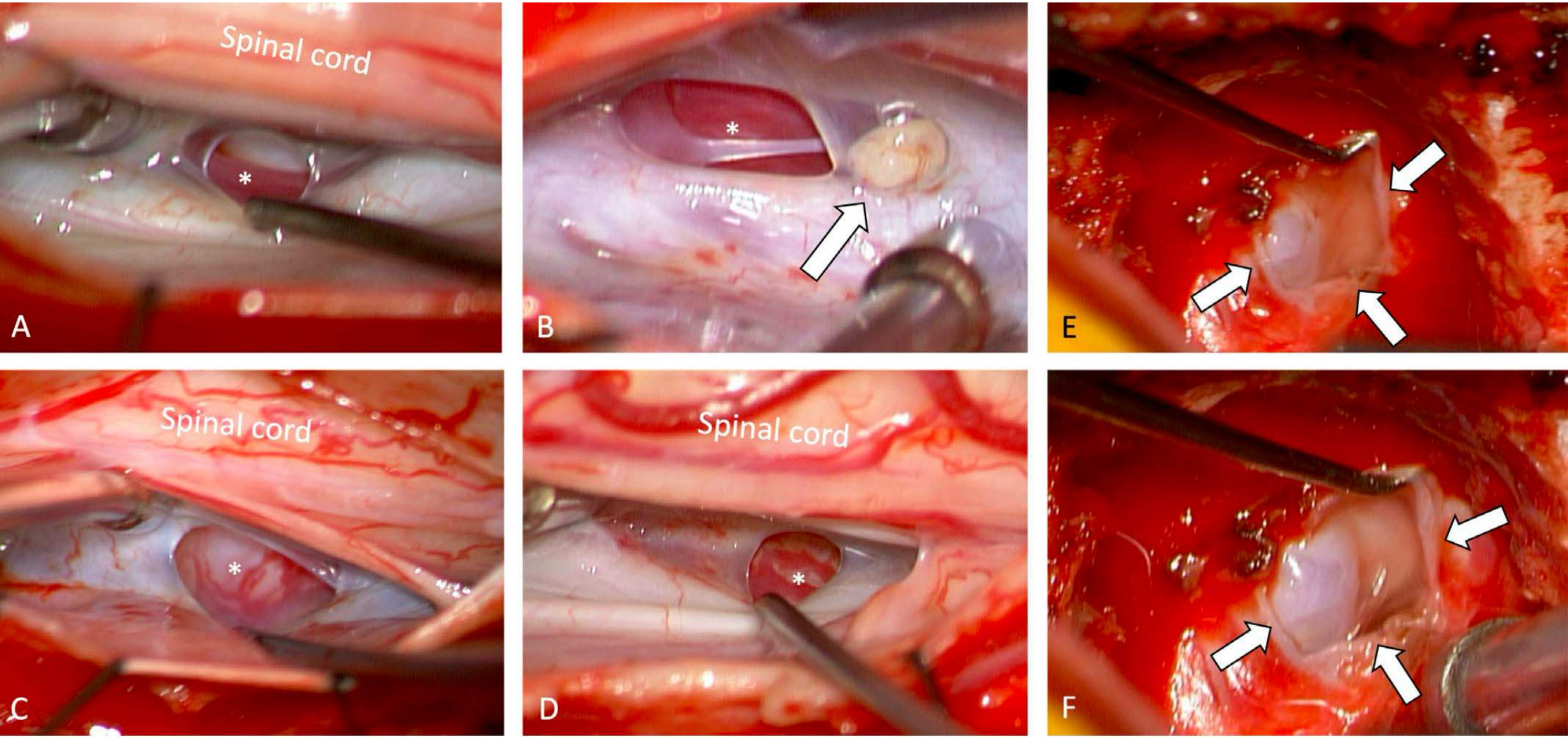




Wilson et al. Werring Ann Neurol 2017

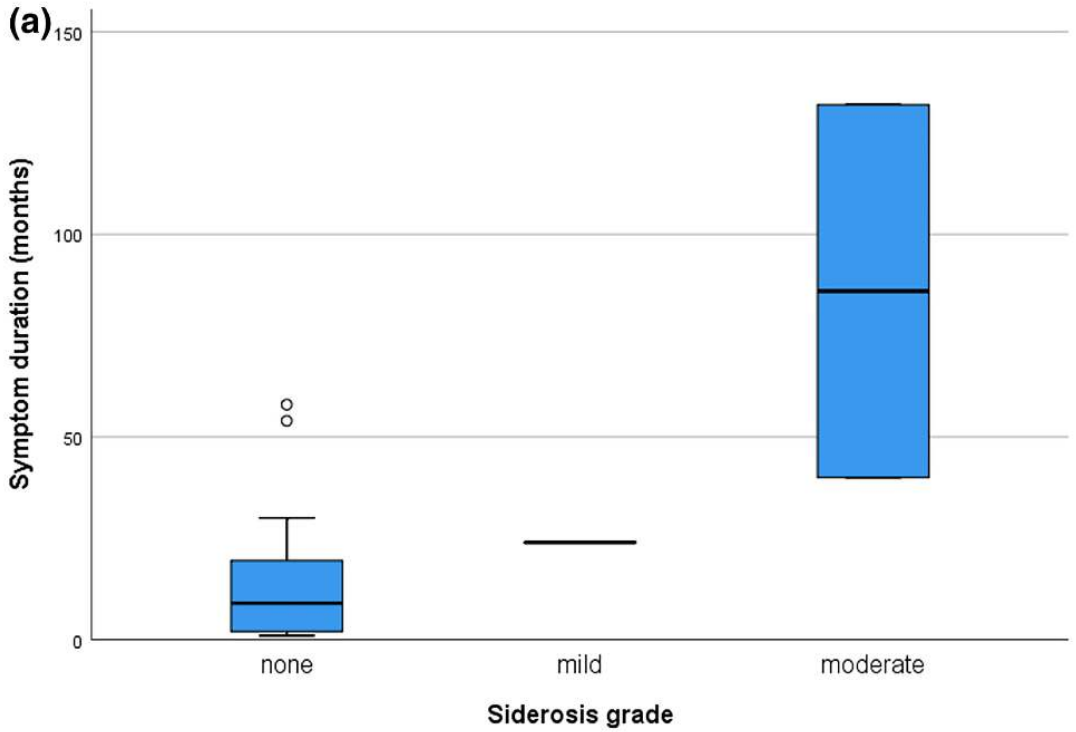


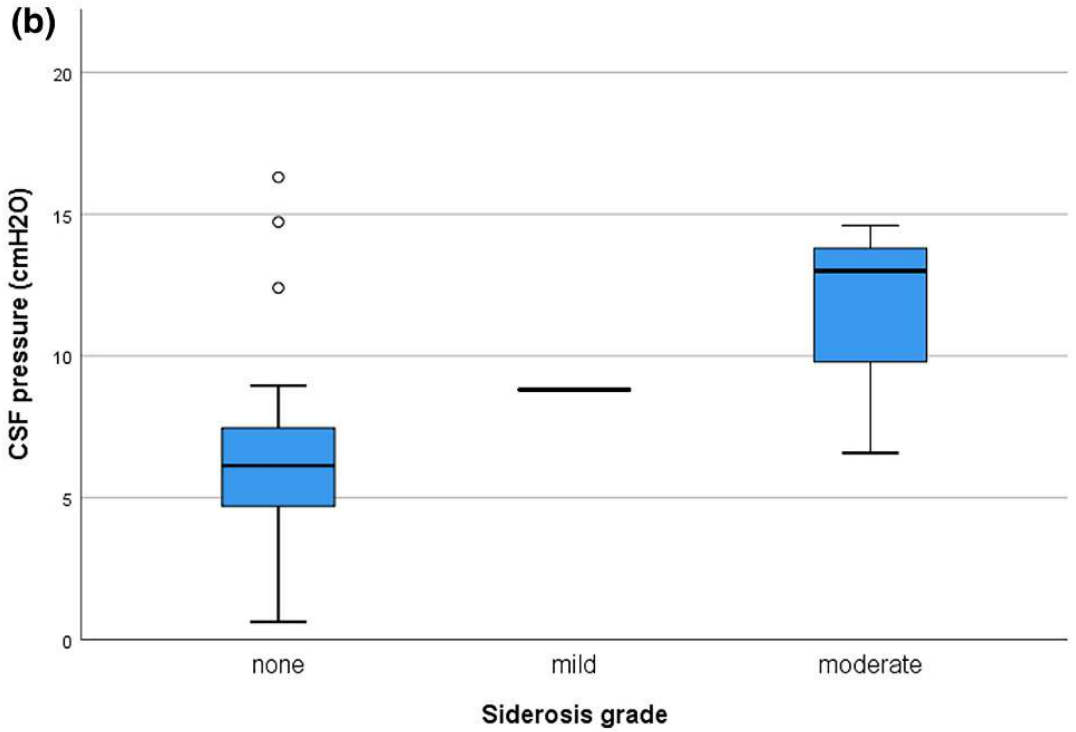
Schievink, Maya et al. Ann Neurol 2023

Cerebrospinal fluid biomarkers of superficial siderosis in patients with spontaneous intracranial hypotension

european journal of neurology

Levin Häni[1,2] | Christian Fung[2] | Christopher Marvin Jesse[1] | Christof Schild[3] | Eike Immo Piechowiak[4] | Tomas Dobrocky[4] | Andreas Raabe[1] | Jürgen Beck[2]





Häni et al. Eur J Neurol 2022

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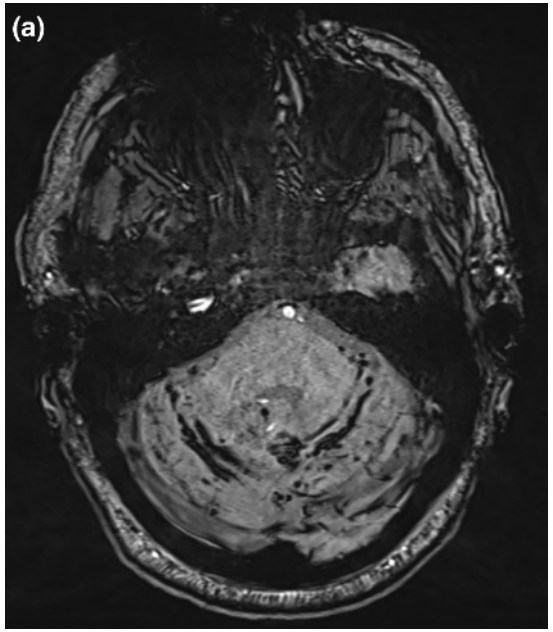

