

# Clinical spectrum of SIH

Matthijs Brouwer | Neurology | 28-06-2025





# Conflicts of interest

None

Research funding

European Research Council

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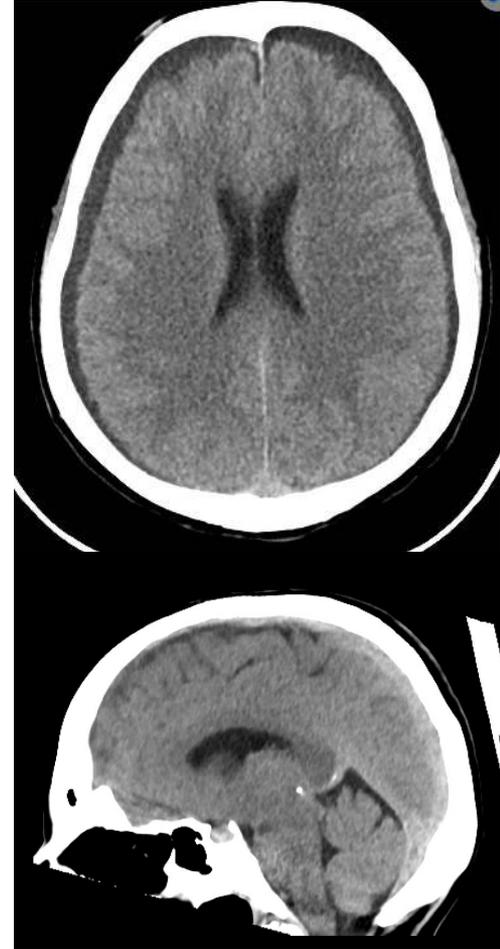
European  
Research  
Council





## 54 y/o woman, presentation ER

- Ref Orthostatic headache, bilateral horizontal diplopia
- His Unremarkable
- A 2 weeks orthostatic headache, woke up with it  
Completely disappears when lying down  
No prior trauma or other significant events  
1 week diplopia when looking left, slow in speech and responding
- Soc Manager, married 2 kids, active runner/sports
- NO E3M6V5, bradyphrenic, limited abduction both eyes (n. VI)
- Plan Cranial CT





# Clinical spectrum of SIH

The NEW ENGLAND JOURNAL of MEDICINE

REVIEW ARTICLE

Allan H. Ropper, M.D., *Editor*

## Spontaneous Intracranial Hypotension

Wouter I. Schievink, M.D.

**Table 1. Clinical Manifestations of Spontaneous Intracranial Hypotension.\***

Manifestations	No. of Patients (%)
<b>Most common</b>	
1. Headache	199 (98.5)
Orthostatic	188 (93.1)
Nonpositional	7 (3.5)
Reverse orthostatic	4 (2.0)
<b>Common</b>	
2. Vestibulocochlear symptoms	
Dizziness or vertigo	102 (50.5)
Muffled hearing or ear fullness	75 (37.1)
Tinnitus	56 (27.7)
Hypoacusis	53 (26.2)
Hyperacusis	12 (5.9)



# Clinical spectrum of SIH

<b>Common</b>	
Nausea and emesis	99 (49.0)
Disequilibrium	86 (42.6)
Posterior neck pain	69 (34.2)
Cognitive impairment	64 (31.7)
Fatigue	49 (24.3)
Phonophobia or photophobia	41 (20.3)
Visual blurring	36 (17.8)
Facial numbness, paresthesias, or pressure	32 (15.8)



# Clinical spectrum of SIH

Least common	
Interscapular pain	22 (10.9)
Dysgeusia	15 (7.4)
Low back pain	8 (4.0)
Behavioral-variant frontotemporal dementia	5 (2.5)
Bibrachial amyotrophy	3 (1.5)
Superficial siderosis†	3 (1.5)
Cerebral venous thrombosis	2 (1.0)
Diplopia with abducens-nerve palsy	2 (1.0)
Spinal cord herniation	2 (1.0)
Coma — GCS score, $\leq 8$ ‡	1 (0.5)
Syringomyelia	1 (0.5)
Hemifacial spasm	1 (0.5)



# But... when do we call it SIH?

\*International Headache Society Criteria (ICHD-III): Headache required

**Table 2. Modified ICHD-III Diagnostic Criteria for Spontaneous Intracranial Hypotension.\***

- A. Any headache attributed to low CSF pressure or CSF leakage that meets criterion C, below
- B. Either or both of the following:
  - Low CSF pressure (<60 mm CSF)
  - Evidence of CSF leakage on imaging
- C. Headache that developed in temporal relation to the low CSF pressure or CSF leakage or that led to its discovery
- D. Headache not better accounted for by another ICHD-III diagnosis



# But... when do we call it SIH?

\*International Headache Society Criteria (ICHD-III): Headache required

However, SIH without headache occurs (>10% of 200 pts)

Presentation:

- (Orthostatic) Tinnitus
- (Orthostatic) Hearing loss
- Gait disturbance
- (Bi)brachial amyotrophy
- Cognitive impairment
- Transient loss of consciousness

Radiological diagnosis → frequently an unexpected finding

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# Clinical presentation: Spontaneous or not?

