

Clinical mimics: bvFTD and BBA: behavioral variant frontotemporal dementia and bibrachial amyotrophy

Wouter I Schievink, MD



Clinical Manifestations of Spontaneous Intracranial Hypotension

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 (%)
Most common	
Headache	199 (98.5)
Orthostatic	188 (93.1)
Nonpositional	7 (3.5)
Reverse orthostatic	4 (2.0)
Common	
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)
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)
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, ≤8‡	1 (0.5)
Syringomyelia	1 (0.5)
Hemifacial spasm	1 (0.5)

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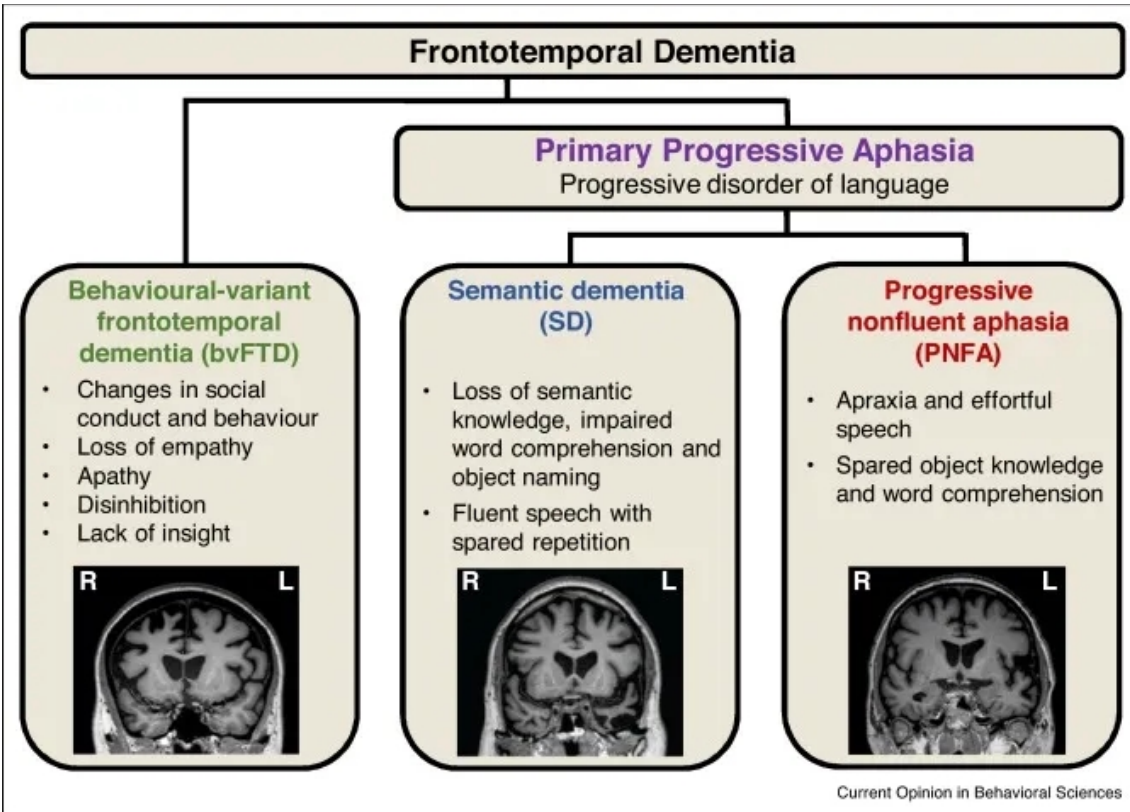
Non-headache manifestations of SIH

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Behavioral – variant frontotemporal dementia

- Progressive deterioration of personality, social comportment, and cognition
- Caused by frontotemporal lobar degeneration
- Incurable
- Second most common cause of dementia in <60 years
- Genetic basis



FTD Types

- Behavioral variant frontotemporal dementia (bvFTD)**
 - Also called frontal variant frontotemporal dementia (fvFTD) or Pick's disease
 - Negatively impacts social skills, emotions, personal conduct, and self-awareness.
 - People with bvFTD might act in inappropriate ways, show a lack of judgment or inhibition, neglect to maintain personal hygiene, do something compulsively or repetitively, or feel euphoric or apathetic.
 - bvFTD is the most common form of frontotemporal dementia.