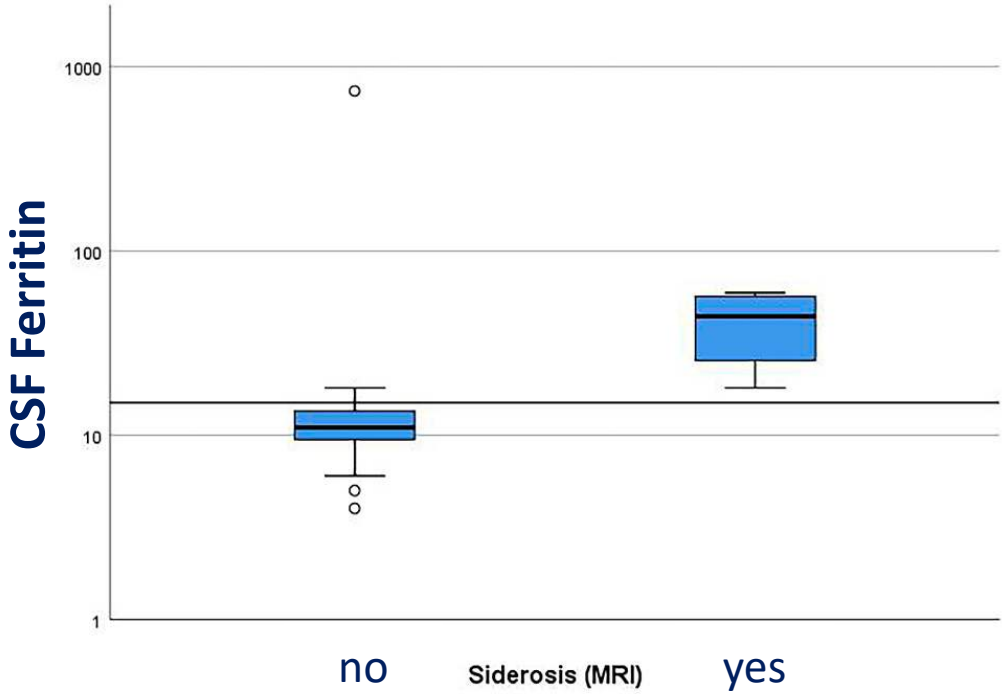

Häni et al. Eur J Neurol 2022

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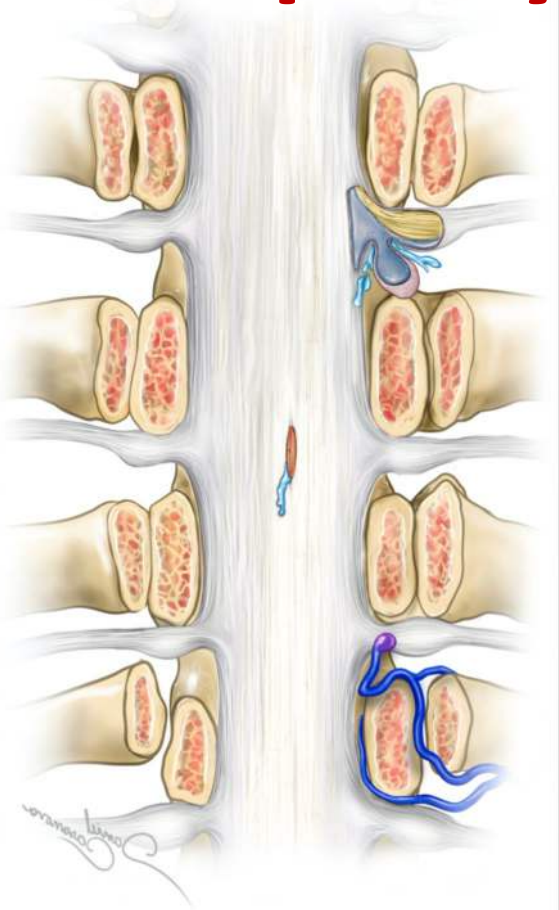
Sequelae of a spinal dural leak

„Duropathy“



Type 2

Type 1

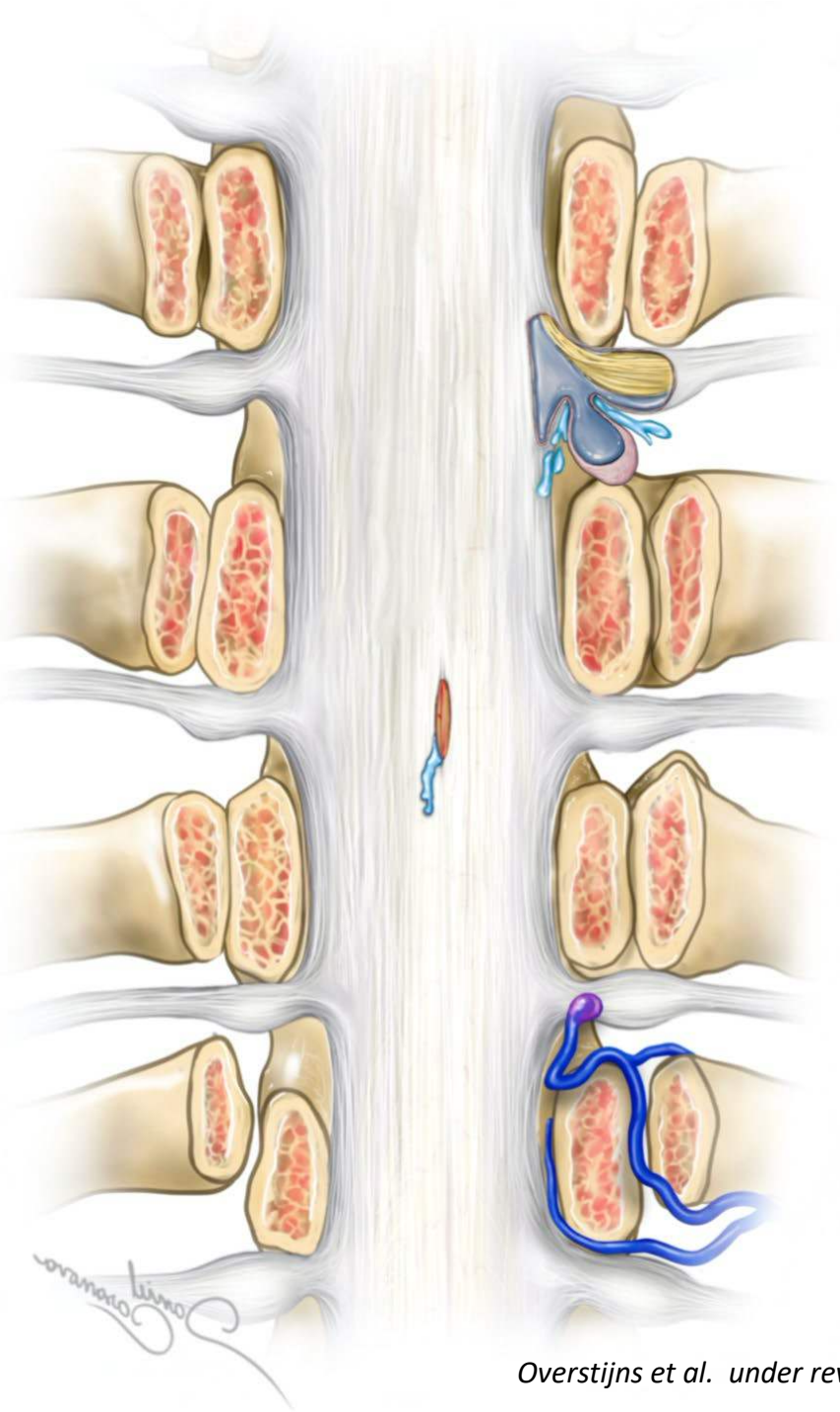
Type 3

Acute

Type ?

Chronic

- **Spontaneous intracranial hypotension**
- **Subdural hematoma**
- Venous sinus thrombosis
- Diffuse nonaneurysmal SAH
- Bibrachial amyotrophy
- Brain sagging dementia
- **Spinal cord herniation**
- **Superficial siderosis (iSS)**



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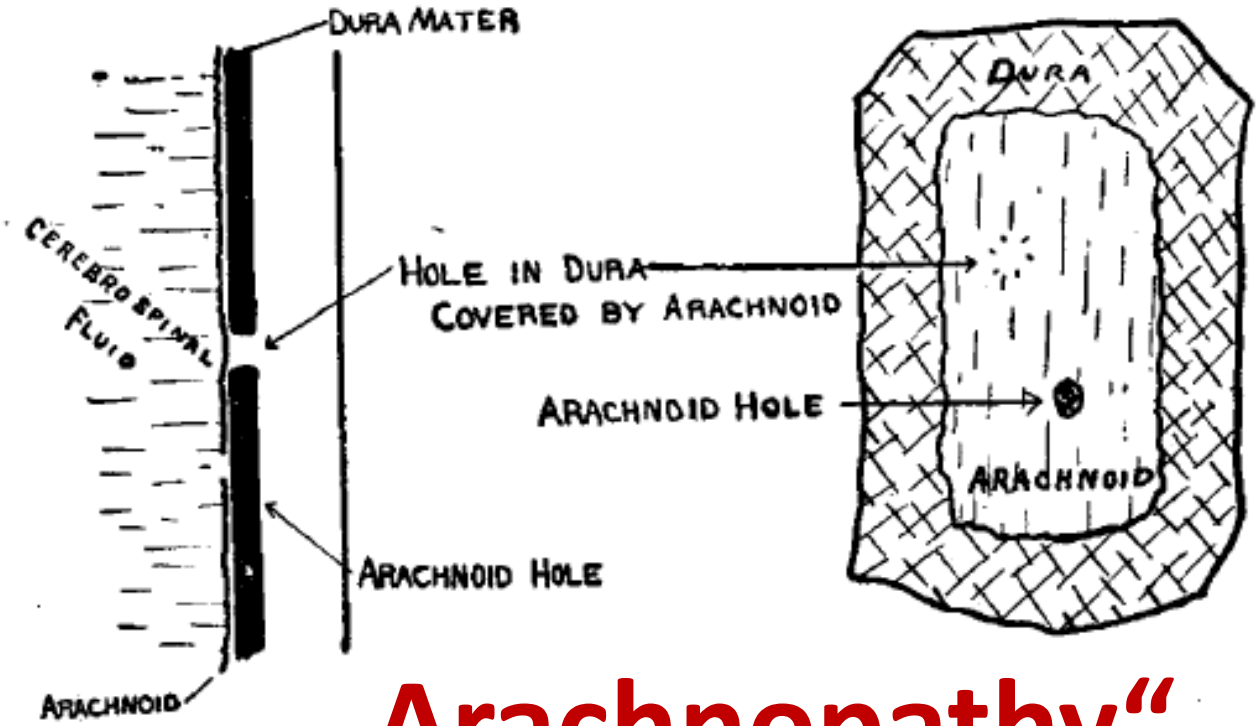

