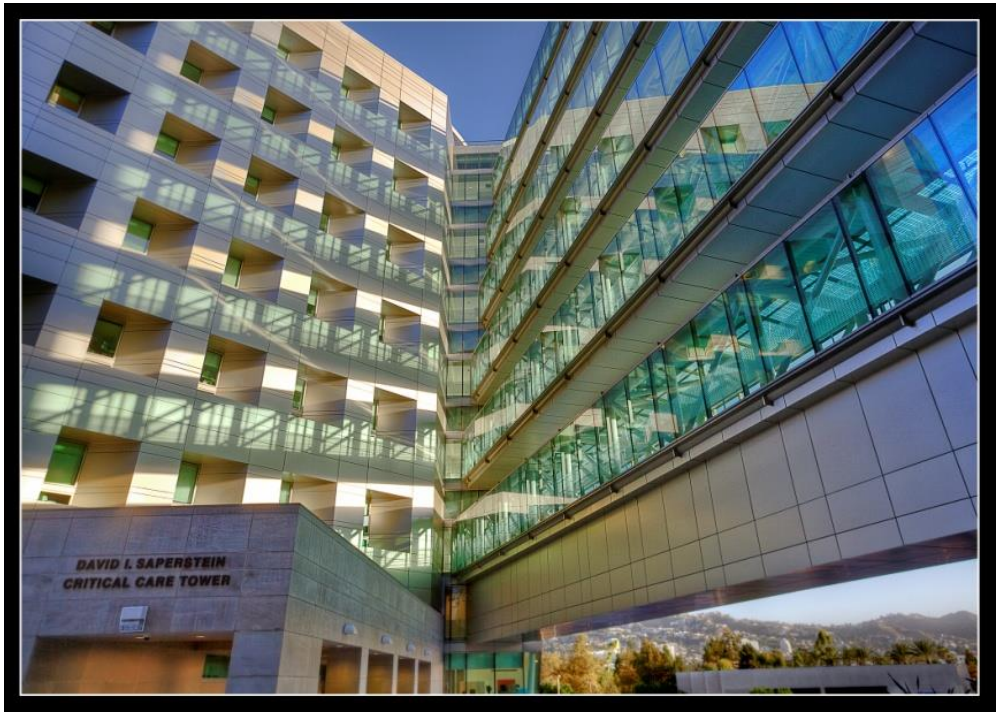
Spontaneous intracranial hypotension
The first 1111 patients: A practical approach



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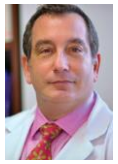
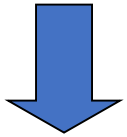


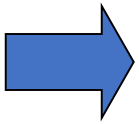
Nomenclature of Spontaneous Intracranial Hypotension

- Aliquorrhoea (Schaltenbrand 1936)

L'hypotension du CSF (Leriche 1917)

- Hypoliquorrhoea

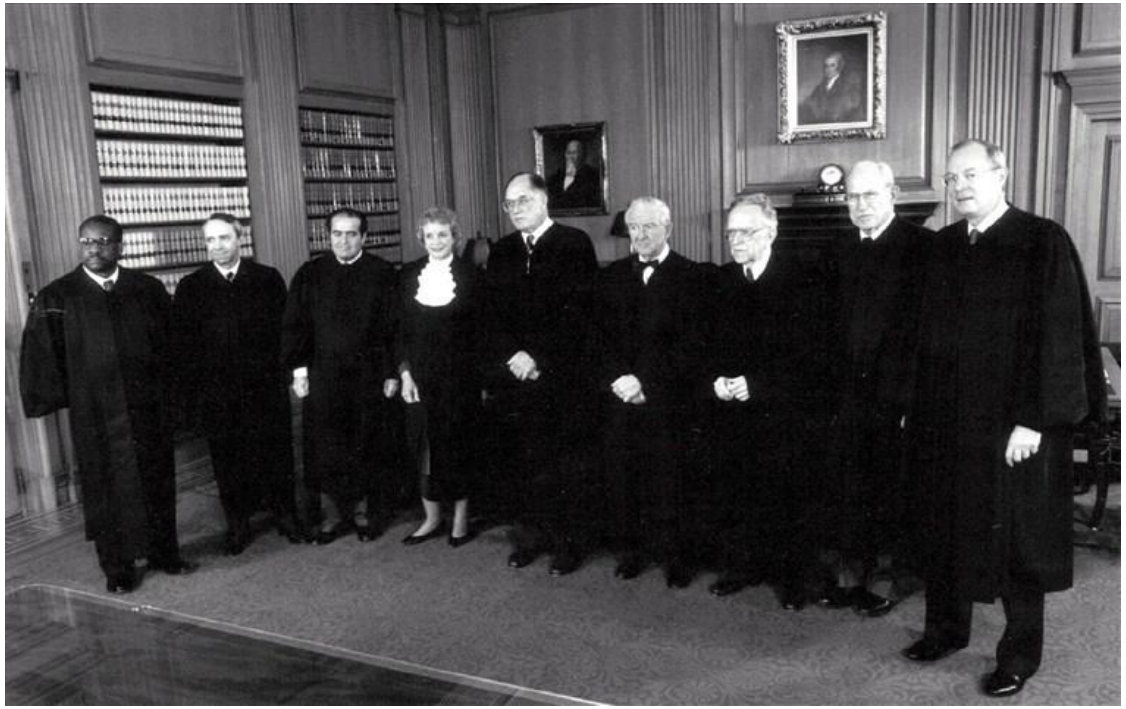


- Intracranial hypotension  CSF hypovolemia

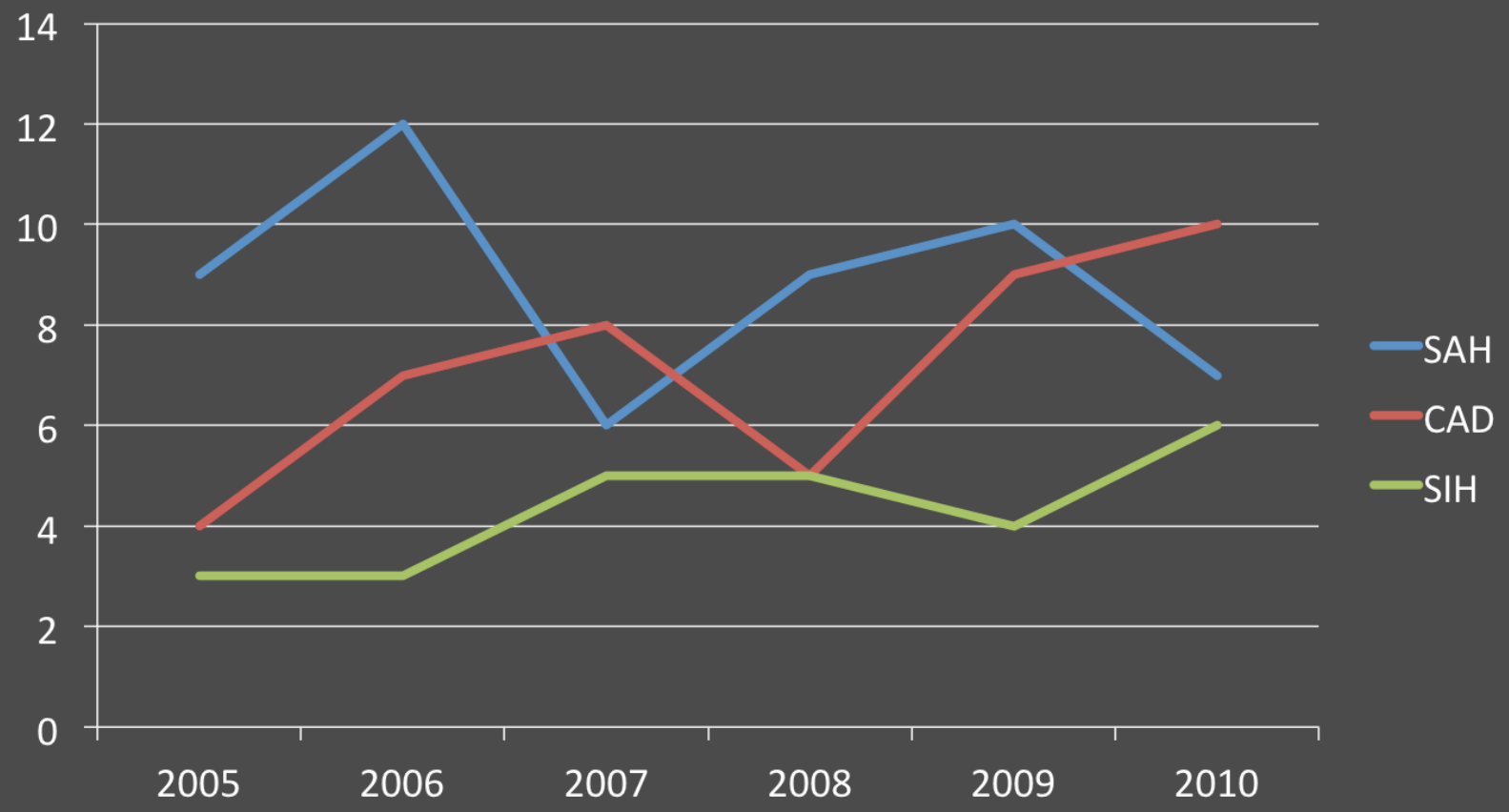
- Spontaneous spinal CSF leak

Spontaneous intracranial hypotension November 1991

November 1991

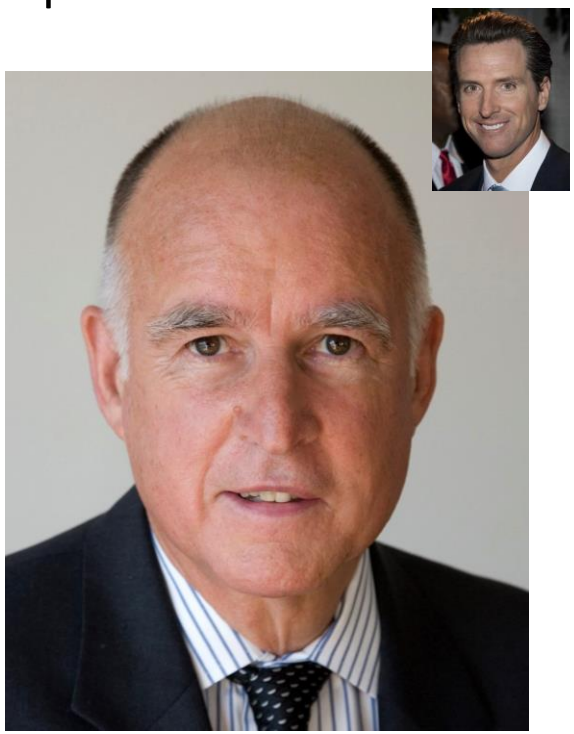


“Epidemiology” of SAH, CAD, and SIH – Cedars-Sinai Emergency Department (2005-2010)

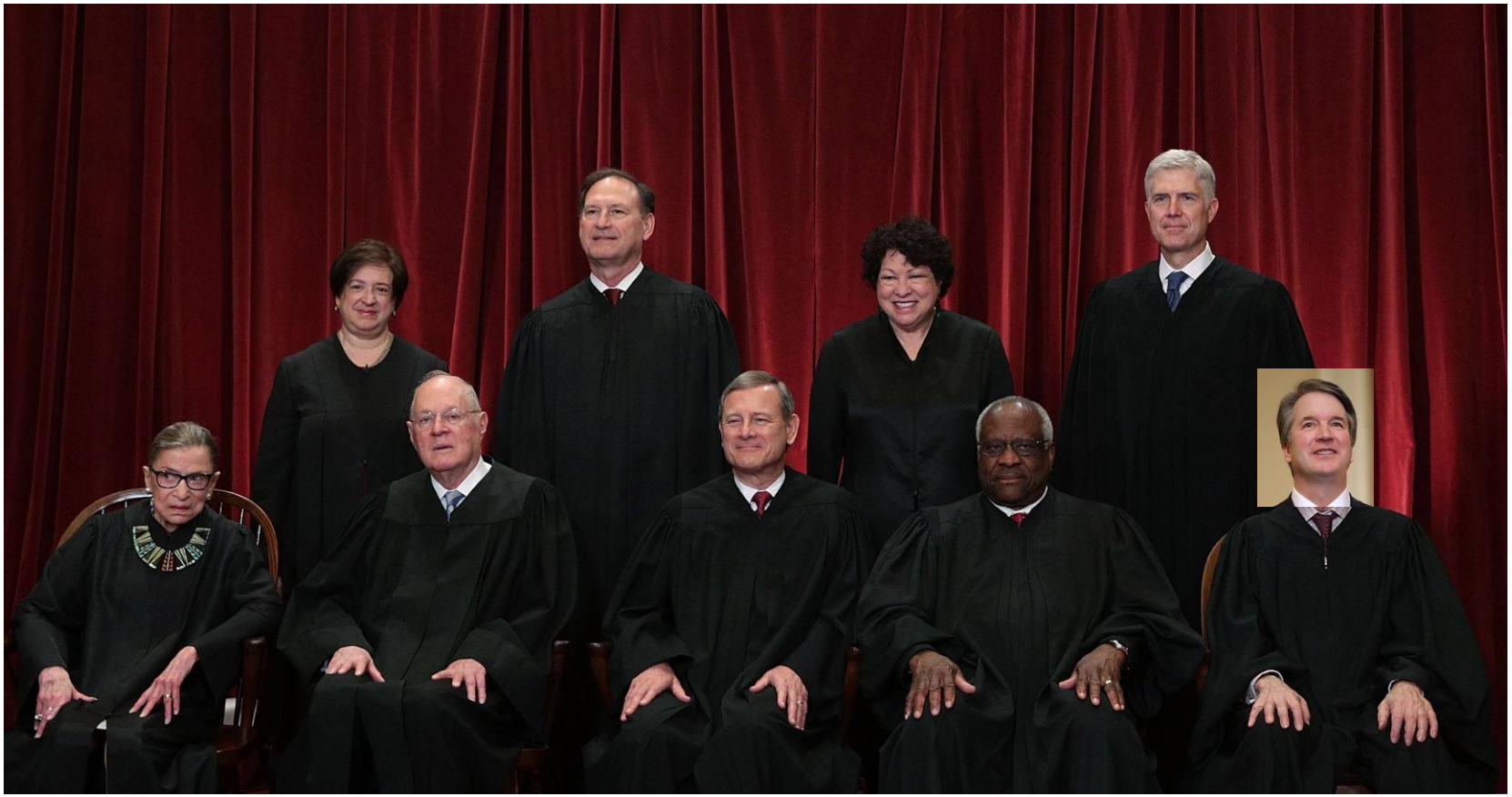


Spontaneous Intracranial Hypotension October 2018

- Routine practice?



October 2018



Spontaneous intracranial hypotension
Cedars-Sinai Medical Center (1-1-2001 to 8-31-2018)

2050 patients evaluated for CSF leak

Spontaneous intracranial hypotension
Cedars-Sinai Medical Center (1-1-2001 to 8-31-2018)

2050 patients evaluated for “CSF leak”

1111 patients met ICHD-III criteria for SIH:

- CSF leak on spinal imaging
- Brain MRI with sagging/meningeal enhancement/SDH
- Opening pressure less than 6.0 cm H_2O

Cephalalgia
An International Journal of Headache

Working group on headache attributed to non-vascular intracranial disorder:
DW Dodick, USA (Chairman) (Dodick.David@mayo.edu)
S Evers, Germany; D Friedman, USA; S Kirby, Canada; B Mokri, USA; J Pascual (Spain); M Peres, Brazil; A Purdy, Canada; K Ravishankar, India; P Sandor, Switzerland; WI Schievink, USA; R Stark, Australia; F Taylor, USA.

Spontaneous intracranial hypotension
Cedars-Sinai Medical Center (1-1-2001 to 8-31-2018)

2050 patients evaluated for “CSF leak”

1111 patients met ICHD-III criteria for SIH:

- CSF leak on spinal imaging
- Brain MRI with sagging/meningeal enhancement/SDH
- Opening pressure less than 6.0 cm H_2O

First 100 patients evaluated: 87 met ICHD-III criteria

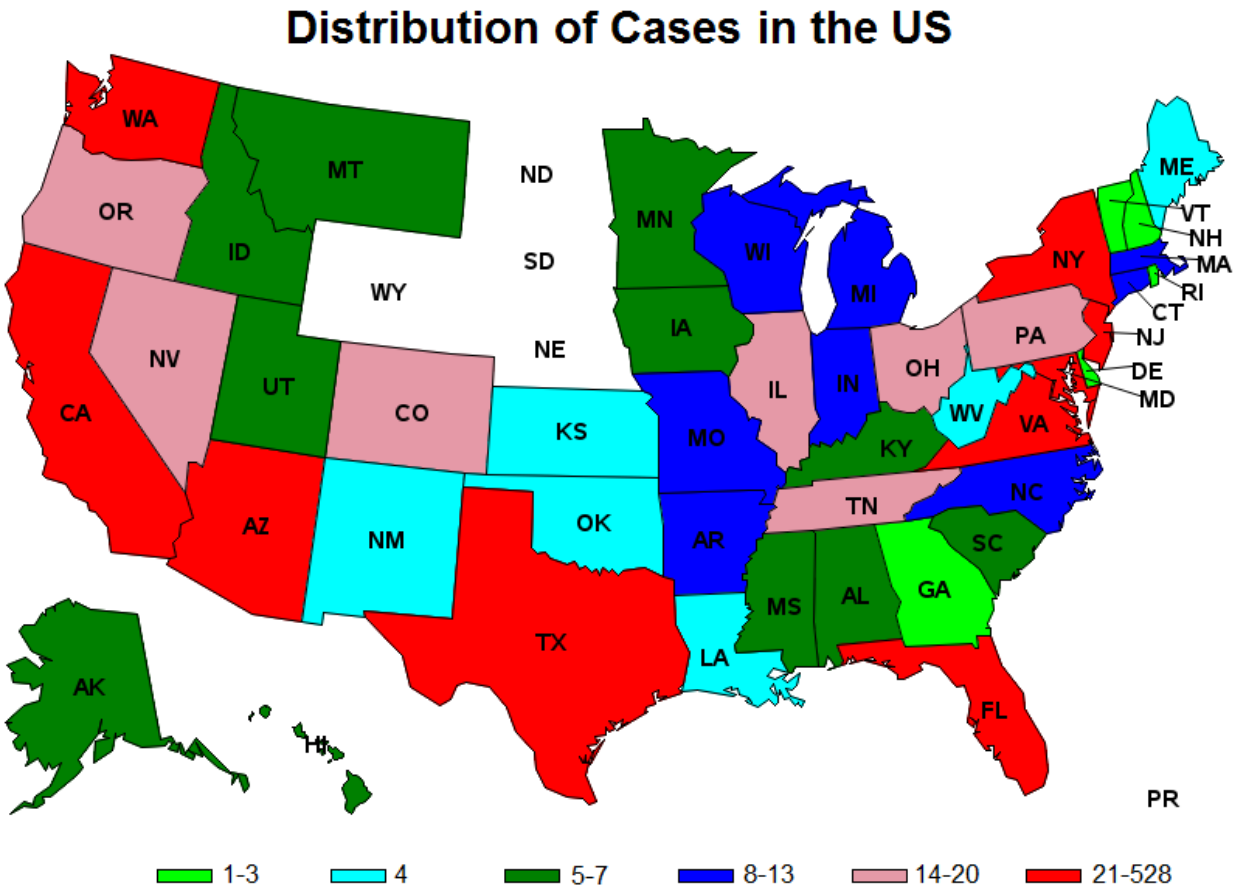
Last 100 patients evaluated: 32 met ICHD-III criteria

Geographic distribution of SIH cases –
Cedars-Sinai Medical Center, Los Angeles, CA

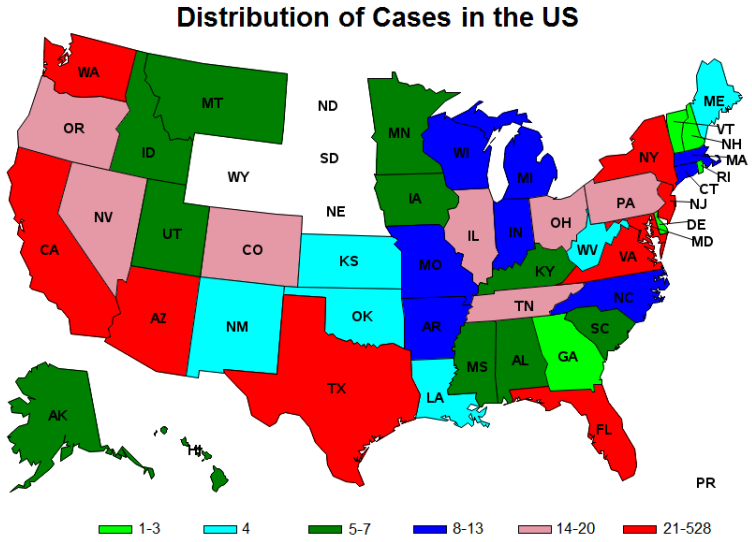
Summary of Cases by Patient Location (N=1111)

Variable	N (%)
Country	
US	1036 (93.2)
non-US	75 (6.8)
For US Cases Only	
LA County	253 (24.4)
California; Non-LA County	275 (26.6)

Geographic distribution of US SIH cases –
Cedars-Sinai Medical Center, Los Angeles, CA



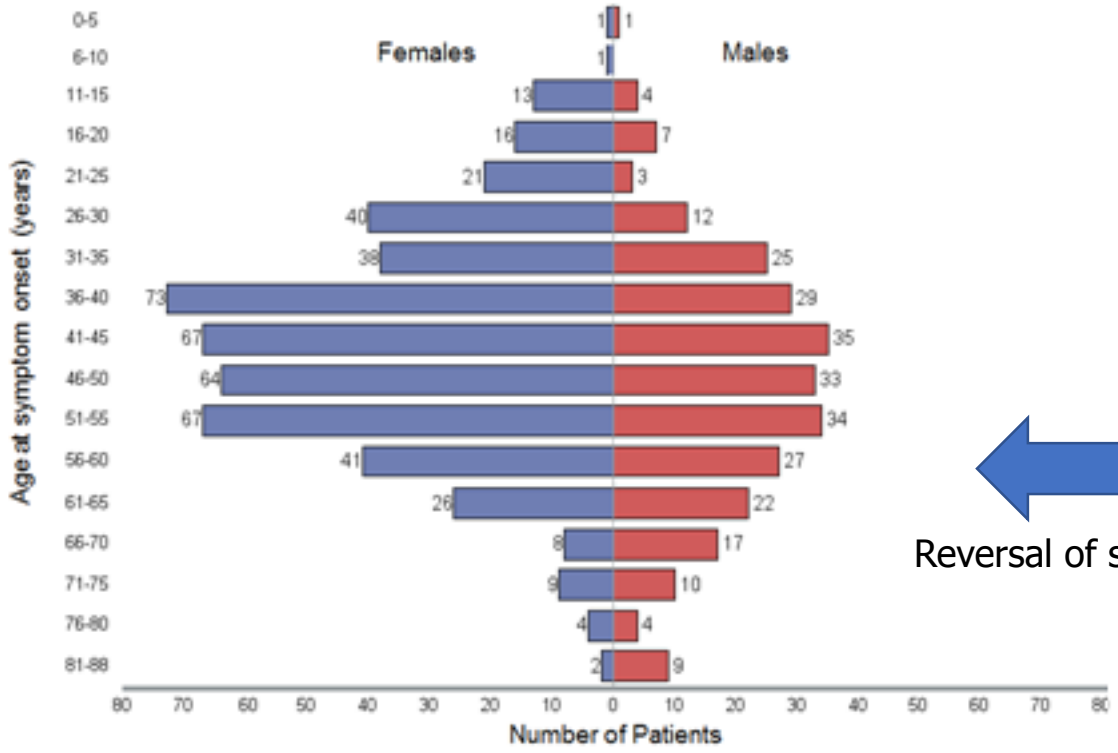
Geographic distribution of US SIH cases – Cedars-Sinai Medical Center, Los Angeles, CA



Age and Sex distribution in SIH (n=1111)

- **Mean age**: 45.6 years
- **Range**: 2 – 88 years
- **Sex**: 1.9:1 F:M ratio

Patients with spontaneous intracranial hypotension: Cedars-Sinai Medical Center



Reversal of sex predilection

J Neurosurg 96:483–489, 2002

Microfibrillopathy in spinal cerebrospinal fluid leaks

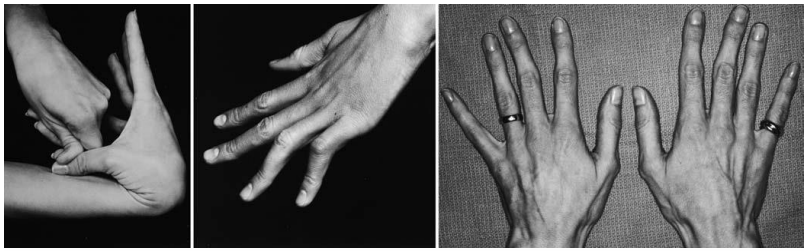


FIG. 1. Photographs demonstrating the hyperextensibility of the wrist and finger joints in one patient (Case 2; *left*) and long slender fingers in two other patients (Cases 3 and 4; *center* and *right*, respectively).