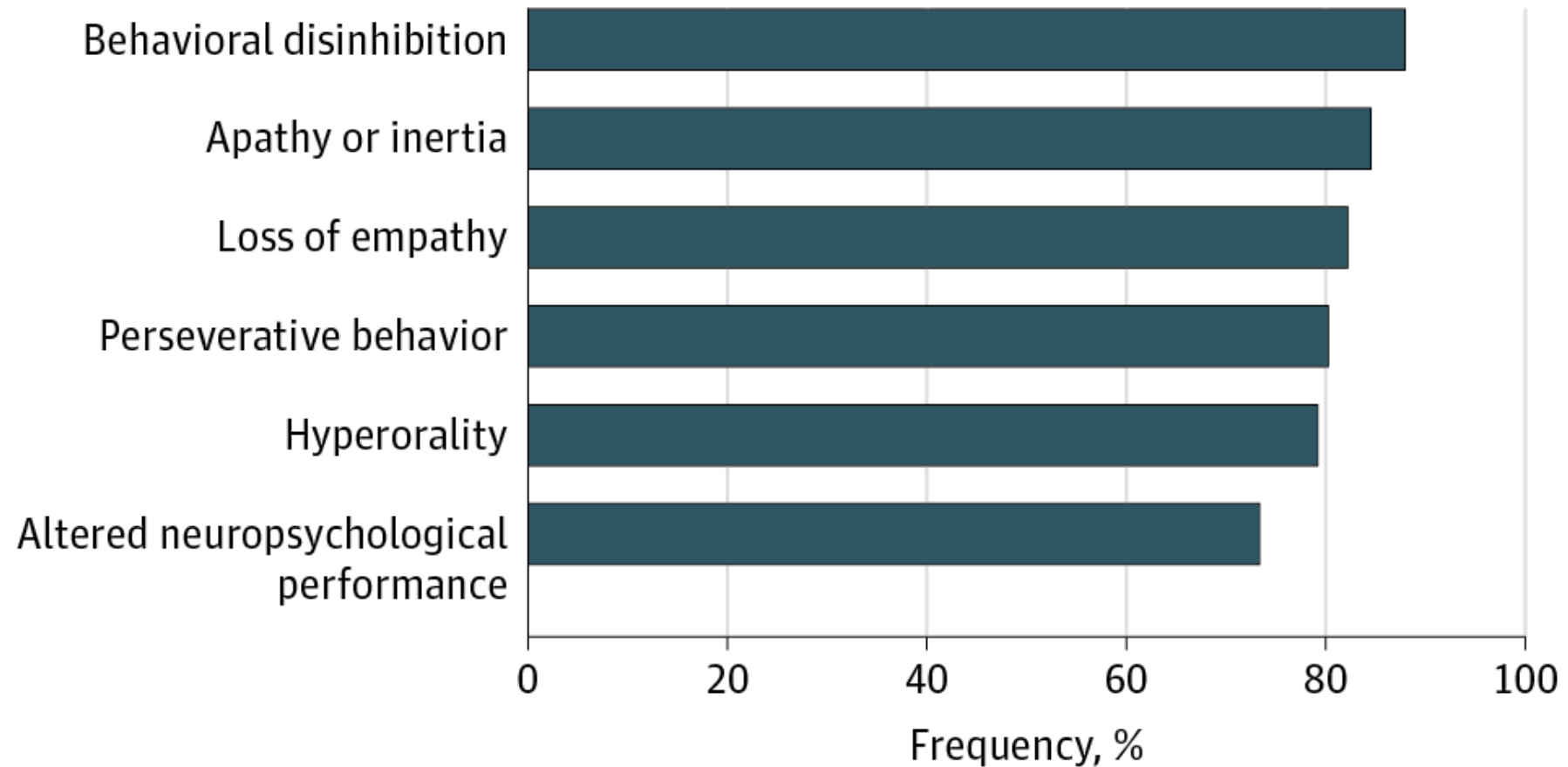
Incurable

NEUROLOGY April 23, 2002; 58 (8) **BRIEF COMMUNICATIONS**

Spontaneous intracranial hypotension causing reversible frontotemporal dementia

M. Hong, G. V. Shah, K. M. Adams, R. S. Turner, N. L. Foster

A Core diagnostic features



From: Sensitivity of revised diagnostic criteria for the behavioural variant of frontotemporal dementia

Brain. 2011;134(9):2456-2477. doi:10.1093/brain/awr179

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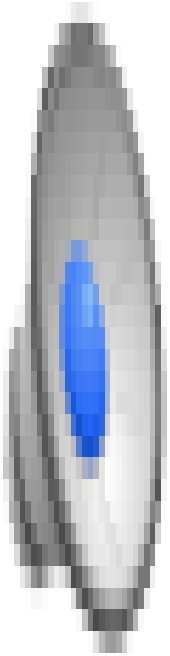
Behavioral disinhibition



Apathy or inertia



Apathy or inertia

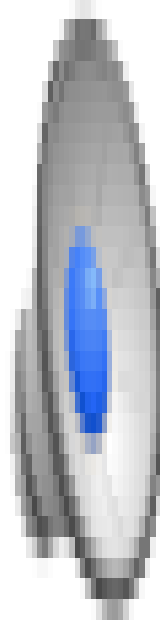


Perseverative behavior

Dr Jeremy Schmahan:
Repetitive obsessive flexion
and breath-holding in
sagging brain syndrome
(ROFBIS)



FTD sagging brain syndrome – Apathy (and its post-operative resolution)



Behavioral variant frontotemporal dementia

Behavioral Variant Frontotemporal Dementia as a Serious Complication of Spontaneous Intracranial Hypotension

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BACKGROUND: Behavioral variant frontotemporal dementia (bvFTD) is a devastating early onset dementia. Symptoms of bvFTD may be caused by spontaneous intracranial hypotension (SIH), a treatable disorder, but no comprehensive study of such patients has been reported.