Spontaneous 65%

Precipitating events 35%

- Positional change
- Valsalva
- Minor (to moderate) trauma

	Precipitating event	
Type of event	Instantaneous or <1 min	>1 min to <24 h
Positional changes*	Picking up empty bottles	
	Picking up toiletries	
	Picking up small refrigerator	
Valsalva maneuver	Heaving/throwing heavy rock	Bench pressing
	Extreme straining at stool	Weight-lifting
	Extreme straining during sex	Lifting heavy crates
	Choking due to laughing while drinking	
	Weight-lifting	
Minor injury	Uncontrollable sobbing/emotional upheaval	
	Yoga stretching	Fall during stunt work
	Twisting back while reaching backward	Assault with neck twisting
	Self-manipulation neck	Children walking on back
	Hit rock while mountain biking	Yoga stretching
		Yoga stretching
		Yoga pulling head up
		Surfing wipe-out
		Tae-bo (kickboxing)
		Rollercoaster ride
	Chiropractic neck manipulation	
	Boogie-boarding accident	
	Forceful flexion hips during sex	



# 54 y/o woman, admitted with SIH

Diagnosis SIH confirmed by MRI

Autologous epidural blood patch → better during 2 days

1 week later

Progressive bradyphrenia, altered mental status (E1M5V3)

Transfer Amsterdam UMC

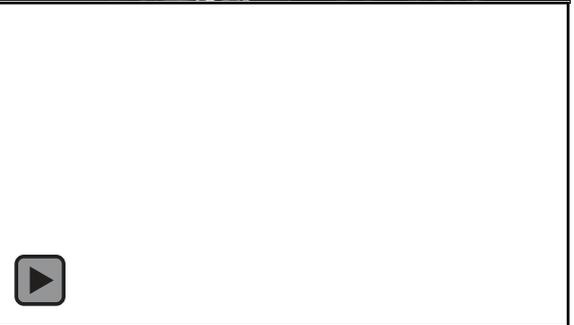
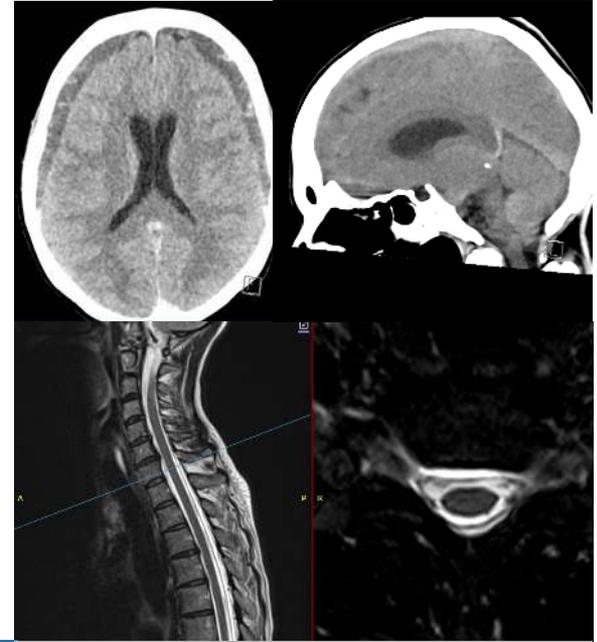
Digital subtraction myelography

Leak Th4-5

Surgical repair

Complete recovery 2 weeks later

Walking in the Alps 6 weeks later





# Common, typical presentation

23 y/o man, outpatient clinic

Lying flat on the ground in the waiting room

Severe orthostatic headache 3 weeks, muffled hearing

Started while swimming lanes

Hypersomnolence, sleeps 20 hours a day, apathy

MRI consistent with SIH

Epidural blood patch 2 levels 35 cc

Complete recovery





# Less common, but typical presentations

36 y/o, male

Progressive paresis both arms since 5 years

Diagnosed segmental spinal muscular atrophy

No headache or arm pain, no sensory symptoms

EMG shows denervation, positive Awaji criteria

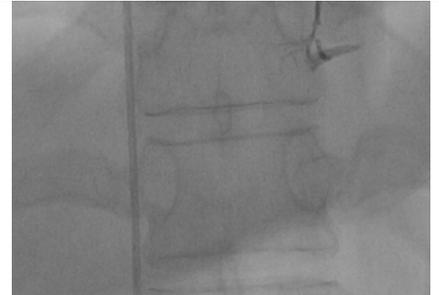
Cranial MRI: subtle signs of intracranial hypotension