Microfibrillopathy in spinal cerebrospinal fluid leaks

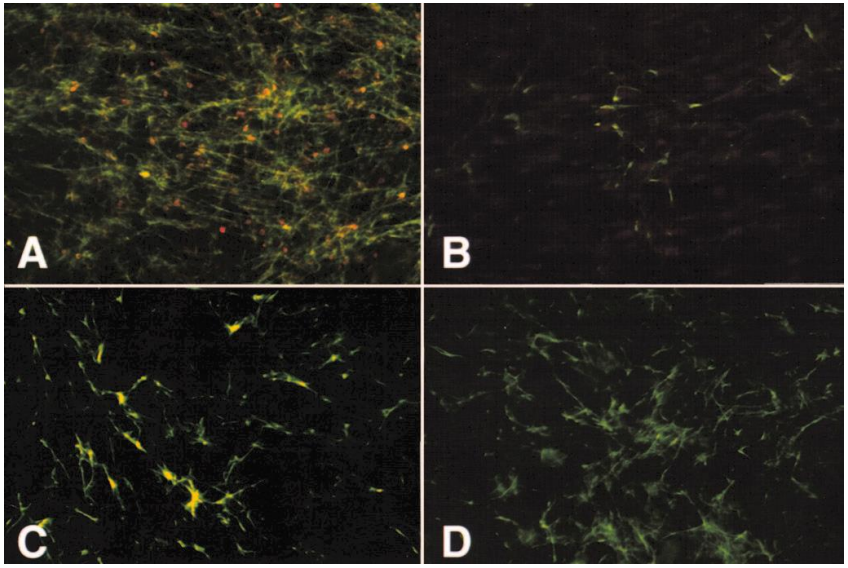


FIG. 4. Photomicrographs demonstrating fibrillin immunostaining of fibroblast cultures in patients whose clinical data are detailed in Table 1. A: Staining of fibroblast cultures from a control patient with a spontaneous spinal CSF leak but no other clinical manifestations of connective tissue abnormalities showing a prominent meshwork of fibrillin-immunostainable fibrils. This pattern is also seen in normal control fibroblasts. B: Case 2. A significant reduction in accumulation of fibrillin-stainable fibrils in cultured cells. This pattern is similar to that observed in more than 95% of people with the Marfan syndrome. C and D: Cases 1 (C) and 3 (D). Fibrillin immunostaining revealing moderate reductions in fibrillin-stainable fibrils. In neither case was the decrease reminiscent of the abnormal pattern seen in Marfan syndrome fibroblast cultures. Original magnification × 50.

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

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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

Connective tissue disorder: 4/20 = 20%

Connective tissue disorders in SIH: First 150 Cedars-Sinai patients

Marfan syndrome	5 / 150
Ehlers-Danlos syndrome III	3 / 150
Polycystic Kidney Disease	1 / 150
Marfan-like	24 / 150

33/150 (22%)

European Journal of Human Genetics (2013) 21, 386–390

www.nature.com/ejhg

ARTICLE

Connective tissue spectrum abnormalities associated with spontaneous cerebrospinal fluid leaks: a prospective study

Eyal Reinstein*,[1], Mitchel Pariani[1], Serguei Bannykh[2], David L Rimoin*,[1] and Wouter I Schievink[3]

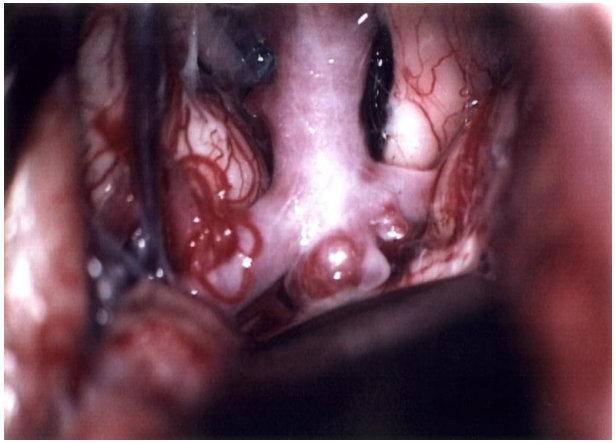
We aimed to assess the frequency of connective tissue abnormalities among patients with cerebrospinal fluid (CSF) leaks in a prospective study using a large cohort of patients. We enrolled a consecutive group of 50 patients, referred for consultation because of CSF leak. All patients have been carefully examined for the presence of connective tissue abnormalities, and based on findings, patients underwent genetic testing. Ancillary diagnostic studies included echocardiography, eye exam, and histopathological examinations of skin and dura biopsies in selected patients. We identified nine patients with heritable connective tissue disorders, including Marfan syndrome, Ehlers–Danlos syndrome and other unclassified forms. In seven patients, spontaneous CSF leak was the first noted manifestation of the genetic disorder. We conclude that spontaneous CSF leaks are associated with a spectrum of connective tissue abnormalities and may be the first noted clinical presentation of the genetic disorder. We propose that there is a clinical basis for considering spontaneous CSF leak as a clinical manifestation of heritable connective tissue disorders, and we suggest that patients with CSF leaks should be screened for connective tissue and vascular abnormalities.
European Journal of Human Genetics (2013) **21,** 386–390; doi:10.1038/ejhg.2012.191; published online 29 August 2012

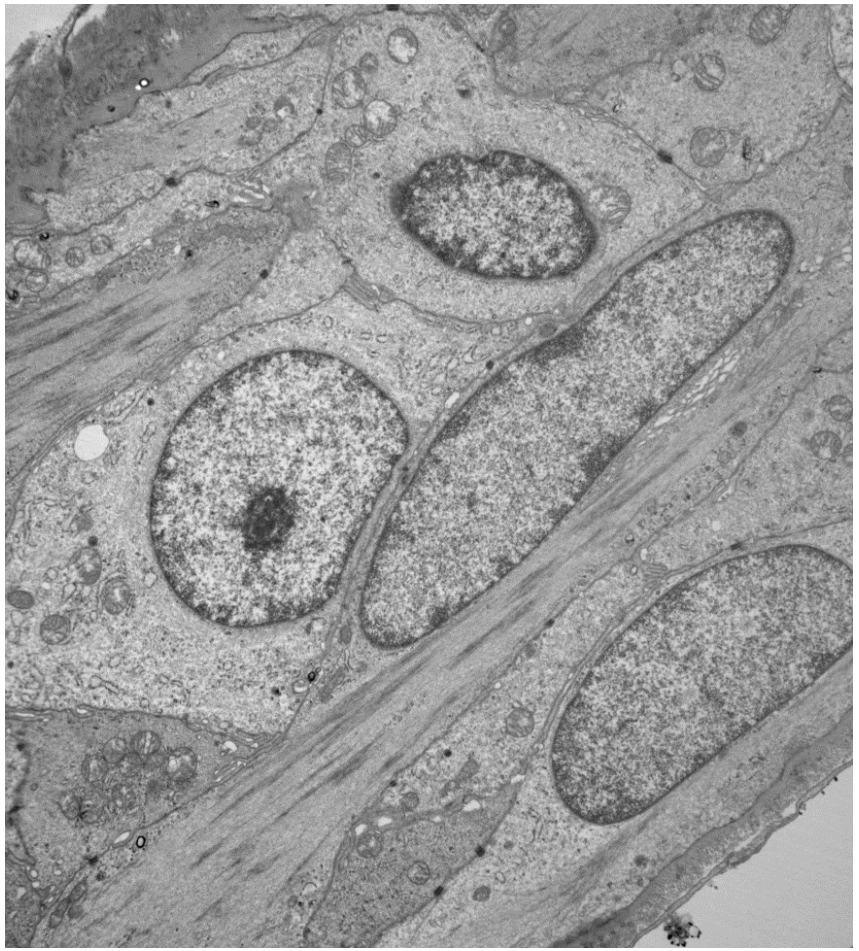
Keywords: hereditary disorders of connective tissue; spontaneous cerebrospinal fluid leak; positional headache screening

17/50 = 34%

Connective tissue disorders in spontaneous intracranial hypotension

- Abnormalities on EM in about 20% of patients
- Aortic dilatation in about 20% of patients
- Intracranial aneurysms in about 10% of patients
- Family history rare (<.5%)





Environmental factors

- More or less trivial trauma (30 - 50%)
- Seasonal variation (peak in spring)
- Bariatric surgery
- Spider bite

Clinical Manifestations

- 90 – 100% Headache
- 50 – 90% Neck Pain

- 25 – 75%
  - Nausea/vomiting
  - Hearing abnormalities
  - Light/noise sensitivity
  - Fatigue

- 1 – 25%
  - Diplopia (**CN VI** or III)
  - Cognitive decline/brain fog
  - Behavioral variant frontotemporal dementia
  - Myelopathy/radiculopathy
  - Tremors/Parkinsonism/ataxia
  - Coma

Headache in spontaneous intracranial hypotension

- Orthostatic headache (95+%)
- Non-positional headache
- Reverse orthostatic headache
- Exertional headache
- Valsalva-induced headache
- Head-shaking headache
- Latter half of the day headache
- Trigeminal neuralgia

Burden of headache

Epidemiology and comorbidity of headache

Rigmor Jensen, Lars J Stovner

Lancet Neurol 2008; 7: 354–61

Danish Headache Center, Department of Neurology, University of Copenhagen, Glostrup Hospital, DK-2600 Glostrup, Denmark (R Jensen MD); and Norwegian National Headache Centre, Department of Neuroscience, Norwegian University of Science and Technology and St Olavs Hospital, Trondheim,

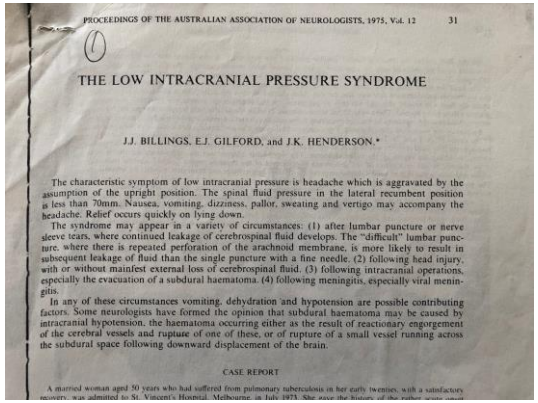
The burden associated with headache is a major public health problem, the true magnitude of which has not been fully acknowledged until now. Globally, the percentage of the adult population with an active headache disorder is 47% for headache in general, 10% for migraine, 38% for tension-type headache, and 3% for chronic headache that lasts for more than 15 days per month. The large costs of headache to society, which are mostly indirect through loss of work time, have been reported. On the individual level, headaches cause disability, suffering, and loss of quality of life that is on a par with other chronic disorders. Most of the burden of headache is carried by a minority who have substantial and complicating comorbidities. Renewed recognition of the burden of headache and increased scientific interest have led to a better understanding of the risk factors and greater insight into the pathogenic mechanisms, which might lead to improved prevention strategies and the early identification of patients who are at risk.

10 million US with chronic, daily, headache

Burden of spontaneous intracranial hypotension

Burden of disease	Patient
	Family
	Care giver
	Co-workers
	Teachers
Economic burden	Treatment costs
	Travel costs
	Disability
	Loss of wages

MRI findings in SIH



PROCEEDINGS OF THE AUSTRALIAN ASSOCIATION OF NEUROLOGISTS, 1975, Vol. 12 31

THE LOW INTRACRANIAL PRESSURE SYNDROME

J.J. BILLINGS, E.J. GILFORD, and J.K. HENDERSON.*

The characteristic symptom of low intracranial pressure is headache which is aggravated by the assumption of the upright position. The spinal fluid pressure in the lateral recumbent position is less than 70mm. Nausea, vomiting, dizziness, pallor, sweating and vertigo may accompany the headache. Relief occurs quickly on lying down.

The syndrome may appear in a variety of circumstances: (1) after lumbar puncture or nerve sleeve tears, where continued leakage of cerebrospinal fluid develops. The "difficult" lumbar puncture, where there is repeated perforation of the arachnoid membrane, is more likely to result in subsequent leakage of fluid than the single puncture with a fine needle. (2) following head injury, with or without mainfest external loss of cerebrospinal fluid. (3) following intracranial operations, especially the evacuation of a subdural haematoma. (4) following meningitis, especially viral meningitis.

In any of these circumstances vomiting, dehydration and hypotension are possible contributing factors. Some neurologists have formed the opinion that subdural haematoma may be caused by intracranial hypotension, the haematoma occurring either as the result of reactionary engorgement of the cerebral vessels and rupture of one of these, or of rupture of a small vessel running across the subdural space following downward displacement of the brain.

CASE REPORT

A married woman aged 30 years who had suffered from pulmonary tuberculosis in her early twenties, with a satisfactory

[Proceedings of the Australian Association of Neurologists.](#)

by Australian Association of Neurologists.

Billings et al 1975 Brain sagging

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

Abstract

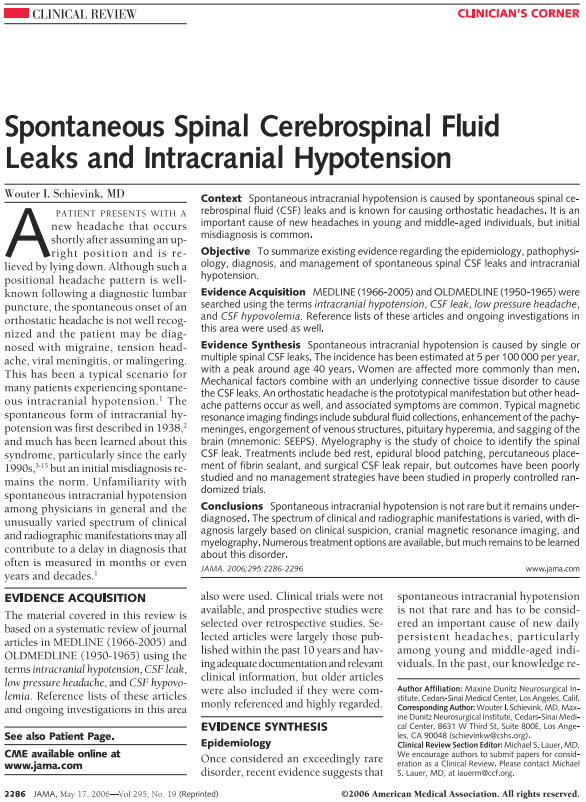
Abstracts: Poster session

First published: August 1991 | https://doi.org/10.1002/ana.410300232 | Cited by: 4

Mokri et al 1991 Meningeal enhancement

MRI findings

- S Subdural fluid collection
- E Enhancement of meninges
- E Engorgement of veins
- P Pituitary hyperemia
- S Sagging of brain



CLINICAL REVIEW

CLINICIAN'S CORNER

Spontaneous Spinal Cerebrospinal Fluid Leaks and Intracranial Hypotension

Wouter I. Schievink, MD

Context Spontaneous intracranial hypotension is caused by spontaneous spinal cerebrospinal fluid (CSF) leaks and is known for causing orthostatic headaches. It is an important cause of new headaches in young and middle-aged individuals, but initial misdiagnosis is common.

Objective To summarize existing evidence regarding the epidemiology, pathophysiology, diagnosis, and management of spontaneous spinal CSF leaks and intracranial hypotension.

Evidence Acquisition MEDLINE (1966-2005) and OLDMEDLINE (1950-1965) were searched using the terms *intracranial hypotension*, *CSF leak*, *low pressure headache*, and *CSF hypovolemia*. Reference lists of these articles and ongoing investigations in this area were used as well.

Evidence Synthesis Spontaneous intracranial hypotension is caused by single or multiple spinal CSF leaks. The incidence has been estimated at 5 per 100 000 per year, with a peak around age 40 years. Women are affected more commonly than men. Mechanical factors combine with an underlying connective tissue disorder to cause the CSF leaks. An orthostatic headache is the prototypical manifestation but other headache patterns occur as well, and associated symptoms are common. Typical magnetic resonance imaging findings include subdural fluid collections, enhancement of the pachymeninges, engorgement of venous structures, pituitary hyperemia, and sagging of the brain (mnemonic: SEEPS). Myelography is the study of choice to identify the spinal CSF leak. Treatments include bed rest, epidural blood patching, percutaneous placement of fibrin sealant, and surgical CSF leak repair, but outcomes have been poorly studied and no management strategies have been studied in properly controlled randomized trials.

Conclusions Spontaneous intracranial hypotension is not rare but it remains underdiagnosed. The spectrum of clinical and radiographic manifestations is varied, with diagnosis largely based on clinical suspicion, cranial magnetic resonance imaging, and myelography. Numerous treatment options are available, but much remains to be learned about this disorder.

JAMA. 2006;295:2286-2296 www.jama.com

A PATIENT PRESENTS WITH A new headache that occurs shortly after assuming an upright position and is relieved by lying down. Although such a positional headache pattern is well-known following a diagnostic lumbar puncture, the spontaneous onset of an orthostatic headache is not well recognized and the patient may be diagnosed with migraine, tension headache, viral meningitis, or malingering. This has been a typical scenario for many patients experiencing spontaneous intracranial hypotension.[1] The spontaneous form of intracranial hypotension was first described in 1938,[2] and much has been learned about this syndrome, particularly since the early 1990s,[3-15] but an initial misdiagnosis remains the norm. Unfamiliarity with spontaneous intracranial hypotension among physicians in general and the unusually varied spectrum of clinical and radiographic manifestations may all contribute to a delay in diagnosis that often is measured in months or even years and decades.[1]

EVIDENCE ACQUISITION

The material covered in this review is based on a systematic review of journal articles in MEDLINE (1966-2005) and OLDMEDLINE (1950-1965) using the terms *intracranial hypotension*, *CSF leak*, *low pressure headache*, and *CSF hypovolemia*. Reference lists of these articles and ongoing investigations in this area also were used. Clinical trials were not available, and prospective studies were selected over retrospective studies. Selected articles were largely those published within the past 10 years and having adequate documentation and relevant clinical information, but older articles were also included if they were commonly referenced and highly regarded.

See also Patient Page.