OBJECTIVE: To describe detailed characteristics of a large cohort of patients with SIH and symptoms of bvFTD.

METHODS: We identified patients with SIH who met clinical criteria for bvFTD. Patients were compared to a cohort of SIH patients without bvFTD.

RESULTS: The mean age for the 21 men and 8 women was 52.9 yr (range, 37–65 yr). All 29 patients with bvFTD symptoms had hypersomnolence. Magnetic resonance imaging showed brain sagging in all patients, cerebrospinal fluid (CSF) opening pressure low in about half of patients, but a spinal CSF leak could not be detected in any patient. All patients underwent epidural blood patching, but 26 patients eventually underwent 1 or more surgical procedures. Overall, a good outcome was obtained in 21 patients (72%); 20 (91%) of 22 patients who had not undergone prior Chiari surgery compared to 1 (14%) of 7 patients who did undergo Chiari surgery ($P < .003$). Compared to SIH patients without symptoms of bvFTD ($n = 547$), those with bvFTD symptoms were older, more often male, less often demonstrated CSF leak on spinal imaging, and more often underwent surgery ($P < .02$).

CONCLUSION: bvFTD in SIH is rare and associated with brain sagging and hypersomnolence. Spinal CSF leaks are rarely detected. bvFTD symptoms are often refractory to the usual percutaneous procedures but most patients can be cured.

KEY WORDS: CSF leak, Dementia, Frontotemporal dementia, Headache, Intracranial hypotension

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RESEARCH ARTICLE

The reversible impairment of behavioral variant frontotemporal brain sagging syndrome: Challenges and opportunities

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Abstract

Introduction: Due to loss of brain buoyancy, spontaneous spinal cerebrospinal fluid (CSF) leaks cause orthostatic headaches but also can cause symptoms indistinguishable from behavioral variant frontotemporal dementia (bvFTD) due to severe brain sagging (including the frontal and temporal lobes), as visualized on brain magnetic resonance imaging. However, the detection of these CSF leaks may require specialized spinal imaging techniques, such as digital subtraction myelography (DSM).

Methods: We performed DSM in the lateral decubitus position under general anesthesia in 21 consecutive patients with frontotemporal dementia brain sagging syndrome (4 women and 17 men; mean age 56.2 years [range: 31–70 years]).

Results: Nine patients (42.8%) were found to have a CSF-venous fistula, a recently discovered type of CSF leak that cannot be detected on conventional spinal imaging. All nine patients underwent uneventful surgical ligation of the fistula. Complete or near-complete and sustained resolution of bvFTD symptoms was obtained by all nine patients, accompanied by reversal of brain sagging, but in only three (25.0%) of the twelve patients in whom no CSF-venous fistula could be detected ($P = 0.0011$), and who were treated with non-targeted therapies.

Discussion: Concerns about a spinal CSF leak should not be dismissed in patients with frontotemporal brain sagging syndrome, even when conventional spinal imaging is normal. However, even with this specialized imaging the source of the loss of spinal CSF remains elusive in more than half of patients.

KEYWORDS

behavioral variant frontotemporal dementia, brain sagging, cerebrospinal fluid leak, frontotemporal dementia, spontaneous intracranial hypotension

The New York Times Magazine

DIAGNOSIS

By Lisa Sanders, M.d.

Nov. 2, 2017

“So, do you like working here?” the middle-aged man bellowed to the young physician at the other end of the hospital coffee shop. The woman, the object of this not-very-subtle pickup line, ignored him. The man’s sister cringed. When had her younger brother, who was 49, turned into such a jerk? He had always been so quiet and shy. She was living across the country in Washington State, so she didn’t see him often, but he had certainly changed.

In his 20s, he had a problem with alcohol. But back then, his drinking made him quieter. And even during the worst of his drinking days, he had always been tidy and well kempt, fastidious in everything he did. That morning, she drove from the airport to pick him up on the way to visit their father, who was in the hospital after heart surgery. She had taken a red-eye flight from Seattle to Philadelphia, but her brother looked worse than she felt: tired, disheveled, dirty. He said he had just showered, but she could tell it wasn’t true.



bvFTD sagging brain syndrome (2005-2022)

51 patients (12 women and 39 men)