„Arachnopathy“

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MAY 11, 1918

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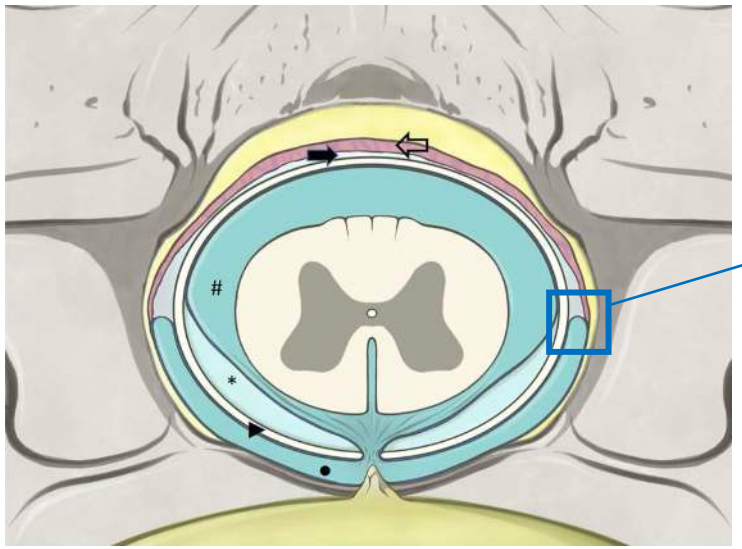
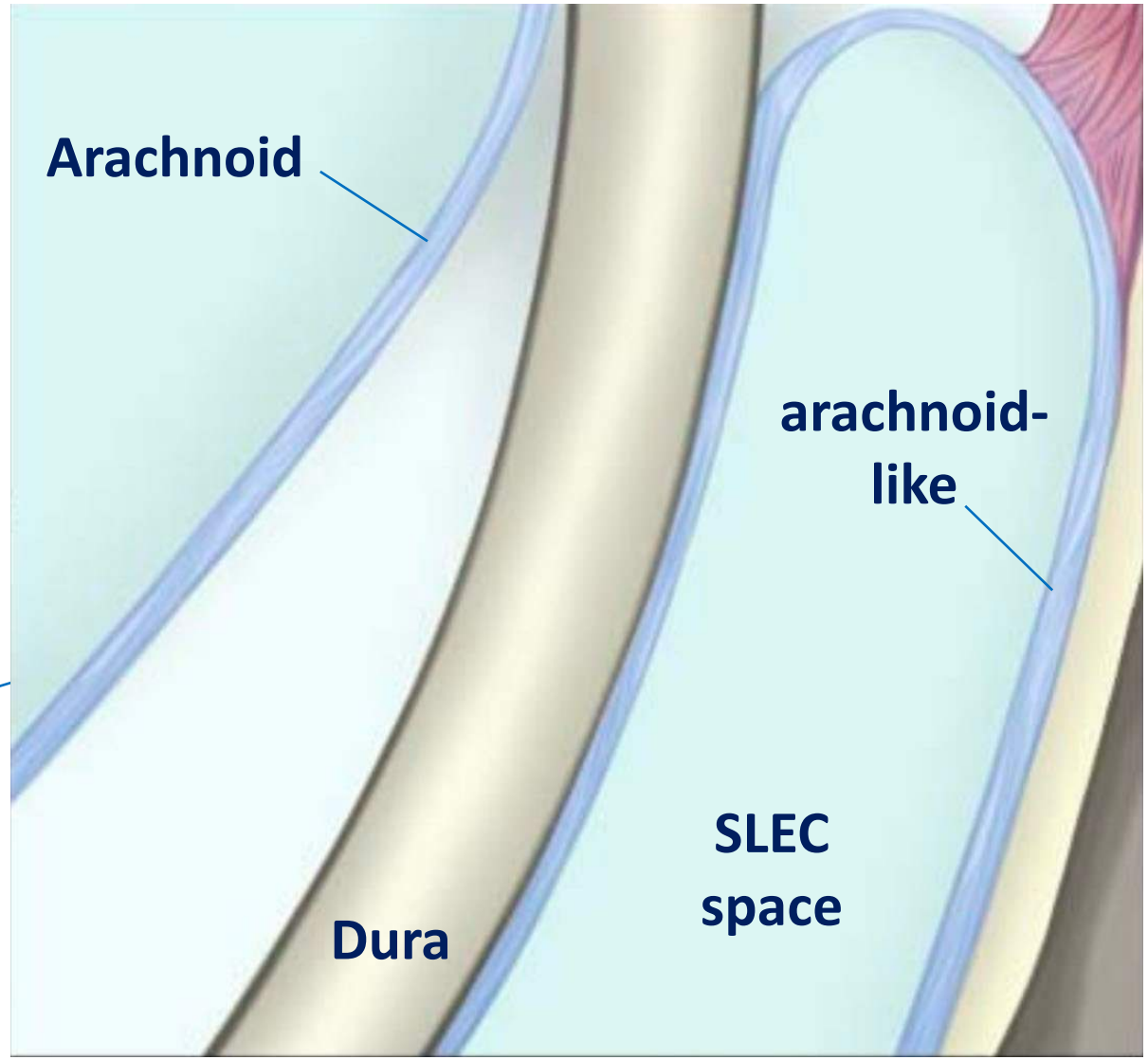
Microsurgical anatomy
-
lateral membranes





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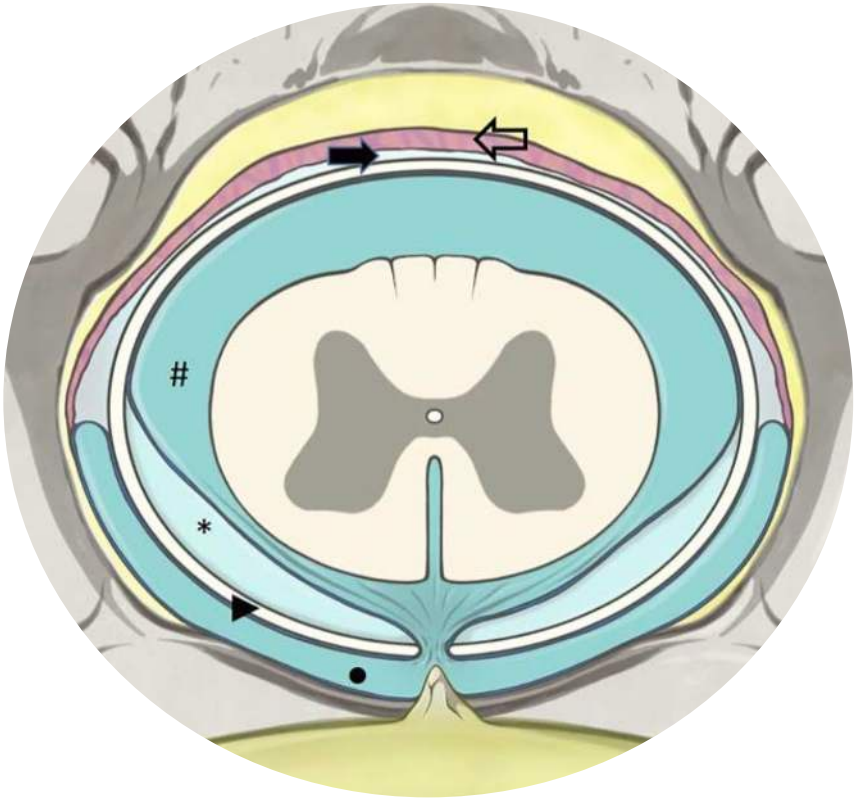
„Membranopathy“

OPEN

Distinct Pattern of Membrane Formation With Spinal Cerebrospinal Fluid Leaks in Spontaneous Intracranial Hypotension

Levin Häni, MD ‡*, Christian Fung, MD‡*, Amir El Rahal, MD‡§, Florian Volz, MD‡, Luisa Mona Kraus, MD‡, Oliver Schnell, MD‡, Roberto Ferrarese, PhD‡, Daniel Erny, MD‖, Marius Schwabenland, MD§, Horst Urbach, MD¶, Niklas Lützen, MD¶, Jürgen Beck, MD‡