CME available online at www.jama.com

EVIDENCE SYNTHESIS

Epidemiology

Once considered an exceedingly rare disorder, recent evidence suggests that spontaneous intracranial hypotension is not that rare and has to be considered an important cause of new daily persistent headaches, particularly among young and middle-aged individuals. In the past, our knowledge re-

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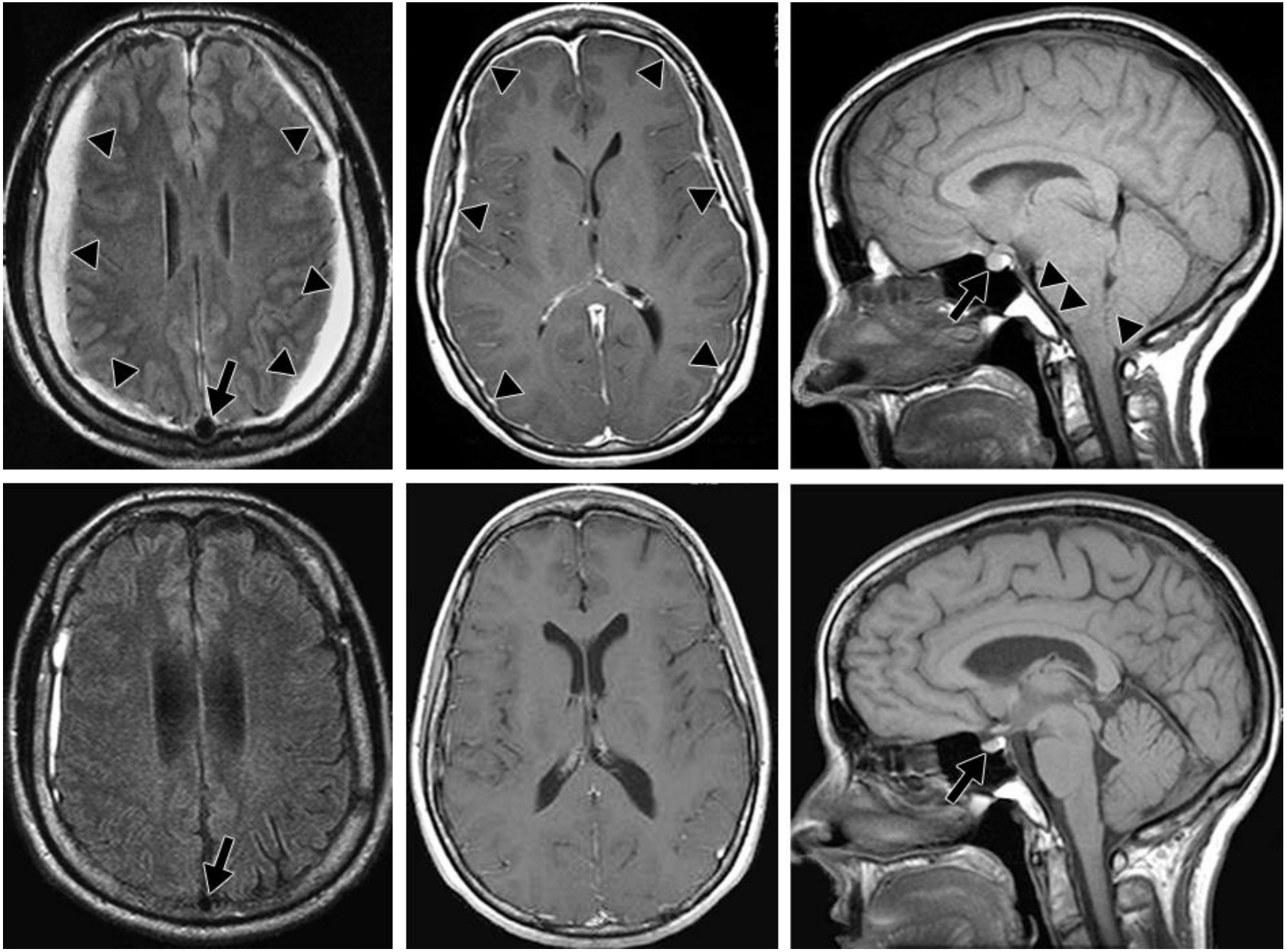
Clinical Review Section Editor: Michael S. Lauer, MD. We encourage authors to submit papers for consideration as a Clinical Review. Please contact Michael S. Lauer, MD, at lauerm@ccf.org.



MRI Findings - Caveats

- Normal MRI in approximately 20% of patients
- MRI often normalizes in spite of persistent CSF leak
- MRI may become abnormal during course of disease

Diagnostic evaluation of SIH – A practical approach

Towards a non-invasive diagnostic work up, i.e., no lumbar puncture and minimizing ionizing radiation

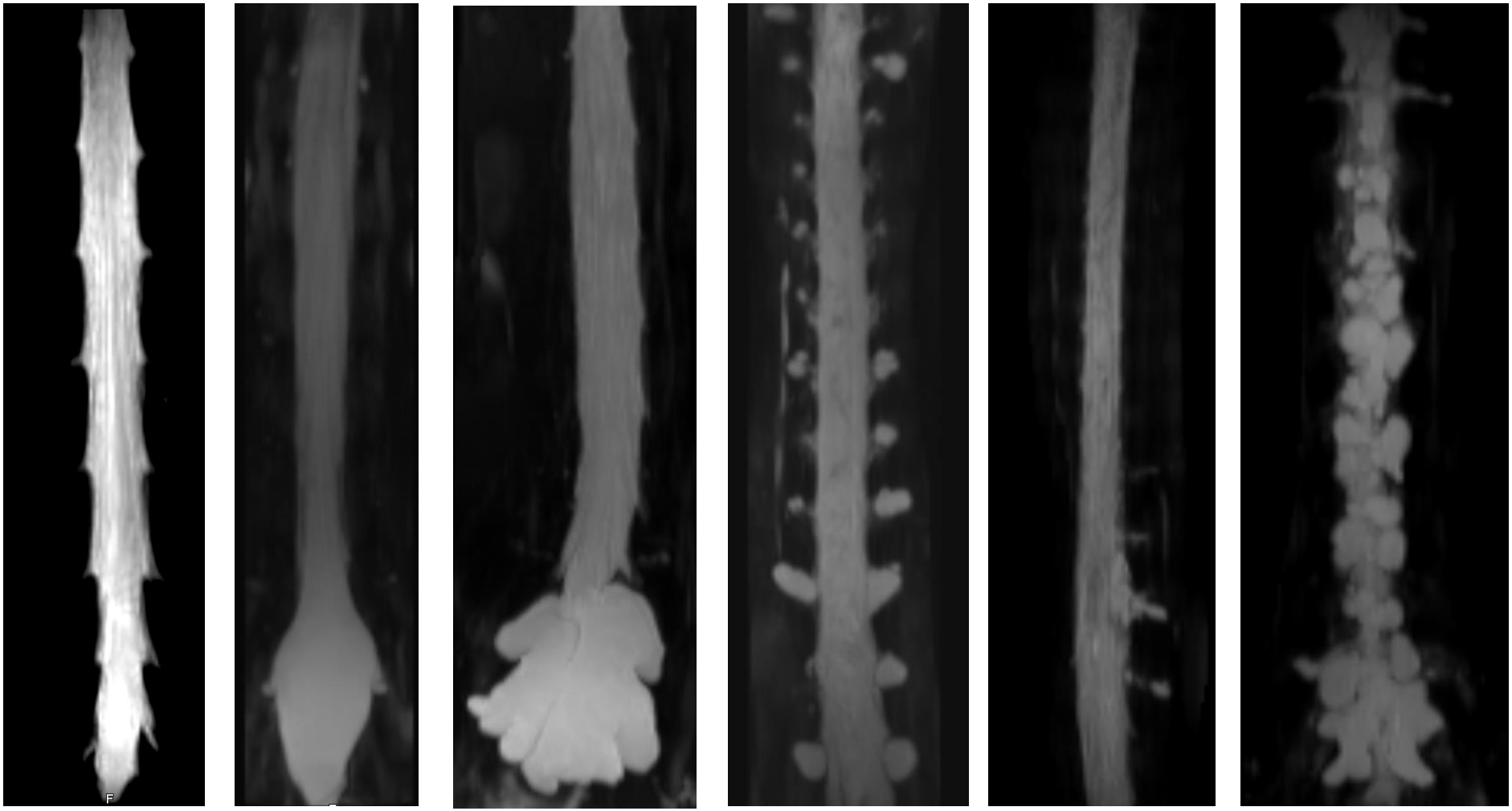
MRI brain

MRI spine/MRMyelography

Epidural blood patching

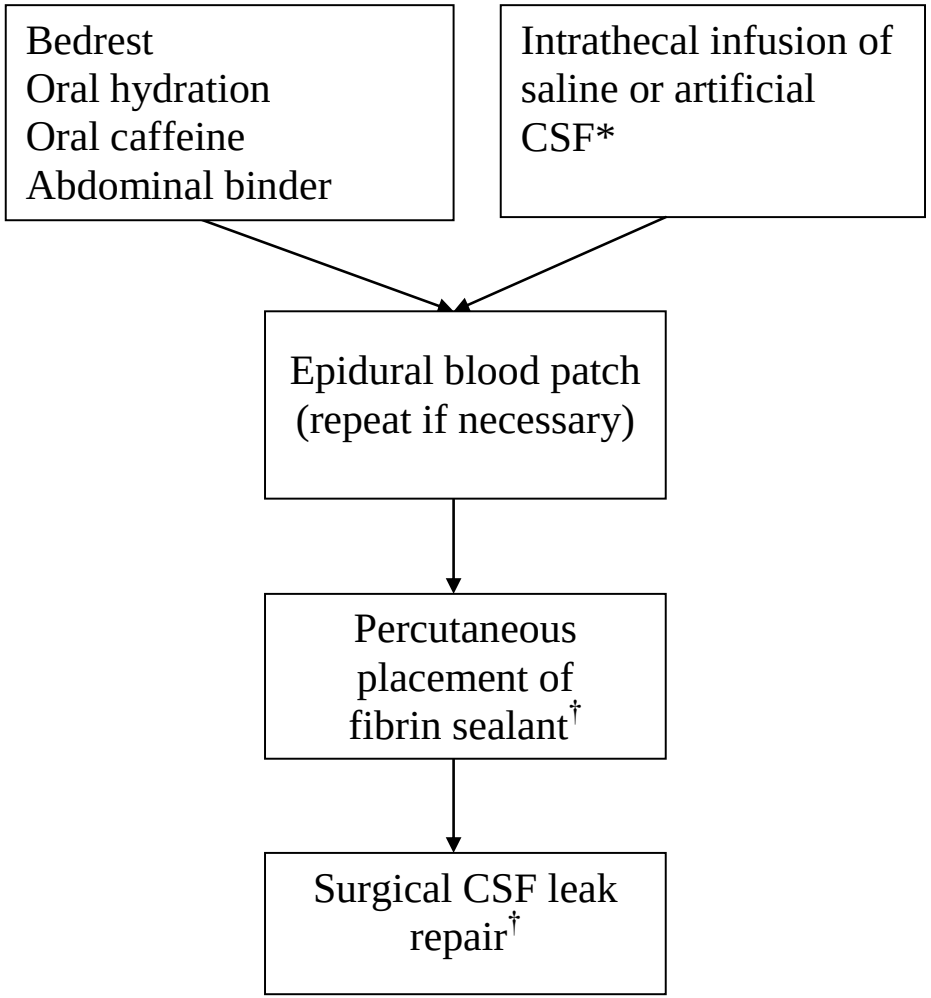
Percutaneous fibrin glue placement

Surgery (+for PDPH)



Detection of spinal CSF Leak – is it necessary?

- MRI/MR-myelography
- ~~Radionuclide Cisternography~~
- ~~CT-Myelography~~/Digital Subtraction Myelography (DSM)
- Dynamic CT-myelography
- Intrathecal gado-enhanced MRI



* When urgent treatment is required such as with coma†

Knowledge of exact site of CSF leak required

Advances in treatment for SIH

- Understanding the anatomy of CSF leaks

Through – advances in imaging
intraoperative observations

Are advances in care available to patients with spontaneous intracranial hypotension?

- Diagnostic delay
- Access to medical care
  - Insurance
  - Across state lines
  - Across national borders
- Appropriate systemic organization
- Appropriate reimbursement (e.g., DSM)

Cause of intracranial hypotension is

- **Spinal CSF leak**

- **Pooling of CSF**

- Inadequate CSF production?
- Rapid CSF absorption?

- ? CSF rhinorrhea–otorrhoea ?

NEVER!

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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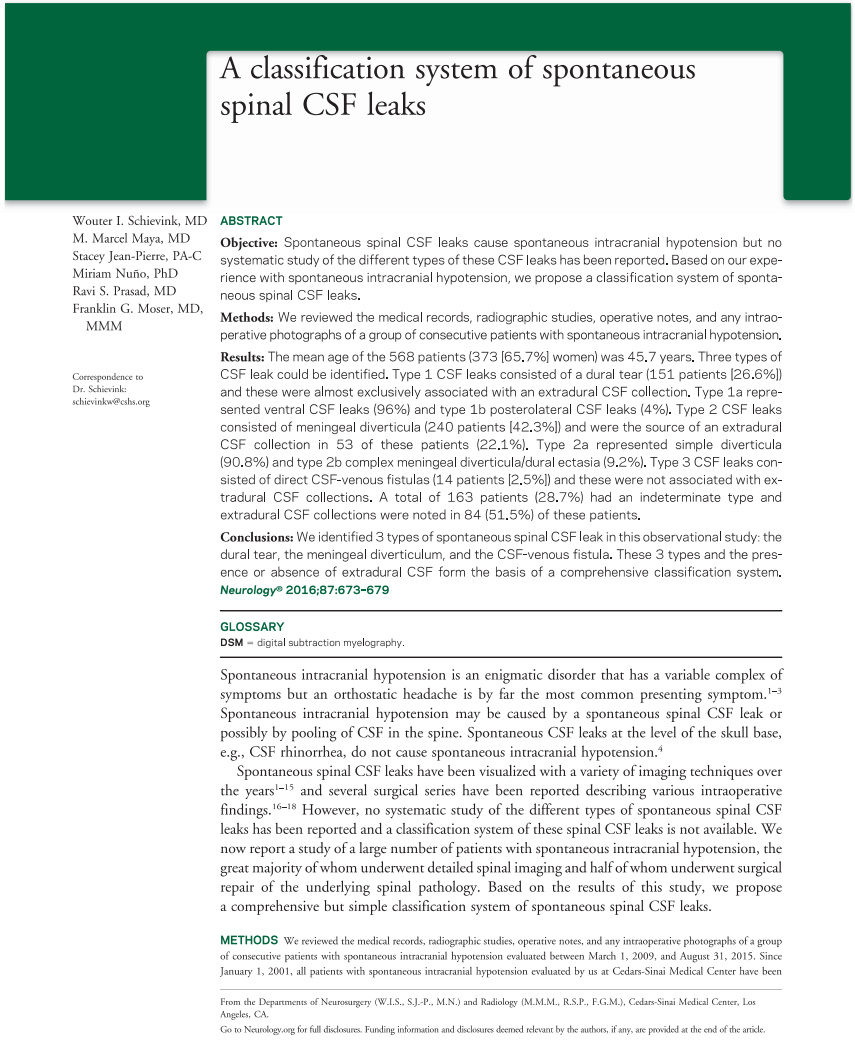
Classification of Spontaneous Spinal CSF Leaks

- Type 1: Dural tear
- Type 2: Arachnoid cyst
- Type 3: CSF-venous fistula

- Type 4: Indeterminate



A classification system of spontaneous spinal CSF leaks

Wouter I. Schievink, MD
M. Marcel Maya, MD
Stacey Jean-Pierre, PA-C
Miriam Nuño, PhD
Ravi S. Prasad, MD
Franklin G. Moser, MD, MMM

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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.

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Neurology® 2016;87:673–679

GLOSSARY

DSM = digital subtraction myelography.

Spontaneous intracranial hypotension is an enigmatic disorder that has a variable complex of symptoms but an orthostatic headache is by far the most common presenting symptom.[1–3] Spontaneous intracranial hypotension may be caused by a spontaneous spinal CSF leak or possibly by pooling of CSF in the spine. Spontaneous CSF leaks at the level of the skull base, e.g., CSF rhinorrhea, do not cause spontaneous intracranial hypotension.[4]

Spontaneous spinal CSF leaks have been visualized with a variety of imaging techniques over the years[1–15] and several surgical series have been reported describing various intraoperative findings.[16–18] However, no systematic study of the different types of spontaneous spinal CSF leaks has been reported and a classification system of these spinal CSF leaks is not available. We now report a study of a large number of patients with spontaneous intracranial hypotension, the great majority of whom underwent detailed spinal imaging and half of whom underwent surgical repair of the underlying spinal pathology. Based on the results of this study, we propose a comprehensive but simple classification system of spontaneous spinal CSF leaks.

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From the Departments of Neurosurgery (W.I.S., S.J.-P., M.N.) and Radiology (M.M.M., R.S.P., F.G.M.), Cedars-Sinai Medical Center, Los Angeles, CA.

Go to Neurology.org for full disclosures. Funding information and disclosures deemed relevant by the authors, if any, are provided at the end of the article.

Classification of Spontaneous Spinal CSF Leaks

- Type 1: Dural tear
- Type 2: Arachnoid cyst
- Type 3: CSF-venous fistula

- Type 4: Indeterminate

A classification system of spontaneous spinal CSF leaks

Wouter I. Schievink, MD
M. Marcel Maya, MD
Stacey Jean-Pierre, PA-C
Miriam Nuño, PhD
Ravi S. Prasad, MD
Franklin G. Moser, MD, MMM

Correspondence to Dr. Schievink: schievinkw@cshs.org

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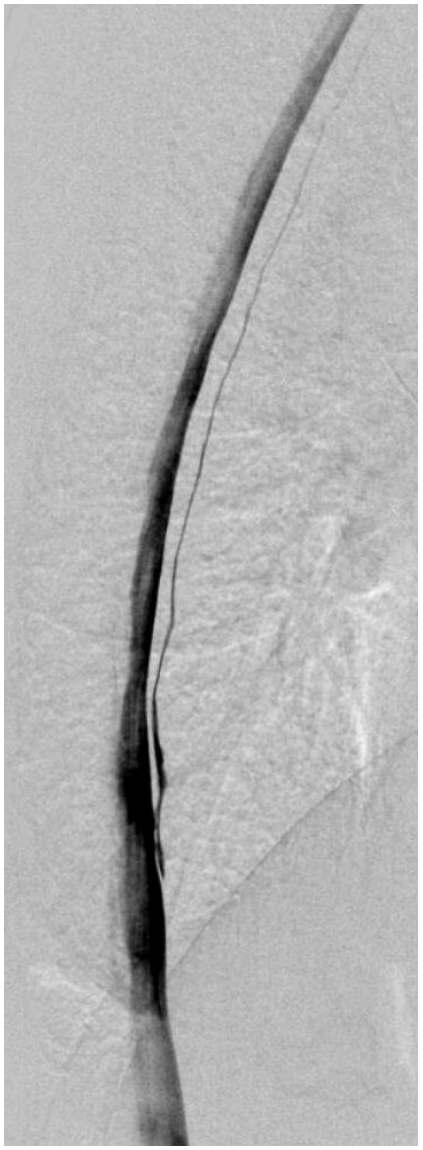
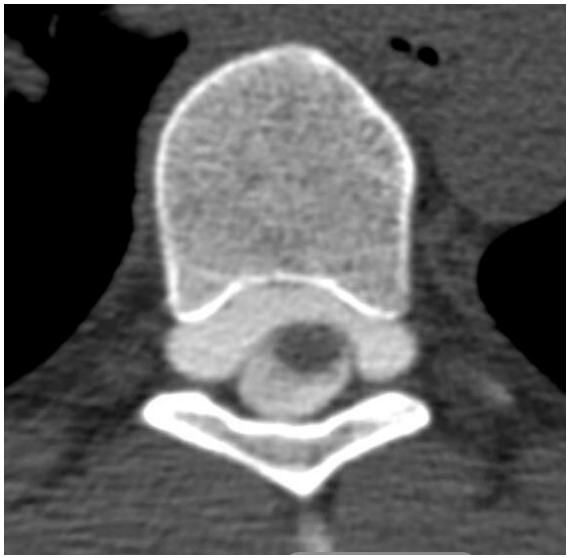
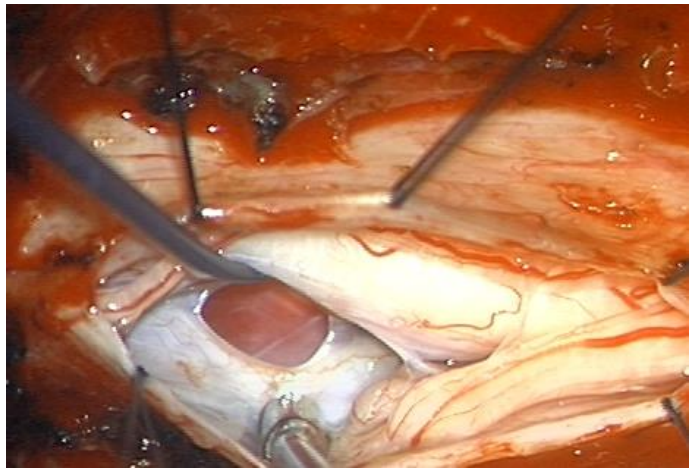
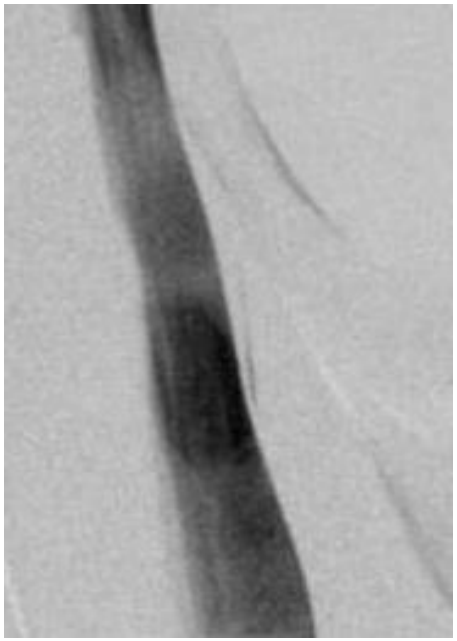
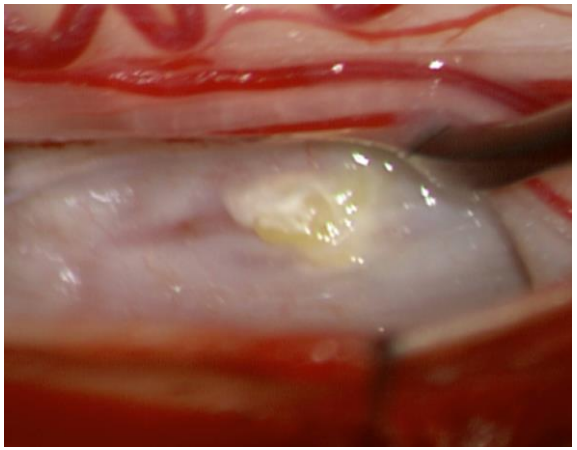