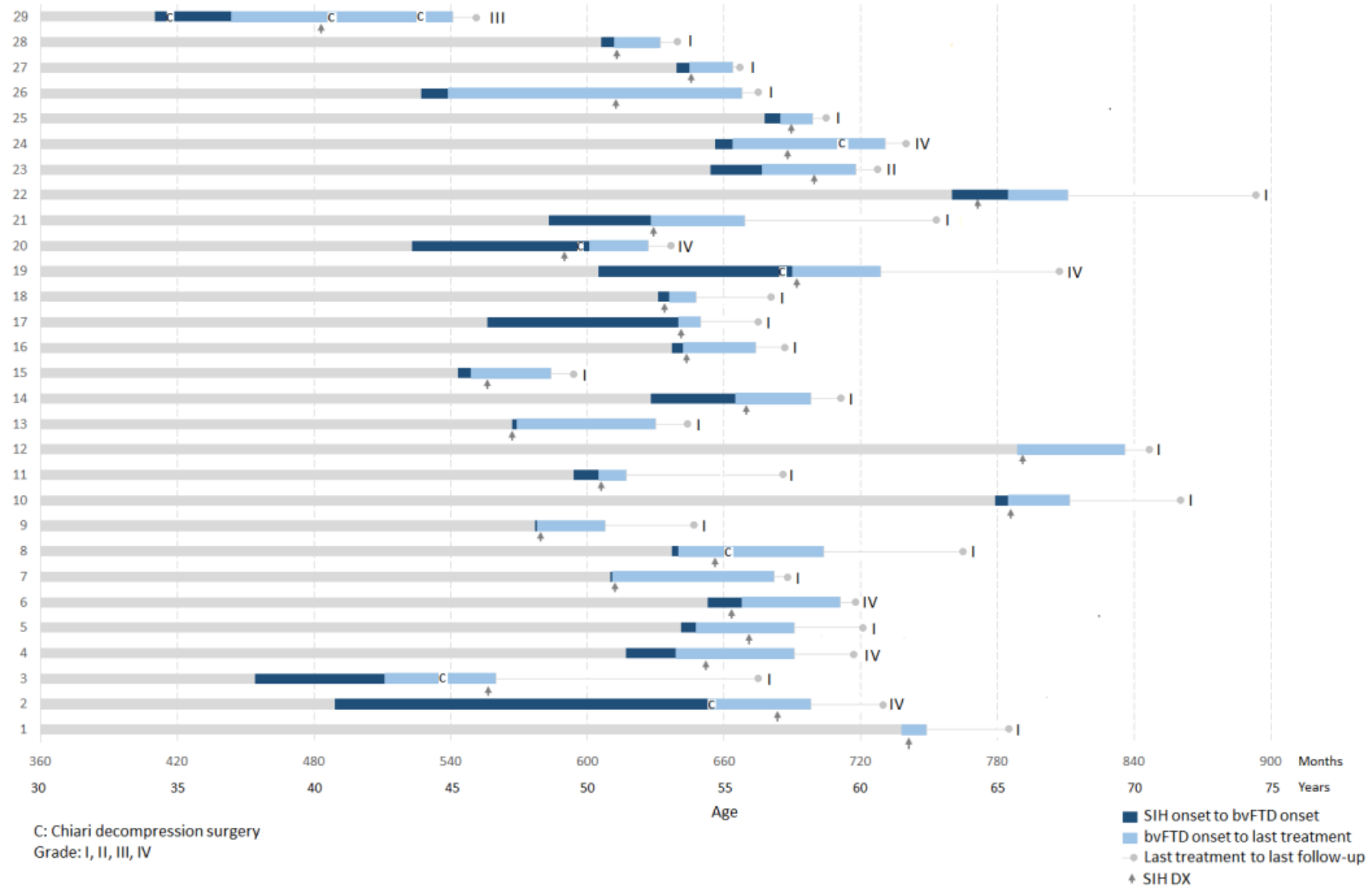
Mean age: 55.5 years (range, 26-70 years)

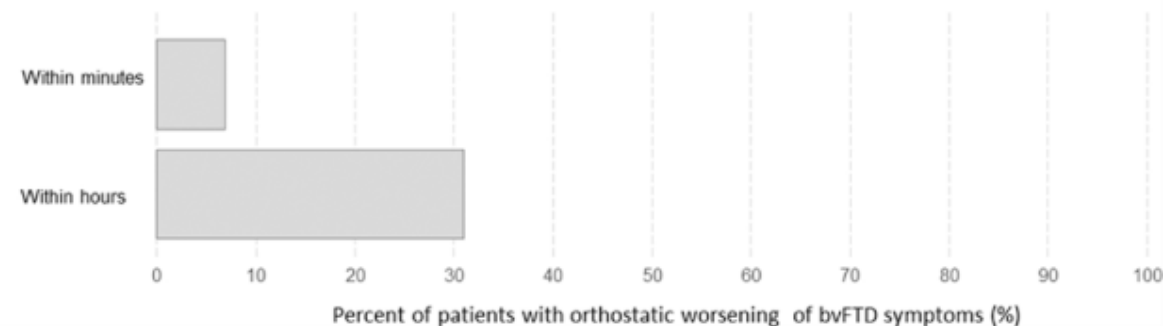
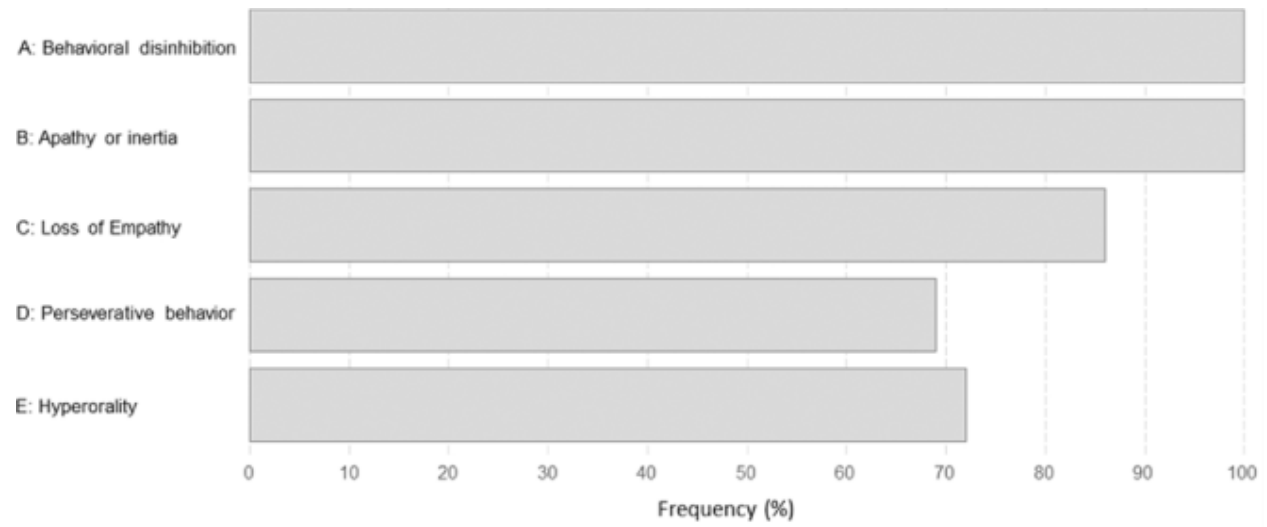
CSF-venous fistula: 13 patients

