Fourth Annual Cedars-Sinai Intracranial Hypotension Symposium

October 2, 2021

Anatomy and pathogenesis of spinal CSF leaks

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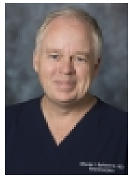
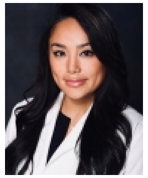
No disclosures

CSF leak program, Cedars-Sinai Medical Center, Los Angeles, CA, USA

- Neurosurgery
  - Wouter I. Schievink, M.D.
  - Rachelle B. Tache, M.S.N., A.P.R.N., N.P.-C.

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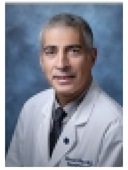

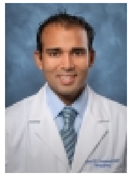
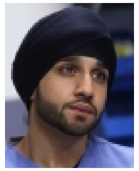

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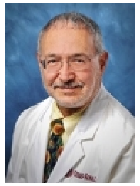

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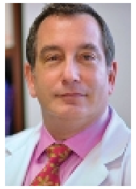


Classification of CSF leaks as a cause of SIH

Classification of CSF leaks as a cause of SIH

- **Spinal**

Lack of causal association between spontaneous intracranial hypotension and cranial cerebrospinal fluid leaks

Clinical article

Wouter I. Schievink, M.D.,[1] Marc S. Schwartz, M.D.,[1,2] M. Marcel Maya, M.D.,[3] Franklin G. Moser, M.D., M.M.M.,[3] and Todd D. Rozen, M.D.[4]

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- CSF rhinorrhea–otorrhea ?

NEVER!

Classification of CSF leaks as a cause of SIH

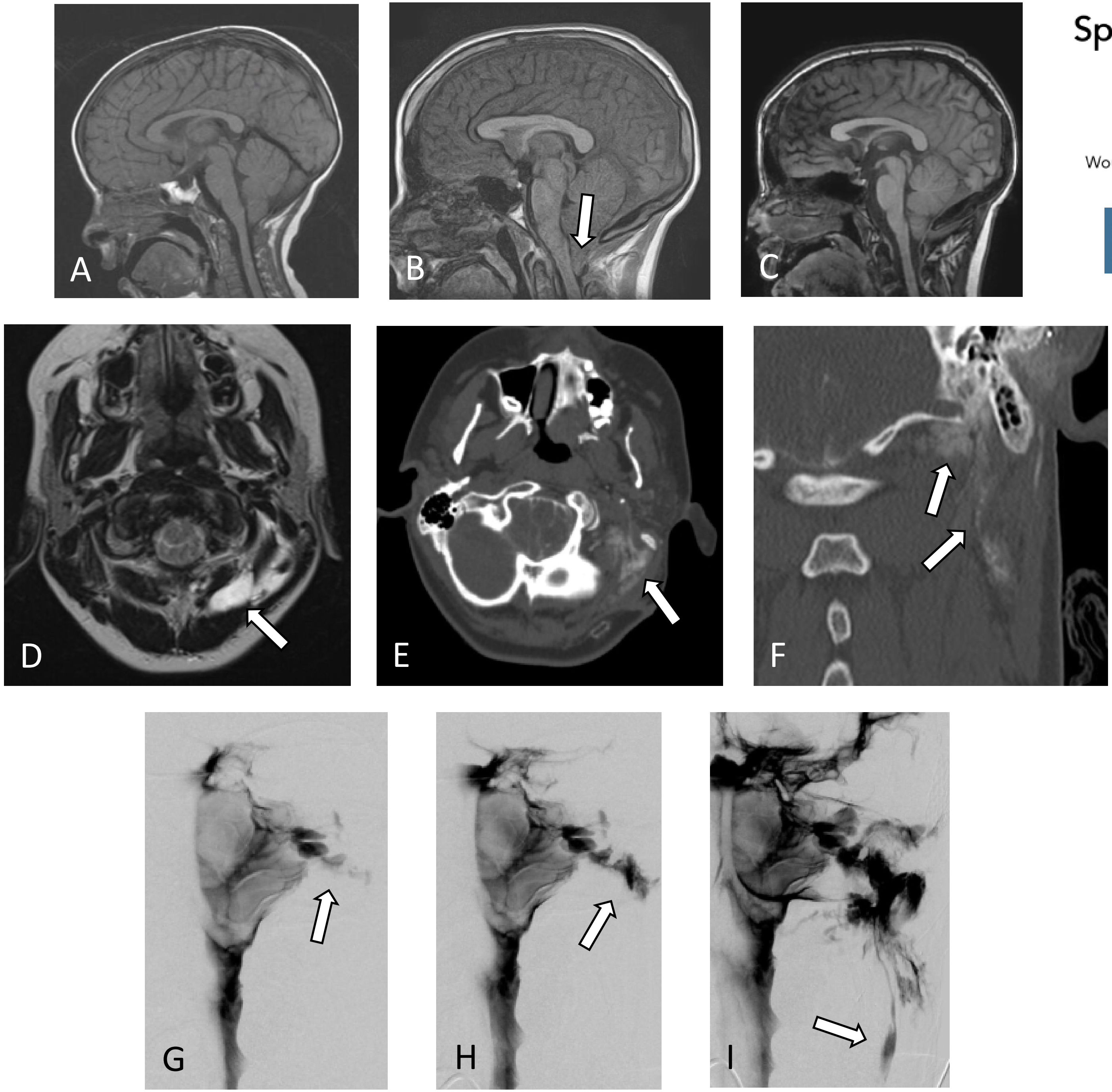
- Spinal

Spontaneous Intracranial Hypotension
Due to Skull-Base Cerebrospinal
Fluid Leak

Spontaneous Intracranial Hypotension Due to Skull-Base Cerebrospinal Fluid Leak

Wouter I. Schievink, MD,[1] L. Madison Michael II MD,[2,3,4] Marcel Maya, MD,[5] Paul Klimo Jr MD,[2,3,4] and Lucas Elijovich, MD[2,3,4,6]

Annals of NEUROLOGY
An Official Journal of the American Neurological Association and the Child Neurology Society



13-year-old boy

Classification of spinal CSF leaks as a cause of SIH

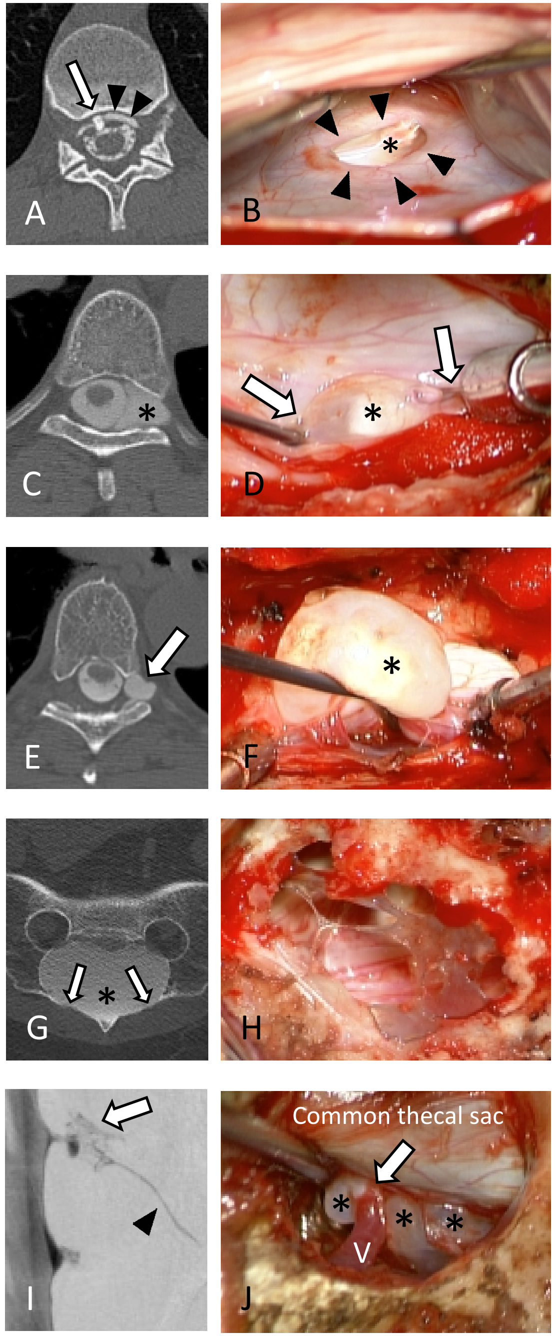
Ventral CSF leak

Lateral CSF leak

Meningeal diverticulum

Dural ectasia

CSF-venous fistula



A classification system of spontaneous spinal CSF leaks

Wouter I. Schievink, MD
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Stacey Jean-Pierre, PA-C
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ABSTRACT

Objective: Spontaneous spinal CSF leaks cause spontaneous intracranial hypotension but no systematic study of the different types of these CSF leaks has been reported. Based on our experience with spontaneous intracranial hypotension, we propose a classification system of spontaneous spinal CSF leaks.

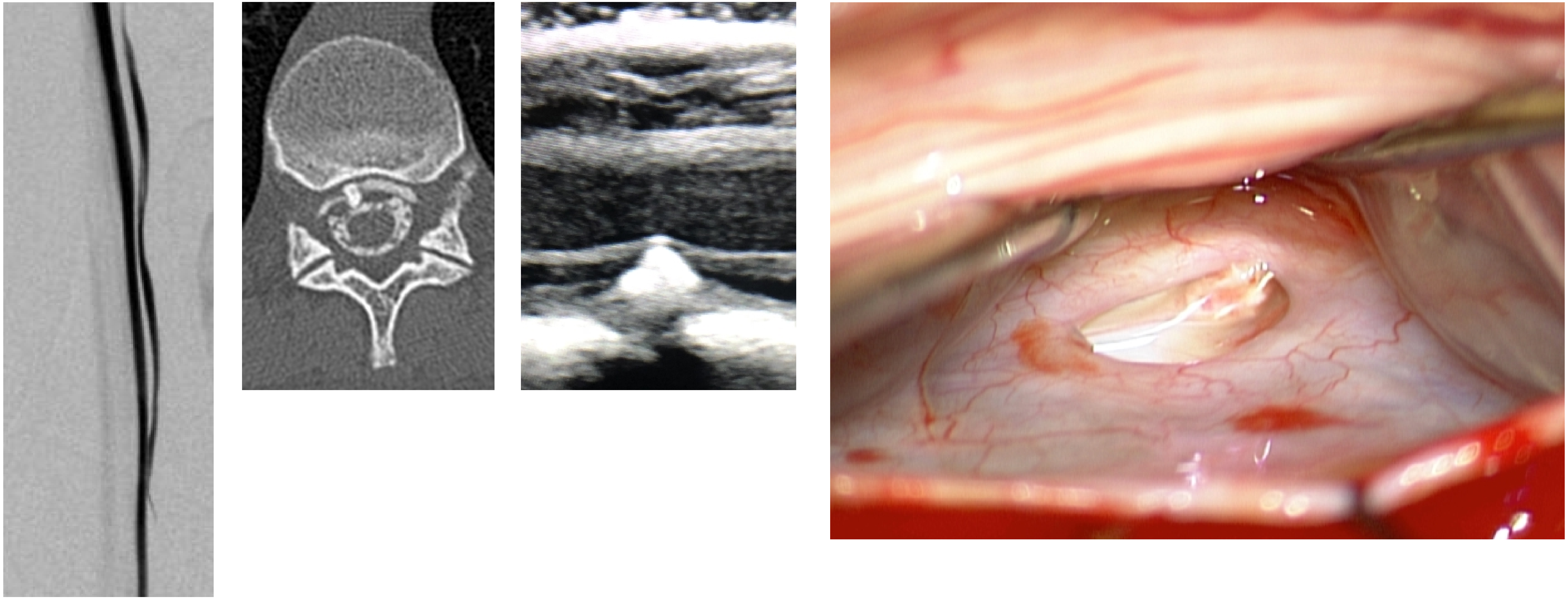
Methods: We reviewed the medical records, radiographic studies, operative notes, and any intraoperative photographs of a group of consecutive patients with spontaneous intracranial hypotension.

Results: The mean age of the 568 patients (373 [65.7%] women) was 45.7 years. Three types of CSF leak could be identified. Type 1 CSF leaks consisted of a dural tear (151 patients [26.6%]) and these were almost exclusively associated with an extradural CSF collection. Type 1a represented ventral CSF leaks (96%) and type 1b posterolateral CSF leaks (4%). Type 2 CSF leaks consisted of meningeal diverticula (240 patients [42.3%]) and were the source of an extradural CSF collection in 53 of these patients (22.1%). Type 2a represented simple diverticula (90.8%) and type 2b complex meningeal diverticula/dural ectasia (9.2%). Type 3 CSF leaks consisted of direct CSF-venous fistulas (14 patients [2.5%]) and these were not associated with extradural CSF collections. A total of 163 patients (28.7%) had an indeterminate type and extradural CSF collections were noted in 84 (51.5%) of these patients.

Conclusions: We identified 3 types of spontaneous spinal CSF leak in this observational study: the dural tear, the meningeal diverticulum, and the CSF-venous fistula. These 3 types and the presence or absence of extradural CSF form the basis of a comprehensive classification system.
Neurology® 2016;87:673-679

Classification of spinal CSF leaks as a cause of SIH

- Type 1: Dural tear
  - 1a: ventral
  - 1b: (postero-)lateral
- Type 2: Meningeal diverticulum
  - 2a: simple
  - 2b: complex/dural ectasia
- Type 3: CSF-venous fistula
- Type 4: Indeterminate



Type 1a – Ventral leak

Type 1a ventral leak

- Microspur/osteophyte in >95% of cases
- Dural tear 1-22 mm in length, >90% 3-6 mm
- Thoracic >90% of cases
- Requires surgery if present for more than 3-6 months
- Significant risk of superficial siderosis if left untreated

Type 1a ventral leak

- Microspur/osteophyte in >95% of cases
- Dural tear 1-22 mm in length, >90% 3-6 mm
- Thoracic >90% of cases
- Requires surgery if present for more than 3-6 months
- Significant risk of superficial siderosis if left untreated

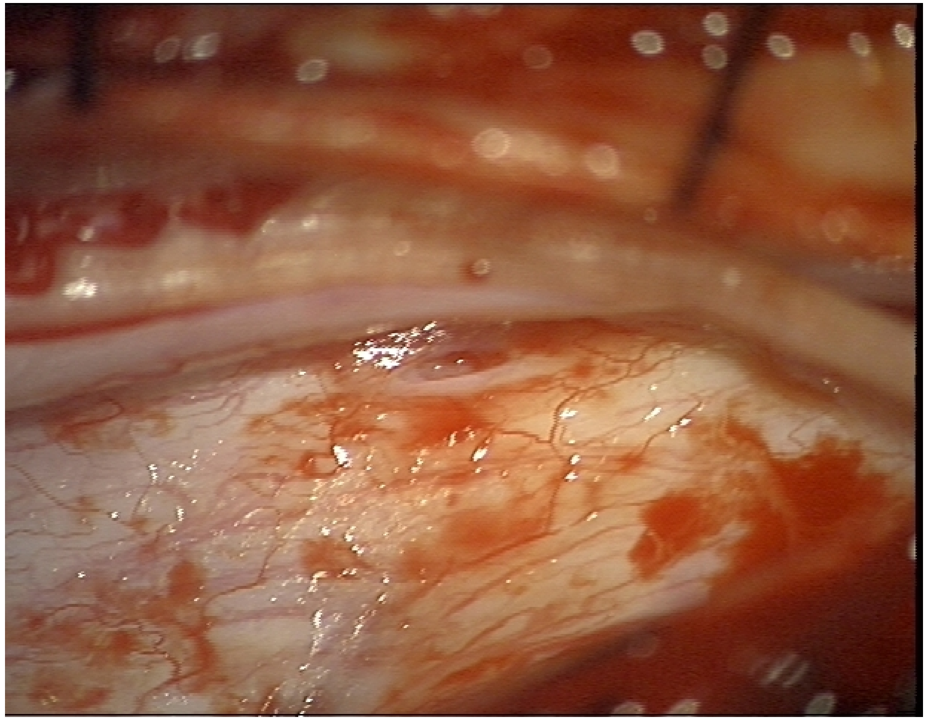
Size of underlying osseous abnormality in ventral spinal CSF leaks

variable

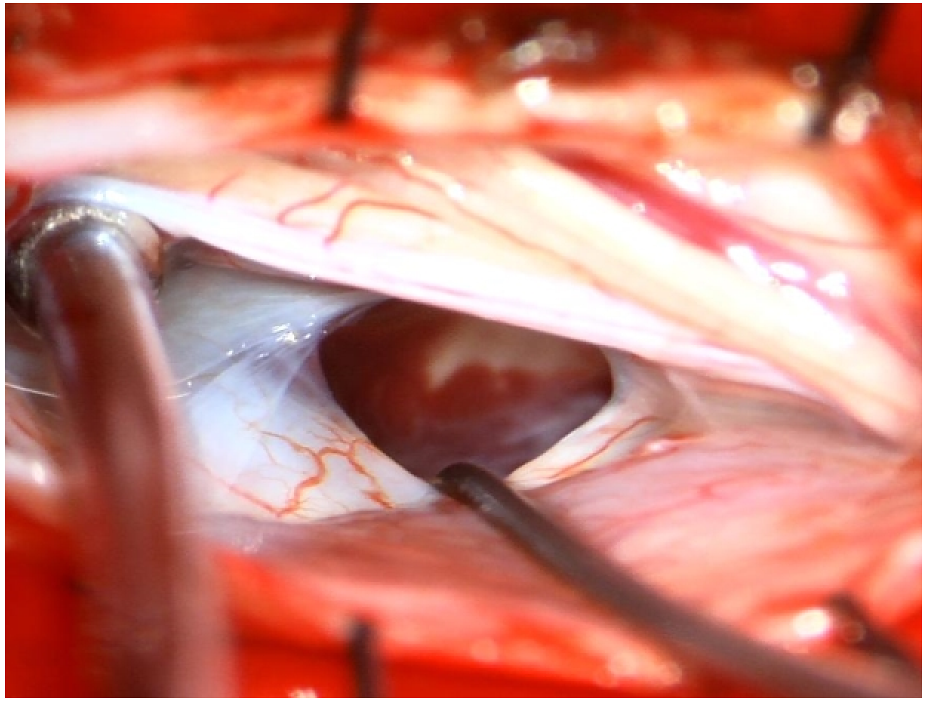
Type 1a ventral leak

- Microspur/osteophyte in >95% of cases
- Dural tear 1-22 mm in length, >90% 3-6 mm
- Thoracic >90% of cases
- Requires surgery if present for more than 3-6 months
- Significant risk of superficial siderosis if left untreated