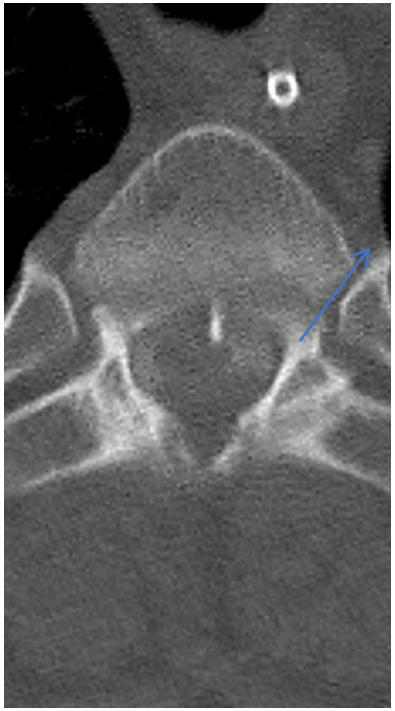


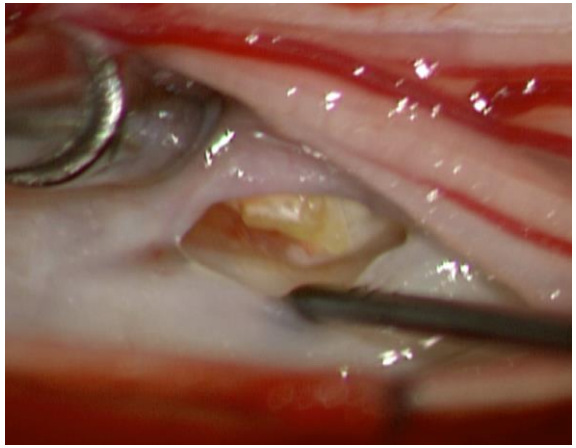


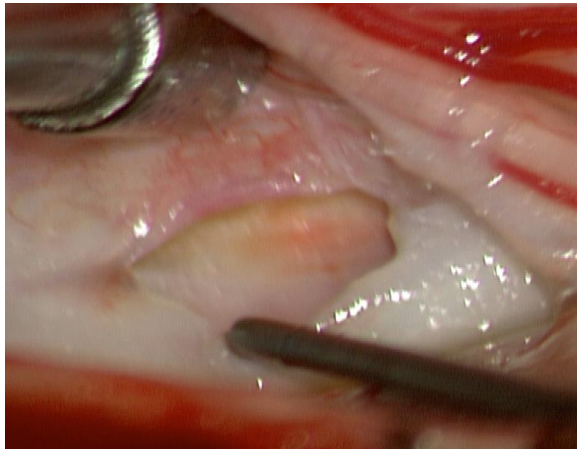
Type 1a

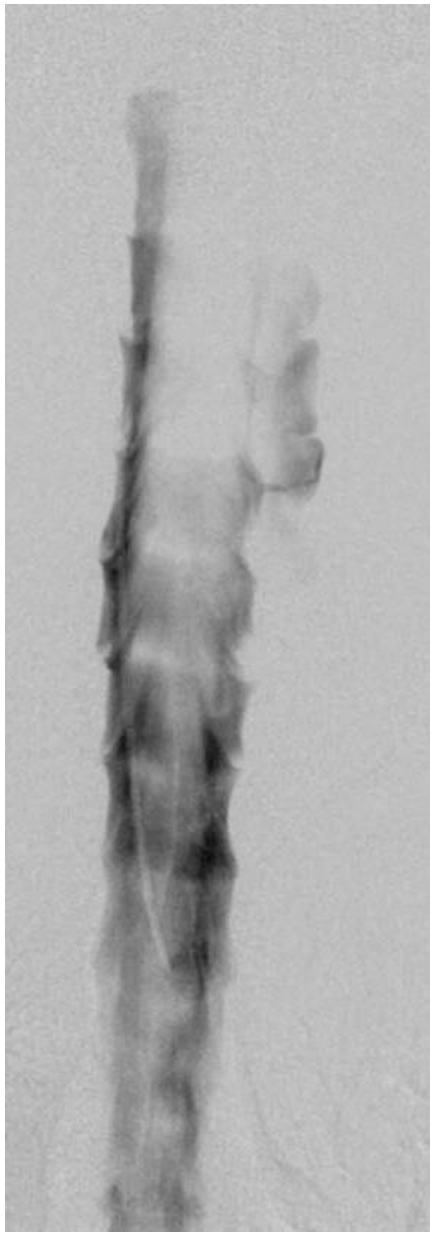


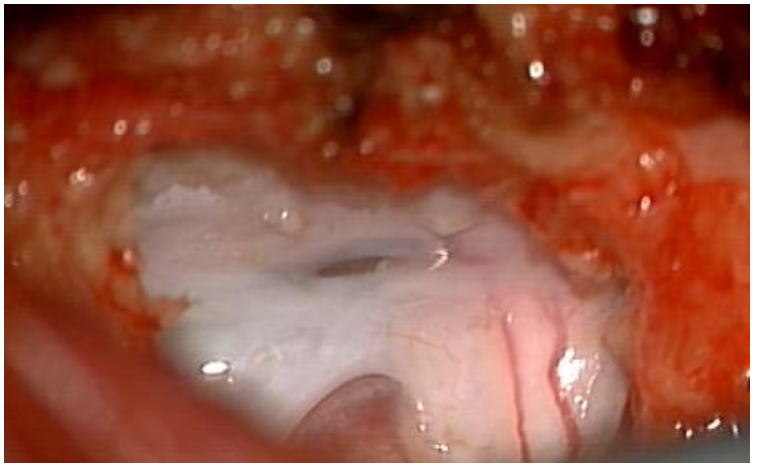












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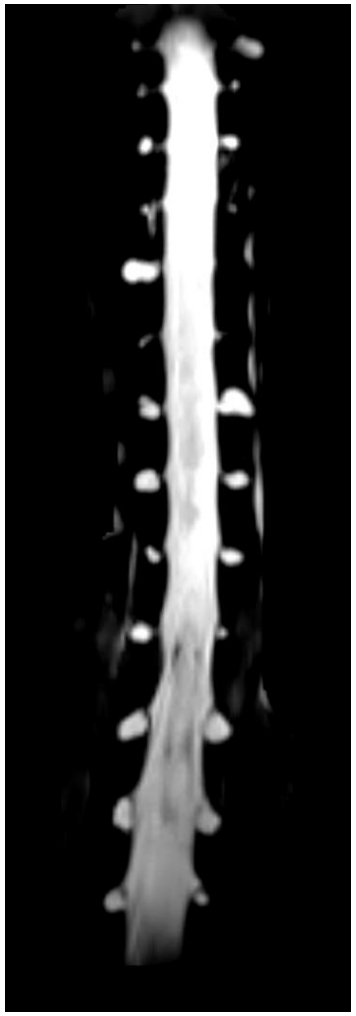

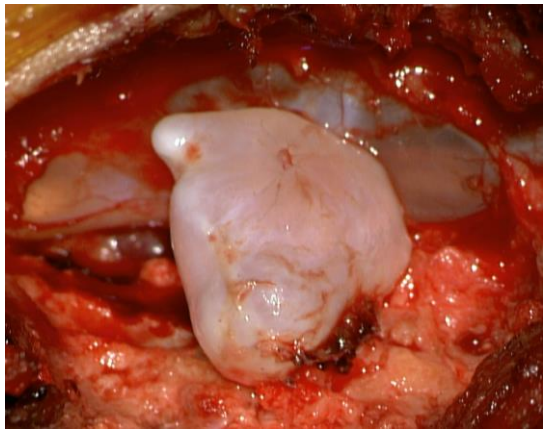

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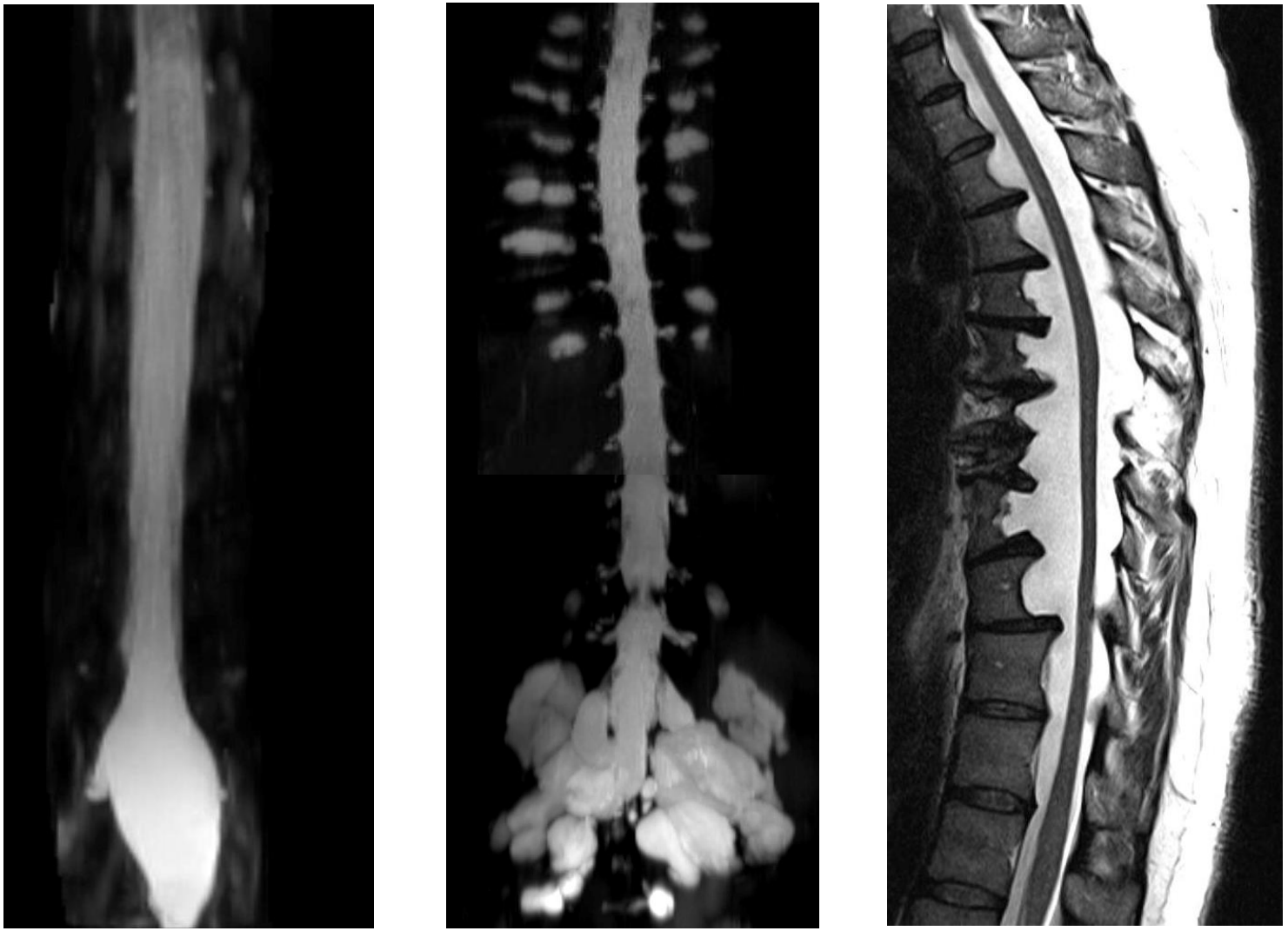

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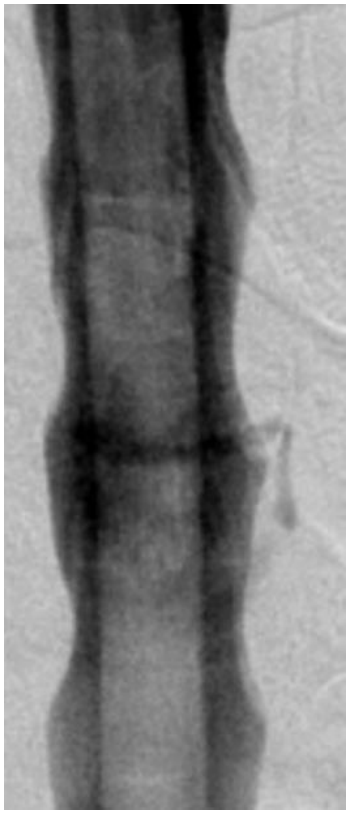
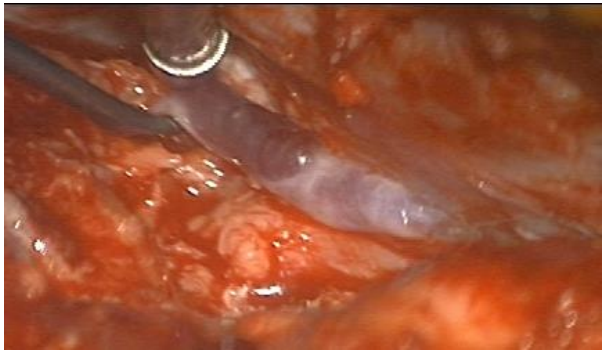

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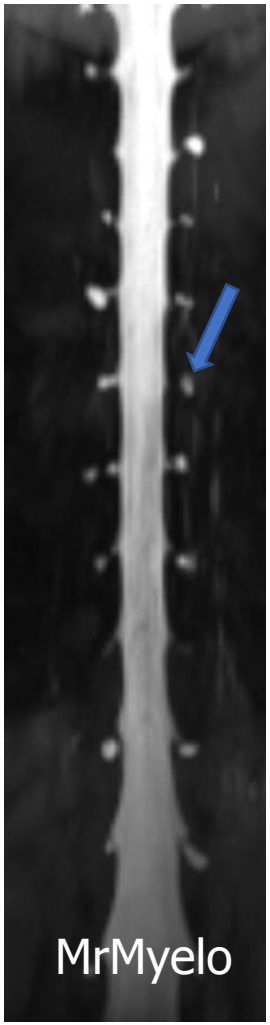
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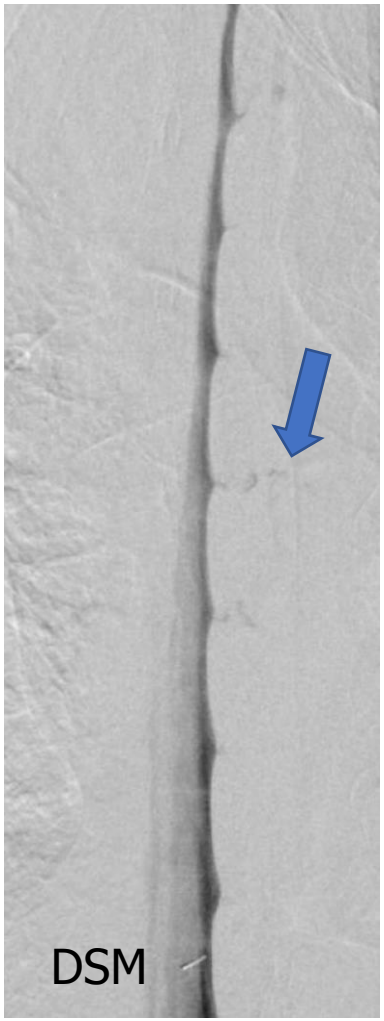
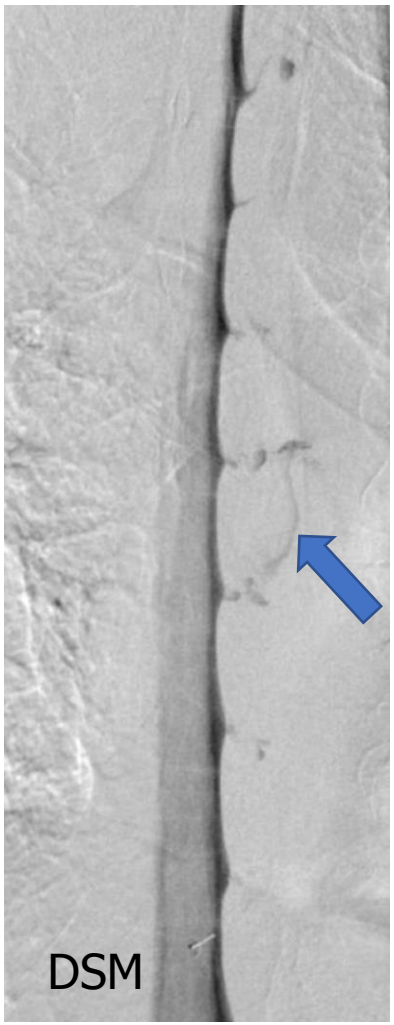
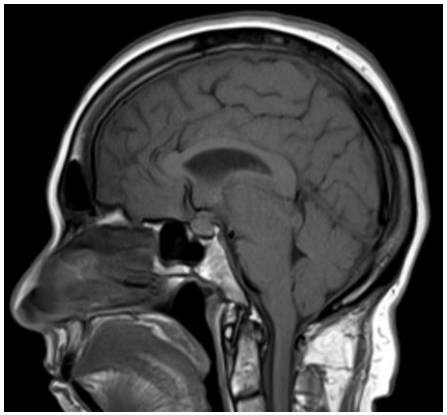
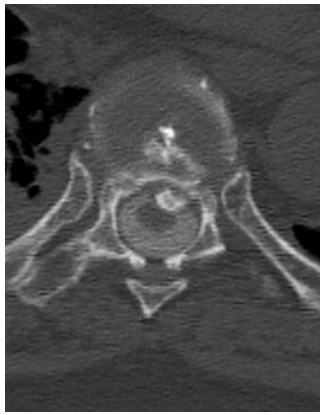
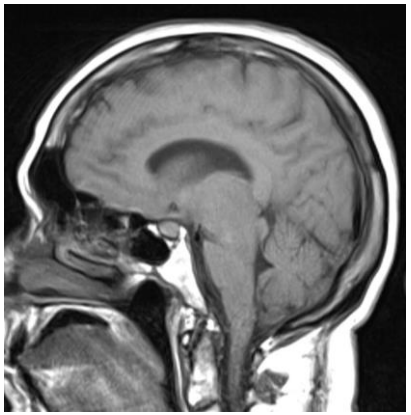
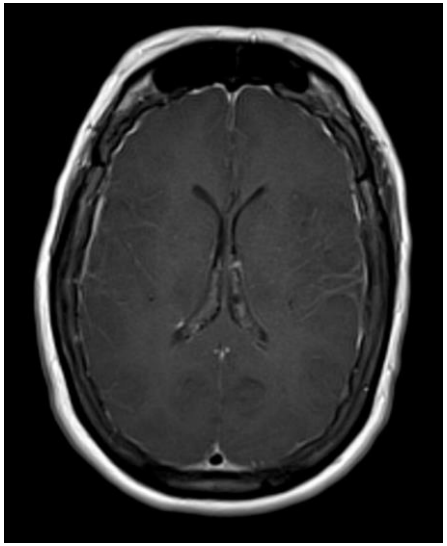