Ventral spinal CSF leak: 1 patient

Nil detected: 37 patients

Frontotemporal dementia in spontaneous intracranial hypotension





From: Behavioral Variant Frontotemporal Dementia as a Serious Complication of Spontaneous Intracranial Hypotension

Oper Neurosurg (Hagerstown). Published online March 08, 2018. doi:10.1093/ons/opy029

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Behavioral variant FTD sagging brain syndrome: Associated symptoms

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%)
Hiccapping	3	(10%)
Abducens nerve palsy	2	(7%)
Coma	1	(3%)
Trigeminal neuralgia	1	(3%)

Sagging brain syndrome – Slowed speech with altered pitch, tremors (and its post-operative resolution)



Behavioral variant frontotemporal dementia in SIH –

Dystonia and
disinhibition
(and its post-
operative
resolution)

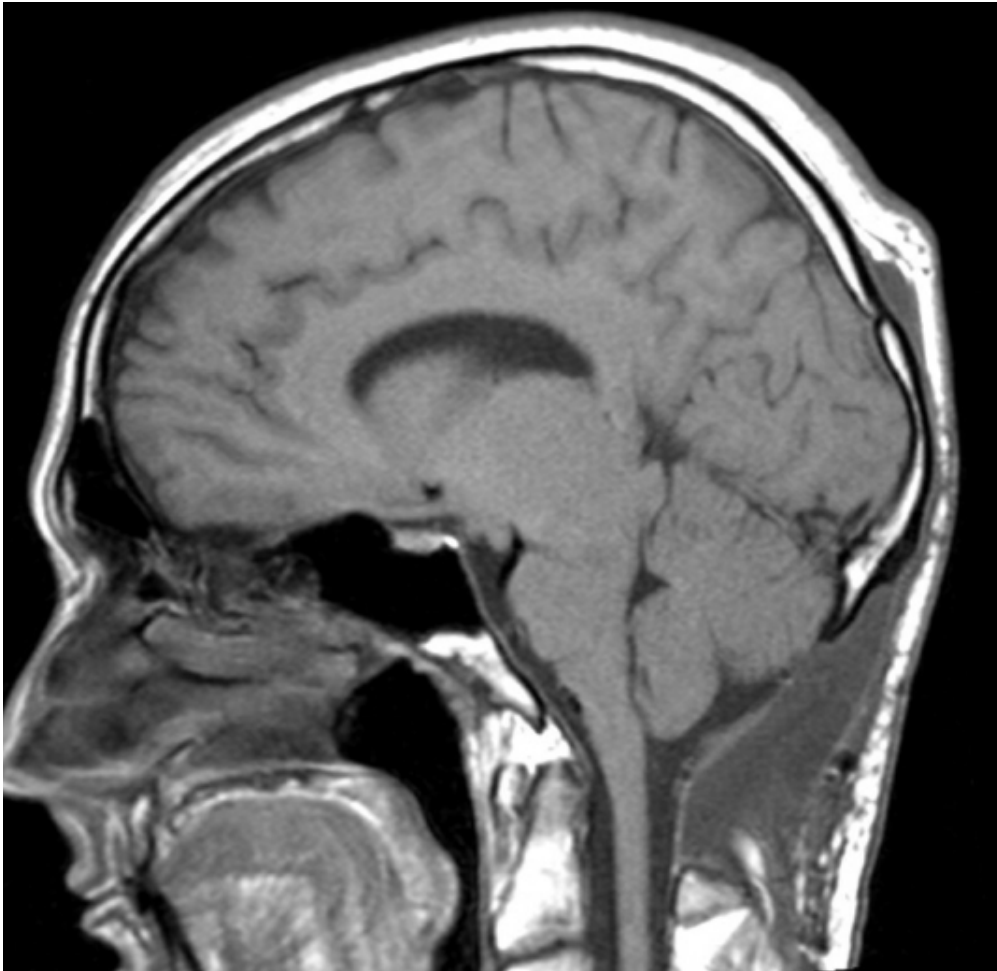


Common barriers to treatment of FTD sagging brain syndrome

- Misinterpretation of brain MRI
- Difficulty in finding source of spinal CSF loss