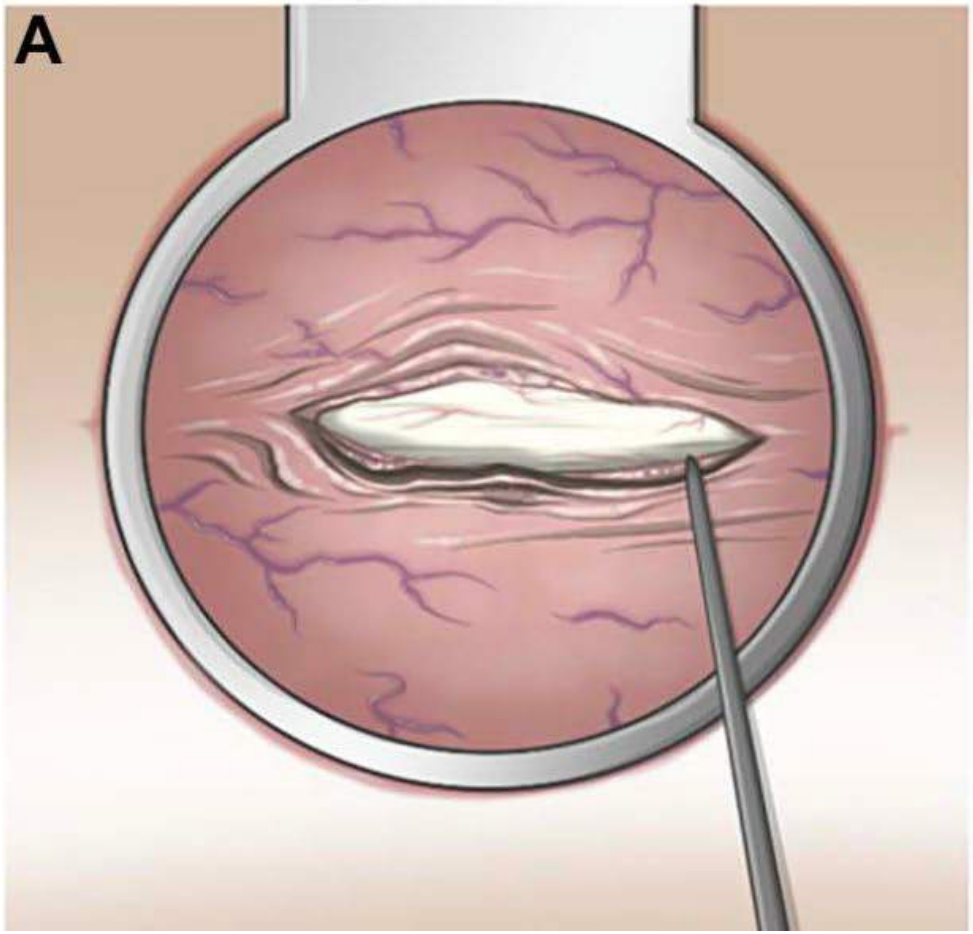


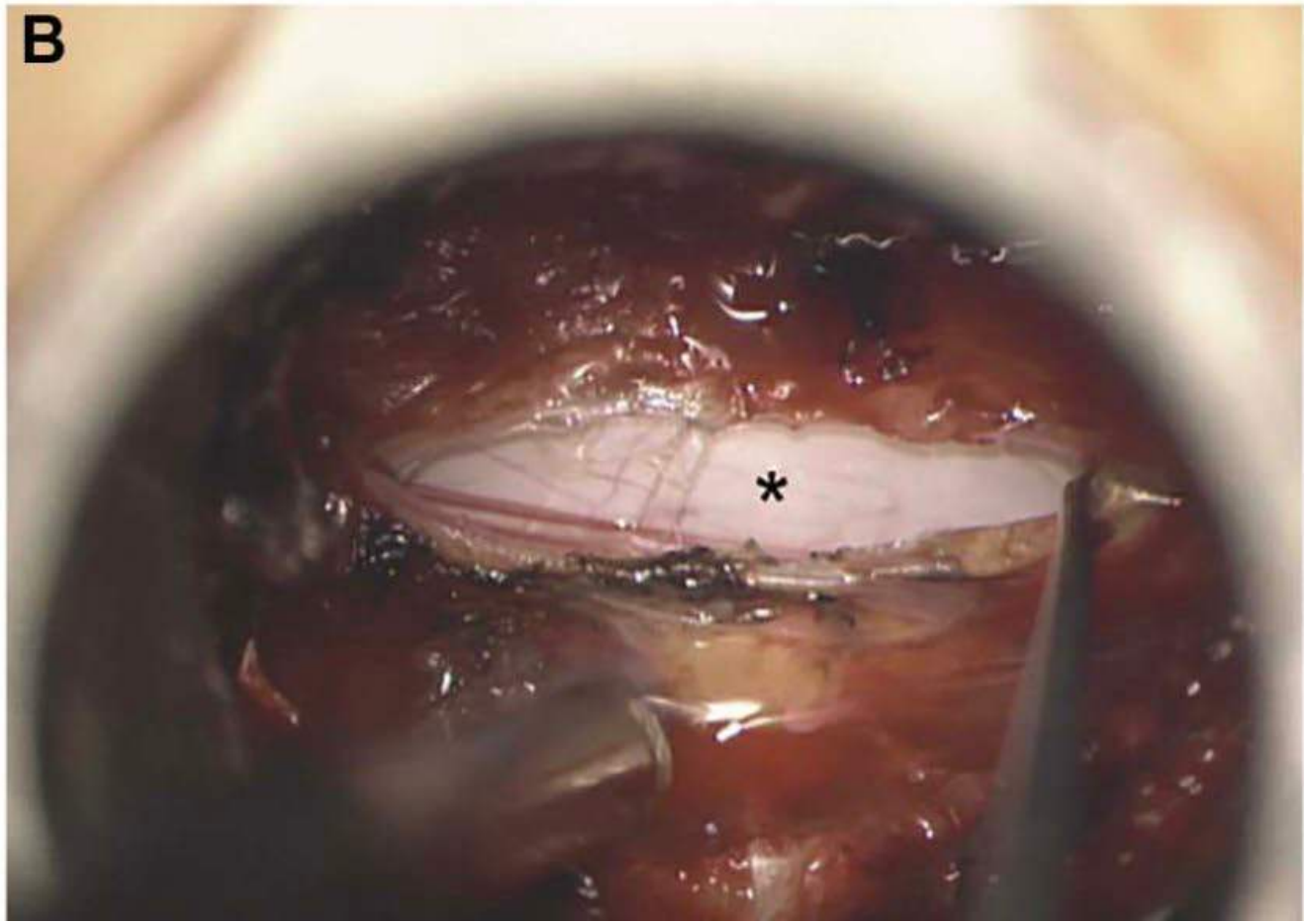
- Dorsal membranes – **neo dura**
- Lateral membranes – herniated arachnoid - SLEC
- Funnel like transdural channel – herniated arachnoid
- Lateral membrane – cysts/diverticula – at axilla of or around the exiting nerve root

- Formation of membranes or herniation of arachnoid
- Specific patho-anatomic feature of SIH
- Paramount importance for diagnosis and treatment

… all about membranes

neo dura





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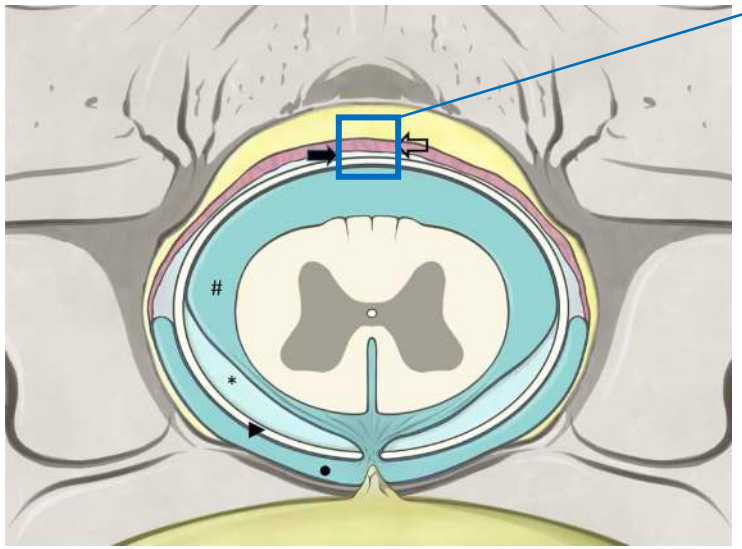
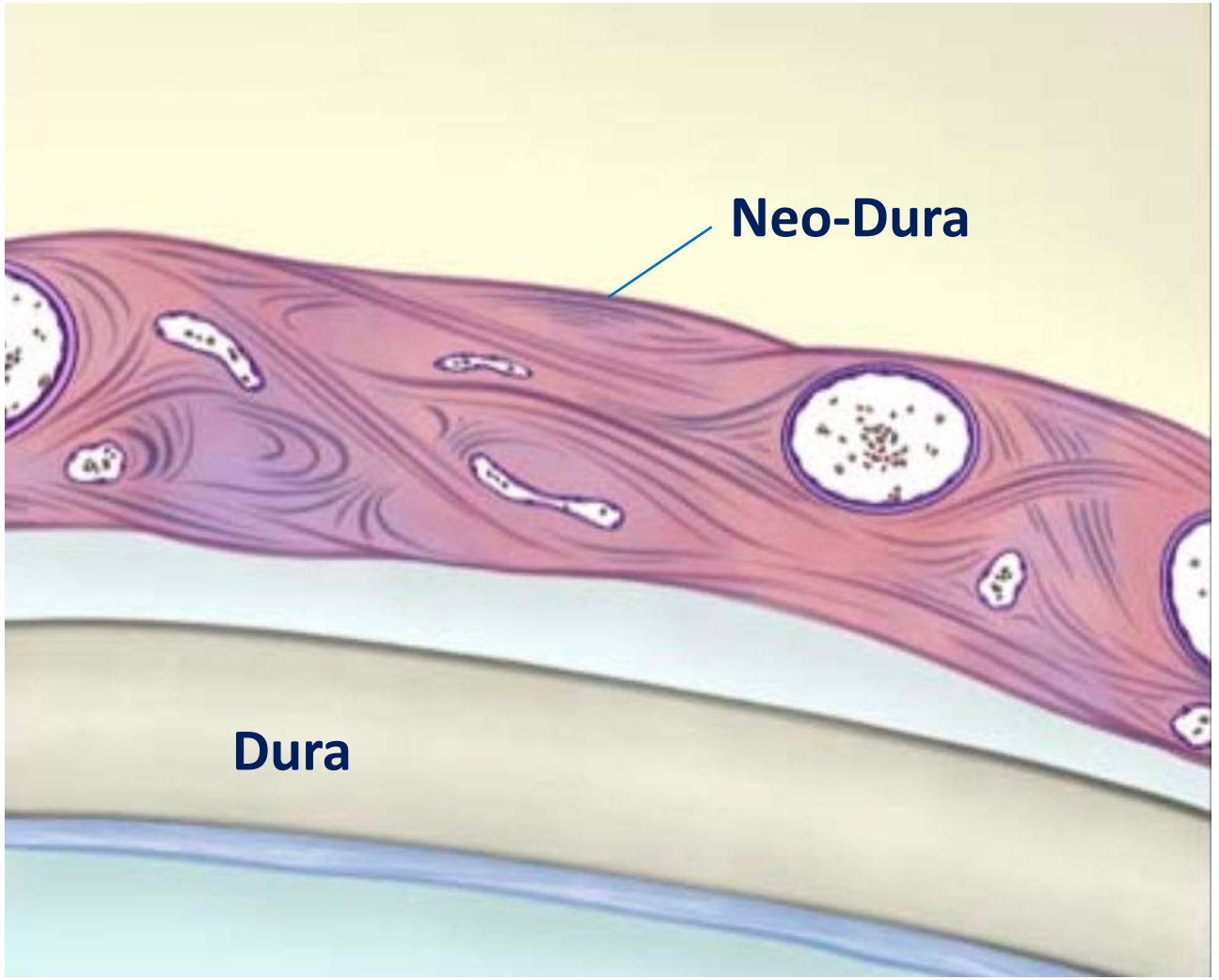
neo-dura





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Infratentorial Superficial Siderosis and Spontaneous Intracranial Hypotension

Wouter I. Schievink, MD [1], M. Marcel Maya, MD,[2] Jennifer Harris, MD,[3]
Javier Galvan, MD,[2] Rachelle B. Taché, NP-C, MSN,[1] and Miriam Nuño, PhD[4]

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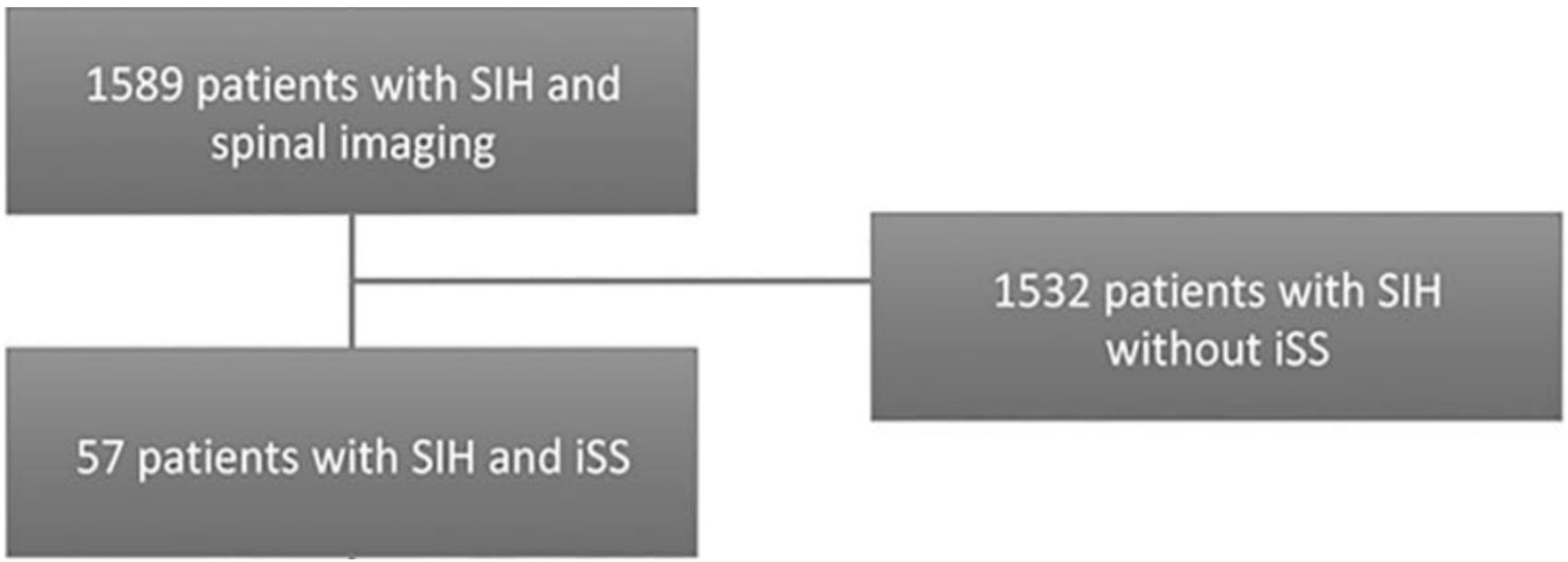