DSM

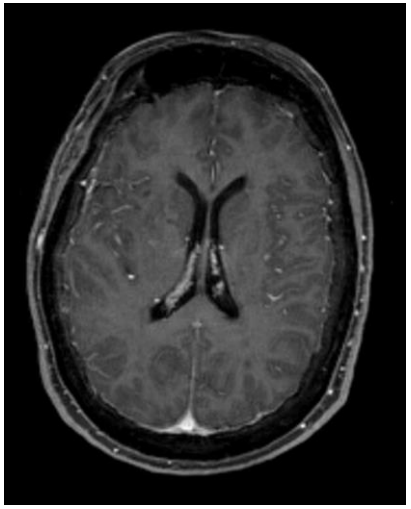

DSM


DSM

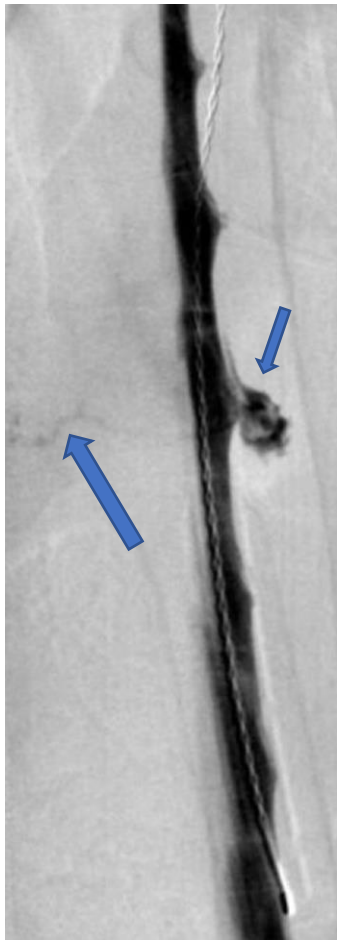
Not from disc Not from largest cyst

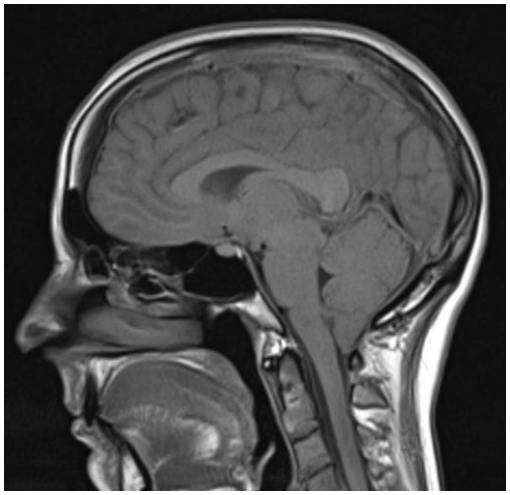



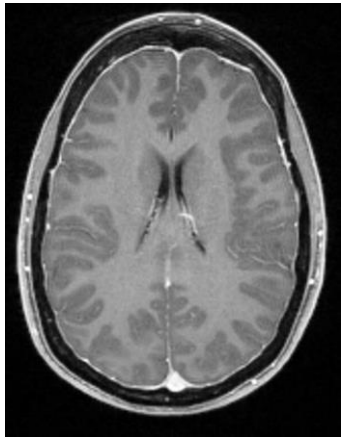



Pre-op DSM: CSF-venous fistula Post-op

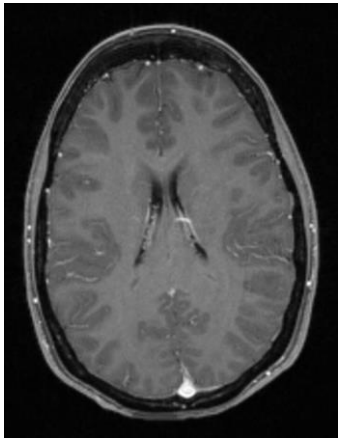


Pre-op

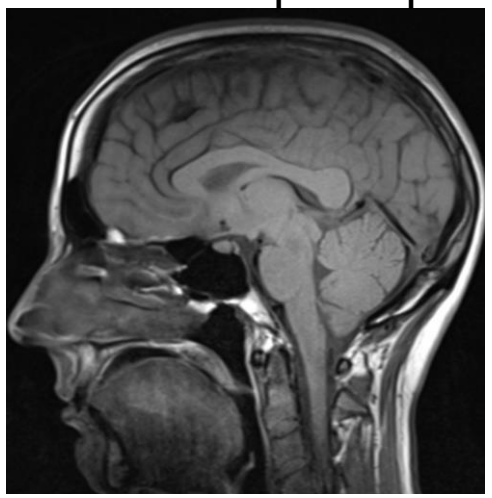


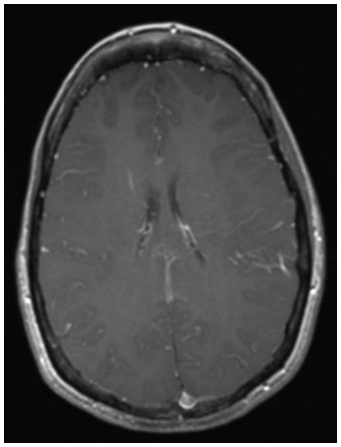


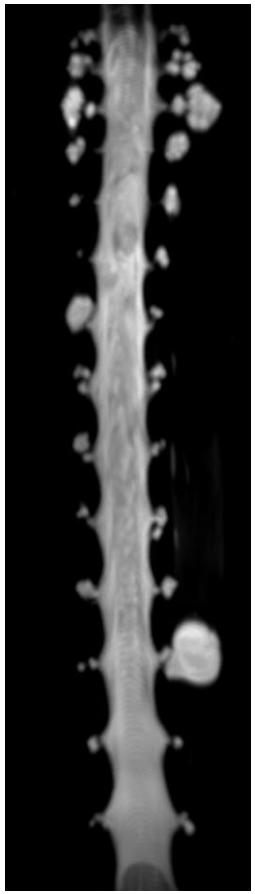
48 hours Post-op

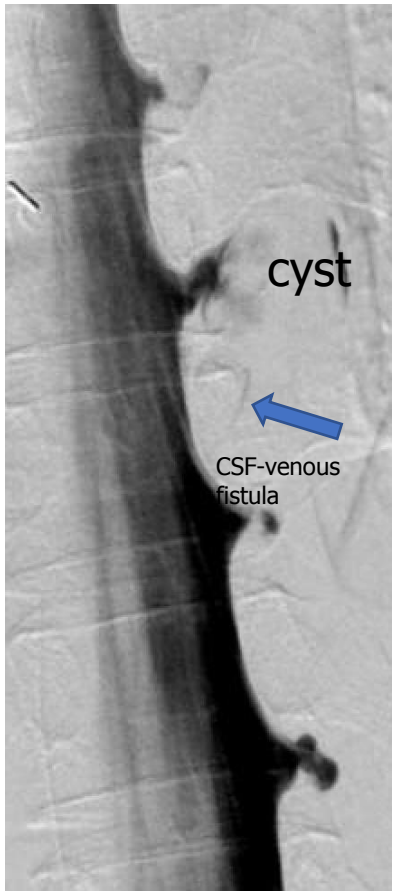


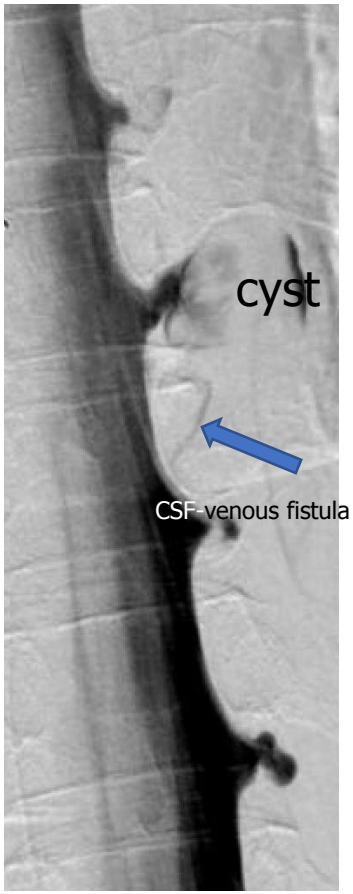
1 month post-op



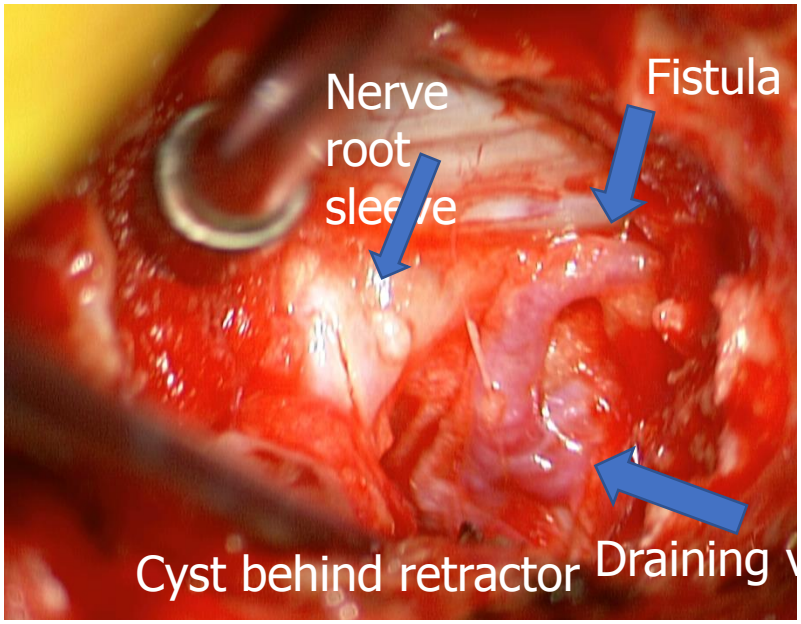








MRmyelogram DSM DSM



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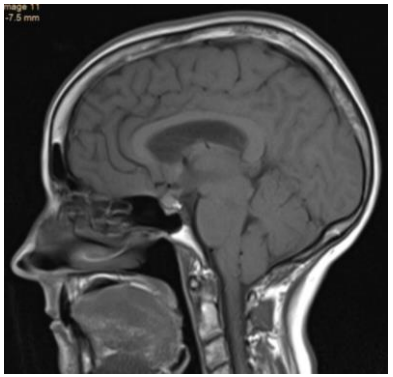
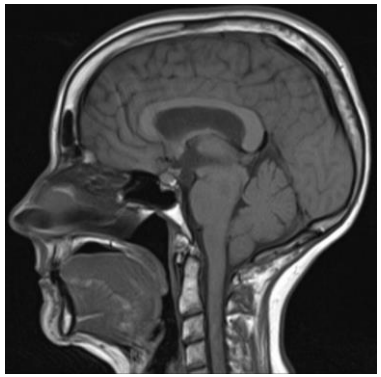
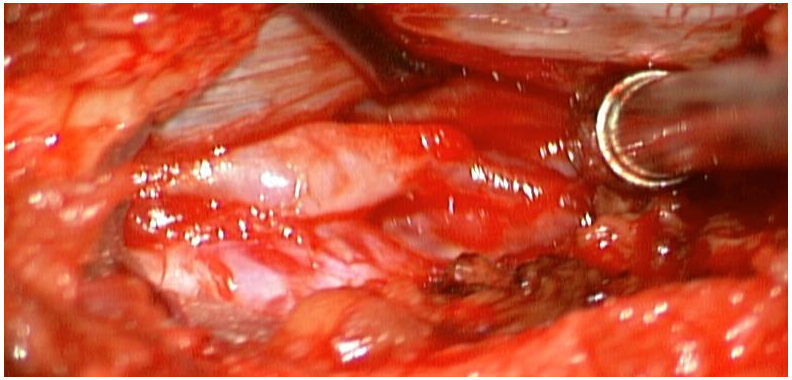
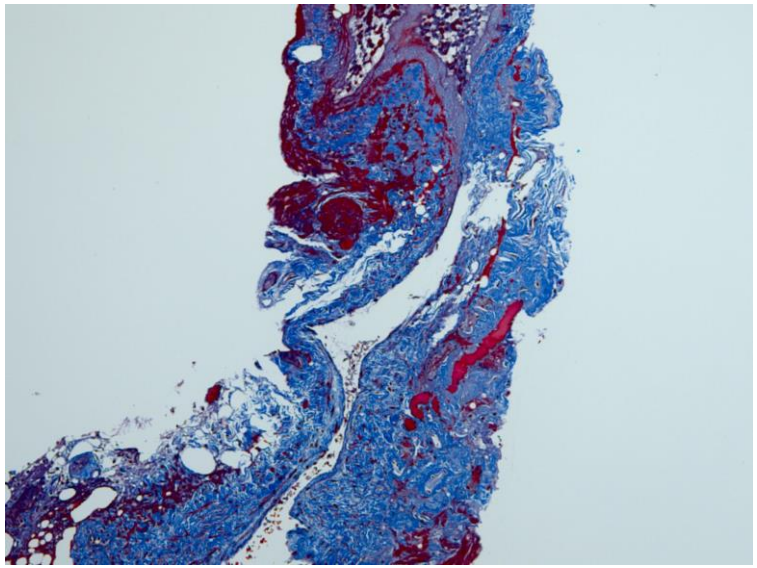


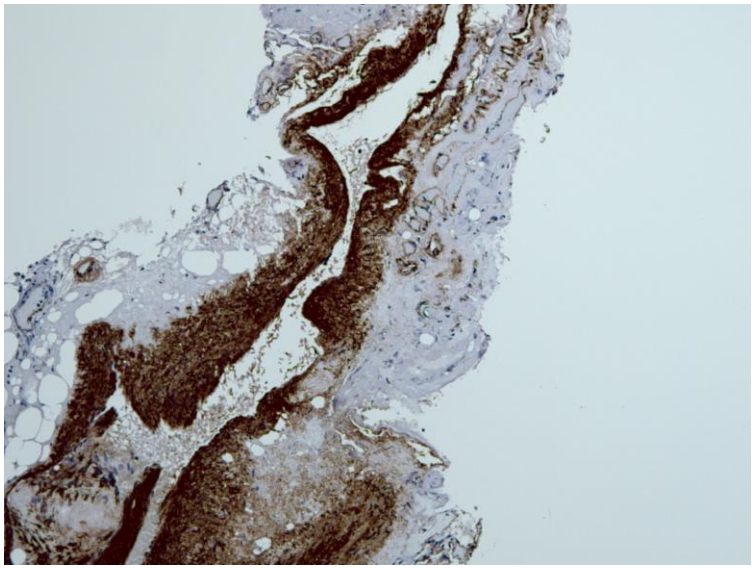


Histopathology of spinal CSF-venous fistula

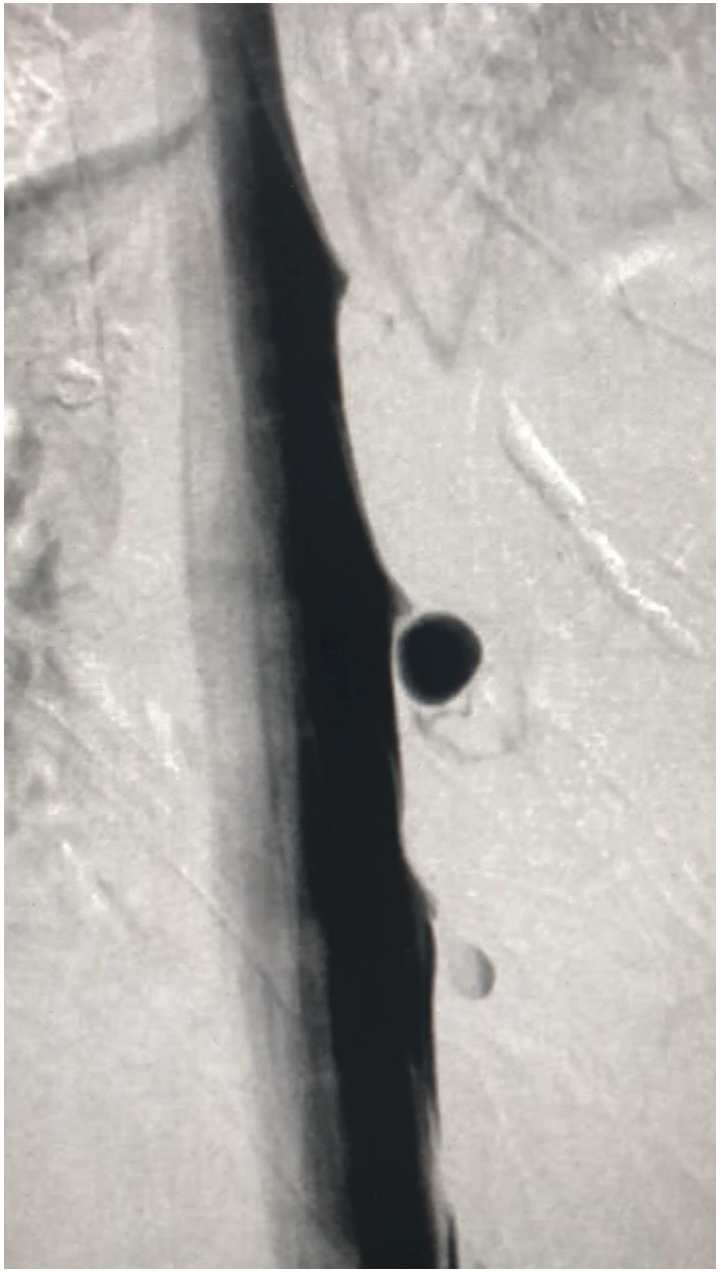




Trichrome stain at 10x (fibrous tissue in blue)

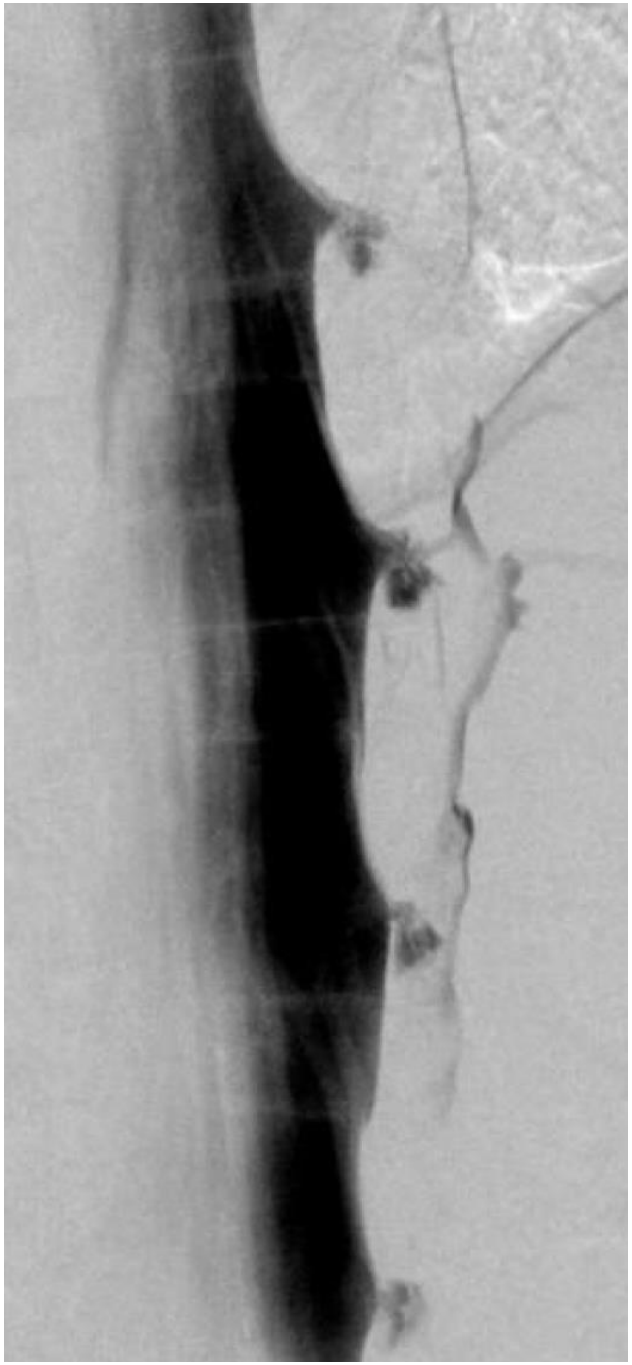


Immunostain for smooth muscle actin 10x (smooth muscle in brown)



Not all CSF-venous fistulas are thoracic





Classification of spontaneous spinal CSF leaks

- Mar 1, 2009-Aug 31, 2015 (n=568)

1	27%
2	42%
3	2.5%
4	29%

A classification system of spontaneous spinal CSF leaks

Wouter I. Schievink, MD
M. Marcel Maya, MD
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Miriam Nuño, PhD
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Correspondence to Dr. Schievink: schievinkw@cshs.org

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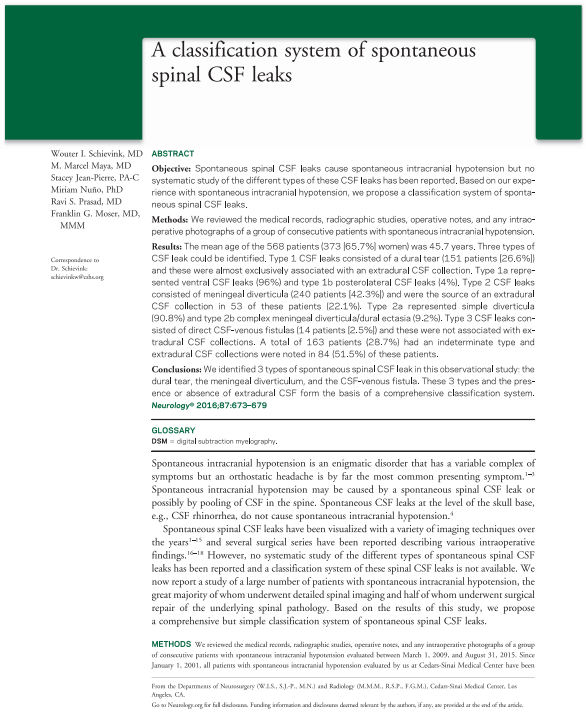
Neurology® 2016;87:673–679

Classification of spontaneous spinal CSF leaks Changes over time

- Mar 1, 2009-Aug 31, 2015 (n=568 new patients)

1	27%	40%	(+48%)
2	42%	17%	(-60%)
3	2.5%	23%	(+840%)
4	29%	19%	(-34%)

Apr 4, 2018-Sept 17, 2018 (n=70 new patients)



A classification system of spontaneous spinal CSF leaks

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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

GLOSSARY

DSM = digital subtraction myelography.

Spontaneous intracranial hypotension is an enigmatic disorder that has a variable complex of symptoms but an orthostatic headache is by far the most common presenting symptom.[1–3] Spontaneous intracranial hypotension may be caused by a spontaneous spinal CSF leak or possibly by pooling of CSF in the spine. Spontaneous CSF leaks at the level of the skull base, e.g., CSF rhinorrhea, do not cause spontaneous intracranial hypotension.[4]

Spontaneous spinal CSF leaks have been visualized with a variety of imaging techniques over the years[5–7] and several surgical series have been reported describing various intraoperative findings.[8–14] However, no systematic study of the different types of spontaneous spinal CSF leaks has been reported and a classification system of these spinal CSF leaks is not available. We now report a study of a large number of patients with spontaneous intracranial hypotension, the great majority of whom underwent detailed spinal imaging and half of whom underwent surgical repair of the underlying spinal pathology. Based on the results of this study, we propose a comprehensive but simple 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 evaluated between March 1, 2009, and August 31, 2015. Since January 1, 2001, all patients with spontaneous intracranial hypotension evaluated by us at Cedars-Sinai Medical Center have been

From the Departments of Neurosurgery (W.I.S., S.J.-P., M.N.) and Radiology (M.M.M., R.S.P., F.G.M.), Cedars-Sinai Medical Center, Los Angeles, CA.

Go to Neurology.org for full disclosures. Funding information and disclosures deemed relevant by the authors, if any, are provided at the end of the article.