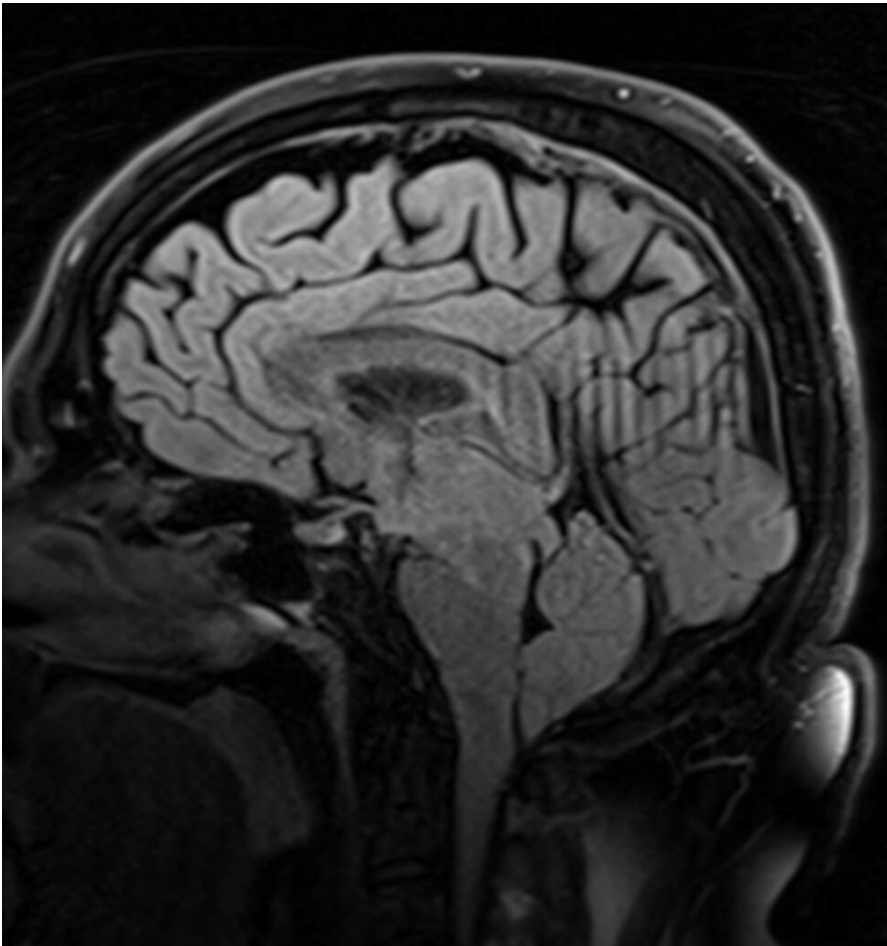
Common barriers to treatment of FTD sagging brain syndrome

- Misinterpretation of brain MRI



Diagnosis: congenital mid brain malformation

Treatment: V-P shunt



Diagnosis: mid brain glioma

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Bibrachial amyotrophy

- Progressive painless weakness and atrophy of shoulder and upper extremity musculature
- Bilateral but may be asymmetric
- Fasciculations
- DDX: Amyotrophic lateral sclerosis, Hirayama disease, man/woman-in-the-barrel syndrome
- Associated with large extradural CSF collection

Bibrachial amyotrophy



Bibrachial amyotrophy



Bibrachial amyotrophy due to SIH (2005-2022)

20 patients (5 women and 15 men)

5 patients without orthostatic headache history

Mean age at SIH onset: 26 years (range, 11-41 years)

5 patients <19 years

Mean interval SIH to BBA: 9 years (range, 1-39 years)