Spinal MRI: no abnormalities

Dynamic subtraction myelography

Endovascular embolisation

Stabilization of symptoms, minor recovery





# Bibrachial amyotrophy

Amsterdam UMC 11 patients in 5 years (10 male; 5% total patient population)

2 of 11 remember initial episode of headache

Cedars-Sinai hospital (conference Hawaii) 20 patients in 16 years

Progressive paresis, muscular atrophy and fasciculations

Initial diagnosis of ALS, progressive or segmental spinal muscular atrophy

Duration of symptoms 2-15 years (diagnostic delay)

9 ventral longitudinal epidural CSF collection, dural tear

1 CSF-venous fistula

1 Myazaki syndrome (overdrainage myelopathy, f)





# Less common, but typical presentations

72 y/o man, 2023 progressive gait disturbance and hearing loss

Medical history 2013 bilateral subdural hematoma

No headache

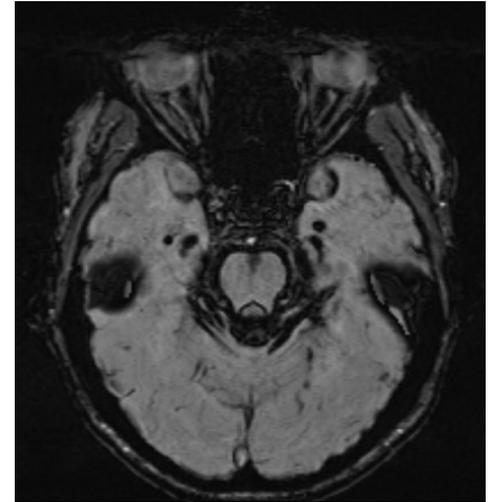
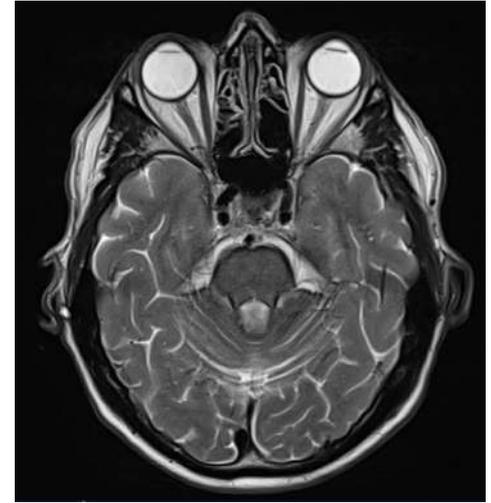
ENT analysis      Cranial MRI - superficial siderosis (SWI images)

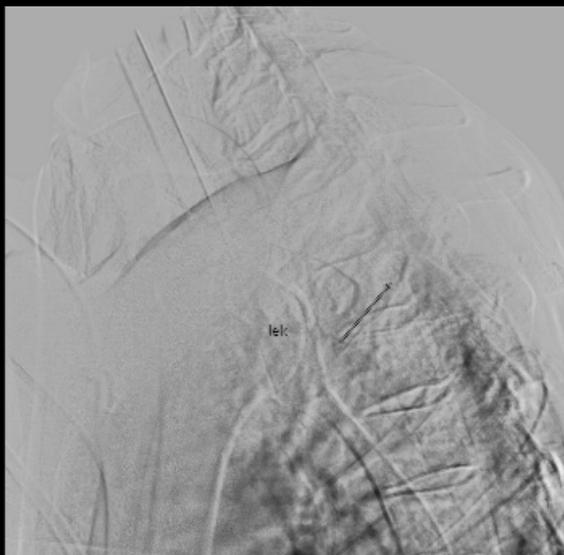
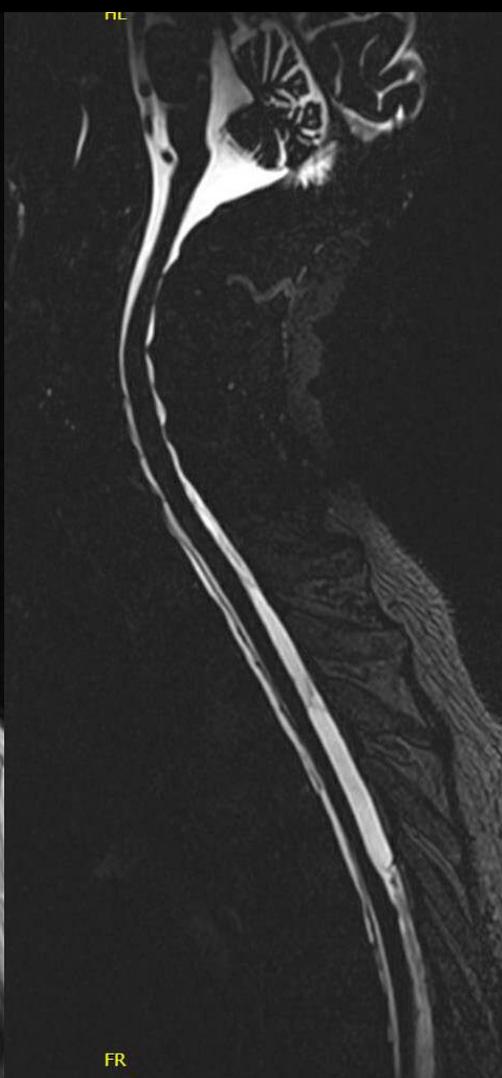
Otherwise no signs of SIH