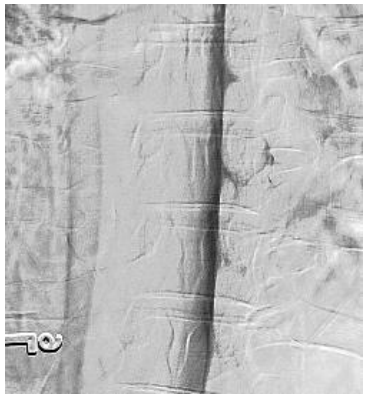
Lateral decubitus DSM vs prone DSM for identifying CSF-venous fistulas

- Prone:

  10/53: 19%

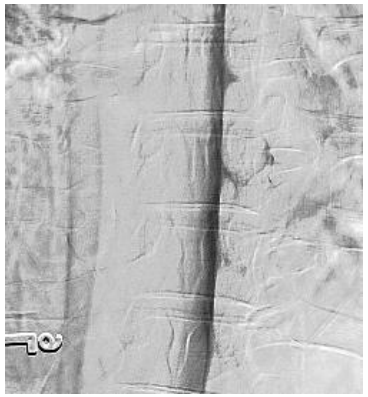
- Lateral decubitus:

  17/23: 74%



Richard Farb, MD

Lateral decubitus DSM vs prone DSM for identifying CSF-venous fistulas



Richard Farb, MD

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- Lateral decubitus:

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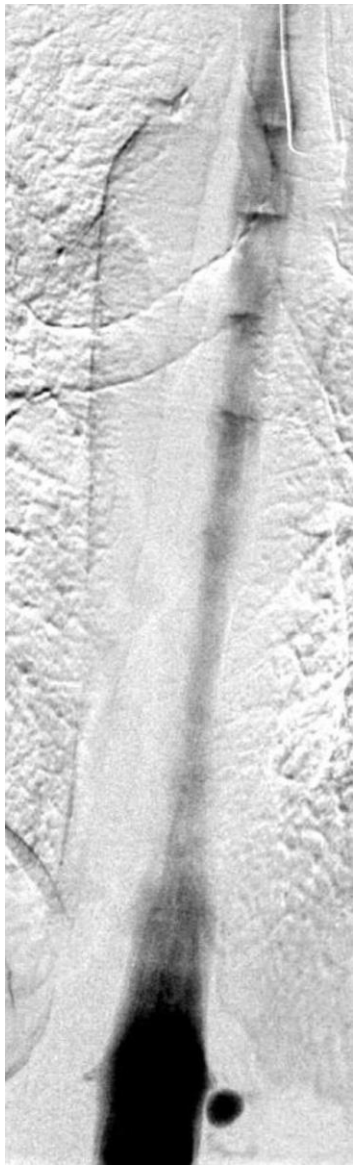
< April 2018: “50% of patients with SIH have negative spine imaging”

> April 2018: “25% of patients with SIH have negative spine imaging”



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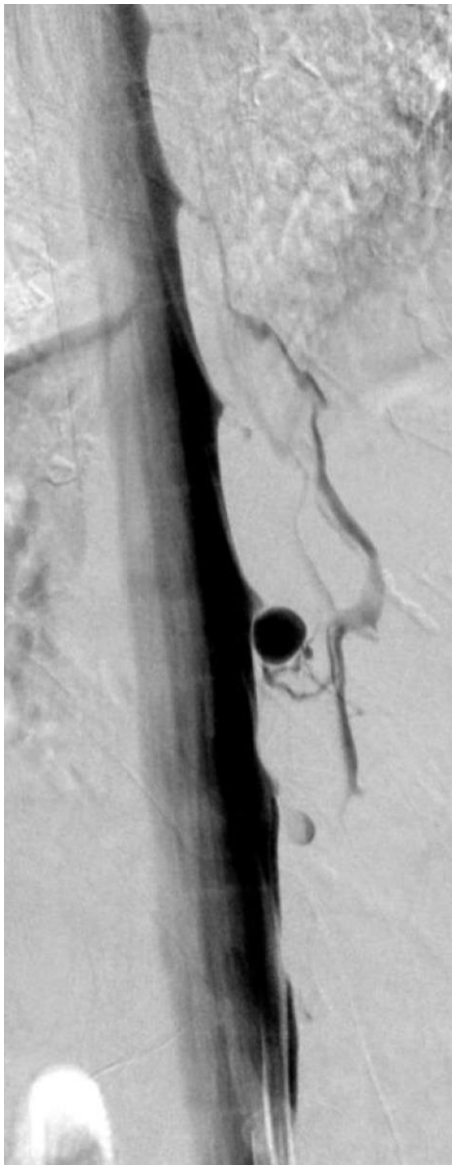

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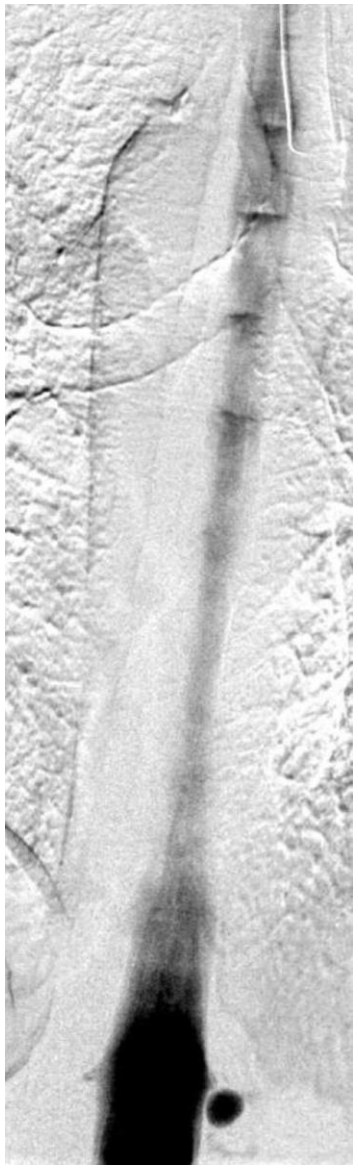

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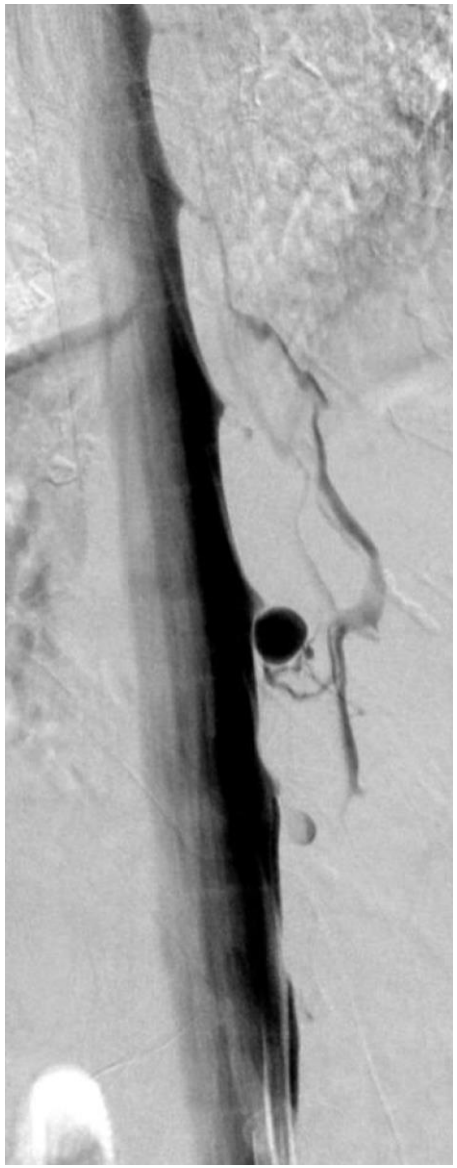

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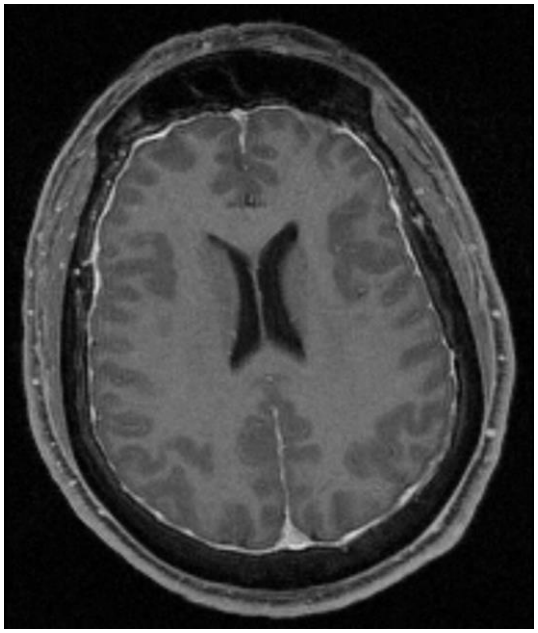
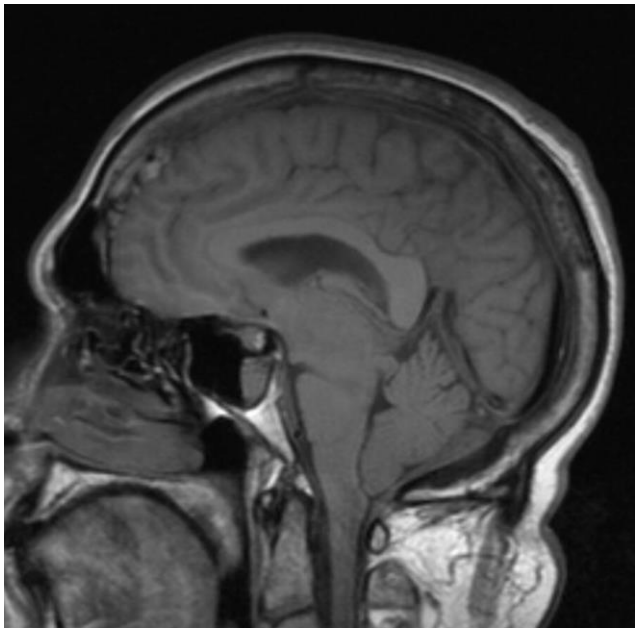
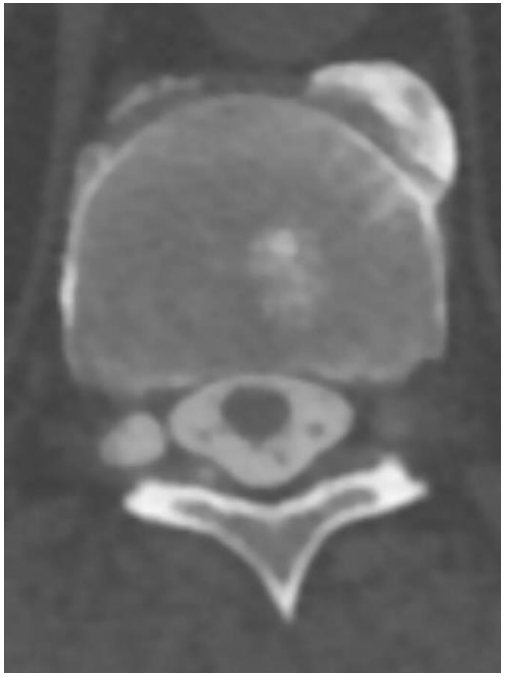
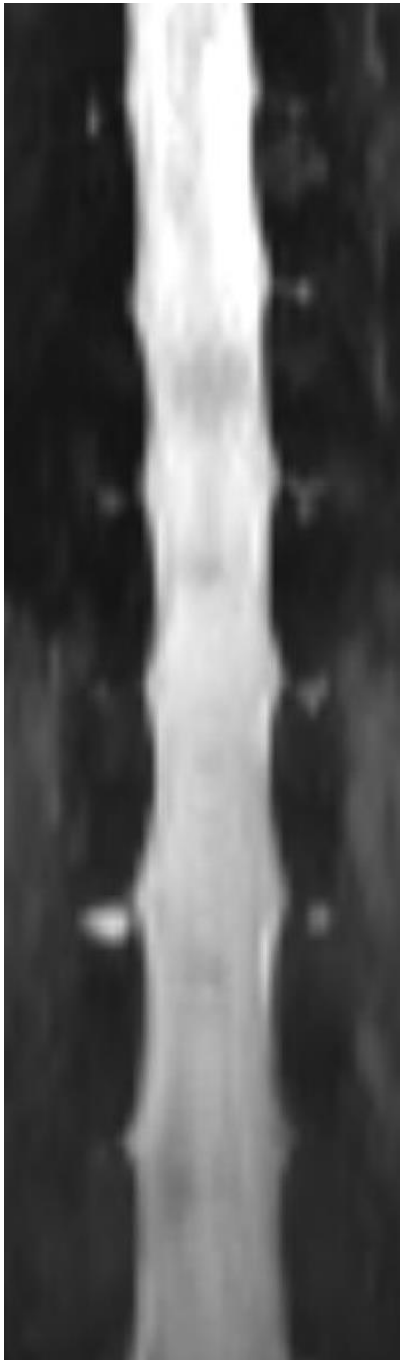
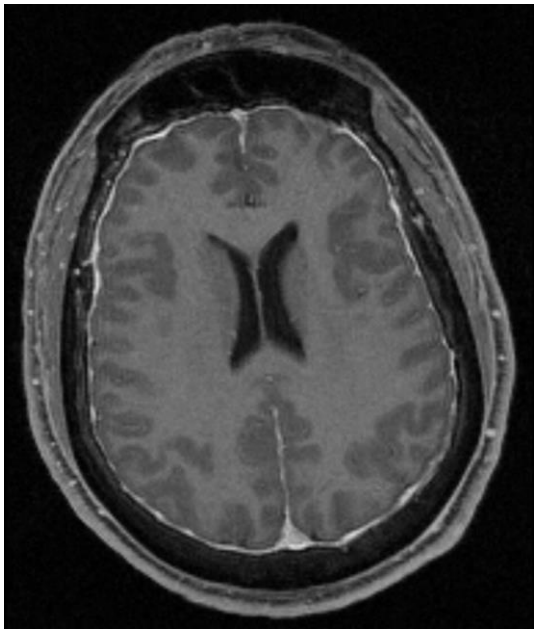
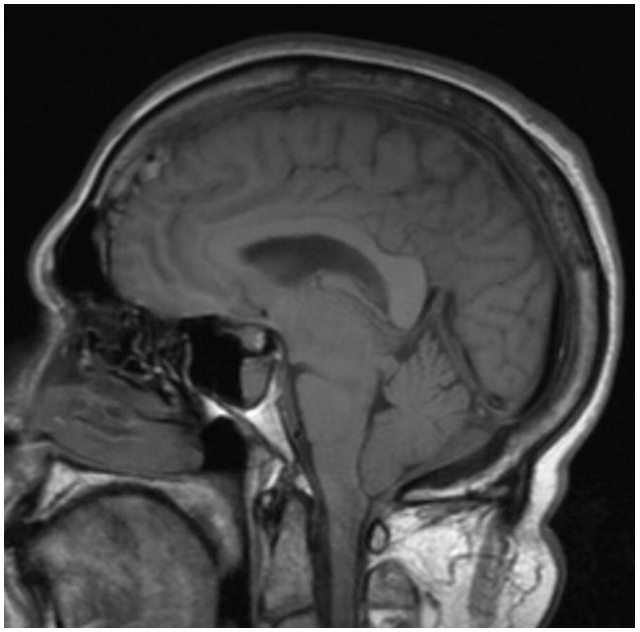


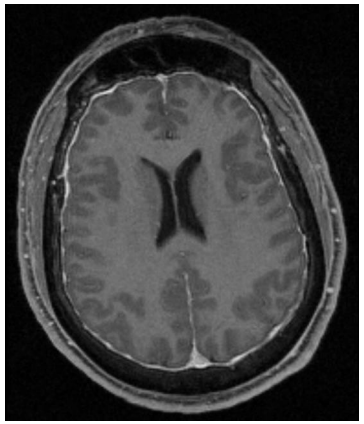


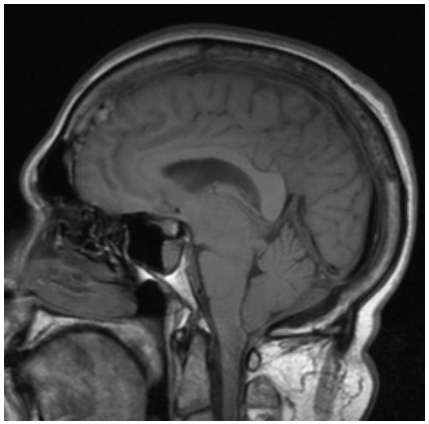


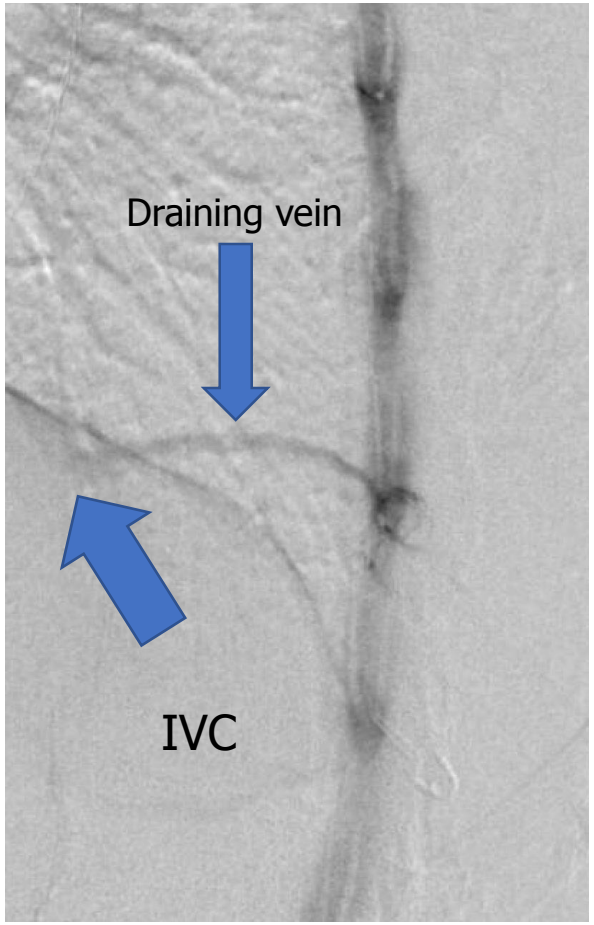


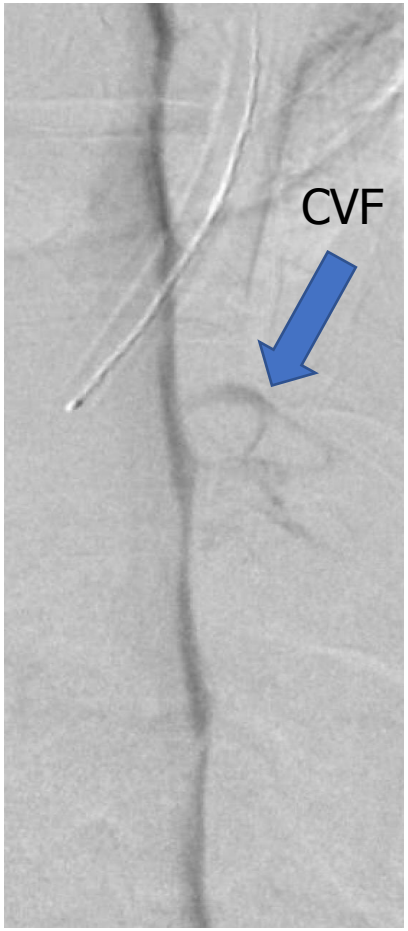









Draining vein
IVC


CVF

False localizing signs in SIH

J Neurosurg 100:639–644, 2004

False localizing sign of C1–2 cerebrospinal fluid leak in spontaneous intracranial hypotension

Wouter I. Schievink, M.D., M. Marcel Maya, M.D., and James Tourje, M.D.

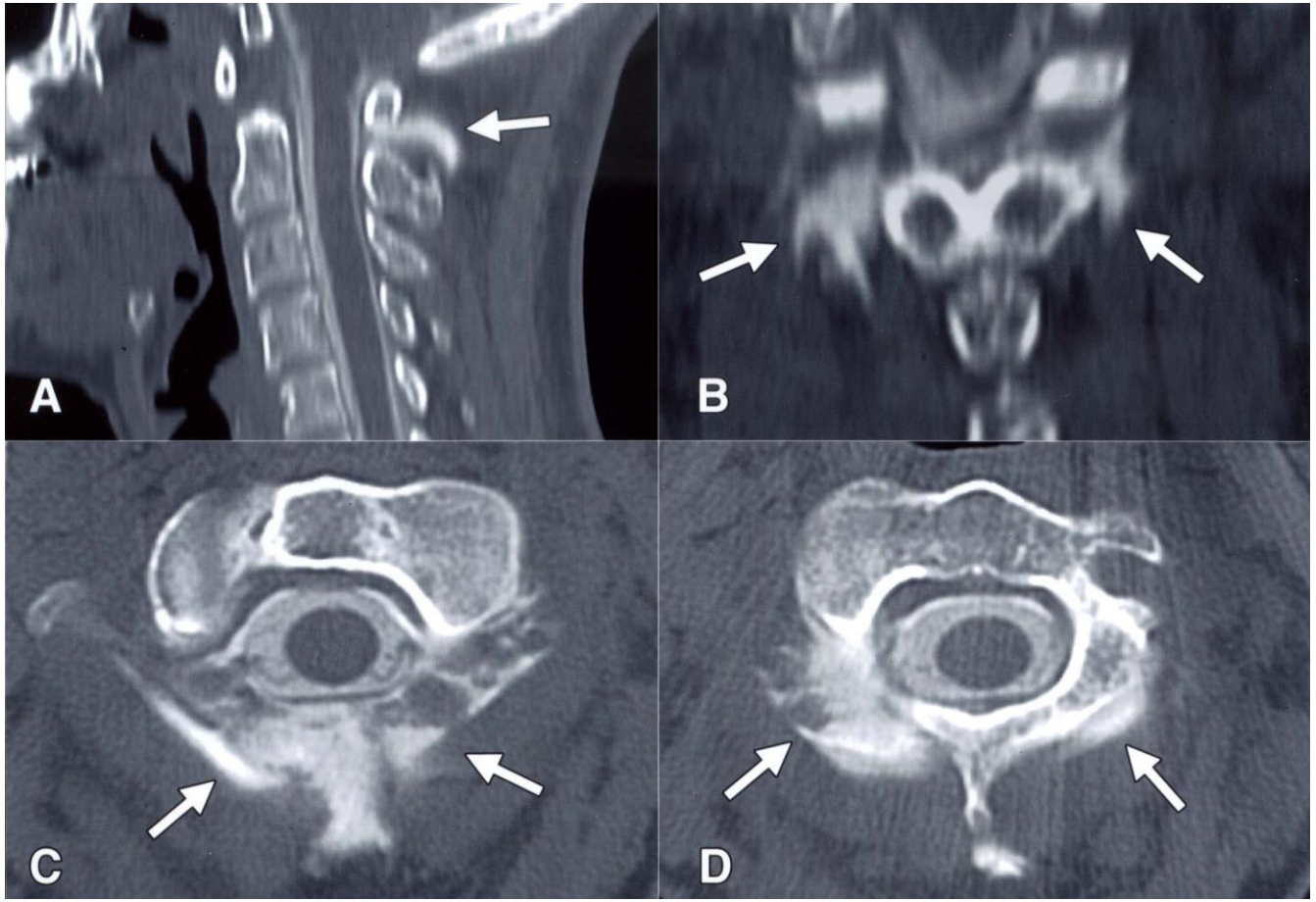
Maxine Dunitz Neurosurgical Institute and Imaging Medical Group, Cedars-Sinai Medical Center, Los Angeles, California

Object. Spontaneous intracranial hypotension due to a spinal cerebrospinal fluid (CSF) leak is an important cause of new daily persistent headaches. Spinal neuroimaging is important in the treatment of these patients, particularly when direct repair of the CSF leak is contemplated. Retrospinal C1–2 fluid collections may be noted on spinal imaging and these are generally believed to correspond to the site of the CSF leak. The authors undertook a study to determine the significance of these C1–2 fluid collections.