Size of ventral dural tears

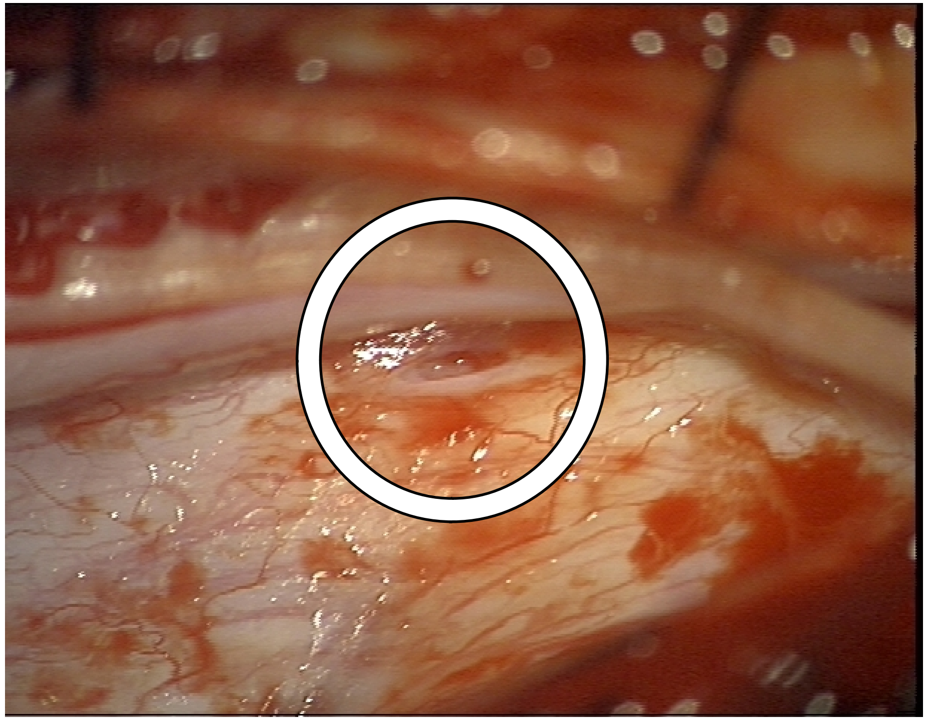


2 mm

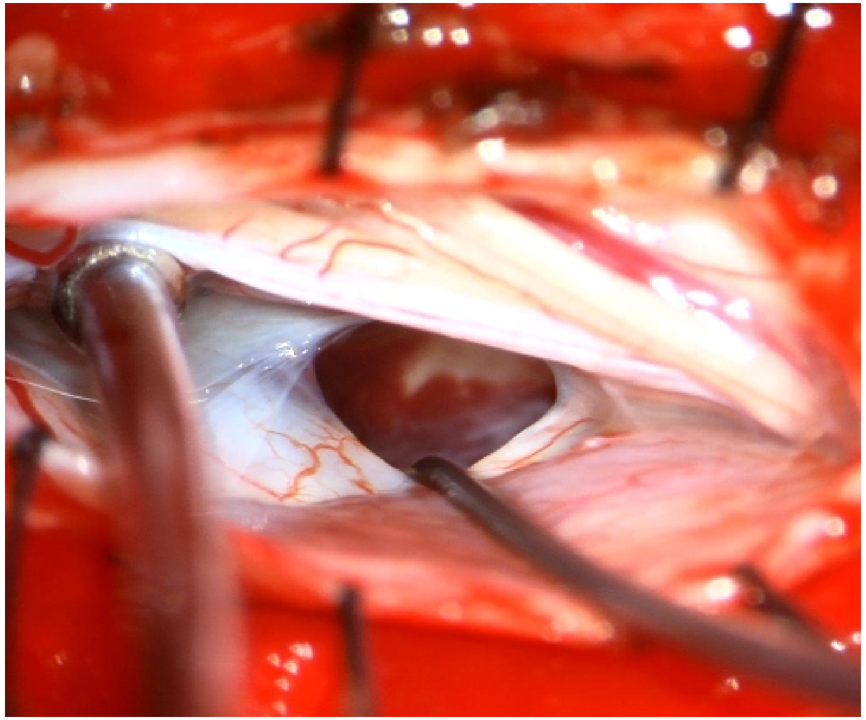


7 mm

Size of ventral dural tears



2 mm



7 mm

Type 1a ventral leak

- Microspur/osteophyte in >95% of cases
- Dural tear 1-22 mm in length, >90% 3-6 mm
- Thoracic >90% of cases
- Requires surgery if present for more than 3-6 months
- Significant risk of superficial siderosis if left untreated

Infratentorial Superficial Siderosis: Classification, Diagnostic Criteria, and Rational Investigation Pathway

Duncan Wilson, MD,[1] Fiona Chatterjee, MD,[2] Simon F. Farmer, PhD,[3] Peter Rudge, FRCP,[3] Mark O. McCarron, MD,[4] Peter Cowley, FRCR, MD,[2] and David J. Werring, FRCP, PhD[1]

Central nervous system infratentorial superficial siderosis (iSS) is increasingly detected by blood-sensitive magnetic resonance imaging (MRI) sequences. Despite this, there are no standardized diagnostic criteria, and the clinical–radiological spectrum, causes, and optimum investigation strategy are not established. We reviewed clinical and radiological details of patients with iSS assessed at a specialist neurological center during 2004–2016 using predefined standardized radiological criteria. All imaging findings were rated blinded to clinical details. We identified 65 patients with iSS, whom we classified into 2 groups: type 1 (classical) and type 2 (secondary) iSS. Type 1 (classical) iSS included 48 patients without any potentially causal radiologically confirmed single spontaneous or traumatic intracranial hemorrhage, of whom 39 (83%) had hearing loss, ataxia, or myelopathy; type 2 (secondary) iSS included 17 patients with a potentially causal radiologically confirmed spontaneous or traumatic intracranial hemorrhage, of whom none had hearing loss, ataxia, or myelopathy. Of the patients with type 1 (classical) iSS, 40 (83%) had a potentially causal cranial or spinal dural abnormality, 5 (11%) had an alternative cause, and 3 (6%) had no cause identified. Intra-arterial digital subtraction angiography did not identify any underlying causal lesions for type 1 iSS. Type 1 (classical) iSS, defined using simple radiological criteria, is associated with a characteristic neurological syndrome. Rational investigation, including spinal MRI, nearly always reveals a potential cause, most often a dural abnormality. Catheter angiography appears to be unhelpful, suggesting that classical iSS is not associated with macrovascular arterial pathology. Recognition of type 1 (classical) iSS should allow timely diagnosis and early consideration of treatment.

ANN NEUROL 2017;81:333–343

Superficial Siderosis: A Clinical Review

Neeraj Kumar, MD

Superficial siderosis of the central nervous system results from subpial hemosiderin deposition due to chronic low-grade bleeding into the subarachnoid space. The confluent and marginal subpial hemosiderin is best appreciated on iron-sensitive magnetic resonance imaging sequences. With widespread use of magnetic resonance imaging, the disorder is increasingly being recognized, including in asymptomatic individuals. Gait ataxia, often with hearing impairment is a common clinical presentation. A clinical history of subarachnoid hemorrhage is generally not present. A macrovascular pathology is generally not causative. The most common etiology is dural disease, often dural tears. Prior or less commonly ongoing symptoms of craniospinal hypovolemia may be present. Common etiologies for dural tears include disc disease and trauma, including surgical trauma. Patients with dural tears due to herniated and calcified discs often have a ventral intraspinal fluid collection due to cerebrospinal fluid leak. A precise identification of the dural tear relies on multimodality imaging. It has been speculated that chronic bleeding from fragile blood vessels around the dural tear may be the likely underlying mechanism. Surgical correction of the bleeding source is a logical therapeutic strategy. Clinical outcomes are variable, although neuroimaging evidence of successful dural tear repair is noted. The currently available data regarding use of deferiprone in patients with superficial siderosis is insufficient to recommend its routine use in patients.

ANN NEUROL 2021;89:1068–1079

RESEARCH ARTICLE

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

Superficial Siderosis and Bibrachial Amyotrophy

Wouter I. Schievink, MD, Marcel Maya, MD, Franklin Moser, MD, MMM, and Miriam Nuño, PhD

Neurology® 2021;97:1-7. doi:10.1212/WNL.0000000000012786

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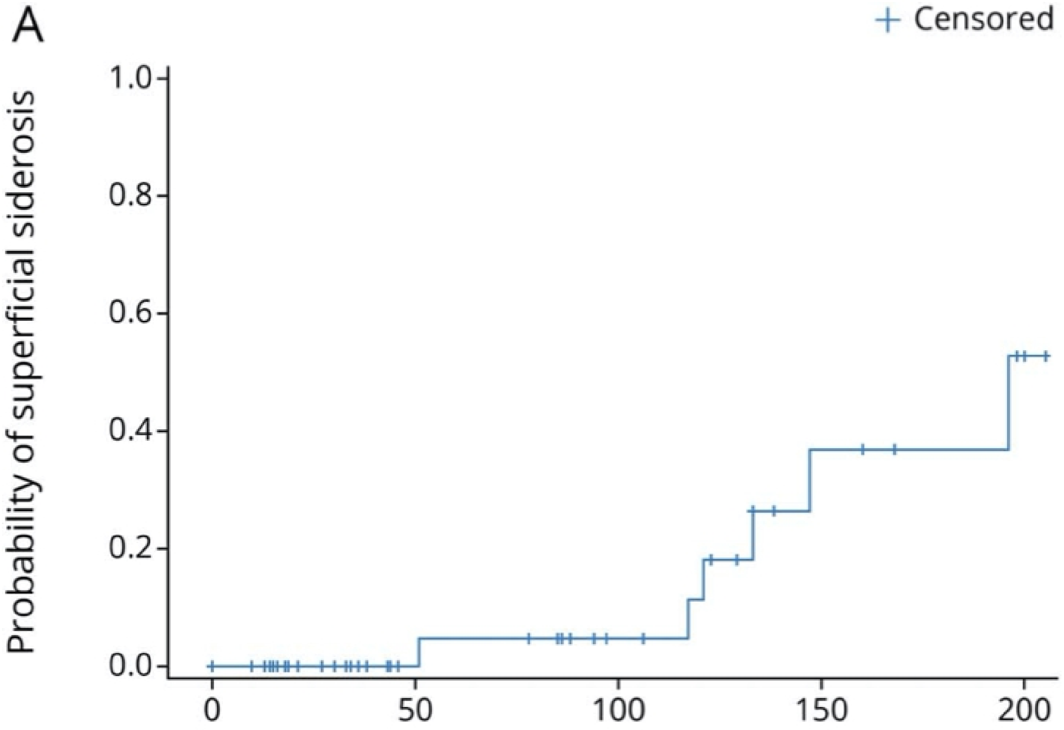
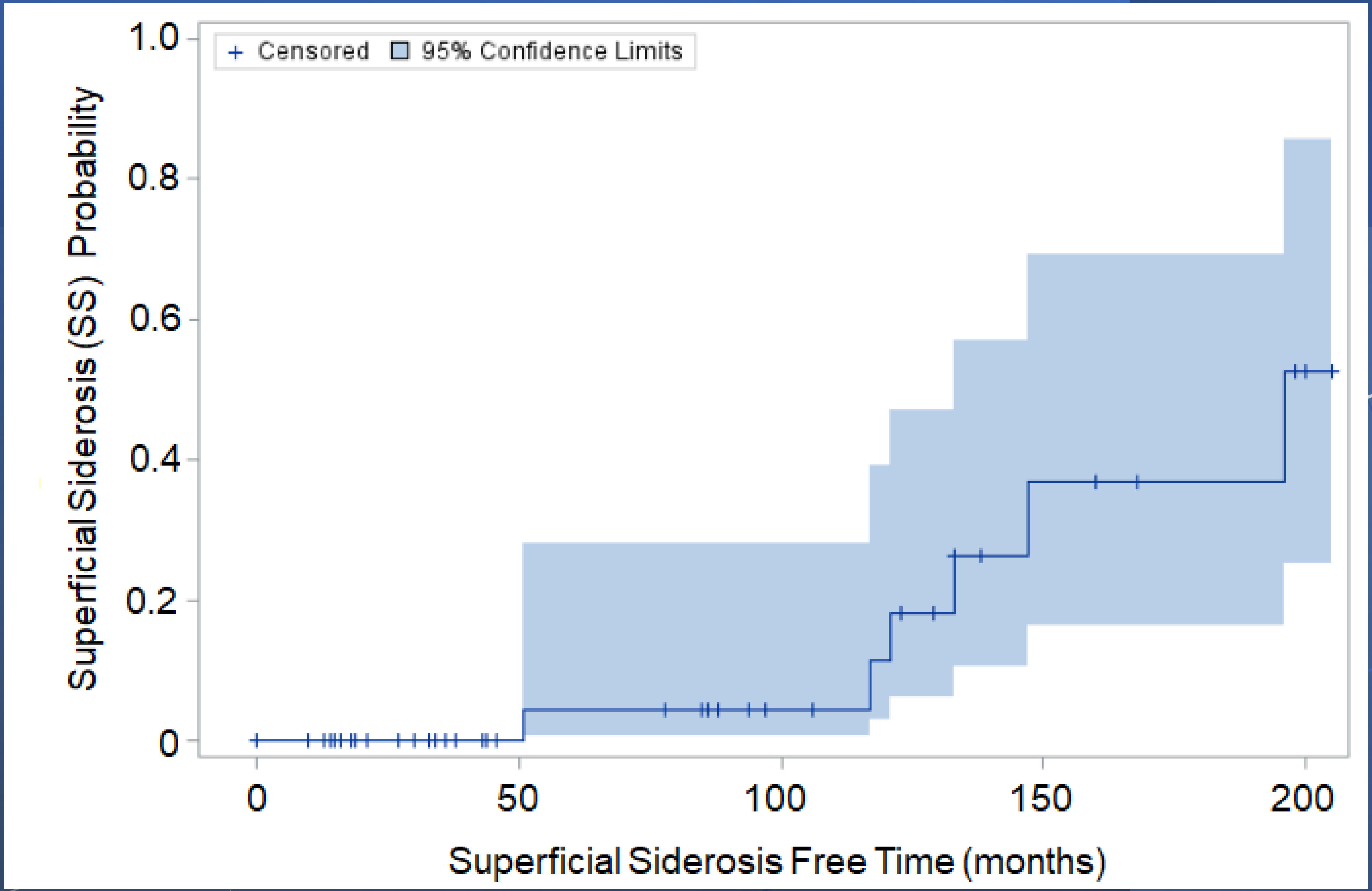
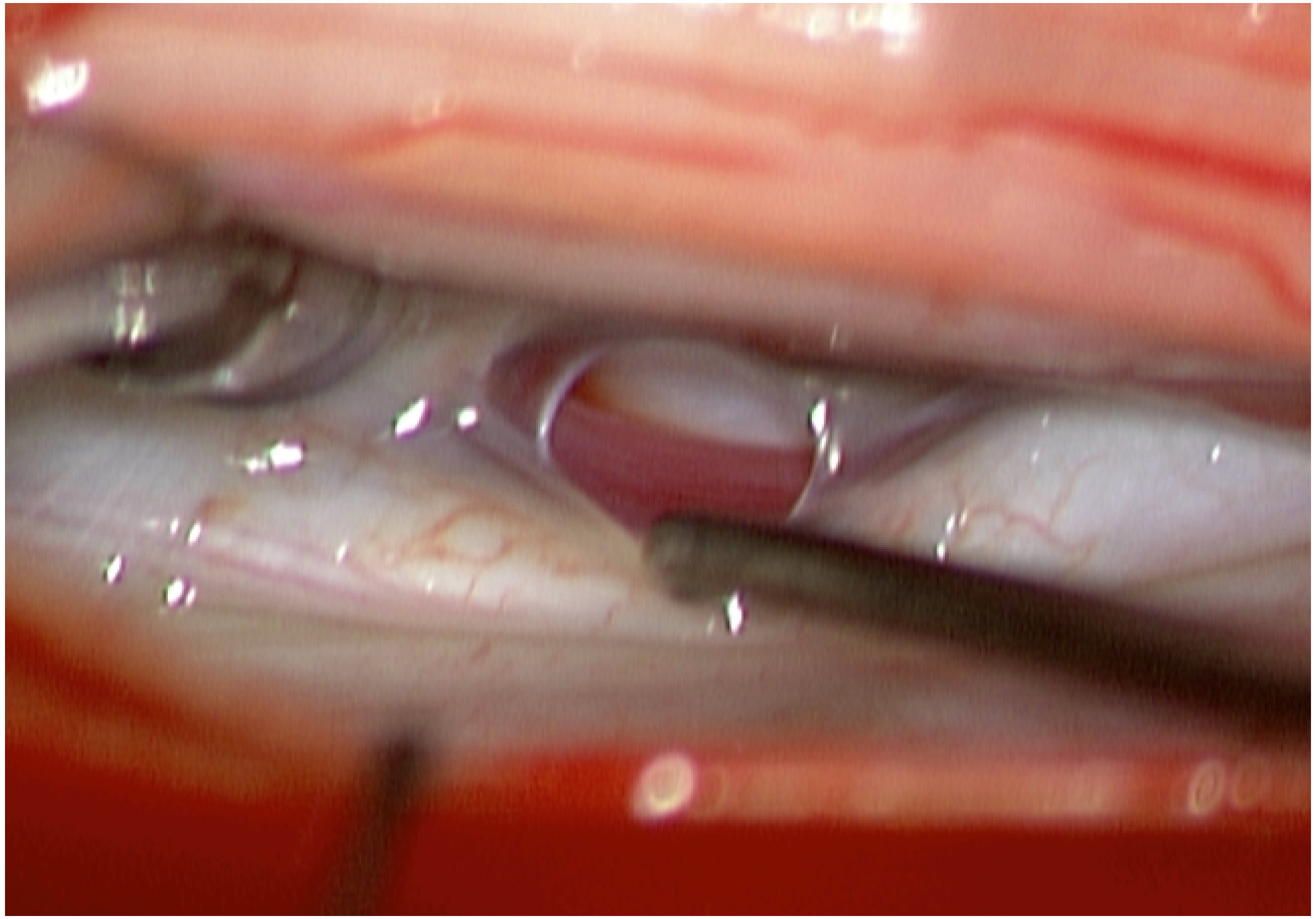
Figure 1 Risk of Serious Long-Term Complications of Persistent Ventral Spinal CSF Leaks