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in 57	**(10.3%)**	of 447 patients with **ventral CSF leaks**
in 2	**(3.9%**)	of 51 patients with **dural ectasia**
in 5	**(2.6%)**	of 194 patients with **CSF-venous fistulas**
in 4	**(0.9%)**	of 457 patients with simple meningeal **diverticula**
in	**(0%)**	none of the 101 patients with **lateral CSF leaks**
in none	**(0%)**	of 339 patients with leaks of **indeterminate origin**

- estimated median latency period **126 months**

- among the 57 patients with iSS 46 **(80.1%)** had a **ventral CSF leak**

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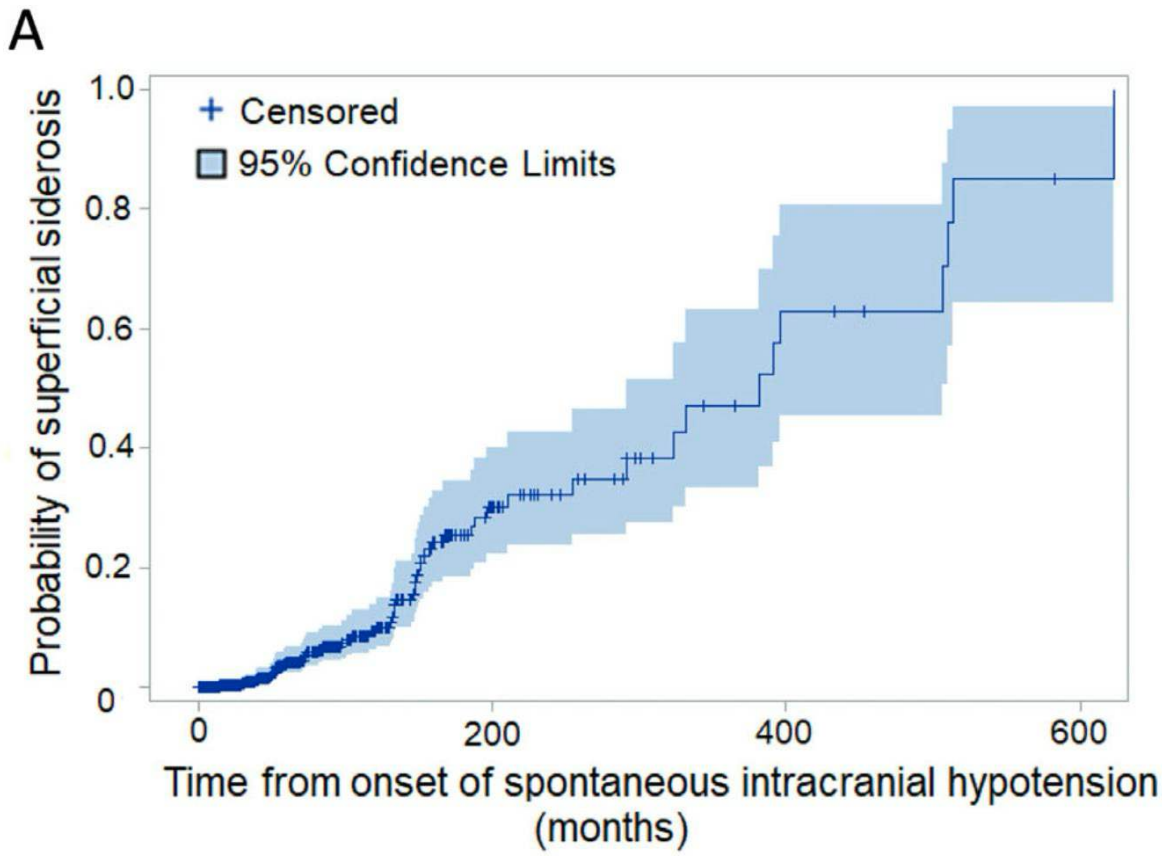
1 after weeks but

latency period was at least

- at least 1 year in 53 patients (98.1%),
- at least 3 years in 49 patients (90.7%)
- at least 5 years in38 patients (70.4%)



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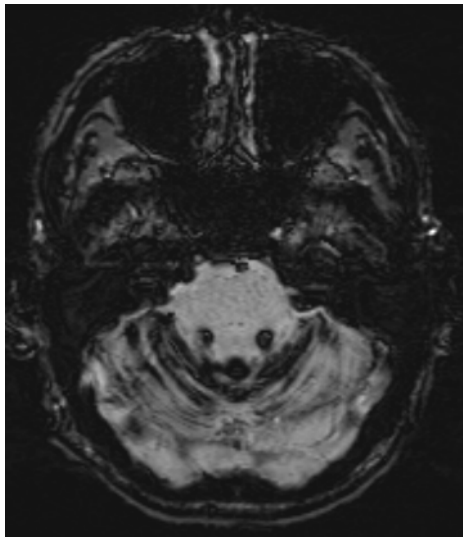
Treatment success - surgery

- **ventral CSF leaks could not be eliminated with percutaneous procedures in any patient**
- **surgical repair resulted in resolution of the CSF leak in all patients (know site)**
- **Surgical repair was associated with low risk (<5%)**

Surgical closure of spinal cerebrospinal fluid leaks improves symptoms in patients with superficial siderosis

european journal of neurology

Amir El Rahal[1,2] | Benedikt Haupt[1] | Christian Fung[1] | Debora Cipriani[1] | Levin Häni[1,3] | Niklas Lützen[4] | Tomas Dobrocky[5] | Eike Piechowiak[5] | Oliver Schnell[1] | Andreas Raabe[3] | Katharina Wolf[1] | Horst Urbach[4] | Luisa Mona Kraus[1] | Florian Volz[1] | Jürgen Beck[1]



SuSi: 1/10 000‘000

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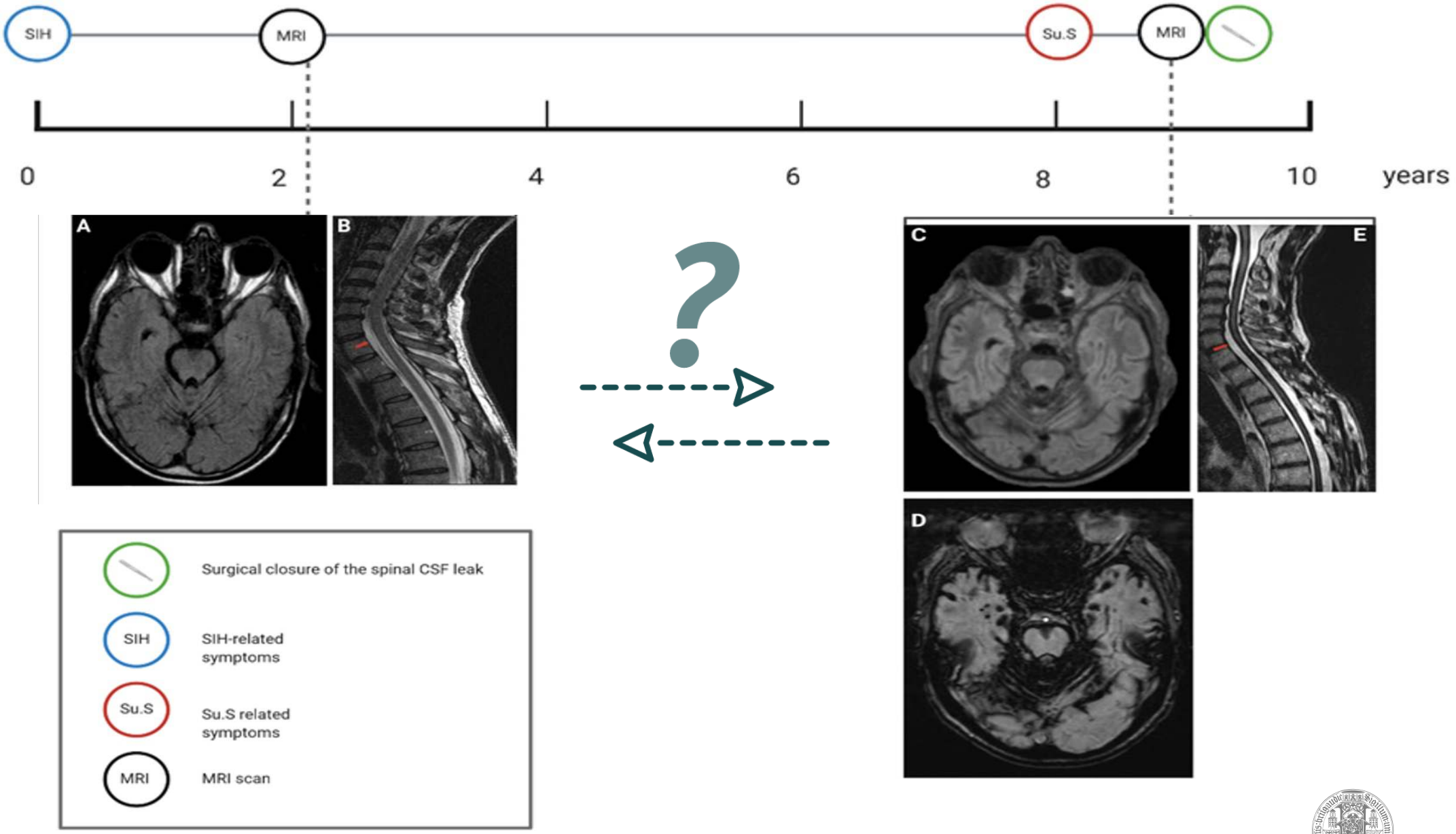