Long-term Risks of Persistent Ventral Spinal CSF Leaks in SIH

Superficial Siderosis and Bibrachial Amyotrophy

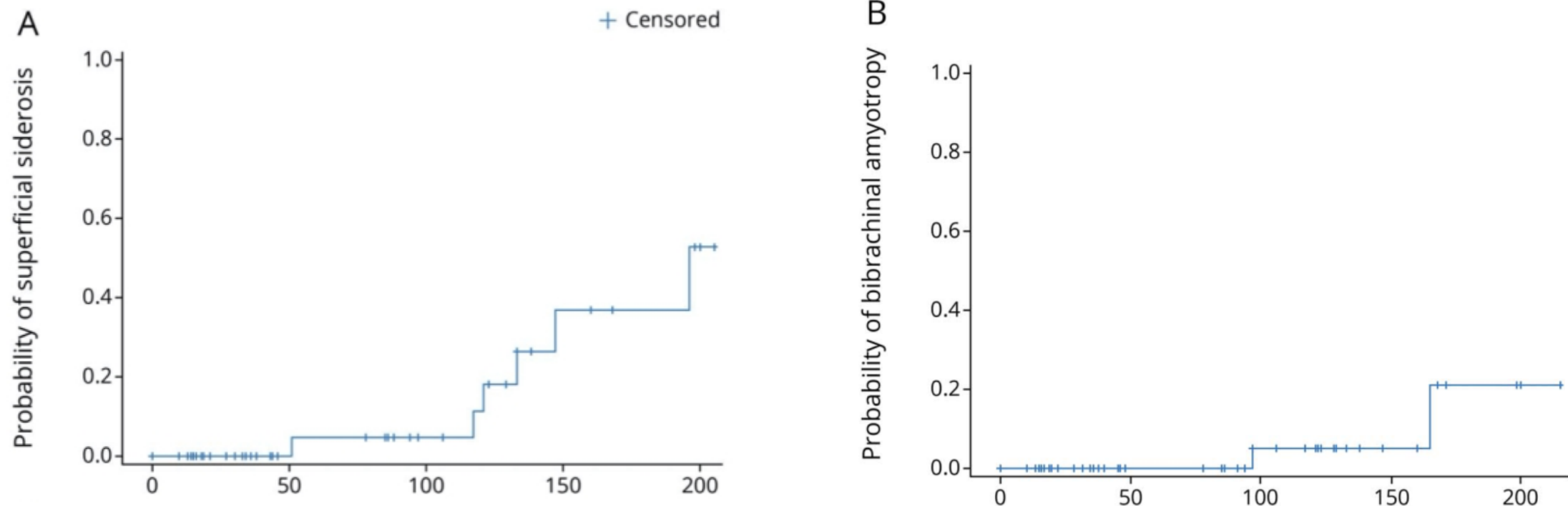
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Neurology® 2021;97:e1964-e1970. doi:10.1212/WNL.00000000000012786

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Figure 1 Risk of Serious Long-Term Complications of Persistent Ventral Spinal CSF Leaks



Common barriers to treatment of bibrachial amyotrophy

- Misinterpretation of spine MRI
- Lack of understanding of relationship of extradural fluid collection and symptoms
- Difficulty localizing the exact leak site

Common barriers to treatment of bibrachial amyotrophy

- Misinterpretation of spine MRI



“No leak”



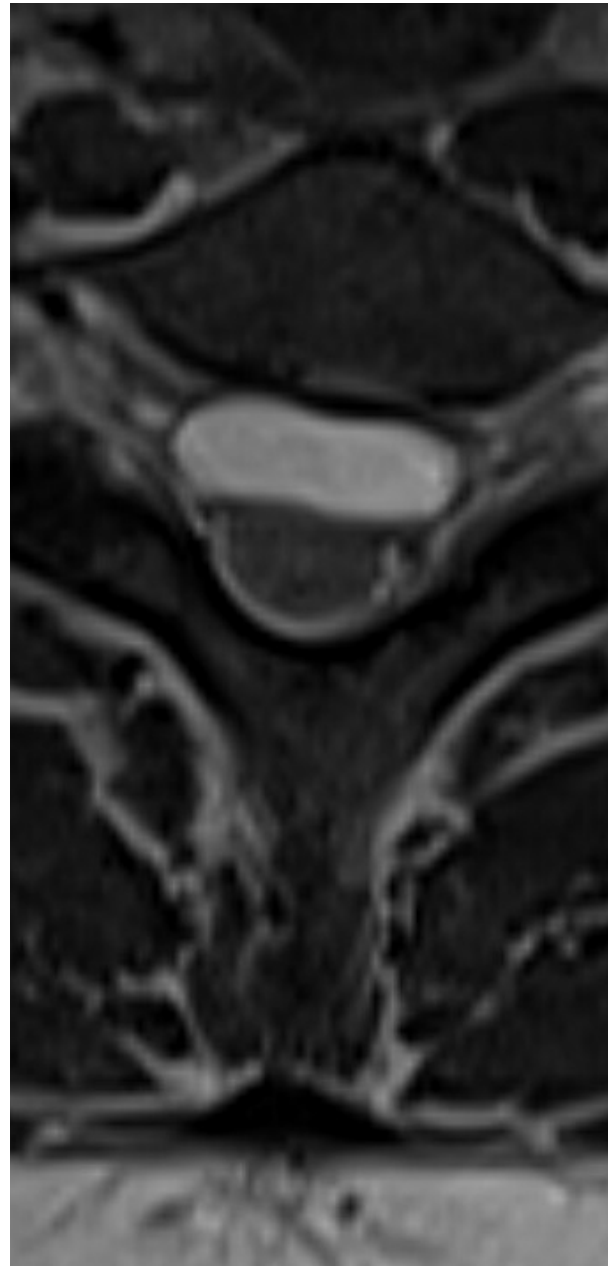
“Degenerative disc disease”



“Epidural hematoma”