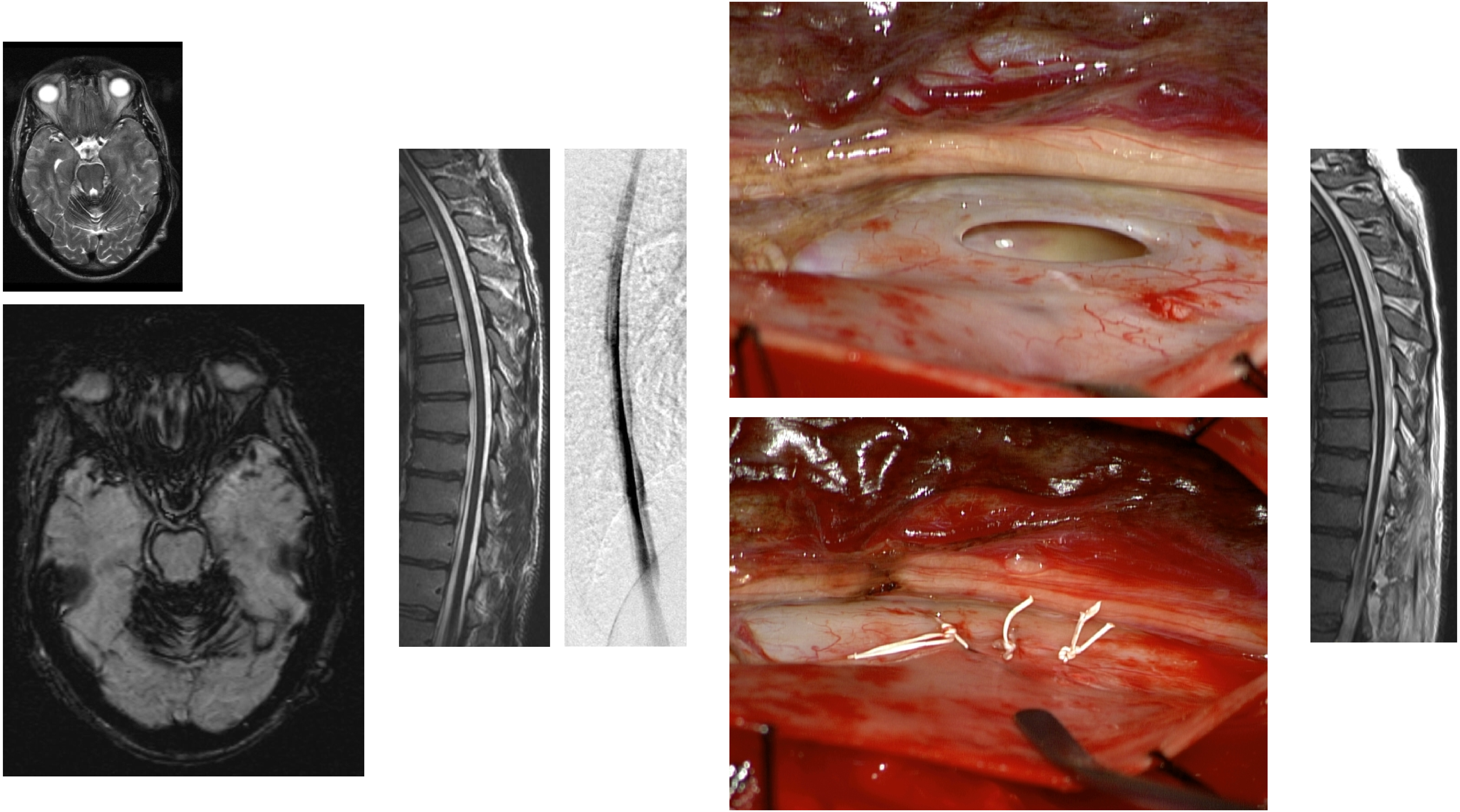


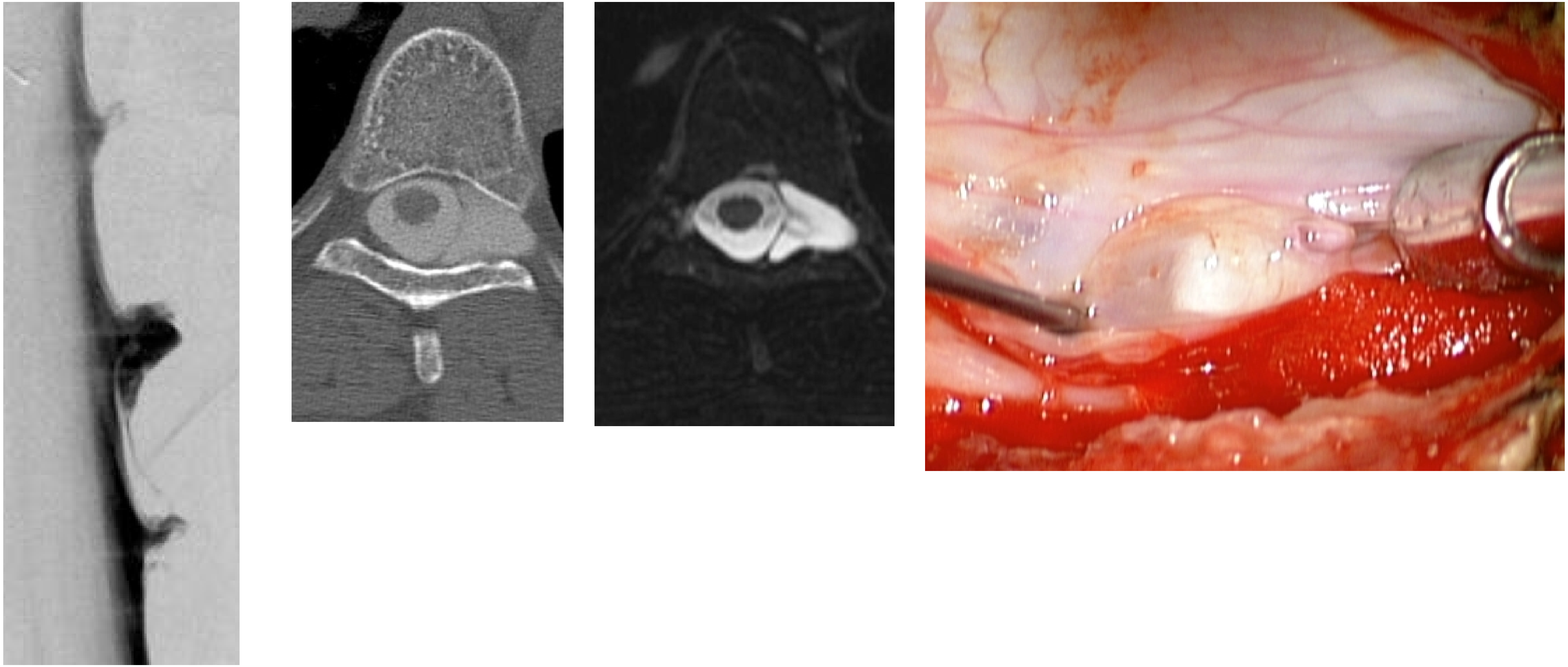


1.0
0.8
0.6
0.4
0.2
0
Superficial Siderosis (SS) Probability
+ Censored
95% Confidence Limits
0
50
100
150
200
Superficial Siderosis Free Time (months)

Ventral epidural venous plexus







Type 1b – Lateral or posterior leak

Type 1b lateral leak

- **No** microspur/osteophyte
- Microfibrillopathy?
- Thoracic >90% of cases
- Percutaneous treatment can be curative at any time
- Less risk of superficial siderosis?

Type 1b lateral leak

- **No** microspur/osteophyte
- Microfibrillopathy?
- Thoracic 95%, cervical 5%
- Percutaneous treatment can be curative at any time
- Less risk of superficial siderosis?

J Neurosurg 96:483–489, 2002

Spontaneous spinal cerebrospinal fluid leaks and minor skeletal features of Marfan syndrome: a microfibrillopathy

IRIS SCHRIJVER, M.D., WOUTER I. SCHIEVINK, M.D., MAURICE GODFREY, PH.D., FREDRIC B. MEYER, M.D., AND UTA FRANCKE, M.D.

Howard Hughes Medical Institute and the Departments of Genetics and Pediatrics, Stanford University School of Medicine, Stanford, California; Cedars–Sinai Neurosurgical Institute, Los Angeles, California; Department of Pediatrics and Center for Human Molecular Genetics, University of Nebraska Medical Center, Omaha, Nebraska; and Department of Neurologic Surgery, Mayo Clinic, Rochester, Minnesota

Object. Spontaneous spinal cerebrospinal fluid (CSF) leaks are increasingly recognized as a cause of postural headaches. The authors examined a group of patients suffering from spontaneous spinal CSF leaks who also had minor skeletal features of Marfan syndrome for abnormalities of fibrillin-containing microfibrils.

Methods. Patients with spontaneous CSF leaks were evaluated for the clinical characteristics of connective tissue disorders. Skin biopsies were obtained in three patients with skeletal manifestations that constitute part of the Marfan syndrome phenotype. Cultured fibroblasts were studied for fibrillin-1 synthesis and incorporation into the extracellular matrix (ECM) by performing quantitative metabolic labeling and immunohistochemical analysis. Among 20 consecutive patients found to have spinal CSF leaks, four (20%) exhibited minor skeletal features of Marfan syndrome, but lacked any ocular or cardiovascular abnormalities. The mean age of these patients (30 years) was lower than that of the 16 patients without skeletal abnormalities (44 years; $p = 0.01$). Abnormalities in fibrillin-1 metabolism and immunostaining were detected in all three patients with the skeletal abnormalities who underwent examination, but not in a control patient without these skeletal manifestations.

Conclusions. Twenty percent of patients who experience spontaneous spinal CSF leaks have minor skeletal features of Marfan syndrome. The authors demonstrated abnormalities in fibrillin-1 protein deposition in all patients examined, but only one person was found to have a fibrillin-1 abnormality typically found in classic Marfan syndrome. The results indicate that there is a heterogeneous involvement of other components of ECM microfibrils at the basis of this cerebrospinal manifestation. In addition, the authors identified a connective-tissue etiological factor in a group of disorders not previously classified as such.

KEY WORDS • fibrillin • headache • intracranial hypotension • cerebrospinal fluid leak • Marfan syndrome • microfibrillopathy • pulse-chase analysis • fibrillin immunofluorescence

FIG. 1. Photographs demonstrating the hyperextensibility of the wrist and finger joints in one patient (Case 2; *left*) and long slender fingers in two other patients (Cases 3 and 4; *center* and *right*, respectively).

ing diverticulum in another (Case 2). In the third patient (Case 3), the exact cause of the leak could not be determined. Although this patient displayed a focal CSF leak at C-7, a meningeal diverticulum was not detected at that site during surgery. Nevertheless, myelography revealed several small meningeal diverticula at other sites. The fourth patient (Case 4) was treated nonsurgically with multiple spinal epidural blood patches. In none of the patients was the characteristic, generalized dural ectasia of Marfan syndrome exhibited on CT myelography studies.

The results of ophthalmological and echocardiographic examinations performed to screen for additional features of Marfan syndrome in Cases 1, 2, and 3 were normal. The fourth patient (Case 4) was treated in an out-patient setting and was unavailable for further studies. All four patients had one or more first-degree relatives with tall stature, but none had a family history of Marfan syndrome, ocular lens dislocations, cardiac valvular disease, aortic aneurysms or dissections, spinal CSF leaks, or other connective tissue abnormalities.

A comparison of clinical and imaging characteristics of patients with and without the minor skeletal features of Marfan syndrome is presented in Table 2. The only significant difference between the two groups was an earlier age at onset of symptoms in those patients who exhibited minor skeletal abnormalities in addition to their CSF leaks.

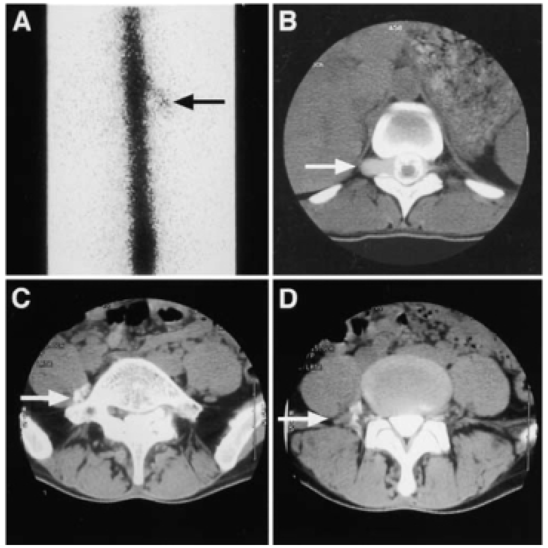
Evaluation of fibrillin synthesis and deposition into the ECM as well as fibrillin immunofluorescence assays was performed on fibroblast samples obtained in three patients (Cases 1, 2, and 3; Table 3), and in one control patient with a spontaneous spinal CSF leak who did not exhibit any features of Marfan syndrome. A search for the *FBN1* mutation was conducted in two patients (Cases 1 and 2).

Pulse-Chase Analysis of Fibrillin-1

The quantitative pulse-chase assay follows the process

Groups III and IV there is normal synthesis, but reduced deposition: 35 to 70% in Group III and less than 35% of control in Group IV. A fifth group, in which both synthesis and deposition are maintained at a level higher than 70%, compared with normal controls, is considered to be within the normal range (100% ± two standard deviations).