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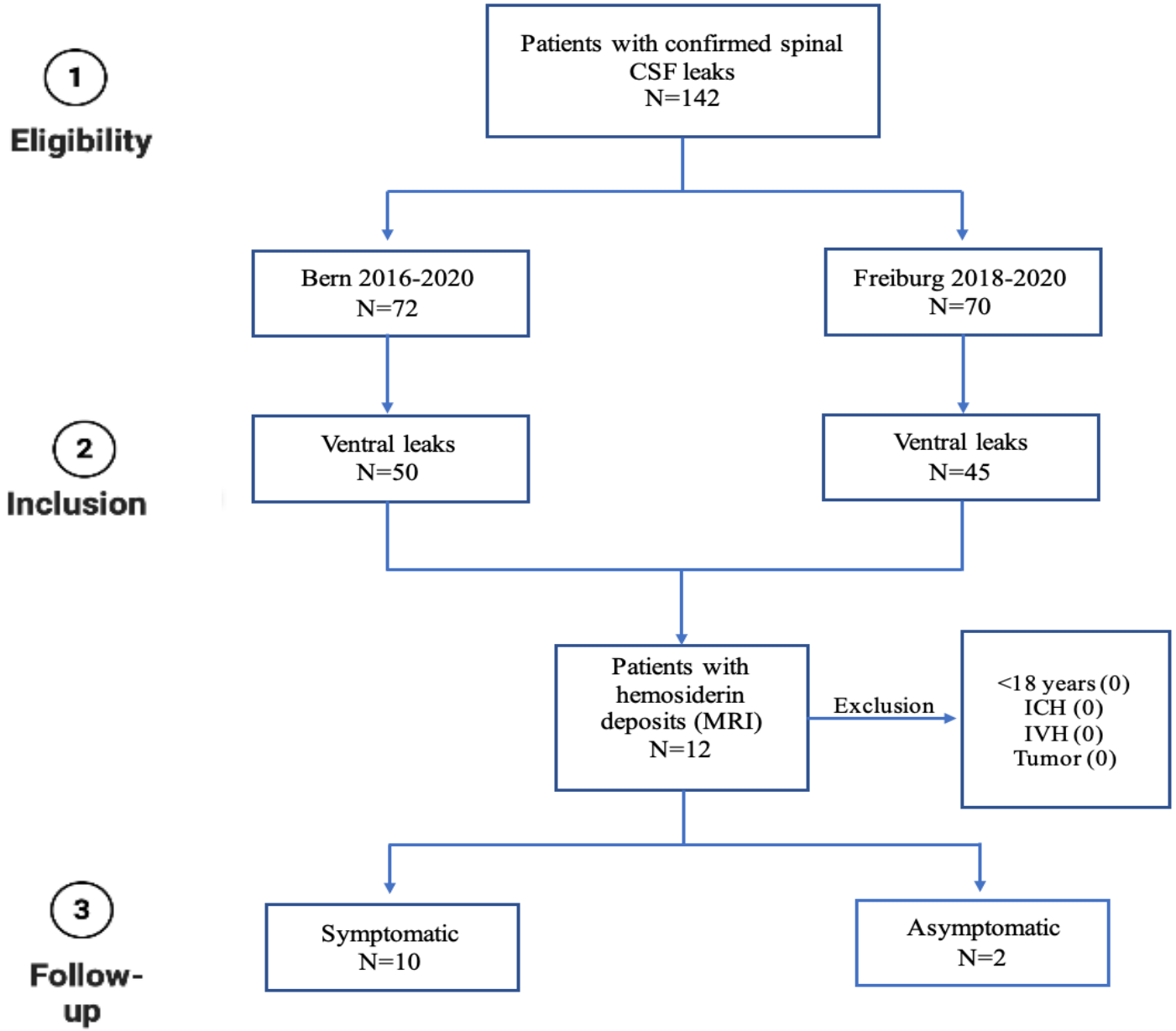
Objectives

- Association of spinal CSF leaks and superficial siderosis

- Does surgery of the spinal CSF leaks improve SS

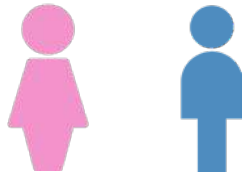

1
Eligibility
Patients with confirmed spinal CSF leaks N=142
Bern 2016-2020 N=72
Freiburg 2018-2020 N=70
2
Inclusion
Ventral leaks N=50
Ventral leaks N=45
Patients with hemosiderin deposits (MRI) N=12
Exclusion
<18 years (0)
ICH (0)
IVH (0)
Tumor (0)
3
Follow-up
Symptomatic N=10
Asymptomatic N=2

Results - Baseline characteristics

- All mainly infratentorial hemosiderin deposits

- All SLEC positive
  SLEC = spinal longitudinal epidural collection

- All CSF leaks were microsurgically sealed



N= 6 50%

N= 6 50%

Median Age – 59 y.o
(IQR +/- 13)

Mean follow up **7 months**

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Results – Outcome

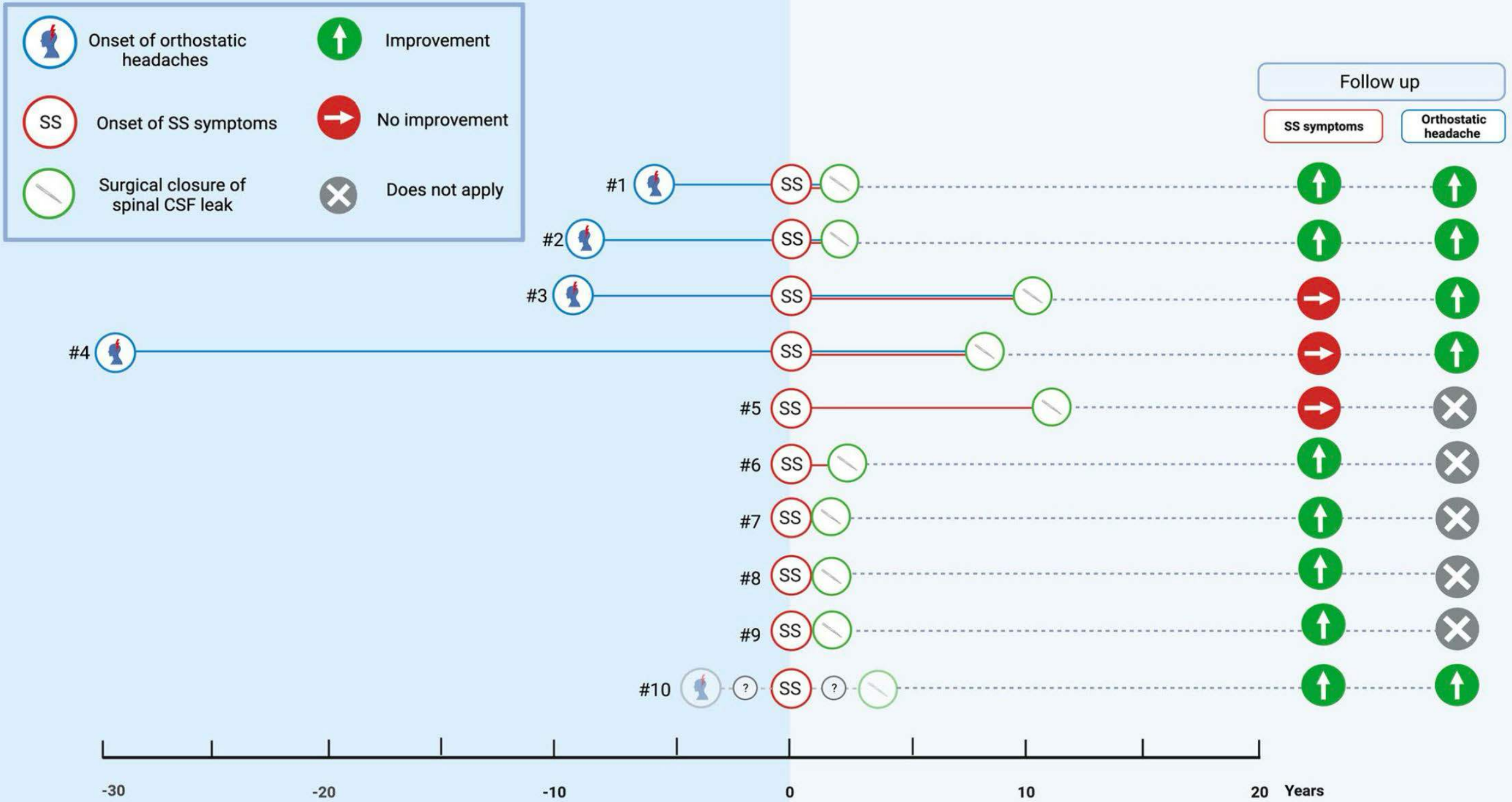
(SuSi)

- Latency SIH to SuSi 9,5 years (7-29 years)
- 70% of patients improved, 30% were stable
- Patients who improved presented after 1-3 years
- Patients who presented after 8-12 years did not improve

(SIH)

- All patient with SIH-symptoms improved
- Follow-up:
  SLEC ultimately resolved in all patients

Fisher's exact test : p= 0.01



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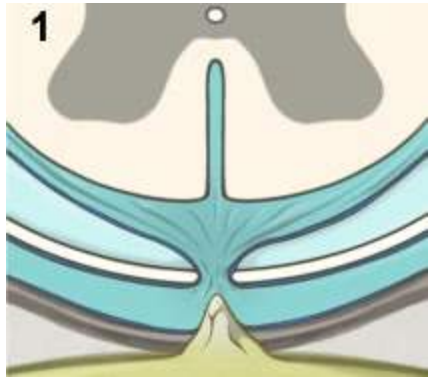
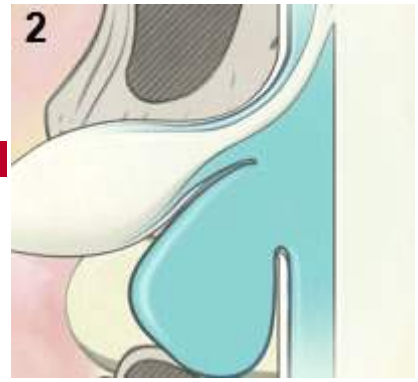
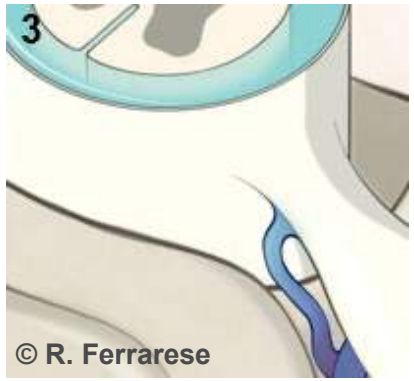
Only ventral leaks (Type I)?



Type 1:
Ventral leak ***(10.3%)***



Type 2:
Lateral leak
„leaking cyst“ ***(3.9%)***



Type 3:
CSF-venous fistula ***(2.6%)***

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Infratentorial Superficial Siderosis and Spontaneous Intracranial Hypotension

Wouter I. Schievink, MD,[1] M. Marcel Maya, MD,[2] Jennifer Harris, MD,[3] Javier Galvan, MD,[2] Rachelle B. Taché, NP-C, MSN,[1] and Miriam Nuño, PhD[4]

>70% of patients with SS Symptoms improved

30% were stable

Long-standing untreated ventral spinal CSF leaks leads to superficial siderosis of the CNS

Timely surgical closure of the leak

Highly associated with ventral leaks:
—> iterative minor epidural bleeding at the leak site

? Duropathy – Arachnopathy – Membranopathy ?