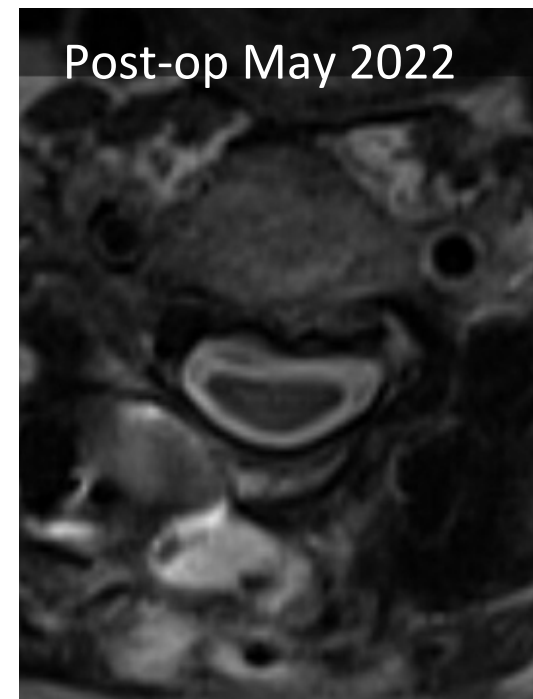
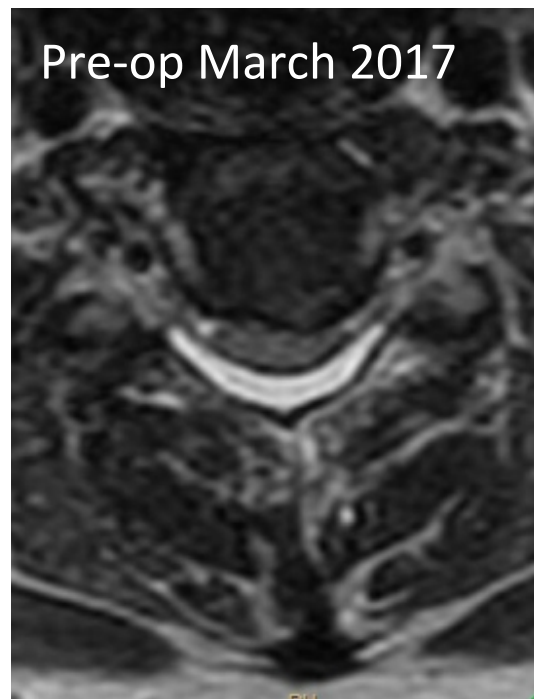
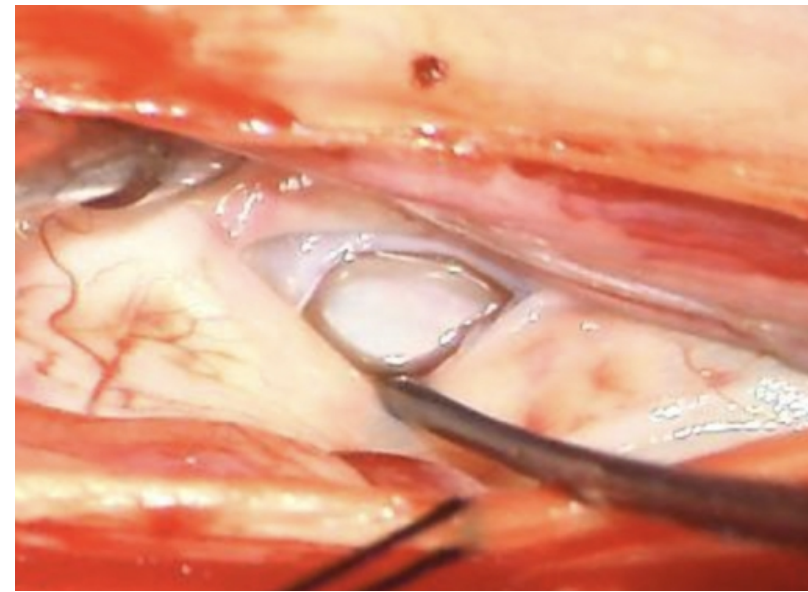
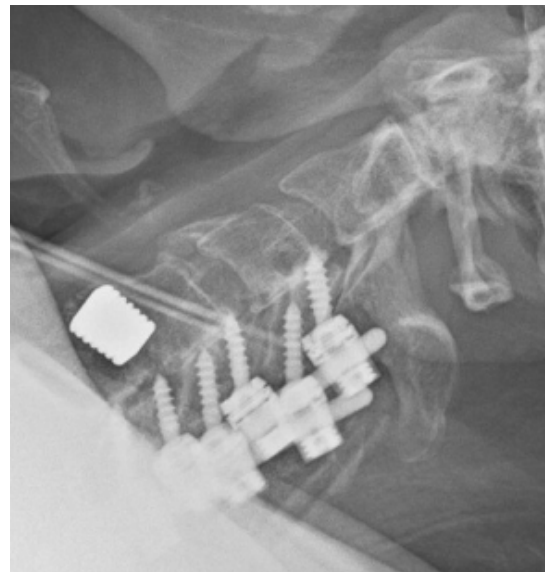
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Common barriers to treatment of bibrachial amyotrophy

- Inability to localize exact leak site



Behavioral – variant frontotemporal dementia

Bibrachial amyotrophy

Both: Insidious onset – headache not prominent – often misdiagnosed

bvFTD: extradural CSF collection rare

BBA: extradural CSF collection universally present

Thank you!



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