Spinal MRI longitudinal epidural CSF collection

Digital subtraction myelography: leak Th2-3

Surgical repair → Hearing did not improve, walking is better







# Superficial siderosis

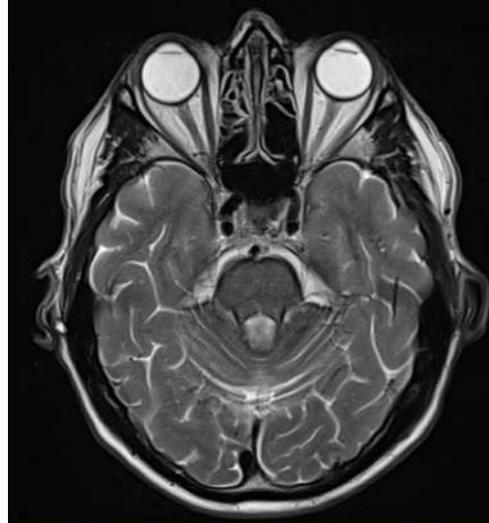
11 patients in 5 year, in 8 proven CSF leak

Main characteristic:

- Older age (70-80)
- Hearing loss
- Cerebellar syndrome
- Pyramidal and other long tract signs

Differential diagnosis

- Vascular malformation
- Neoplasm
- Idiopathic → probably at least some undiagnosed SIH





# Less common, but typical presentation

58 y/o woman, referral

3 month headache and cognitive decline

Apathy, lack of initiative, needs help in ADL from 80+ y/o parents

Desoriented, loses her way, loss of emotions

NE      Bradyphrenia, does not know what day it is

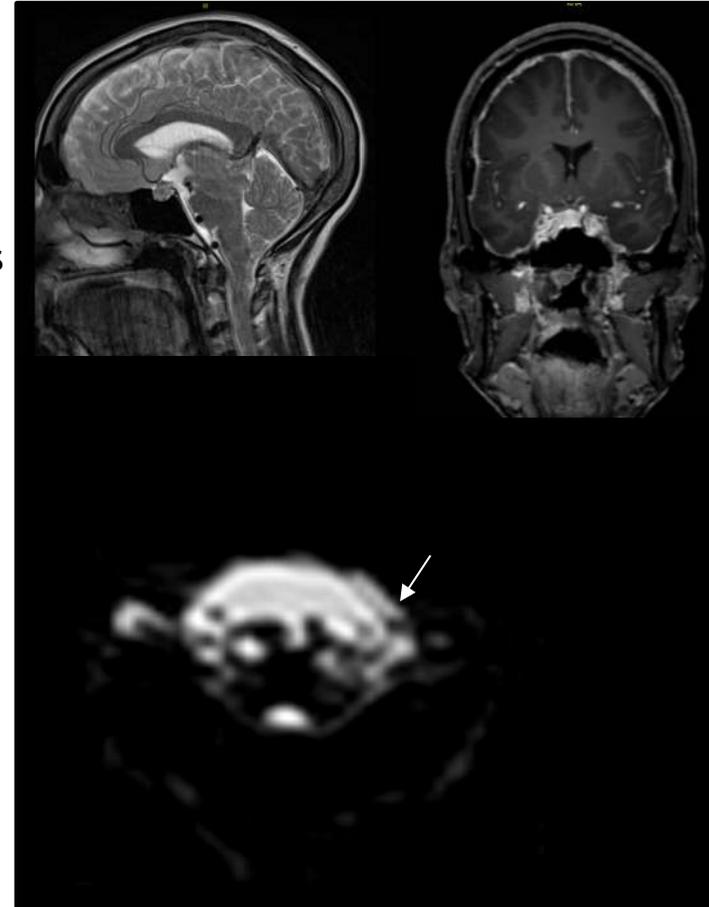
Repeating words 3/3, recall 0/3, MOCA 22/30