In our biochemical evaluation of three patients with spontaneous spinal CSF leaks and minor features of Marfan syndrome, one control patient who experienced a spontaneous spinal CSF leak but did not have skeletal anom-

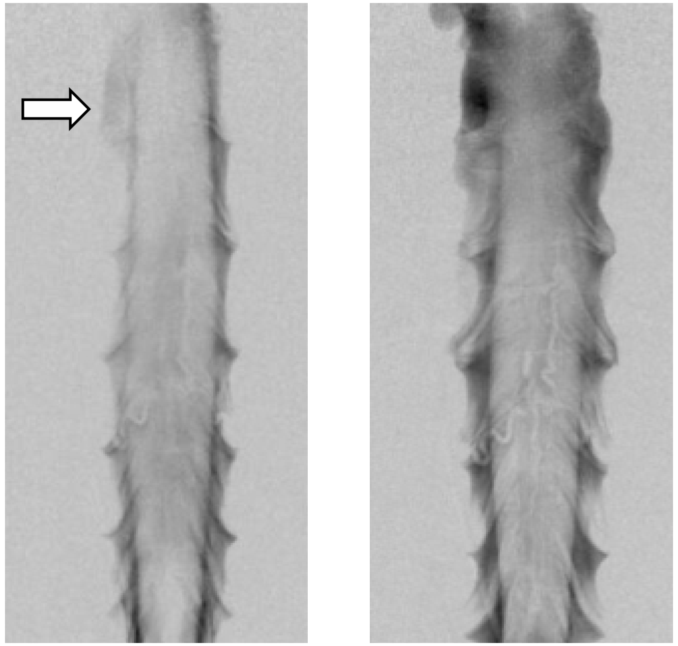


Type 1b lateral leak

- **No** microspur/osteophyte
- Microfibrillopathy?
- Thoracic >90% of cases
- Percutaneous treatment can be curative at any time
- Less risk of superficial siderosis?

Pre-op

Post-op



Lateral leak

Neurosurgical forum

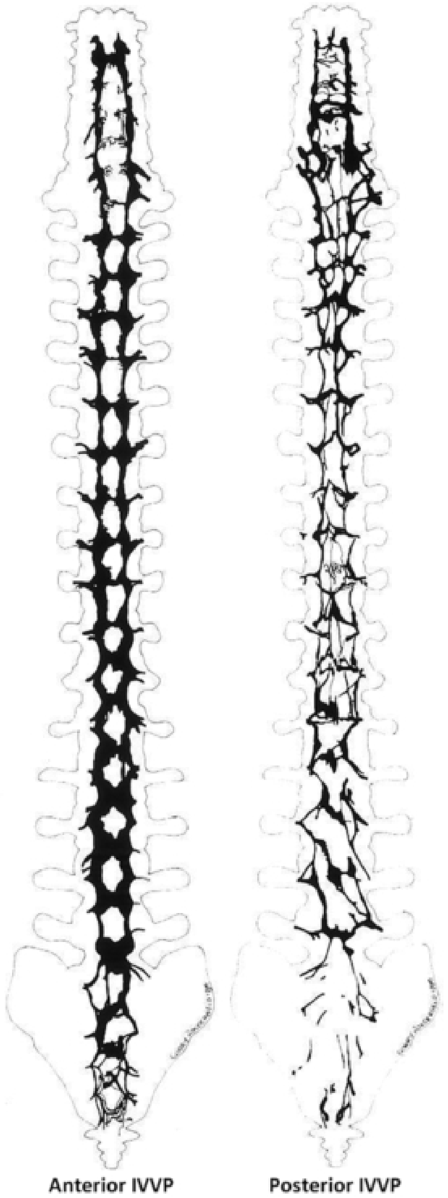
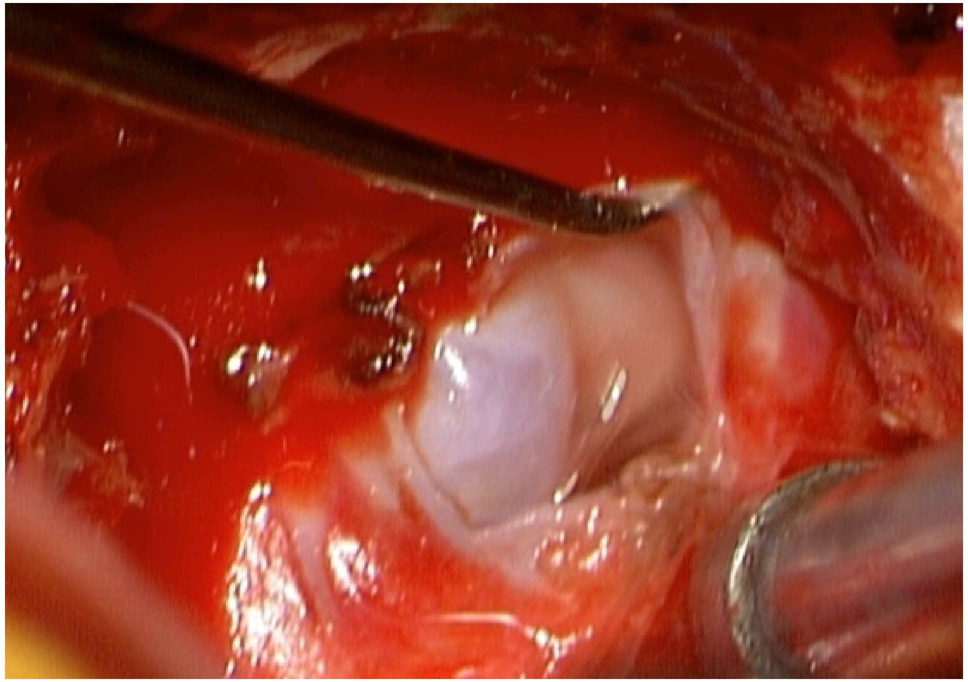
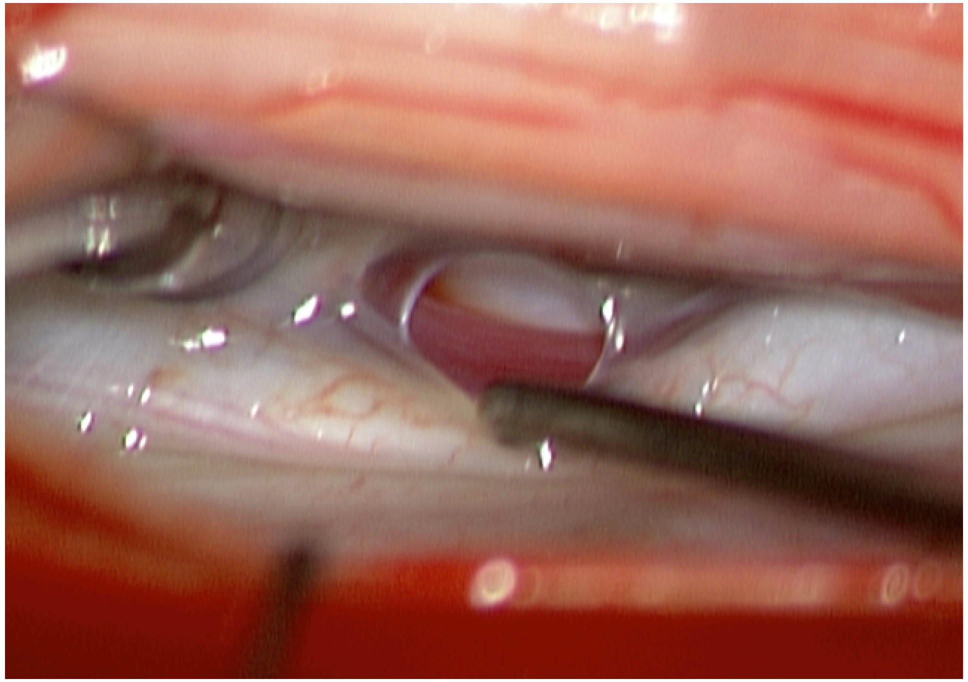


Fig. 2. Schematized representation of the anterior and the posterior IVVPs. Reprinted with permission from Blackwell Publishing. Groen RJ, Groenewegen HJ, van Alphen HA, et al: Morphology of the human internal vertebral venous plexus: a cadaver study after intravenous Araldite CY 221 injection. **Anat Rec 249:**285–294, 1997

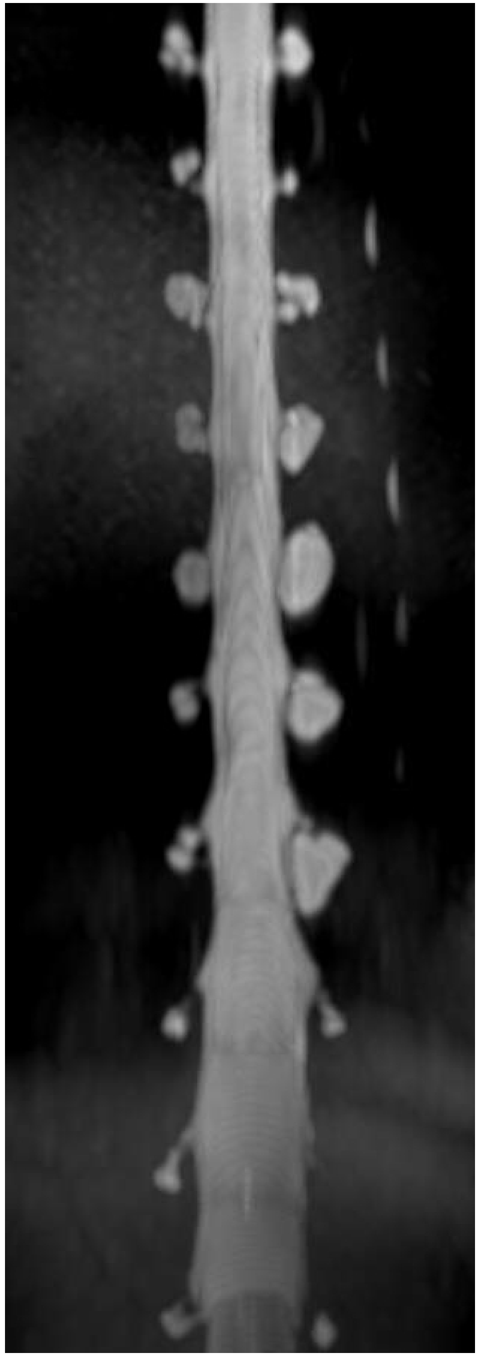
2 reasons

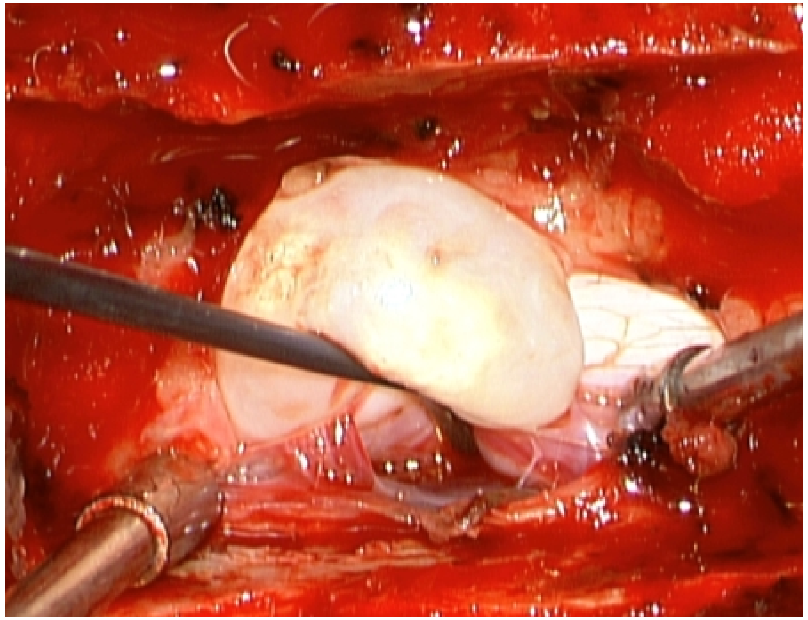


1b
Lateral

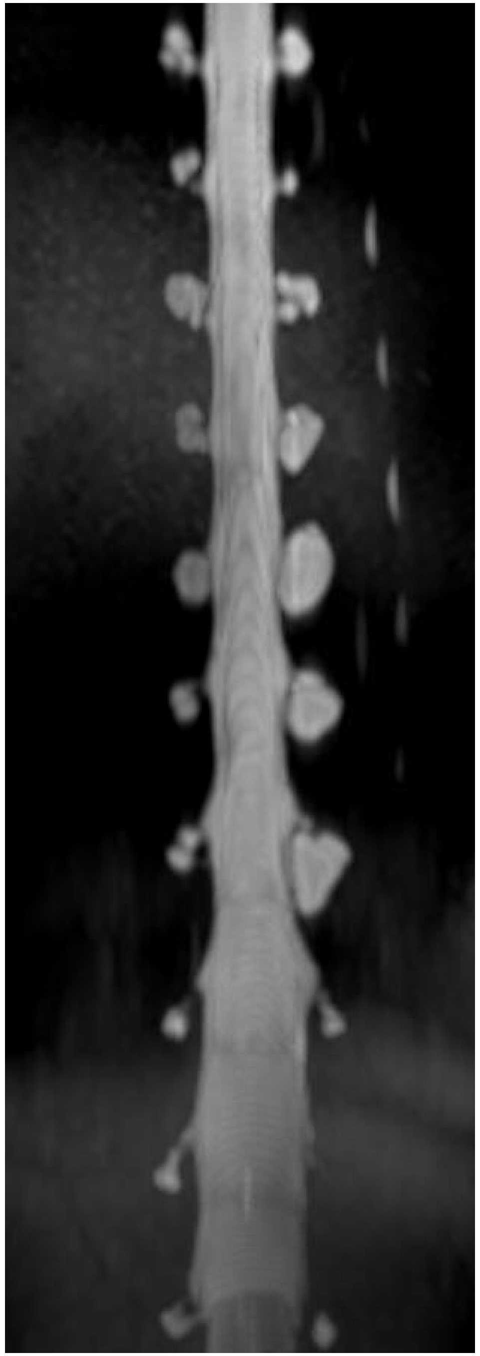


1a
Ventral

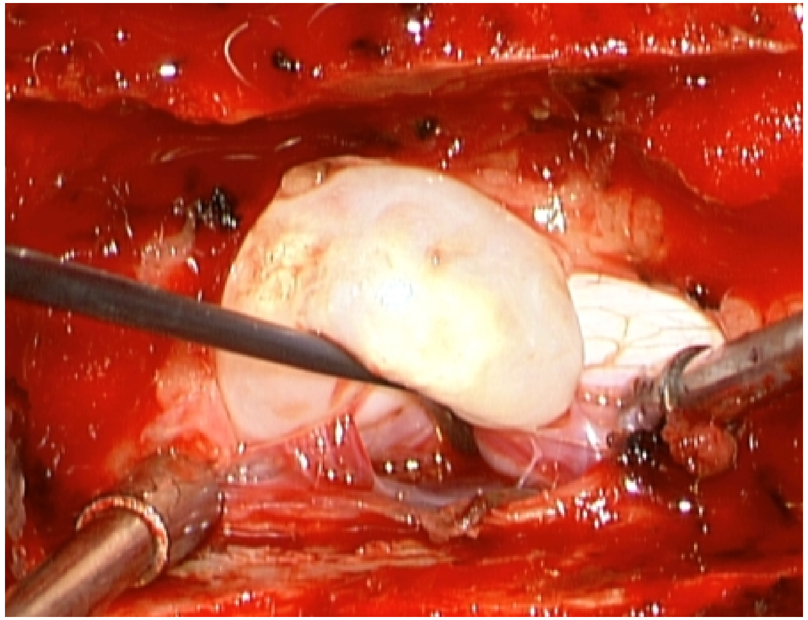




Type 2a – simple meningeal diverticulum



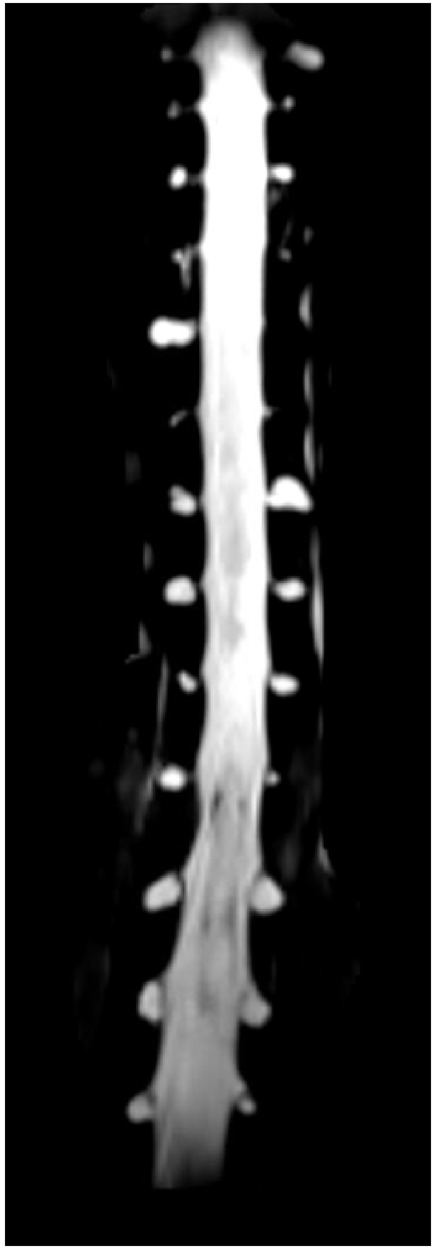
2a



1b

Type 2a simple meningeal diverticulum

- Very common at cervicothoracic junction and lower thoracic spine
- Underlying connective tissue disorder?
- How often is it really a CSF-venous fistula



Spinal Meningeal Diverticula in Spontaneous Intracranial Hypotension: Analysis of Prevalence and Myelographic Appearance

P.G. Kranz, S.S. Stinnett, K.T. Huang, and L. Gray

Table 1: Demographic data

	Control	SIH	*P* Value
Age (mean) (SD) (yr)	55.56 (13.85)	50.37 (13.27)	
Min/median/max	35/58/77	30/52/81	.186[a]
Sex			
Male (No.) (%)	12 (67)	5 (26)	
Female (No.) (%)	6 (33)	14 (74)	.014[b]

Note:—Min indicates minimum; max, maximum.
[a] *P* value based on the Wilcoxon rank sum test of the difference between medians.
[b] *P* value based on a χ^2 test of the difference in proportions.