Methods. The patient population consisted of a consecutive group of 25 patients (18 female and seven male) who were evaluated for surgical repair of a spontaneous spinal CSF leak. The mean age of the 18 patients was 38 years (range 13–72 years). All patients underwent computerized tomography myelography. Three patients (12%) had extensive retrospinal C1–2 fluid collections; the mean age of this woman and these two men was 41 years (range 39–43 years). The actual site of the CSF leak was located at the lower cervical spine in these patients and did not correspond to the site of the retrospinal C1–2 fluid collection.

Conclusions. A retrospinal fluid collection at the C1–2 level does not necessarily indicate the site of the CSF leak in patients with spontaneous intracranial hypotension. This is an important consideration in the treatment of these patients because therapy may be inadvertently directed at this site.

C1-2 false localizing sign



False localizing signs in SIH

False localizing sign of cervico-thoracic CSF leak in spontaneous intracranial hypotension

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

Correspondence to Dr. Schievink: schievinkw@cshs.org

ABSTRACT

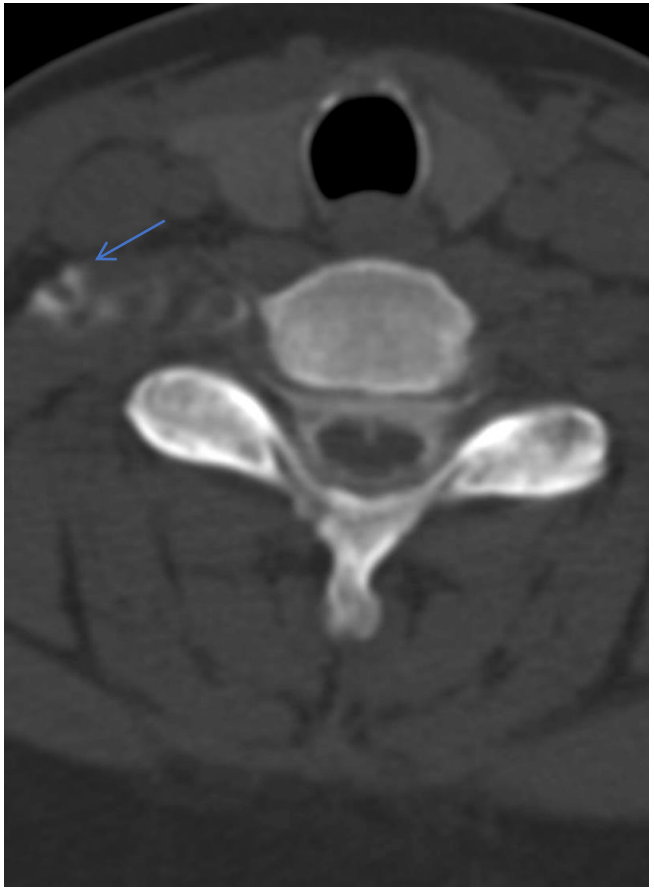
Objective: Spontaneous spinal CSF leaks are an important cause of new-onset headaches. Such leaks are reported to be particularly common at the cervico-thoracic junction. The authors undertook a study to determine the significance of these cervico-thoracic CSF leaks.

Methods: The patient population consisted of a consecutive group of 13 patients who underwent surgery for CSF leak repair based on CT myelography showing CSF extravasation at the cervico-thoracic junction but without any evidence of an underlying structural lesion.

Results: The mean age of the 9 women and 4 men was 41.2 years. Extensive extrathecal longitudinal CSF collections were demonstrated in 11 patients. At surgery, small leaking arachnoid cysts were found in 2 patients. In the remaining 11 patients, no clear source of CSF leakage could be identified at surgery. Resolution of symptoms was achieved in both patients with leaking arachnoid cysts, but in only 3 of the 11 patients with negative intraoperative findings. Postoperative spinal imaging was performed in 9 of the 11 patients with negative intraoperative findings and showed persistence of the longitudinal intraspinal extradural CSF. Further imaging revealed the site of the CSF leak to be ventral to the thoracic spinal cord. Five of these patients underwent microsurgical repair of the ventral CSF leak with resolution of symptoms in all 5 patients.

Conclusions: Cervico-thoracic extravasation of dye on myelography does not necessarily indicate the site of the CSF leak. Treatment directed at this site should not be expected to have a high probability of sustained improvement of symptoms. ***Neurology*® 2015;84:2445–2448**

Cervico-thoracic false localizing sign



False localizing signs in SIH – type 1

J Neurosurg 100:639–644, 2004

False localizing sign of C1–2 cerebrospinal fluid leak in spontaneous intracranial hypotension

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False localizing sign of cervico-thoracic CSF leak in spontaneous intracranial hypotension

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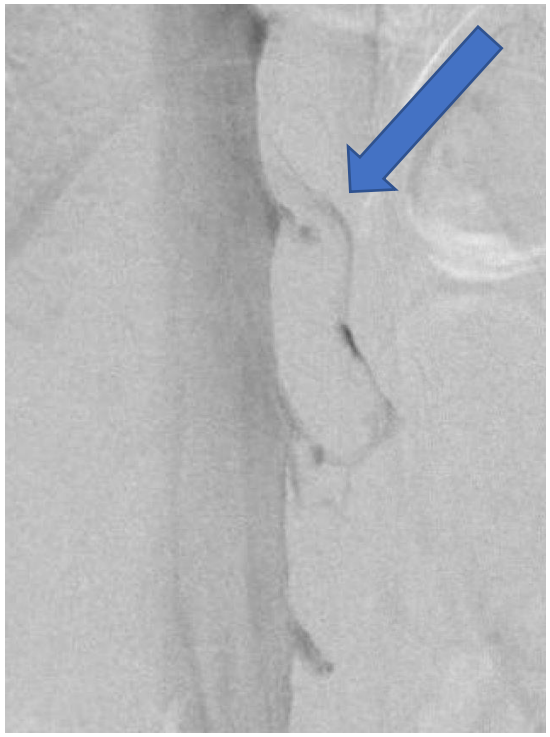
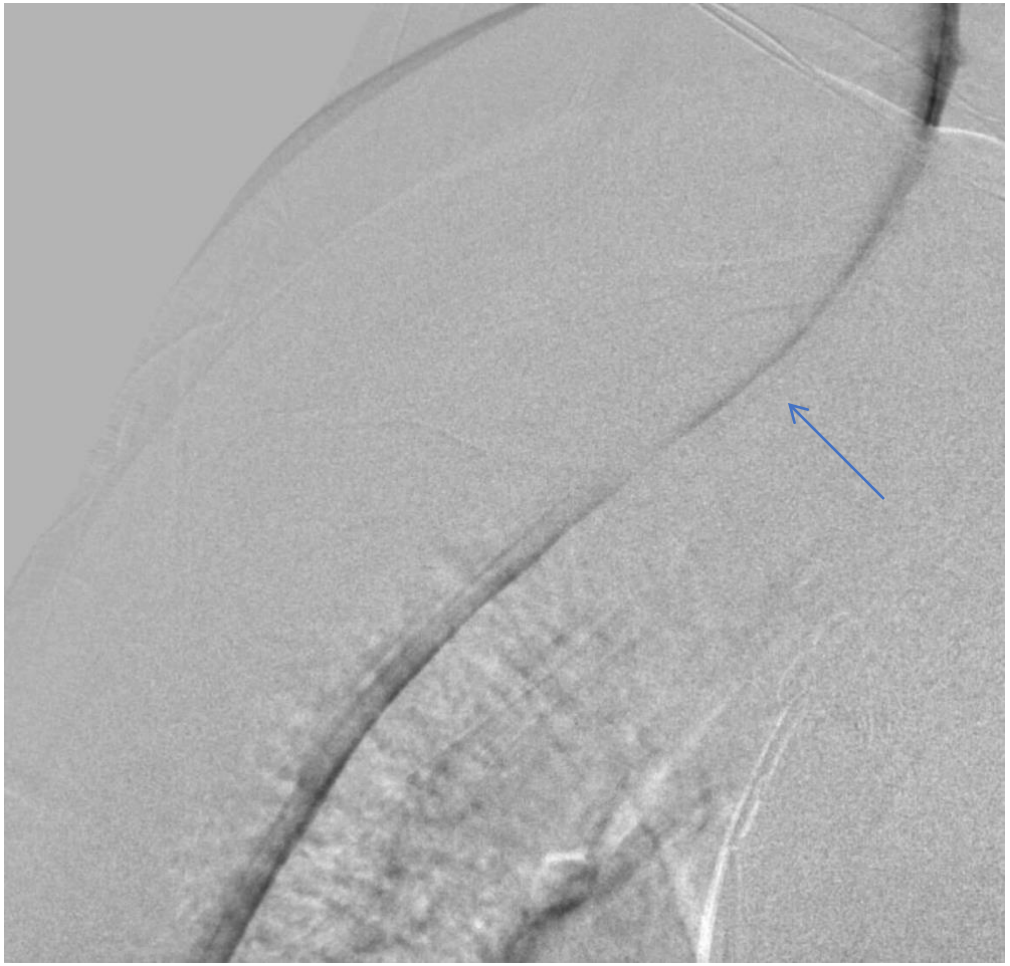
False localizing signs in CSF-venous fistulas

Laterality



Spinal level

Ventral spinal CSF leaks – Digital subtraction myelography



The value of digital subtraction myelography

Dorsal

Ventral

Looking for a ventral CSF leak

- **DSM**
- **DSM #2**
- **DSM #3**
- Dynamic CT-myelogram
- Bony spicule

Bony spicule as cause of type 1a ventral CSF leak

J Neurosurg 89:483–484, 1998

Cervical bone spur presenting with spontaneous intracranial hypotension

Case report

A. GIANCARLO VISHTEH, M.D., WOUTER I. SCHIEVINK, M.D., JONATHAN J. BASKIN, M.D., AND VOLKER K. H. SONNTAG, M.D.

Division of Neurological Surgery, Barrow Neurological Institute, Mercy Healthcare Arizona, Phoenix, Arizona

Spontaneous intracranial hypotension due to a spinal cerebrospinal fluid (CSF) leak is a rare but increasingly recognized cause of postural headaches. The exact cause of these CSF leaks often remains unknown. The authors treated a 32-year-old man with a unique cause of spontaneous intracranial hypotension. He suffered an excruciating headache that was exacerbated by his being in an upright position. The results of four-vessel cerebral angiography were negative; however, magnetic resonance (MR) imaging of the brain revealed pachymeningeal enhancement and hindbrain herniation. A presumptive diagnosis of spontaneous intracranial hypotension was made. Myelography revealed extrathecal contrast material ventral to the cervical spinal cord as well as an unusual midline bone spur at C5–6. The patient's symptoms did not resolve with the application of epidural blood patches, and he subsequently underwent an anterior approach to the C5–6 spur. After discectomy, a slender bone spur that had pierced the thecal sac was found. After its removal, the dural rent was closed using two interrupted prolene sutures. The patient was discharged home 2 days later. On follow up his symptoms had resolved, and on MR imaging the pachymeningeal enhancement had resolved and the cerebellar herniation had improved slightly.

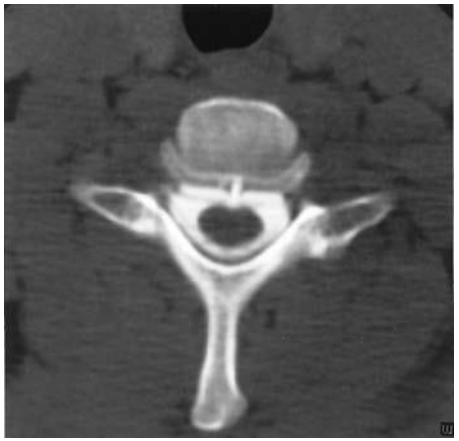
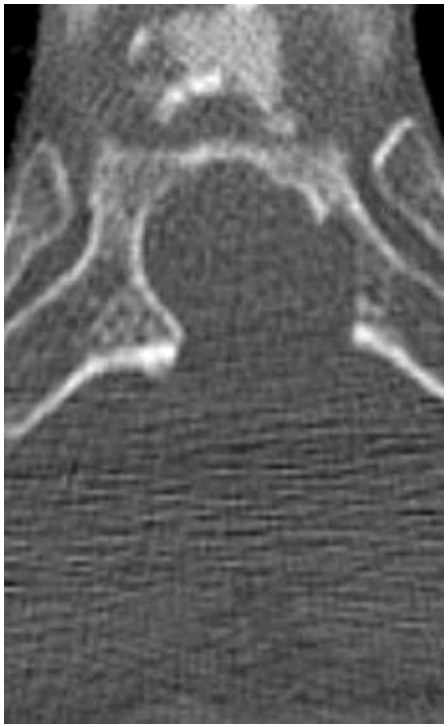


FIG. 1. Postmyelography axial CT obtained at the C5–6 level, showing the bone spur protruding into the thecal sac and associated extrathecal contrast.

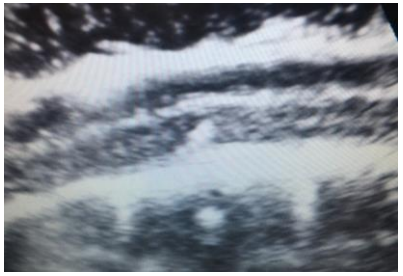
Bony spicules in SIH

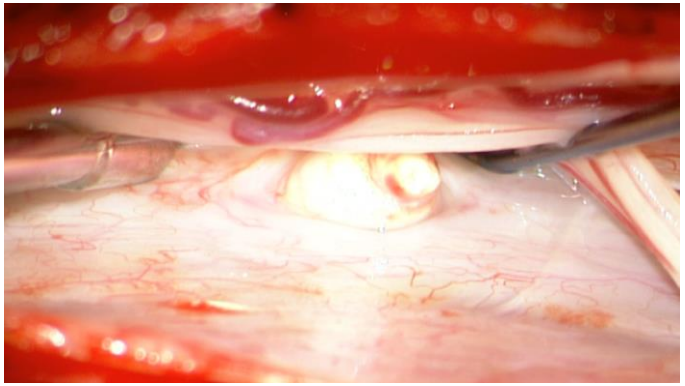
Pre-op

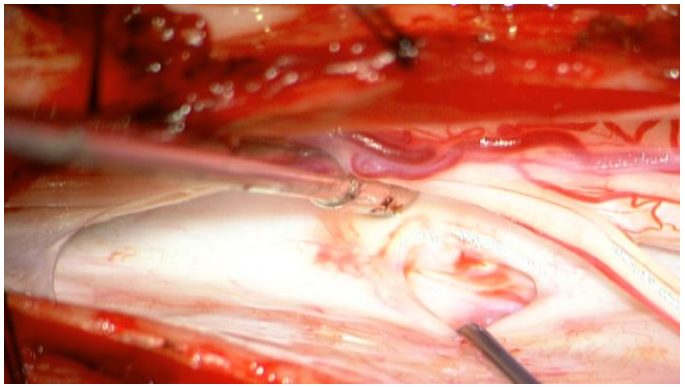


Post-op

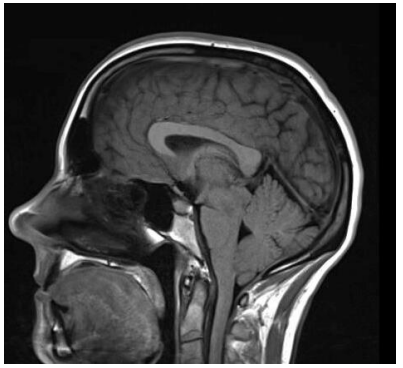
Intra-op

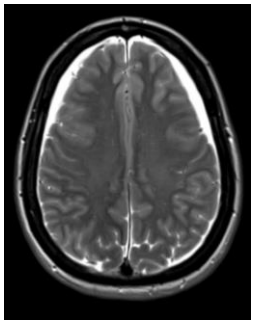


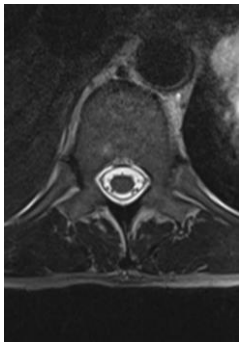




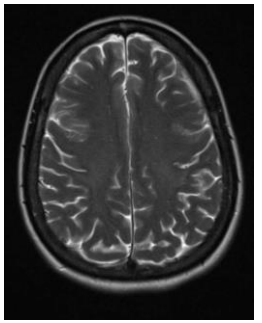
Pre-op







Post-op



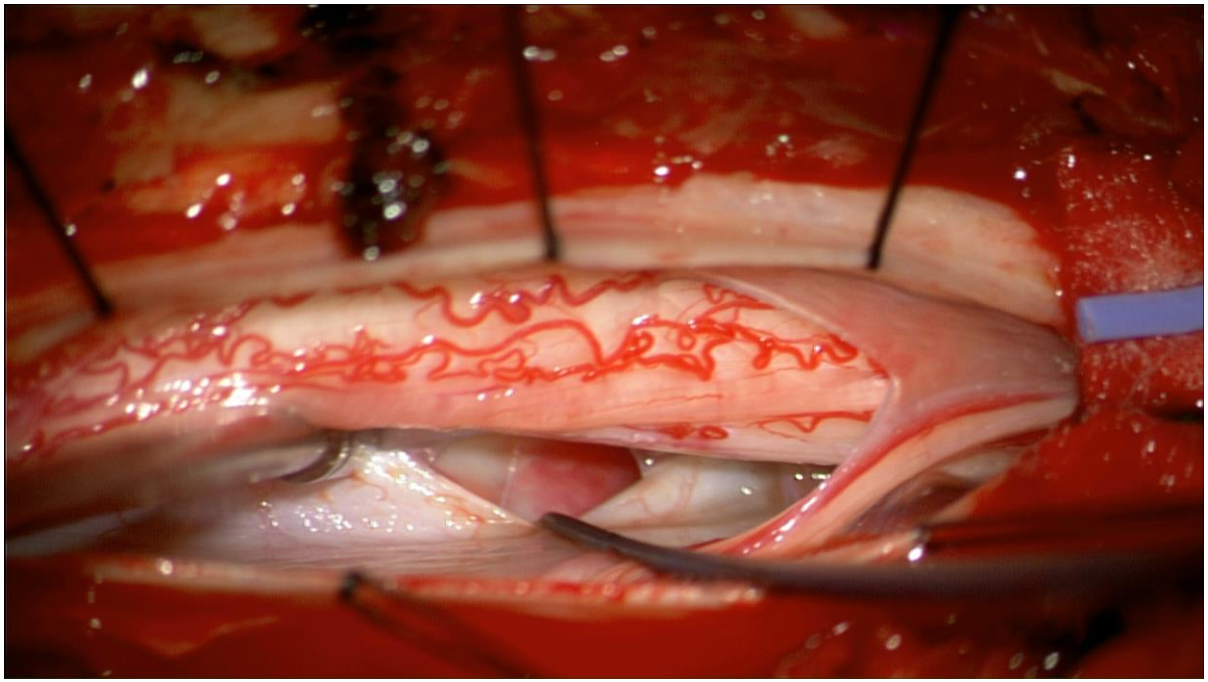
Caveats in identification of bony spicule as source of ventral type 1a CSF leak

- Present in approximately 85% of patients with type 1a CSF leak
- Multiple spicules – not always the largest
- Absorption of spicule over time