Brain Sagging dementia

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ARTICLES | April 18, 2011

Frontotemporal brain sagging syndrome

An SIH-like presentation mimicking FTD

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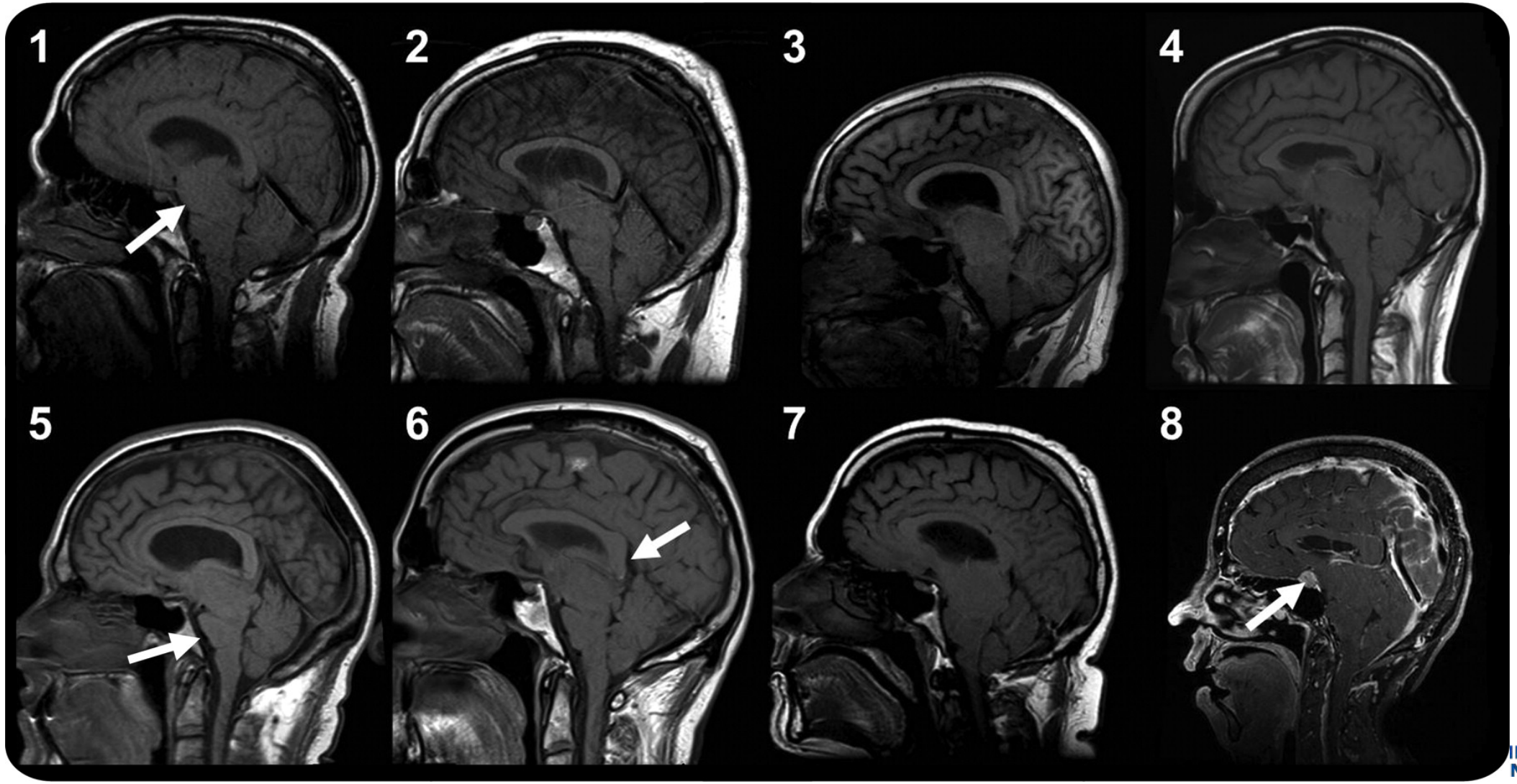


Reversible Dementia

- **bvFTD catastrophic disease – young patients – no cure**

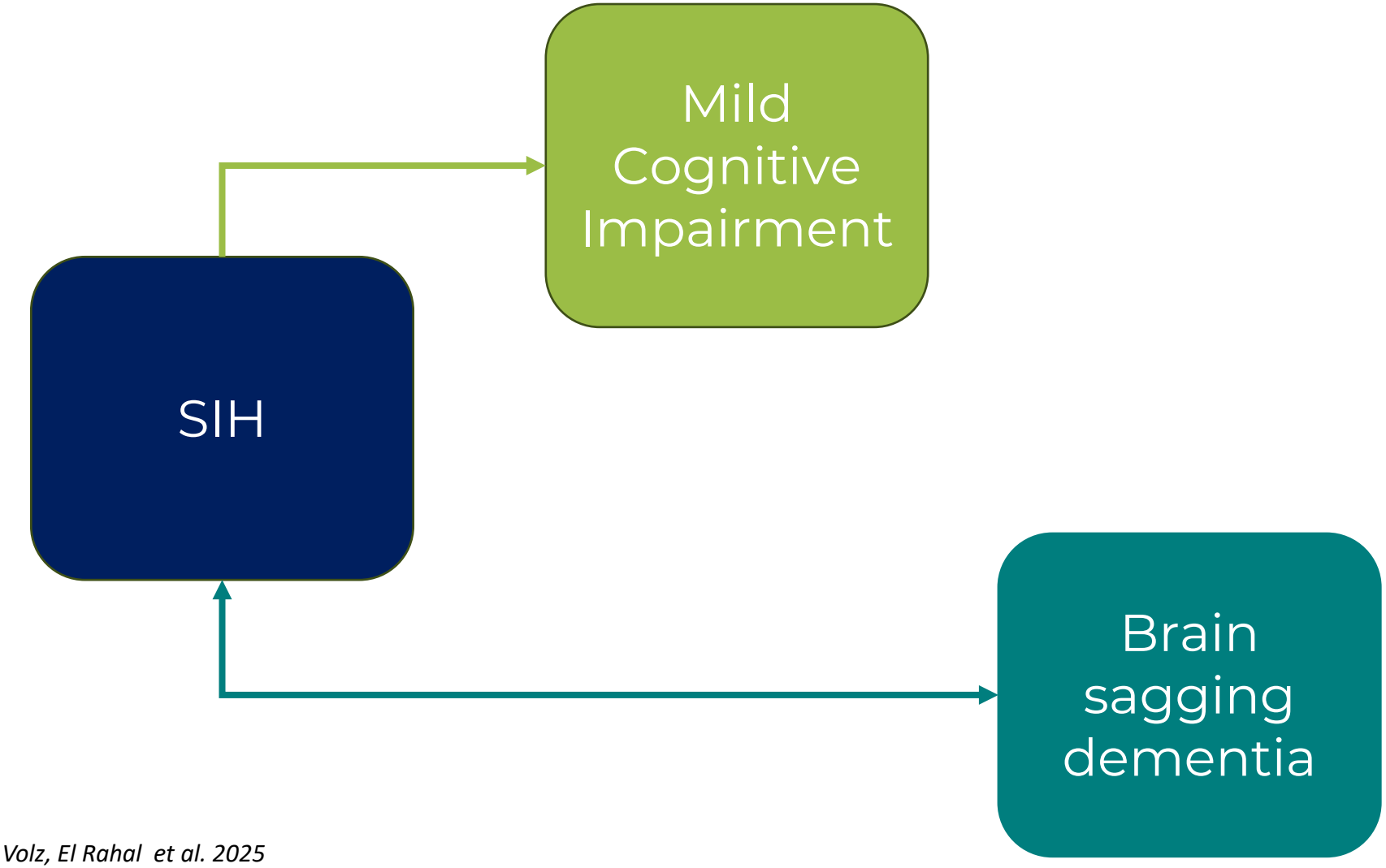
TABLE 1 Frequency of diagnostic criteria of bvFTD in 21 patients with behavioral variant frontotemporal brain sagging syndrome.

A Behavioral disinhibition:	21/21	100%
B Apathy or inertia:	21/21	100%
C Loss of empathy:	15/21	71.4%
D Perseverative behavior:	15/21	71.4%
E Hyperorality:	16/21	76.2%
F Executive dysfunction:	14/15	93.3%

43% with CSF-venous fistula – after surgery: complete or near complete recovery

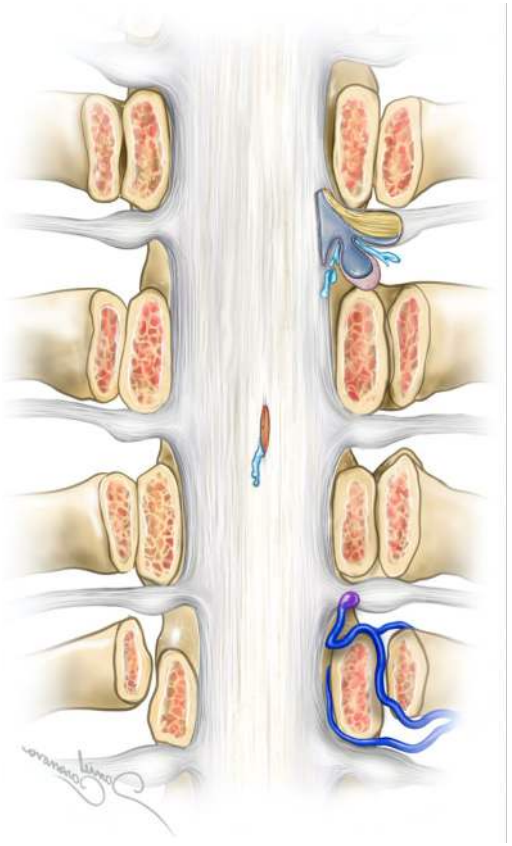
Schievink et al. Alzheimer's Dement. 2022