Table 2: Prevalence of spinal meningeal diverticula by sex

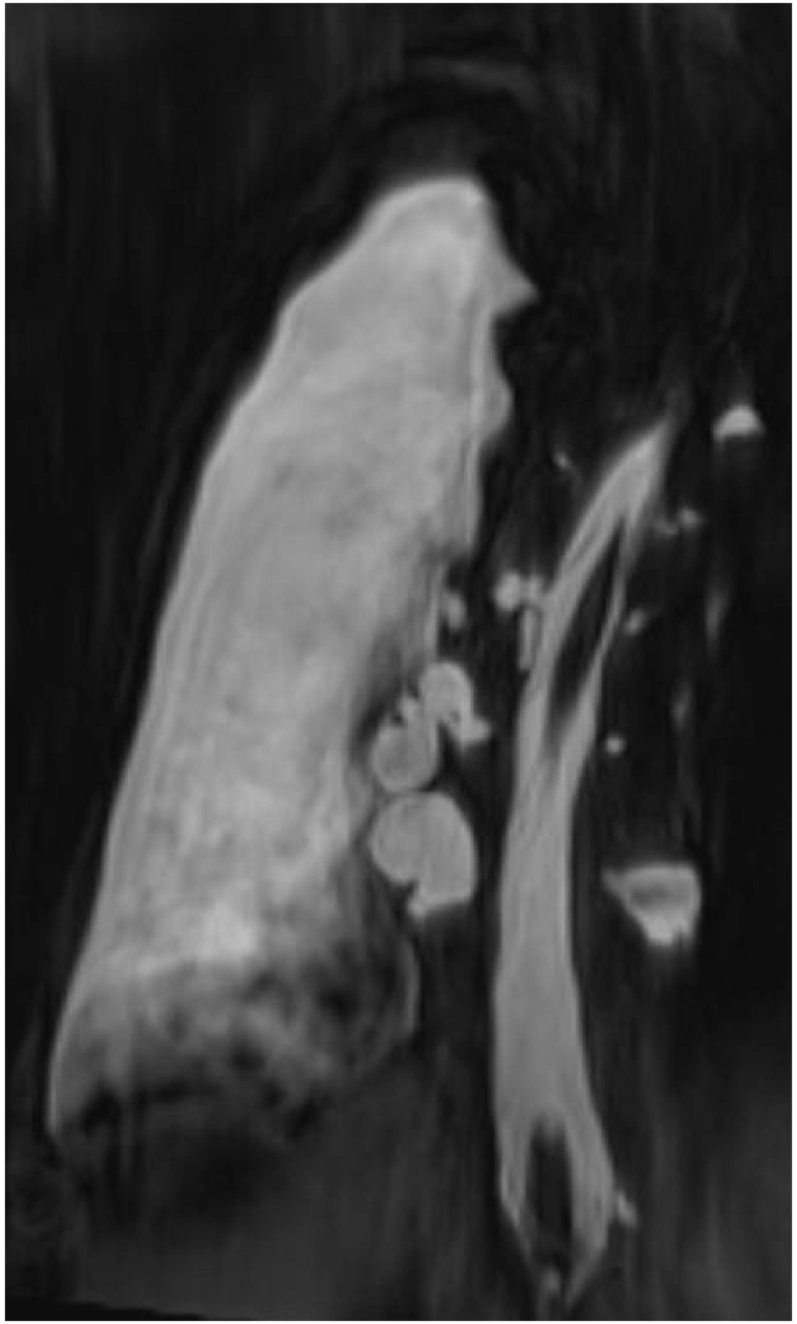
Diverticula per Patient	Male	Female	*P* Value[a]
Mean (SD)	1.6 (2.3)	6.5 (7.9)	.166
Min/median/max	0/1/7	0/1.5/23	

Note:—Min indicates minimum; max, maximum.
[a] *P* value based on the Wilcoxon rank sum test of the difference between medians.

Table 3: Prevalence of spinal meningeal diverticula and prominent nerve sheaths

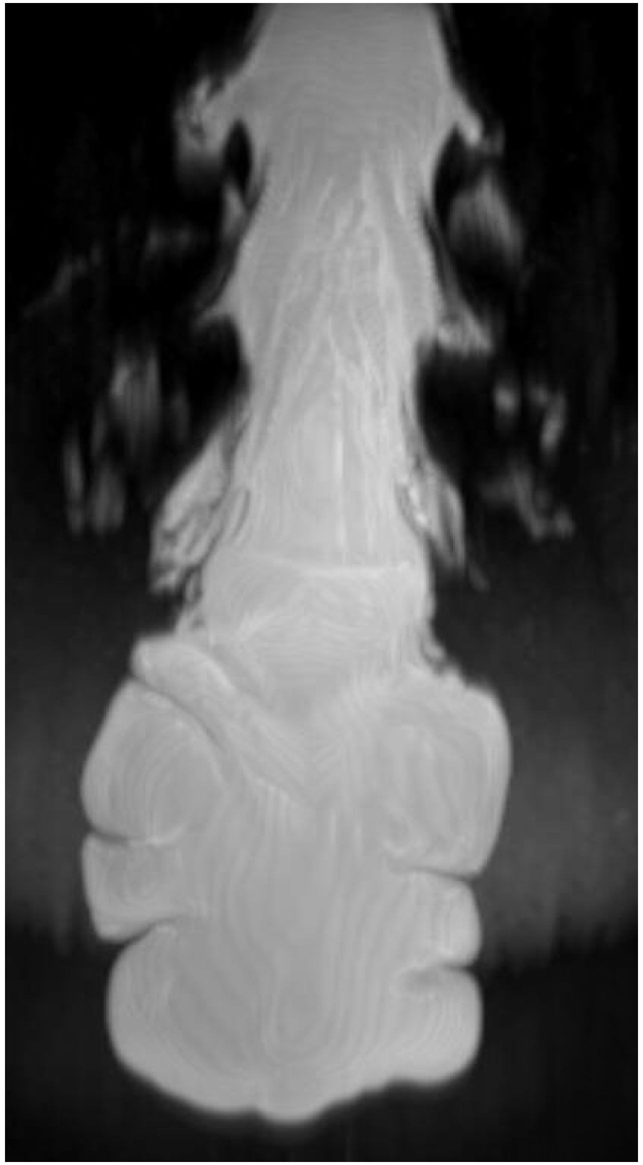
	Control	SIH	*P* Value
Patients with diverticula present (No.) (%)	8 (44)	13 (68)	.141[a]
Diverticula per patient (mean)	2.2 (3.3)	6.3 (8.0)	.099[b]
Min/median/max	0/0/10	0/2/23	
Patients with prominent nerve sheaths present (No.) (%)	14 (78)	17 (89)	.405[c]
Prominent nerve sheaths per patient (mean)	2.6 (3.1)	6.1 (4.2)	.004[c]
Min/median/max	0/1.5/13	0/5/15	

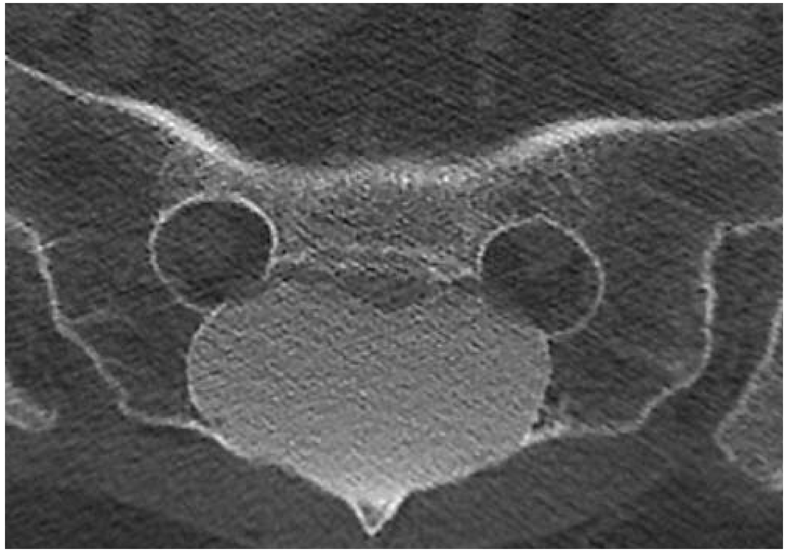
Note:—Min indicates minimum; max, maximum.
[a] *P* value based on a χ^2 test of the difference between proportions.
[b] *P* value based on the Wilcoxon rank sum test of the difference between medians.
[c] *P* value based on the Fisher exact test of the difference between proportions.

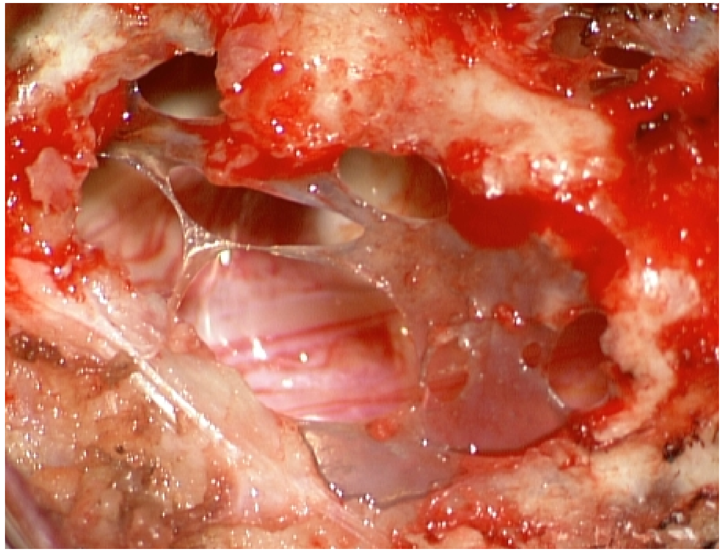


Meningeal diverticula with extradural CSF

CSF hydrothorax





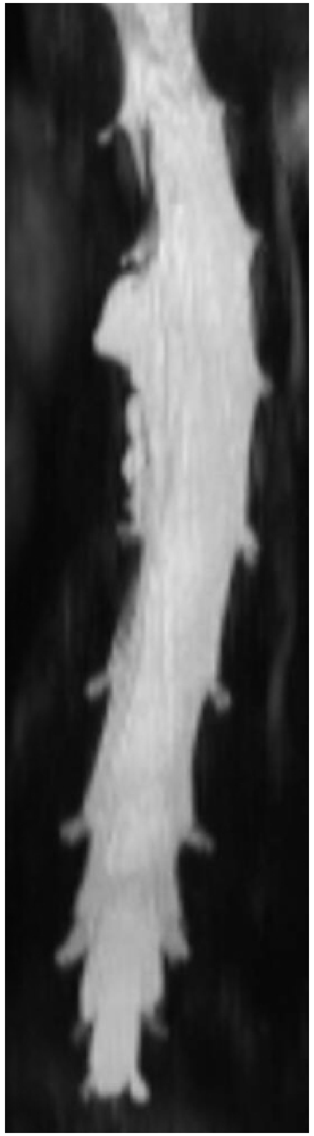


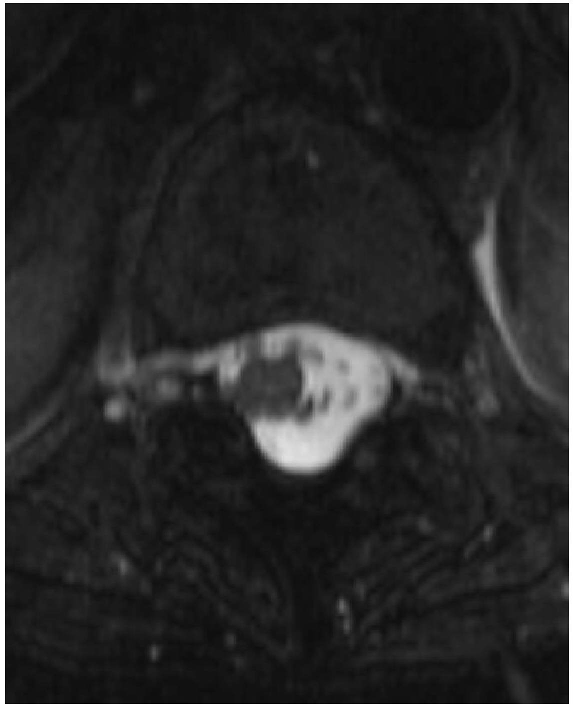
Type 2b – dural ectasia

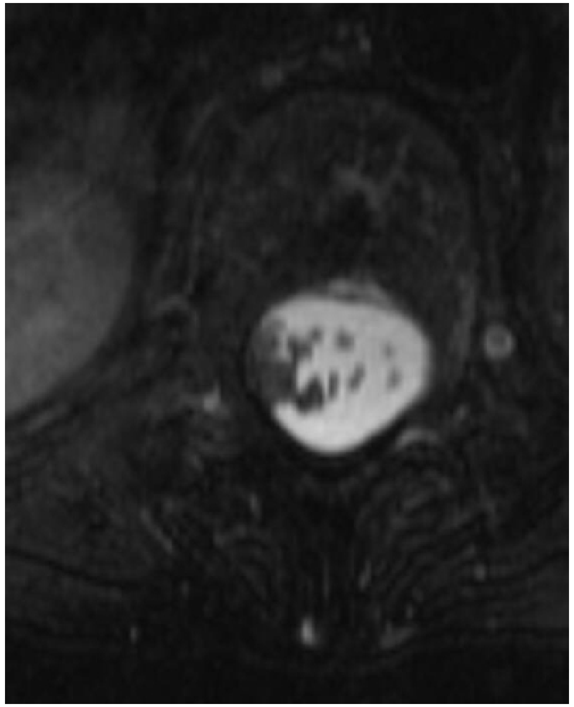
Type 2b - dural ectasia

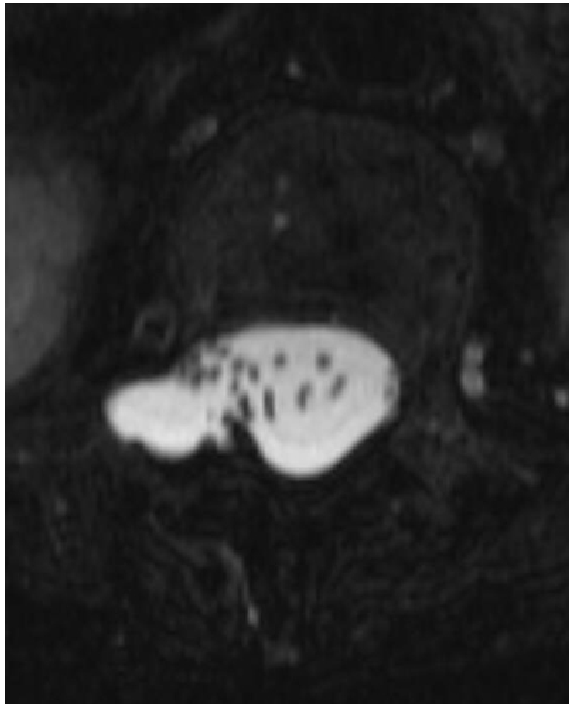
- An underlying connective tissue disorder
  - Ankylosing spondylitis
  - Marfan syndrome
  - Neurofibromatosis type 1

Ankylosing spondylitis (M Bechterew)