NPE      Severely reduced imprinting

Epidural blood patch → MOCA 30/30

DSM      Leak dural cyst L1

Surgical repair





# Cognitive disorders in CSF leaks

“Brain sagging dementia”

Fatigue / hypersomnolence

Behavioral disinhibition

Apathy, loss of empathy, hyperorality

Perseverative (repetitive) behaviour

Often clinical picture similar to behavioral variant  
of frontotemporal dementia

Reversible cognitive disorders

**TABLE 1. Manifestations of Spontaneous Intracranial Hypotension in 29 Patients With Symptoms of Behavioral Variant Frontotemporal Dementia.**

Hypersomnolence	29	(100%)
Headache	27	(93%)
Orthostatic	20	(69%)
Nonpositional	4	(14%)
Reverse orthostatic	3	(10%)
Auditory symptoms	21	(72%)
Dysequilibrium/gait dysfunction	19	(66%)
Tremors	15	(52%)
Posterior neck pain	13	(45%)
Nausea/emesis	10	(34%)
Dysphagia/dysarthria	9	(31%)
Orofacial dyskinesia	6	(21%)
Blurred vision	5	(17%)
Dysgeusia	4	(14%)
Impotence/erectile dysfunction	4	(14%)
Incontinence	4	(14%)
Hiccupping	3	(10%)
Abducens nerve palsy	2	(7%)
Coma	1	(3%)
Trigeminal neuralgia	1	(3%)



## Less common, not very typical presentation

80 yo woman

4 y history of episodes of vertigo and hearing loss

No headache, one episode of diplopia during weeks

MRI consistent with SIH

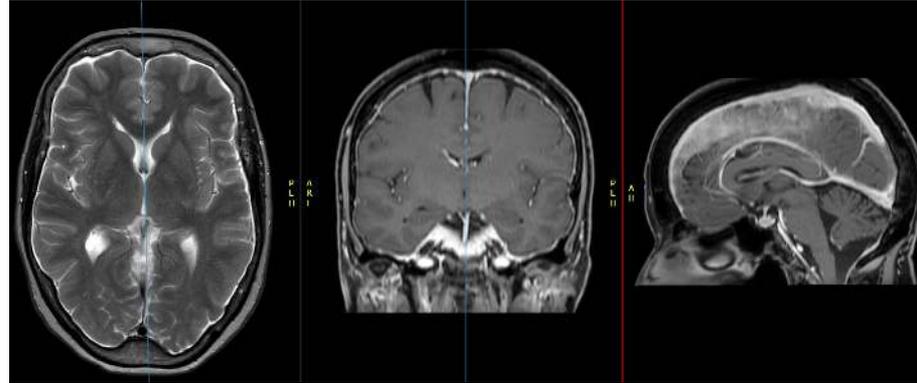
Epidural blood patch:

- substantial return of hearing for 1 week

2x myelography no leak

- again short periods of improved hearing

New epidural blood patch, direct improvement, 50% reduction symptoms after 4 weeks





# Less common, not very typical presentation

77 y/o woman

3 y history orthostatic pulsatile tinnitus and hearing loss

No headache, symptoms not present when lying down

MRI consistent with SIH

Epidural blood patch:

- Improved MRI, no clinical response

Myelography CSF venous fistula Th5 - embolisation not possible

Neurosurgical treatment

Gradually improving





# Conclusion clinical spectrum of SIH

Typical, common

(Sub)Acute Orthostatic headache

- accompanied by
- and sometimes

Hearing and equilibrium disorders

Fatigue, somnolence, apathy, difficulties concentrating

Typical, uncommon

Bibrachial amyotrophy, superficial siderosis, brain sagging dementia

- often without
- often

Headache

Slowly progressive, long time to diagnosis

Less typical, uncommon

Isolated orthostatic hearing and equilibrium disorders

➔ If the MRI fits it is SIH 😊

# Clinical spectrum of SIH

Enjoy the rest of the course!

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