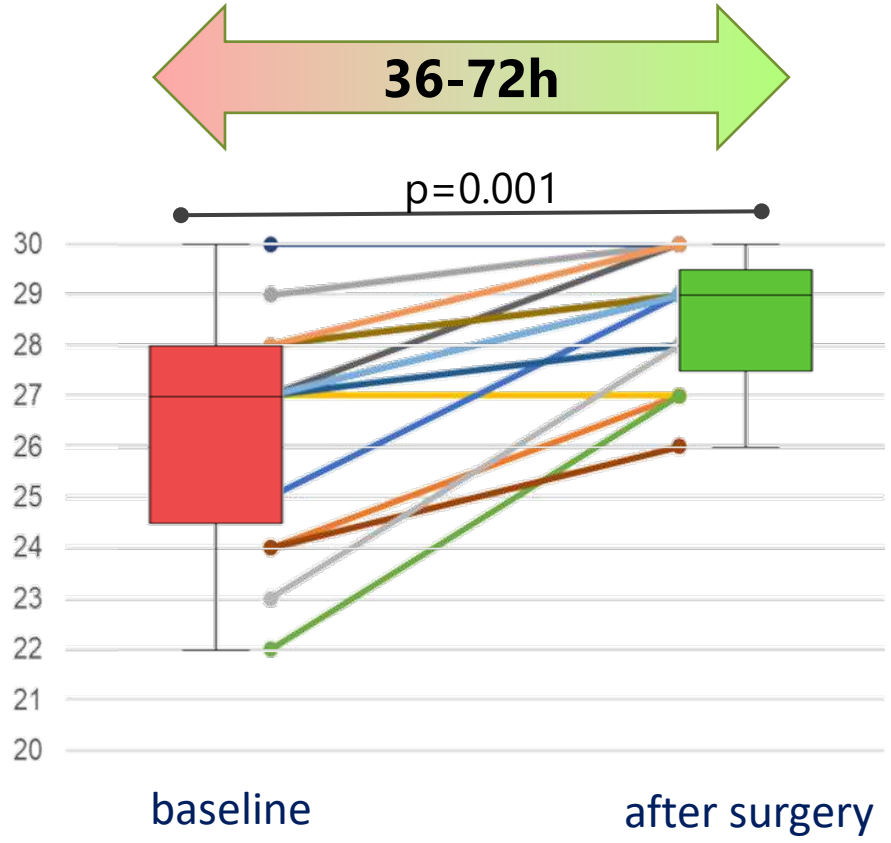
Spontaneous Intracranial Hypotension



Wolf, Volz, El Rahal et al. 2025

Cognitive impairement in SIH





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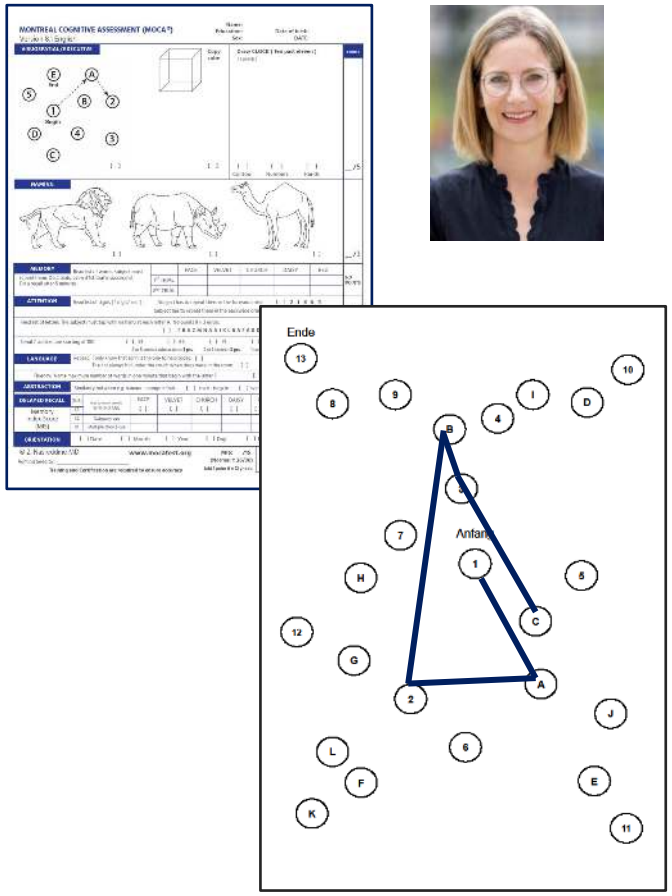
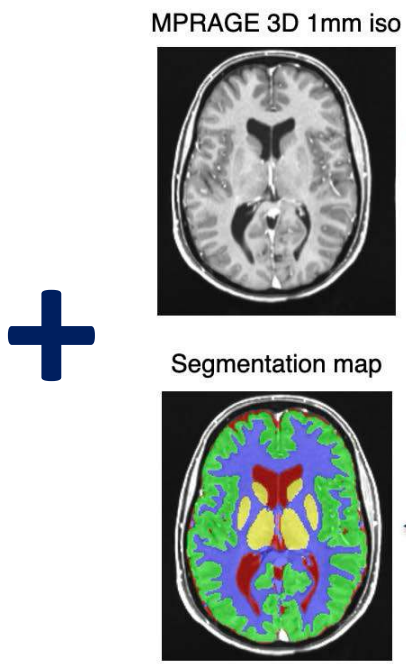
Cognitive Impairment and SIH





Standard Clinical Testing

Wolf K, El Rahal A, Beck J et al, Headache 2025

CNN - Volumetry and brain segmentation

El Rahal A, Wolf K, Volz F Beck J et al, under revision

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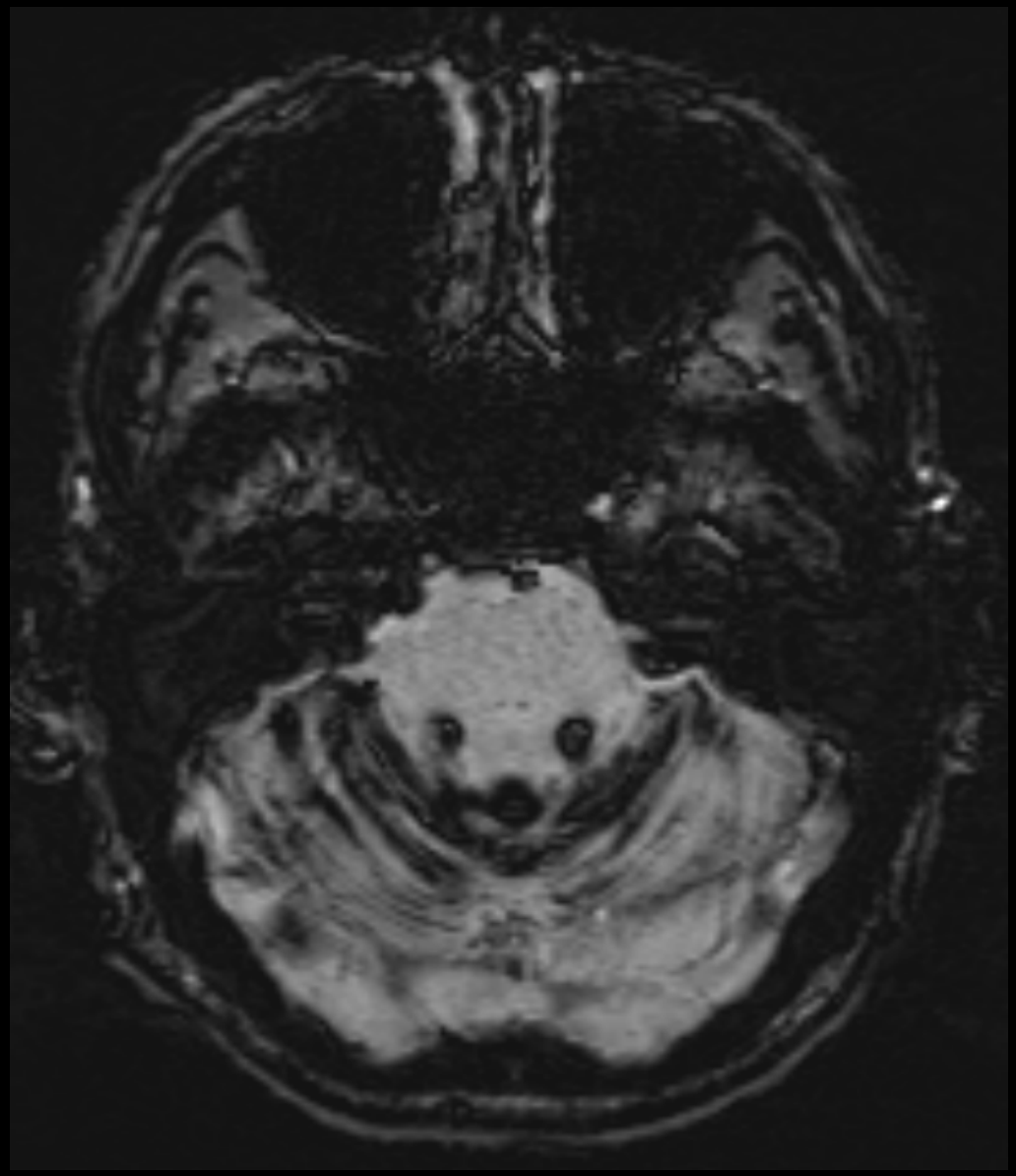

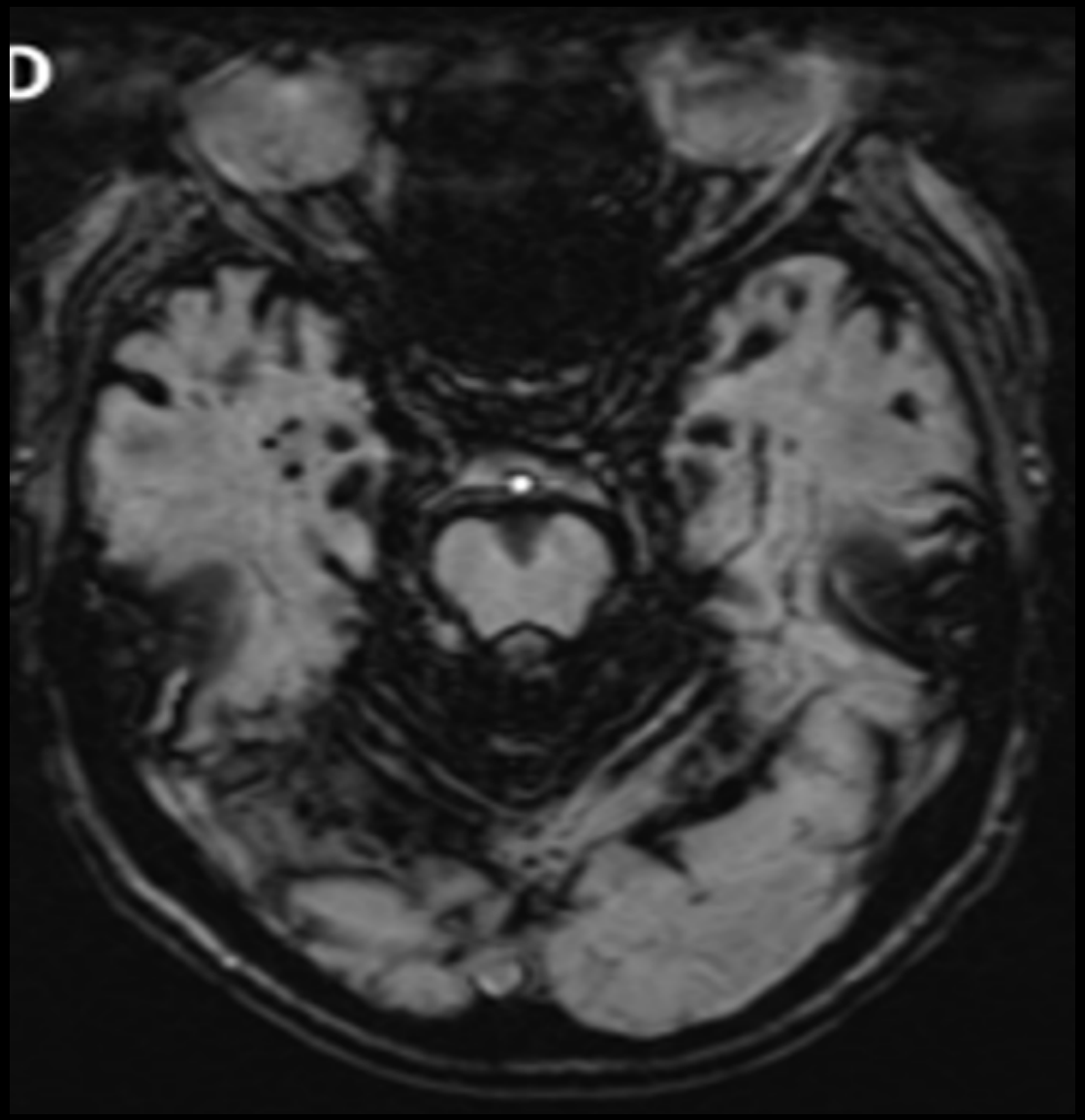

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Thank you

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