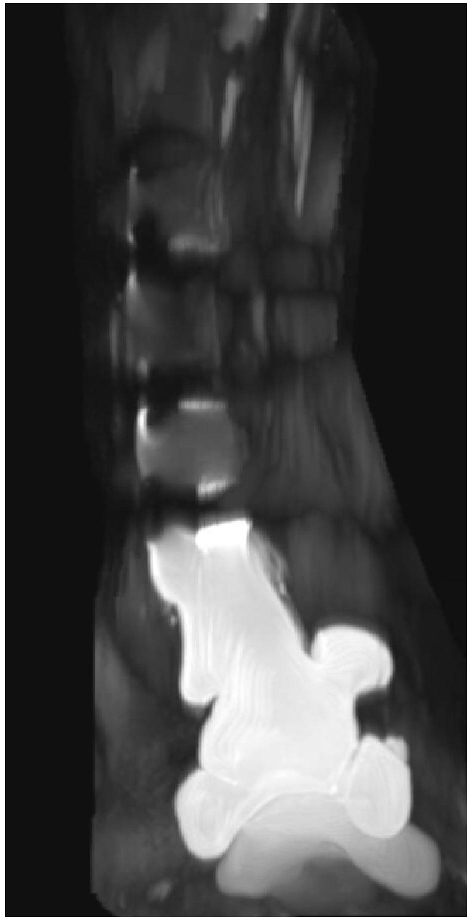


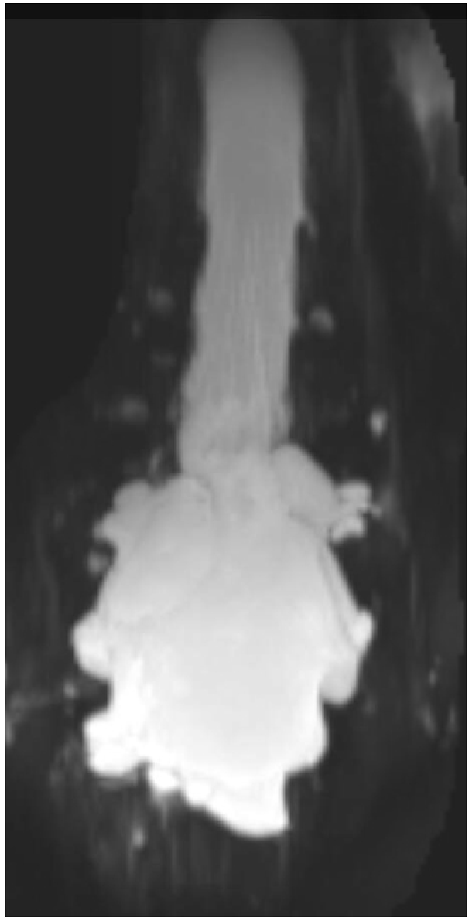
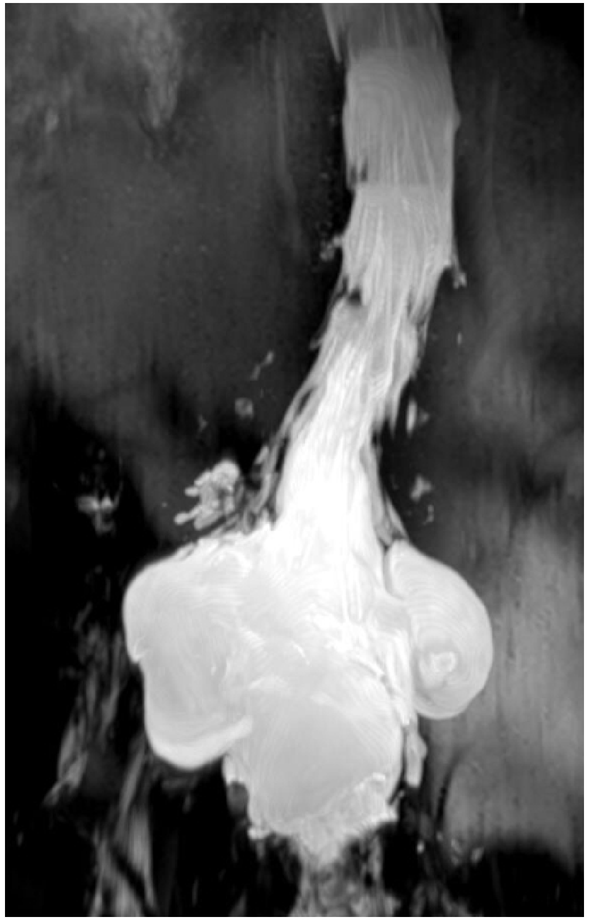




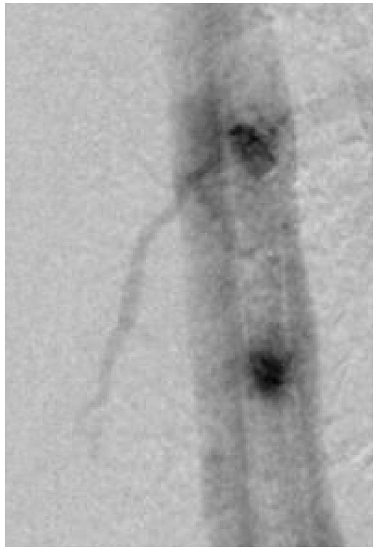


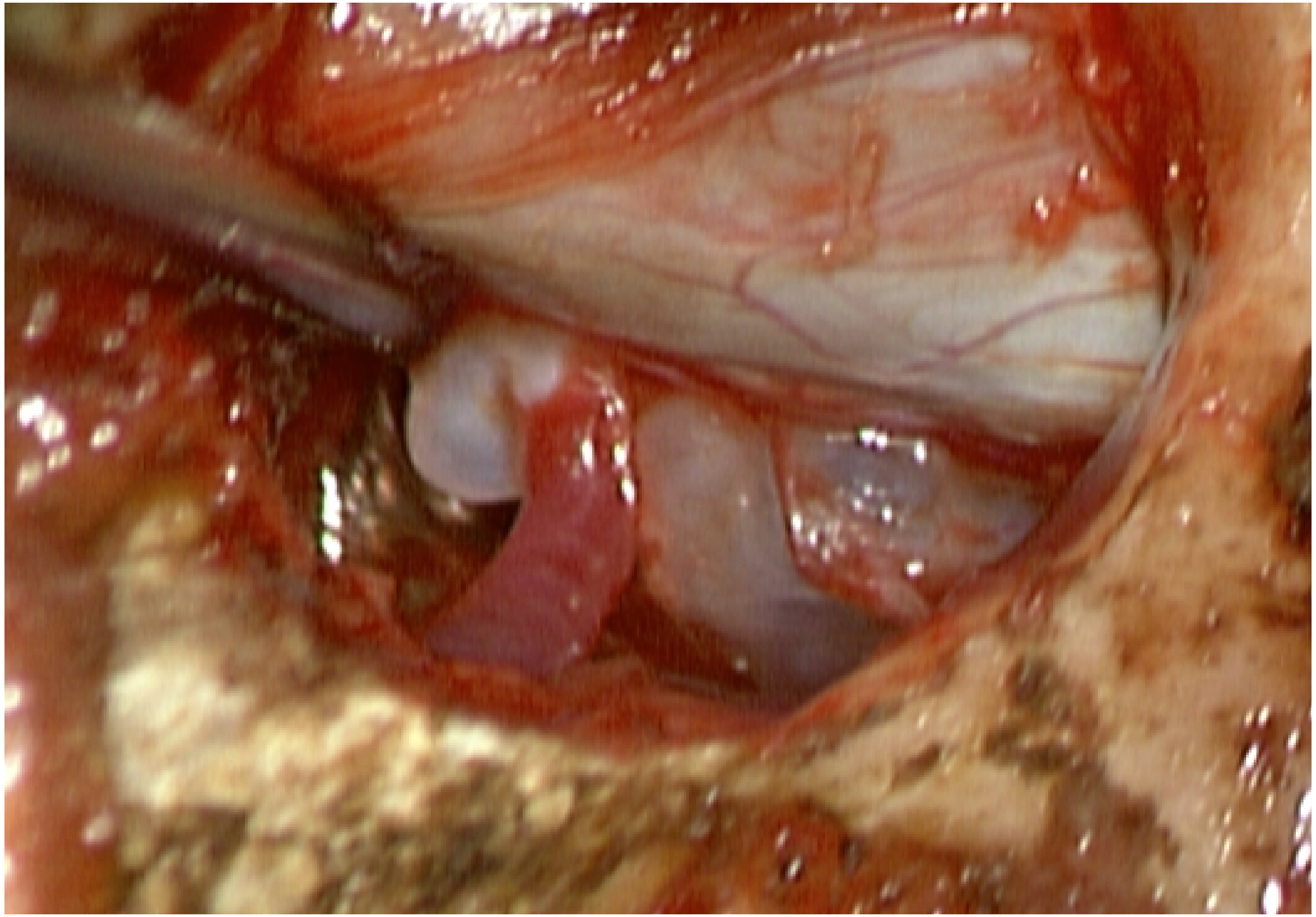
Marfan syndrome



Neurofibromatosis type 1





Type 3 – CSF-venous fistula

Type 3 – CSF-venous fistula

Wouter I. Schievink, MD
Franklin G. Moser, MD, MMM
M. Marcel Maya, MD

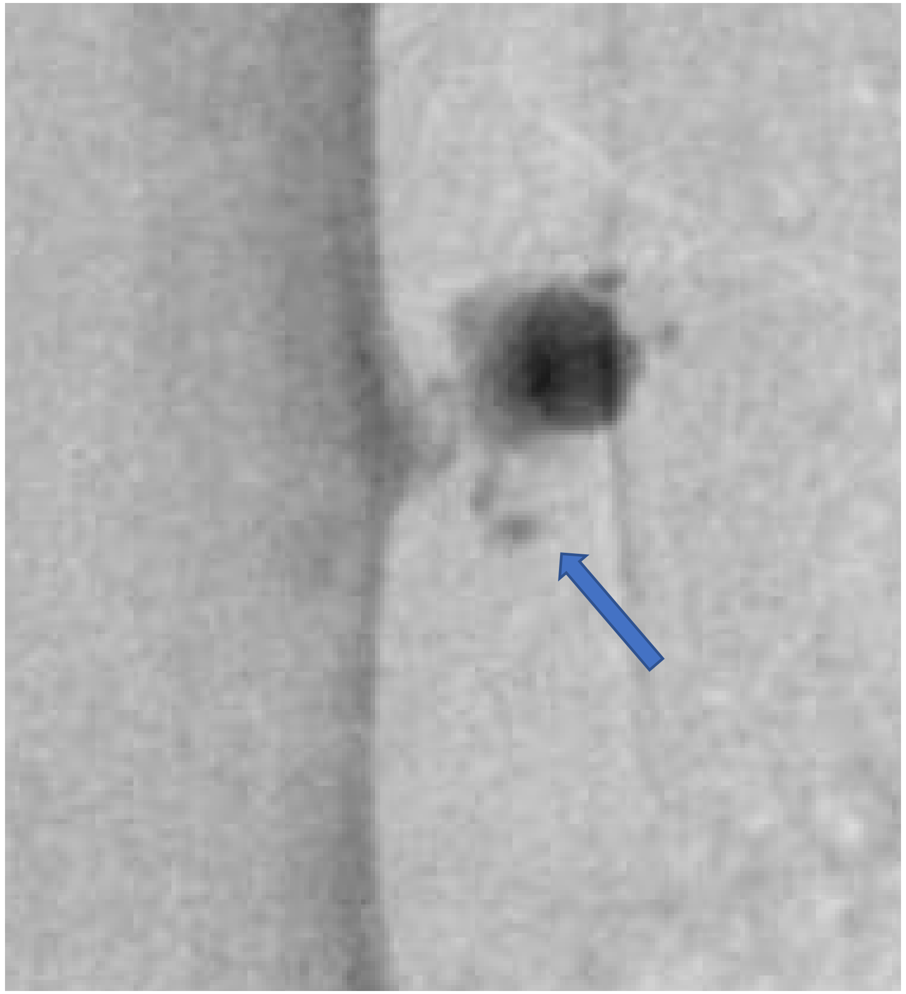
CSF-VENOUS FISTULA IN SPONTANEOUS INTRACRANIAL HYPOTENSION

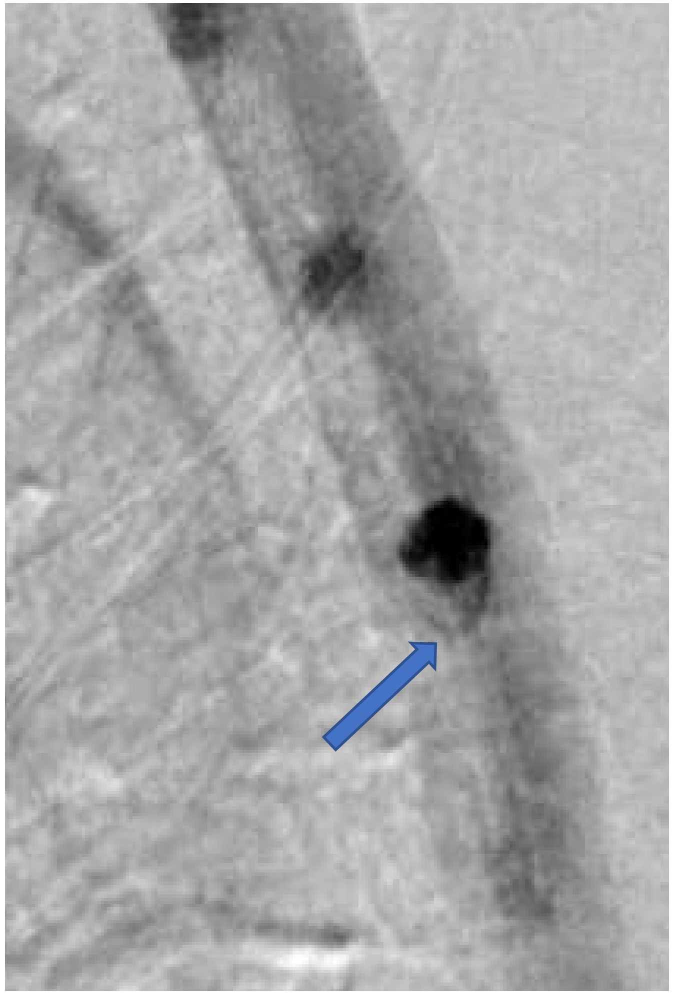
Spontaneous intracranial hypotension (SIH) is an important cause of new daily persistent headaches.[1]

Neurology. 2014 Jul 29;83(5):472-3

- Ruptured arachnoid granulation
- Associated with meningeal diverticula or arise from common thecal sac (different types)
- Associated with venous or veno-lymphatic vascular malformations
- Any spinal level but >90% thoracic
- How common?

Spinal CSF–venous fistulas: Are they "shy"





Spinal CSF–venous fistulas: Are they "shy"

Not always

Type 3 – CSF-venous fistula

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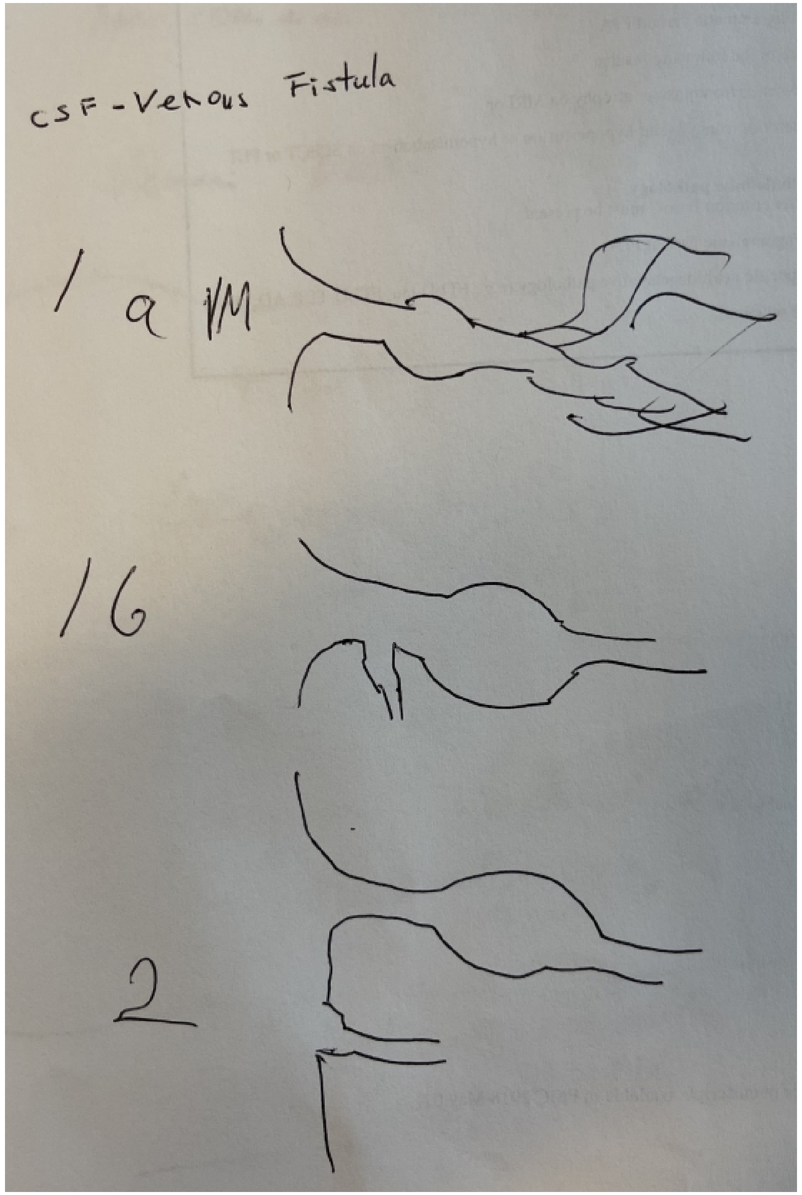
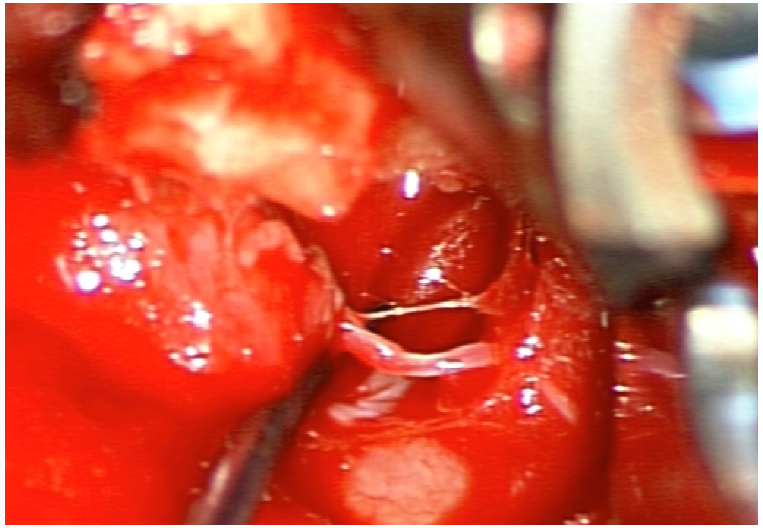
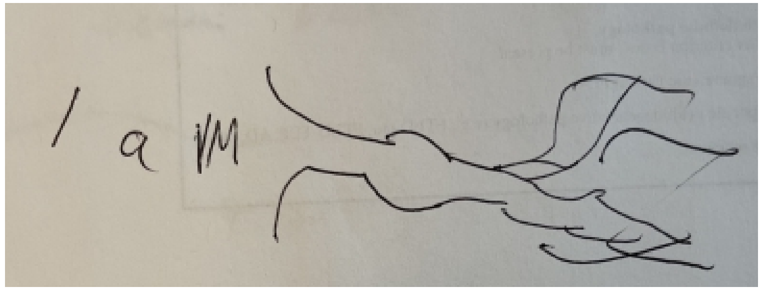
Types of spontaneous spinal CSF-venous fistulas

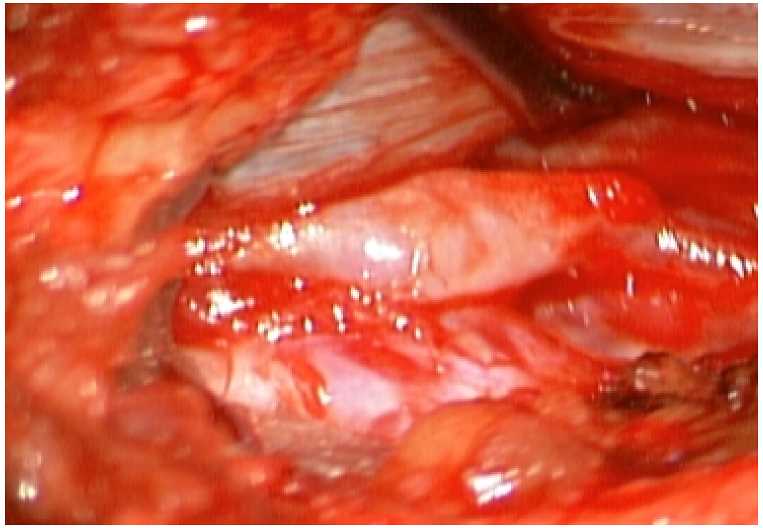
* based on fistula origin

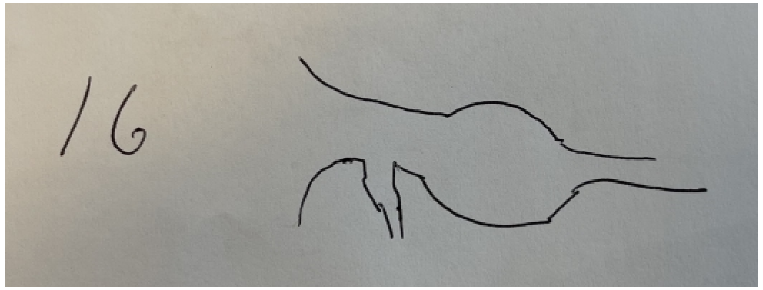




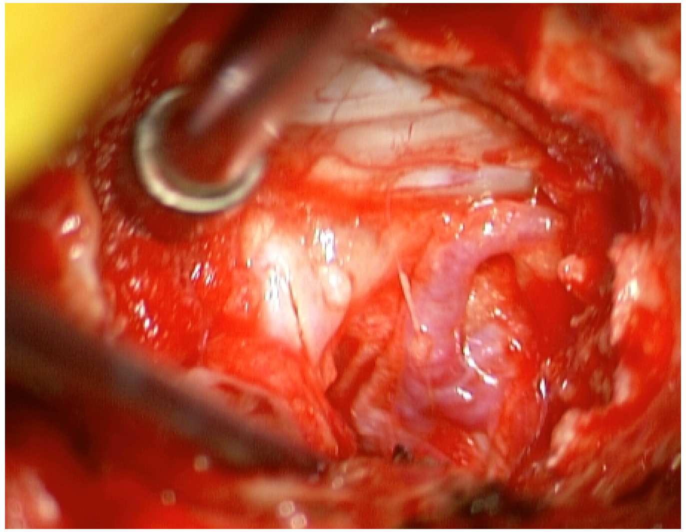


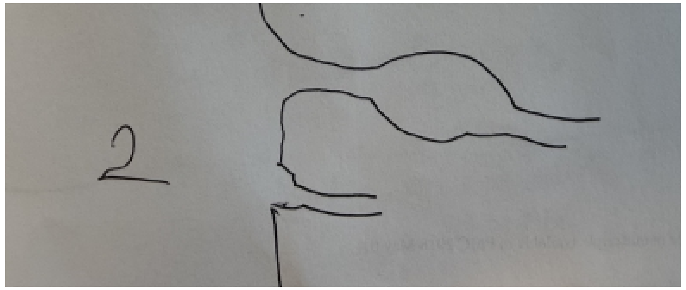
75%





15%





10%

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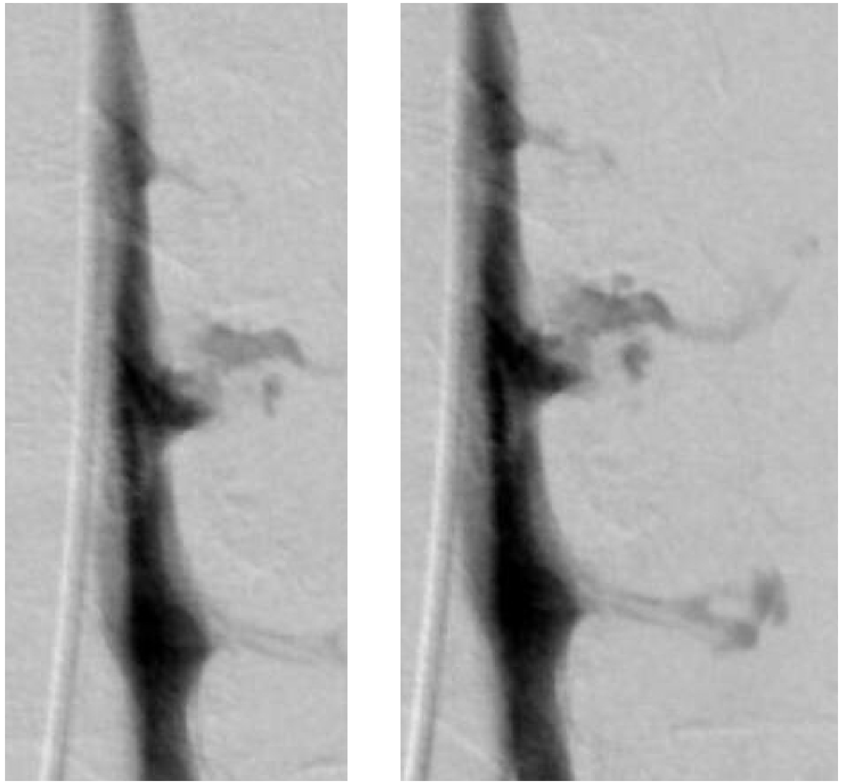
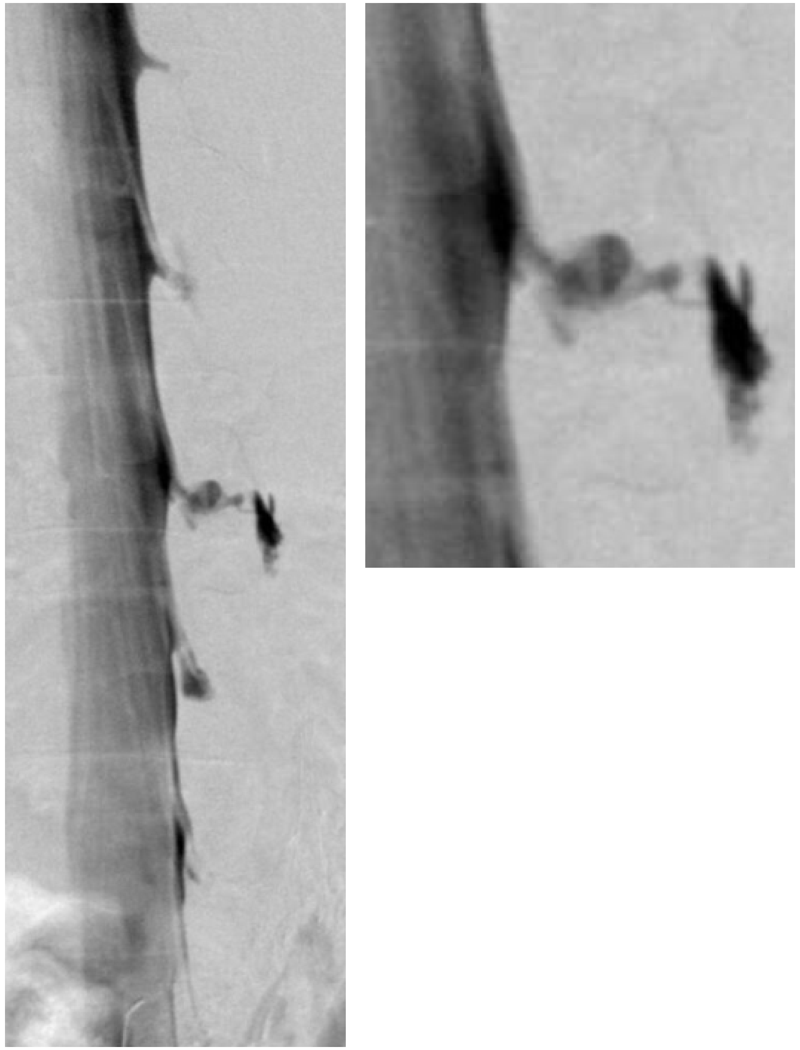
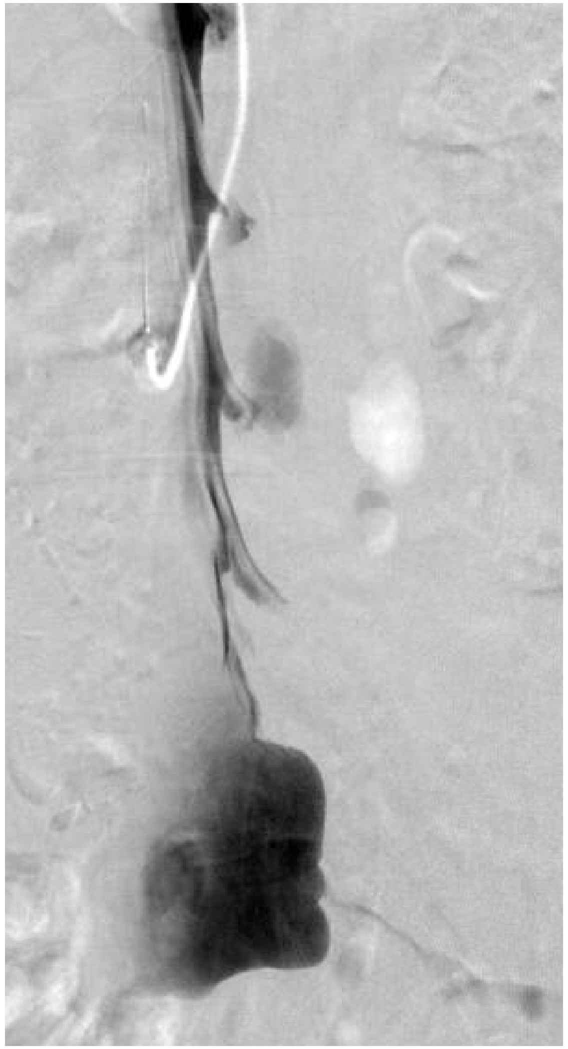


Cervical



Lumbar



Sacral

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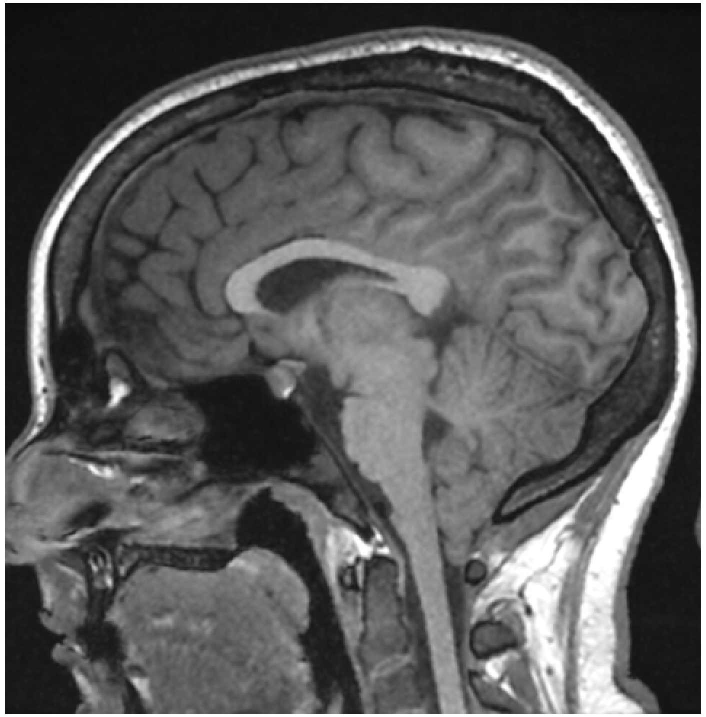
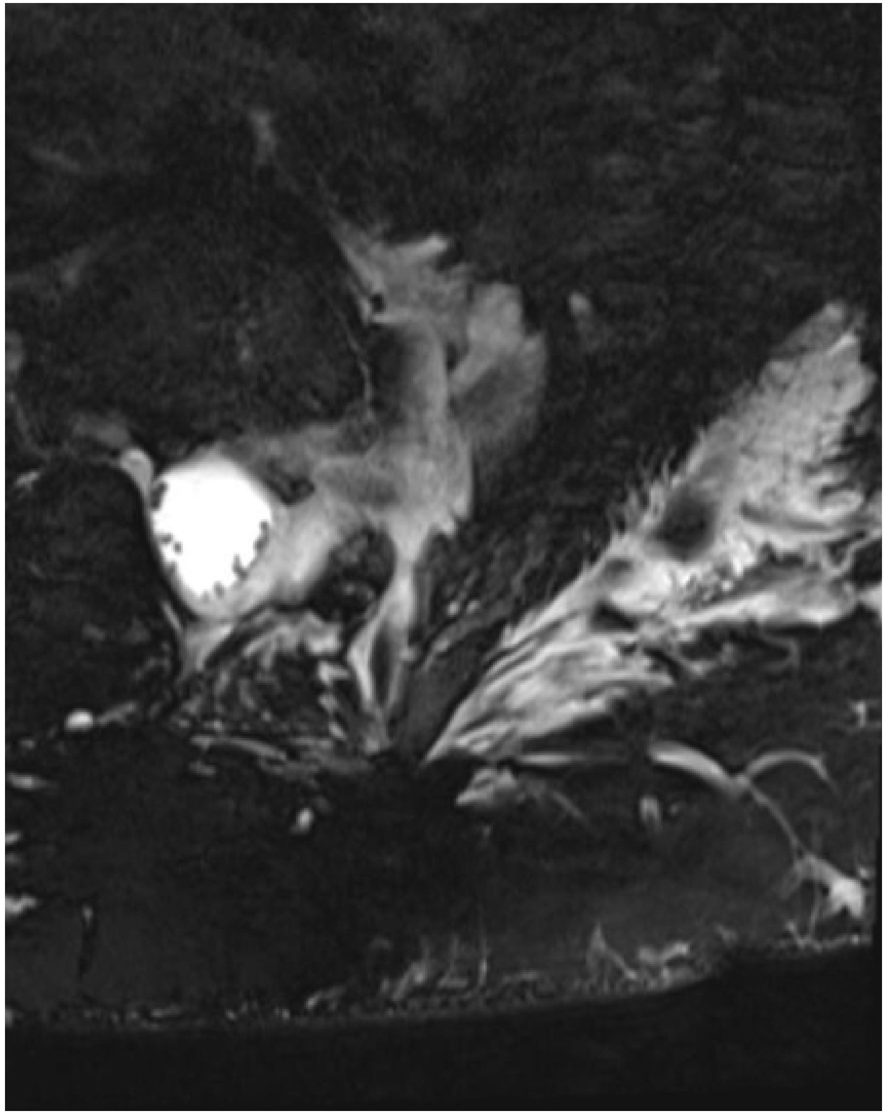
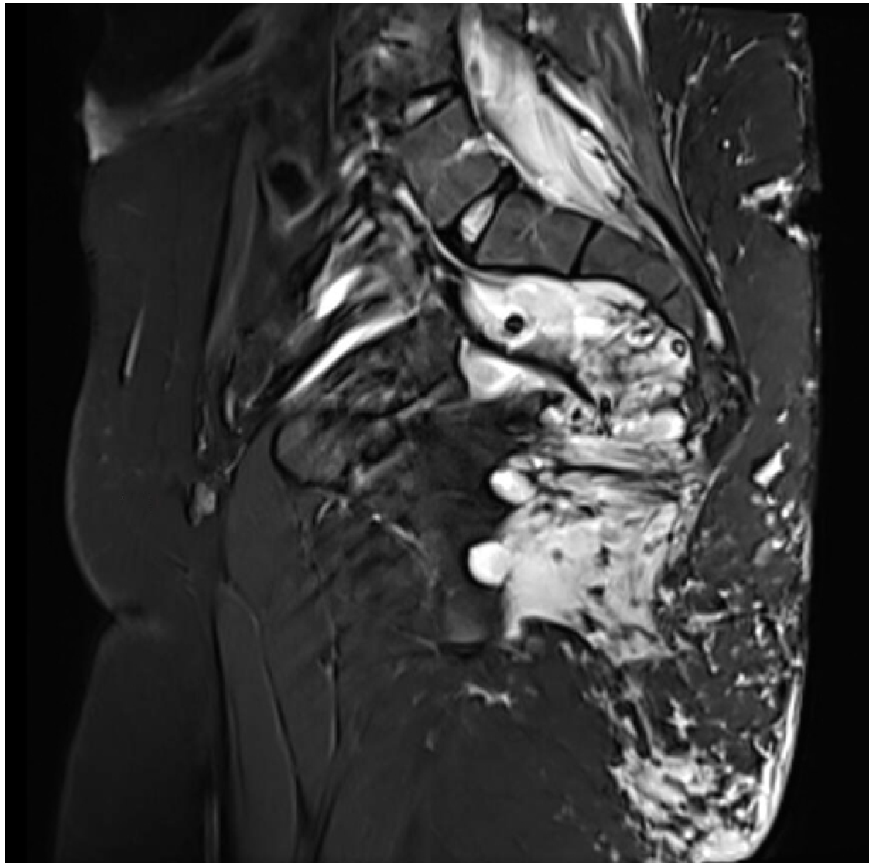
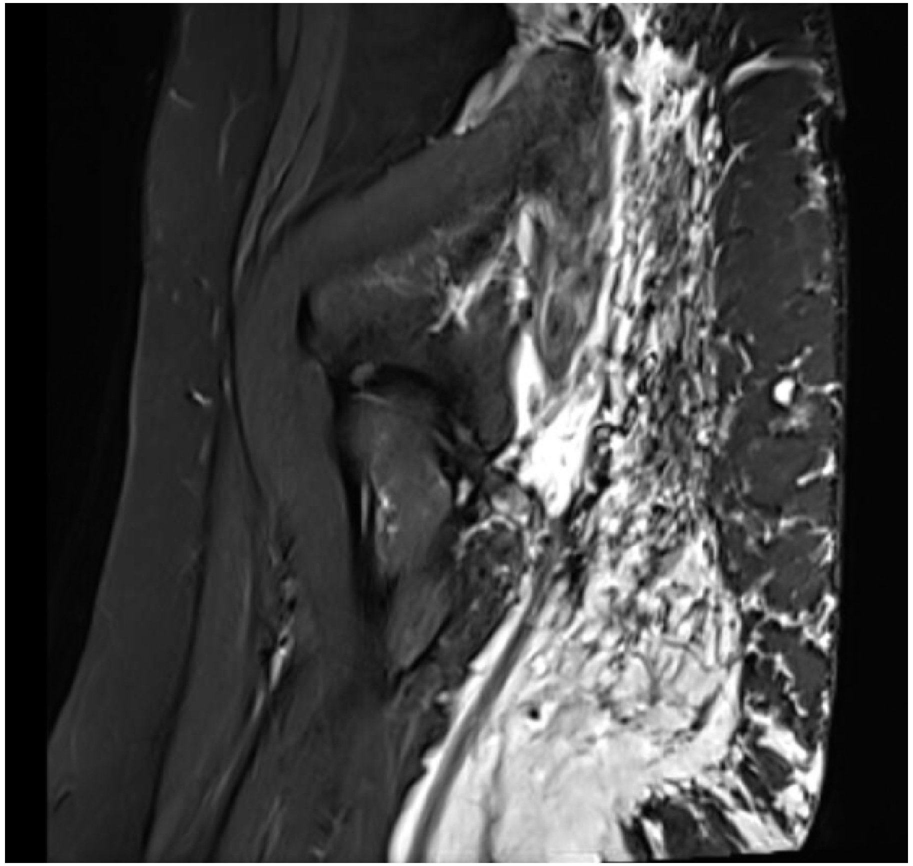
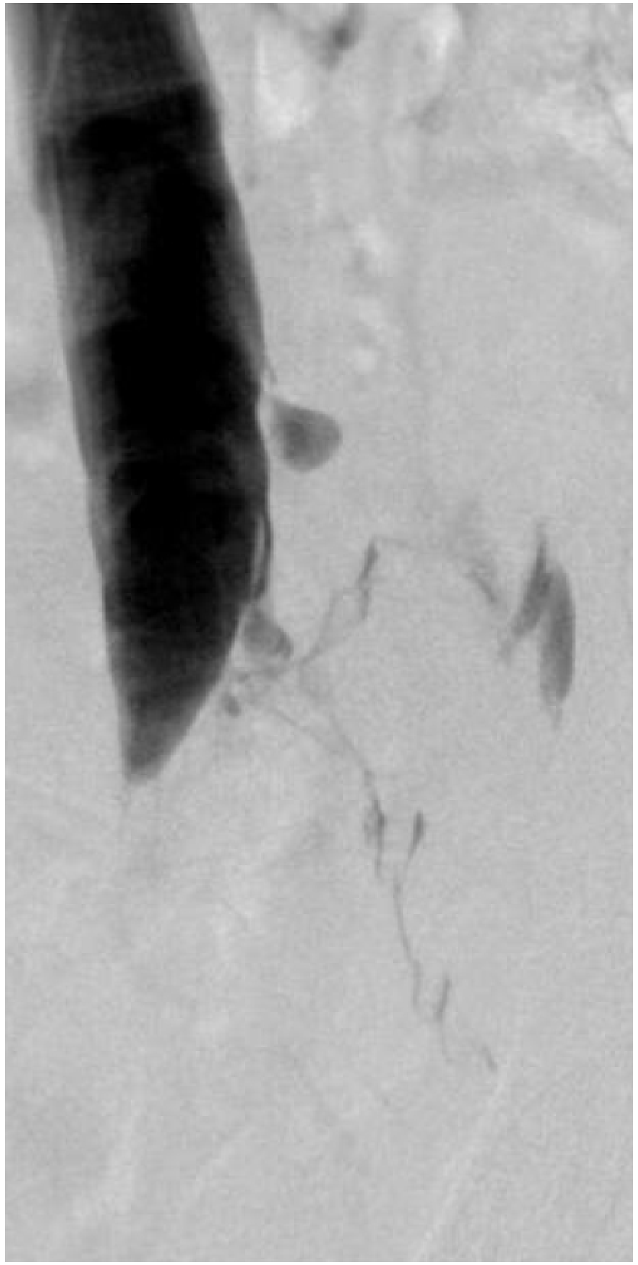
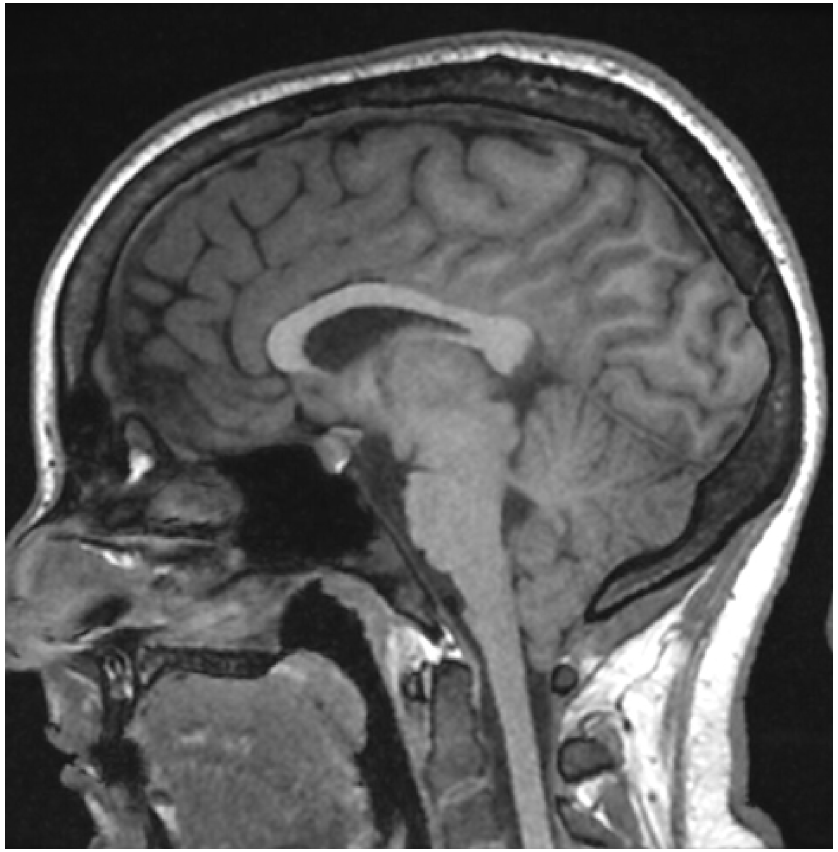


17 year-old girl





Yield of DSM in identifying CSF-venous fistulas

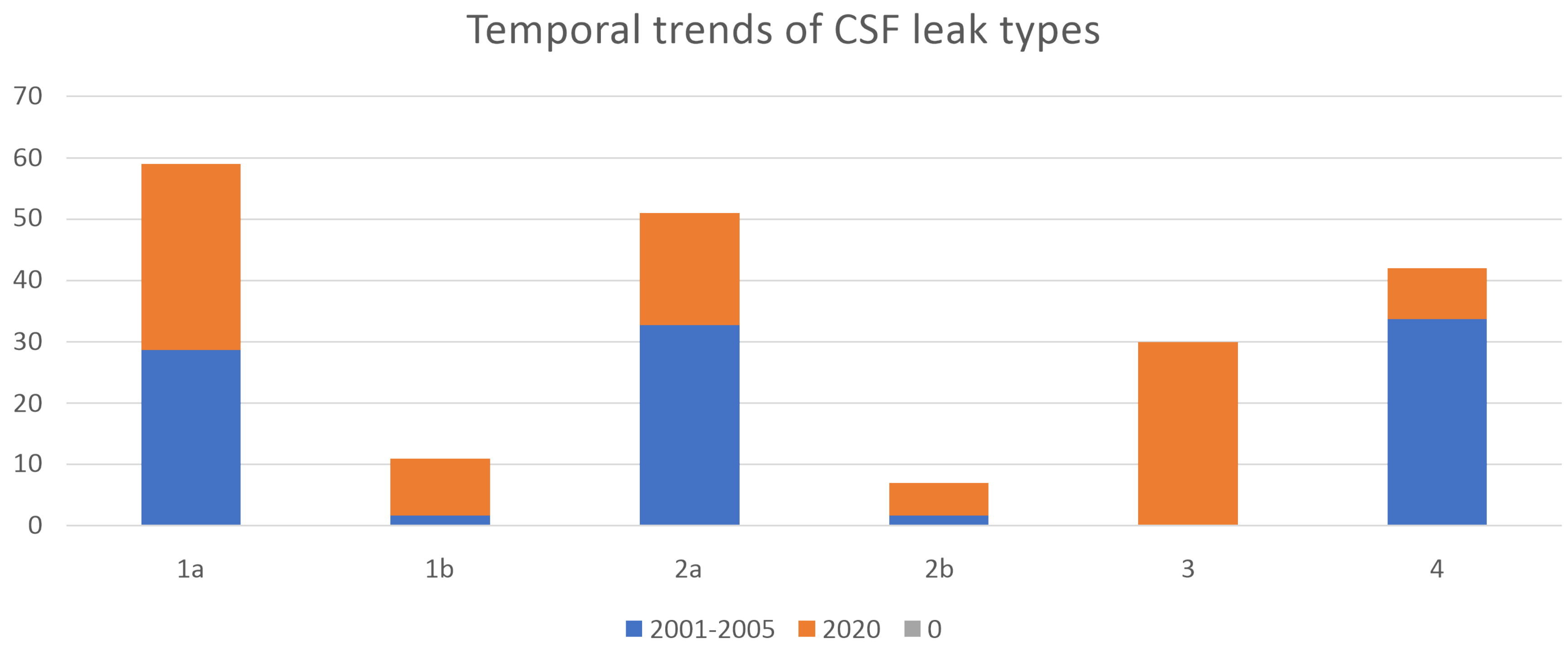
- Positive brain MRI: 80+%
- Negative brain MRI: 10% (20% if meningeal diverticula present)
- Brain sagging/bvFTD: 40%

How common are these different types?

- Temporal trends!

How common are these different types?

- Temporal trends!



How common are these different types?

- Referral bias!

How common are these different types?

Original Article

Cephalalgia An International Journal of Headache — International Headache Society

Cephalalgia
0(0) 1–5

DOI: 10.1177/03331024211048510
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Incidence of spontaneous intracranial hypotension in a community. Beverly Hills, California, 2006–2020

Wouter I Schievink[1], M Marcel Maya[2], Franklin G Moser[2], Paul Simon[3] and Miriam Nuño[4]

Abstract [AQ1]

Background: Spontaneous intracranial hypotension is diagnosed with an increasing frequency, but epidemiologic data are scarce. The aim of this study was to determine the incidence rate of spontaneous intracranial hypotension in a defined population.

Methods: Using a prospectively maintained registry, all patients with spontaneous intracranial hypotension residing in Beverly Hills, California, evaluated at our Medical Center between 2006 and 2020 were identified in this population-based incidence study. Our Medical Center is a quaternary referral center for spontaneous intracranial hypotension and is located within 1.5 miles from downtown Beverly Hills.

Results: A total of 19 patients with spontaneous intracranial hypotension were identified. There were 12 women and seven men with a mean age of 54.5 years (range, 28 to 88 years). The average annual incidence rate for all ages was 3.7 per 100,000 population (95% confidence interval [CI]: 2.0 to 5.3), 4.3 per 100,000 for women (95% CI, 1.9 to 6.7) and 2.9 per 100,000 population for men (95% CI, 0.8 to 5.1).

Conclusion: This study, for the first time, provides incidence rates for spontaneous intracranial hypotension in a defined population.

Table 1: Clinical and radiographic characteristics of 19 patients with spontaneous intracranial hypotension.

Variable	
Age in years	
mean	54.5
range	28–88
Female sex, no (%)	12 (63.2)
Race, no (%)	
Non-Hispanic White	17 (89.5)
Asian	2 (10.5)
Clinical manifestations, no (%)	
Headache	17 (89.5)
Orthostatic headache	12 (63.2)
Non-orthostatic headache	5 (26.3)
Vestibulo-cochlear symptoms	10 (52.6)
Nausea/vomiting	9 (47.4)
Neck pain/stiffness	6 (31.6)
Phono/photophobia	5 (26.3)
Face pressure	3 (15.8)
Blurred vision	3 (15.8)
Cognitive difficulties	3 (15.8)
Coma	1 (5.3)
Bibrachial amyotrophy	1 (5.3)
Syringomyelia	1 (5.3)
Brain MRI findings, no (%)	
Normal	2 (10.5)
Abnormal (see reference 2)	17 (89.5)
S Subdural fluid collections	8 (42.1)
E Enhancement of meninges	14 (73.7)
E Engorgement of venous structures	3 (15.8)
P Pituitary enlargement	5 (26.3)
S Sagging of brain	12 (63.2)
Spinal CSF leak type, no (%) (see reference 9)	
1a	5 (26.3)
2a	9 (47.4)
4	5 (26.3)

Thank you!

schievinkw@cshs.org