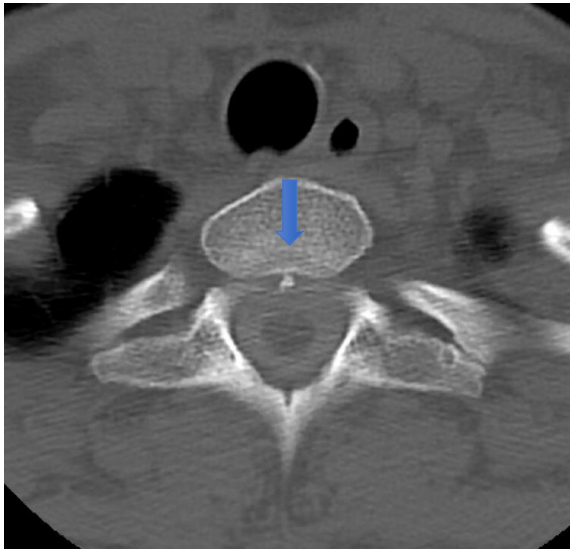
Not always the largest spicule

Not always the largest disc herniation

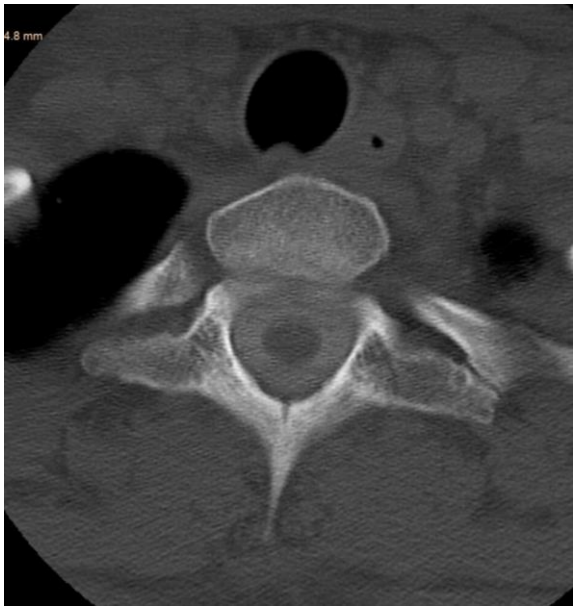
Not always the largest disc herniation

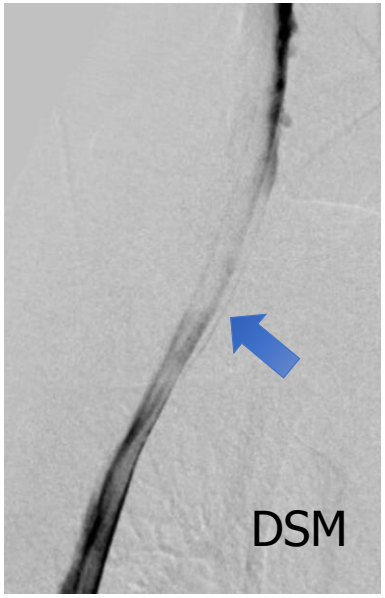


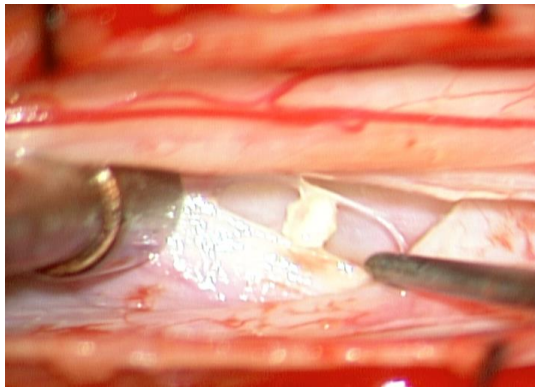
Absorption of spicule over time

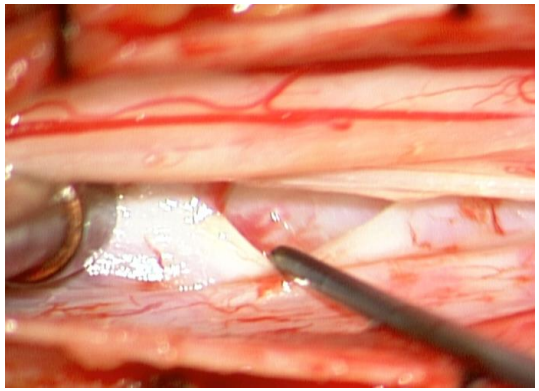


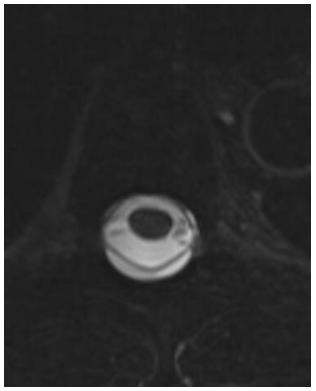
8 months



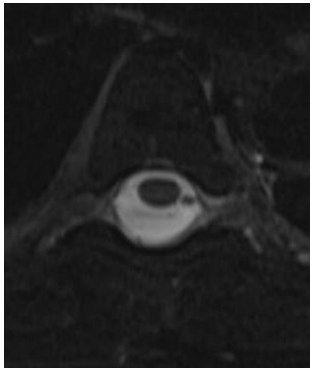








Pre-op



Post-op

Surgical Repair

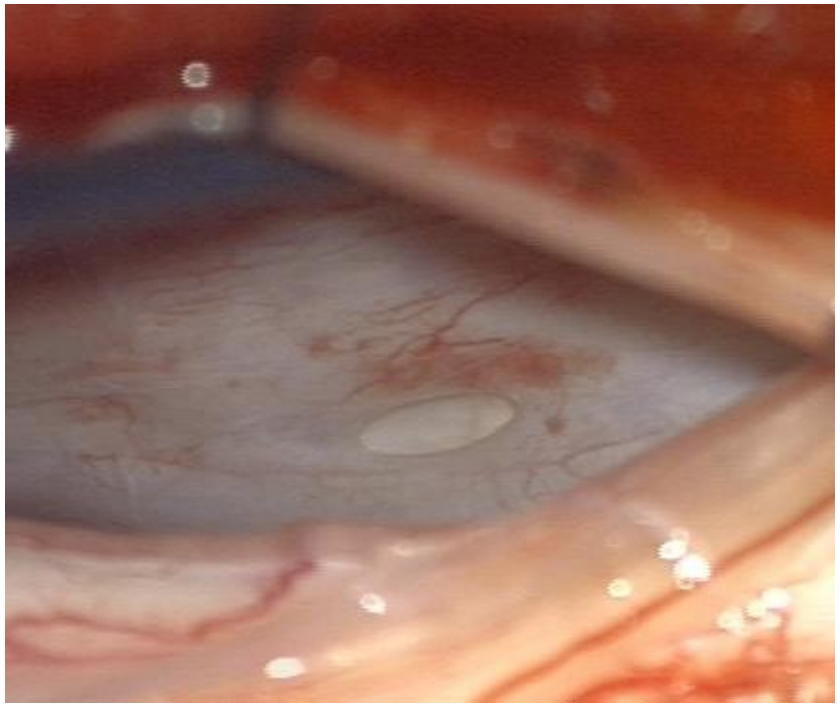
- Suturing
- Muscle graft
- Clipping of cyst

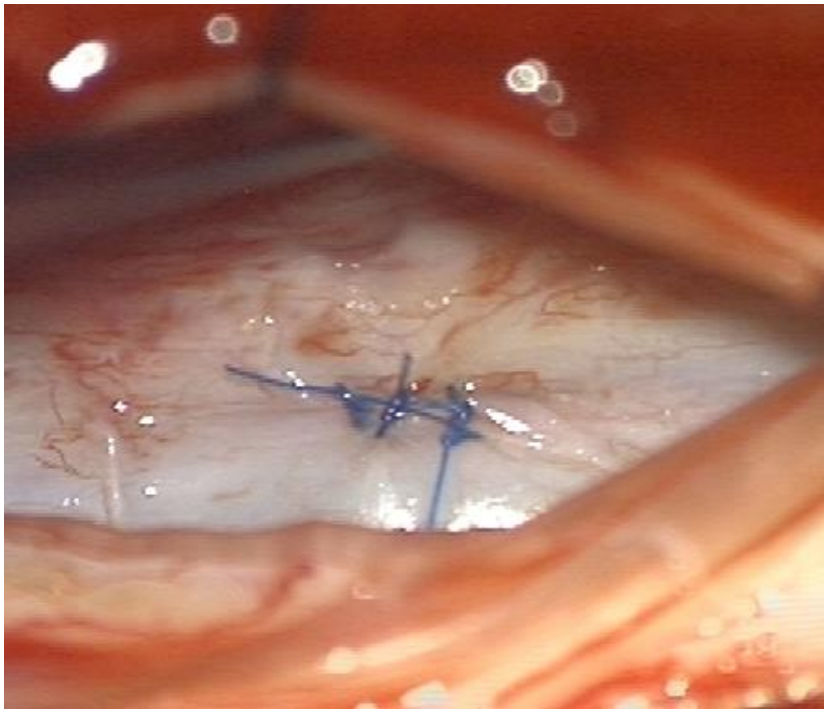
2018

- Fibrin glue/blood
- Gel foam

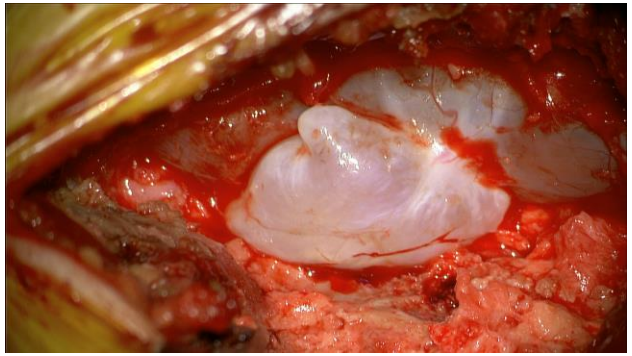
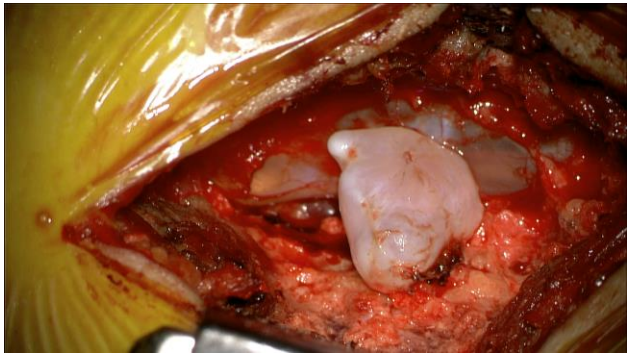
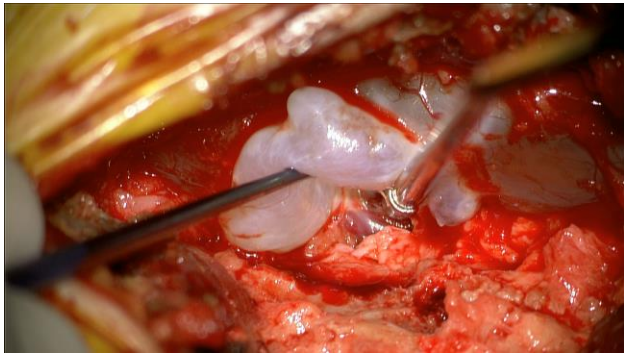
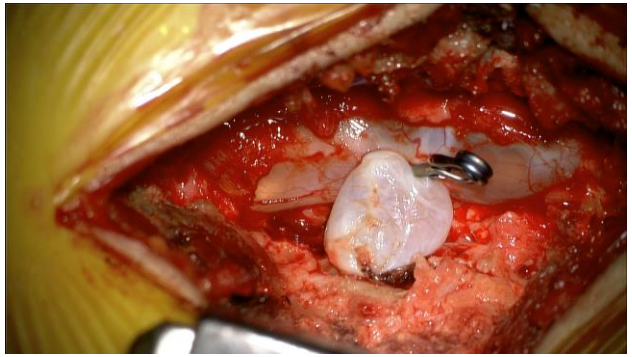
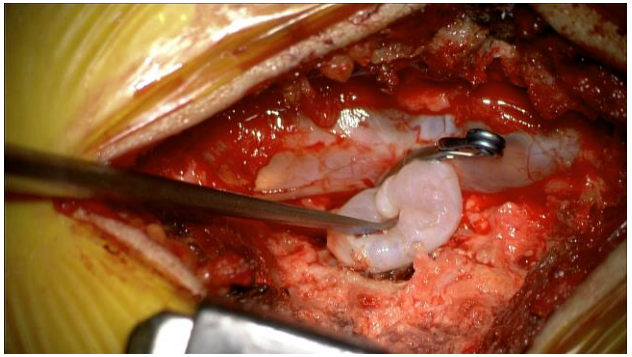
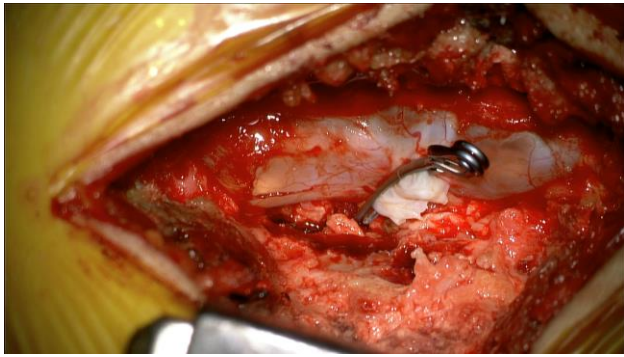
20th century

Ventral leak – surgical repair

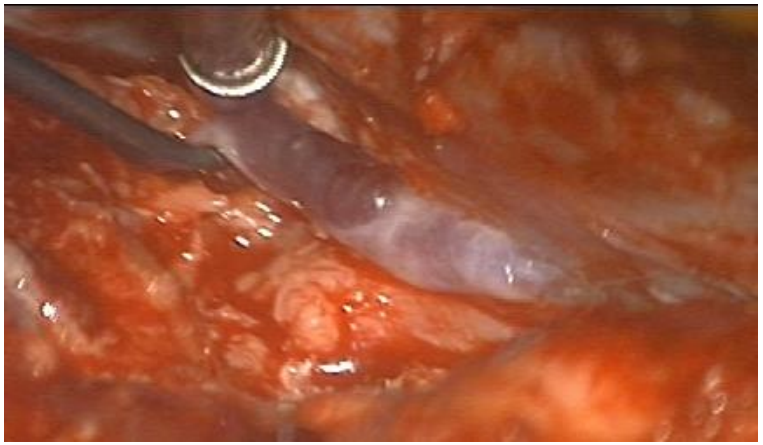


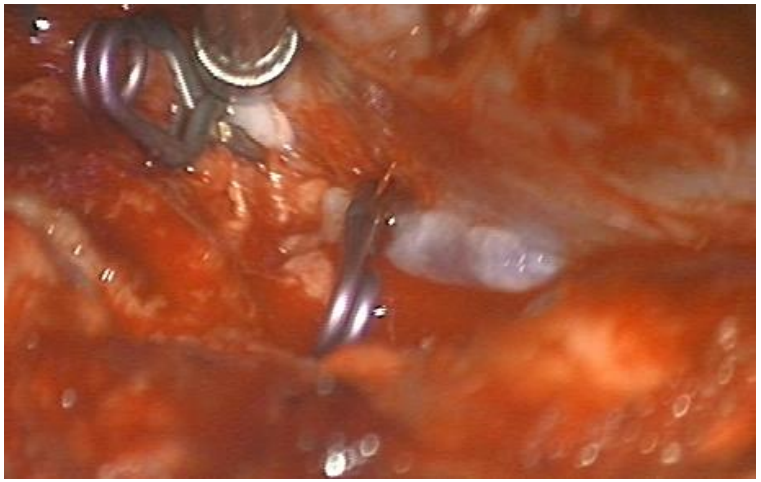


Arachnoid cysts – surgical repair

CSF–venous fistula – surgical repair





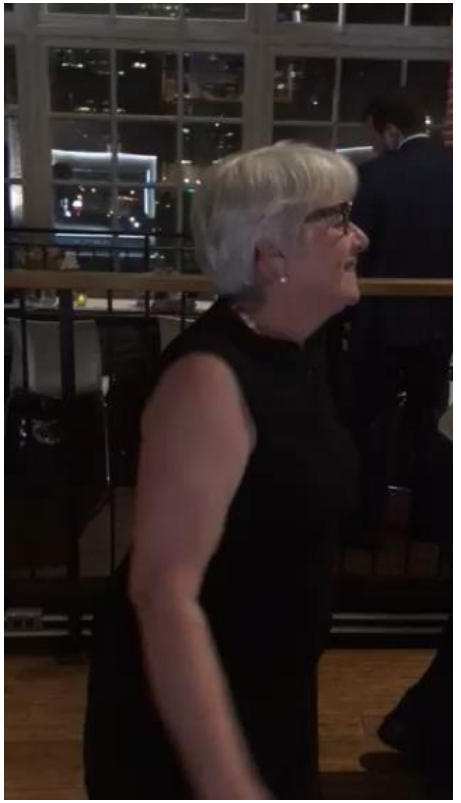
Surgery for spontaneous intracranial hypotension

Surgery	- cyst + leak:	95% cure rate
	- cyst only:	75% cure rate
	- CSF-venous fistula:	95% cure rate
	- ventral/intradural:	95% cure rate
	- ventral/extradural:	25% cure rate

Surgery for spontaneous intracranial hypotension

Surgery	- cyst + leak:	95+% cure rate
	- cyst only:	75% cure rate
	- CSF-venous fistula:	95+% cure rate
	- ventral/intradural:	95+% cure rate
	- ventral/extradural:	25% cure rate

Post-op recovery

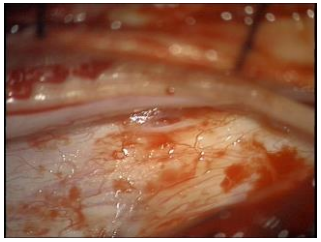
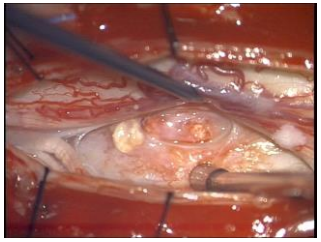
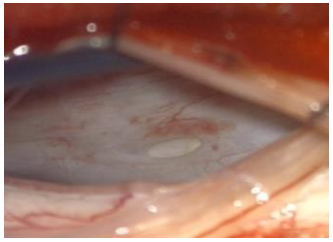
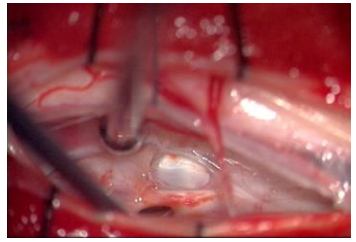
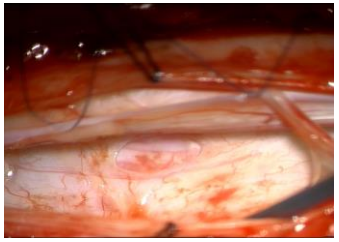
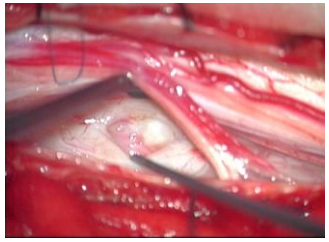
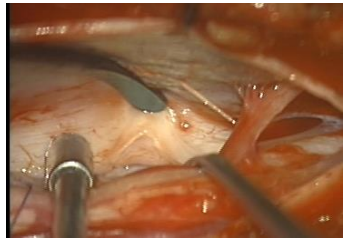
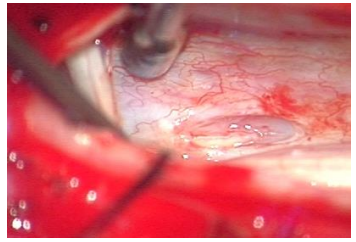
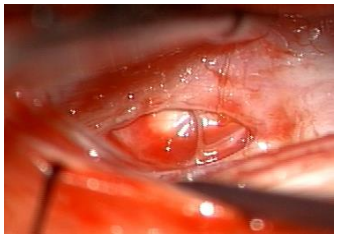


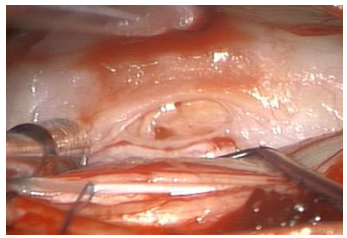
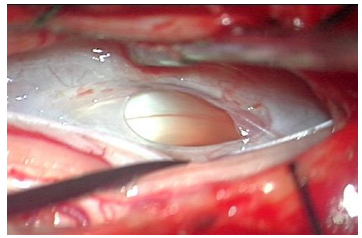
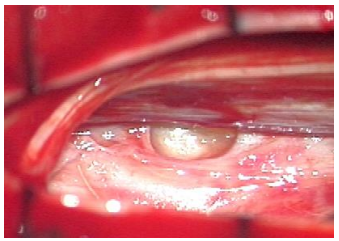
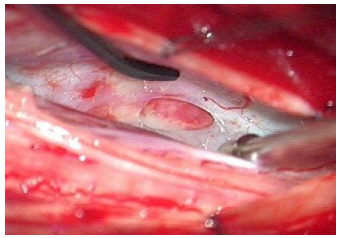
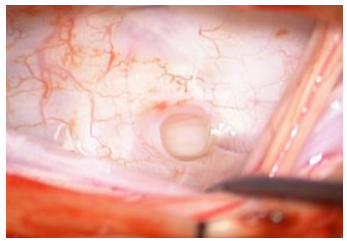
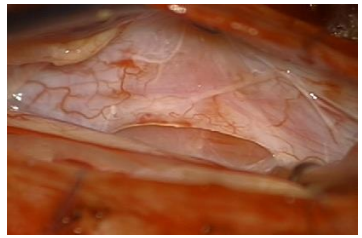
Persistent symptoms in spite of resolution of spinal CSF leak

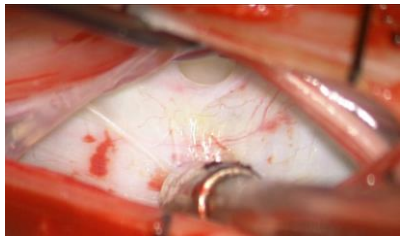
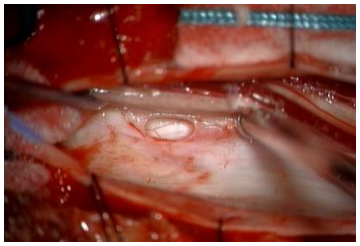
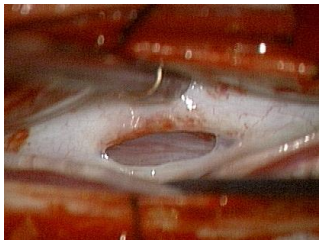
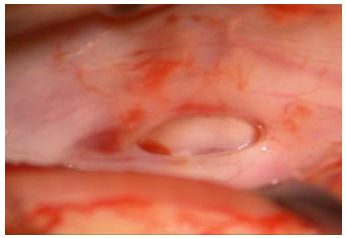
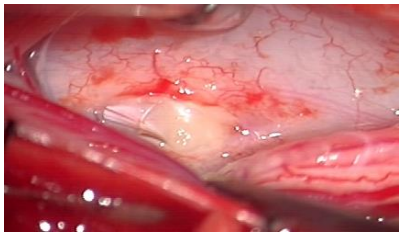
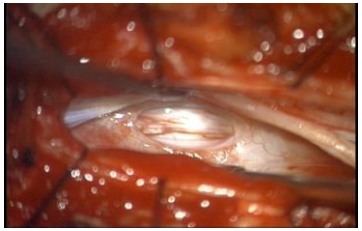
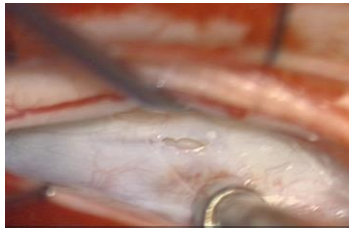
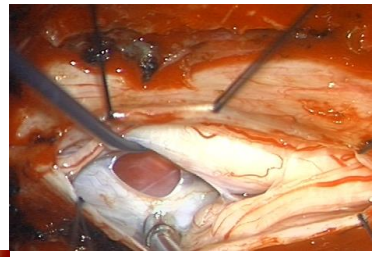
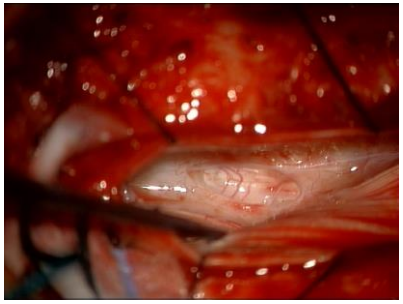
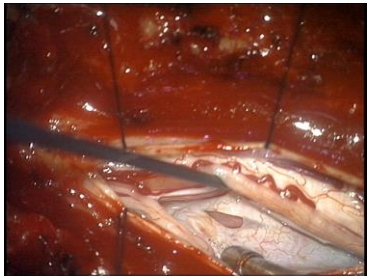
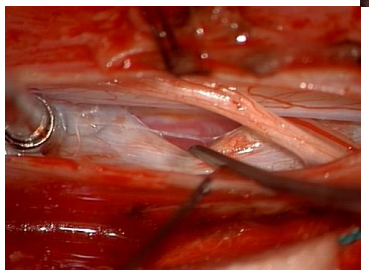
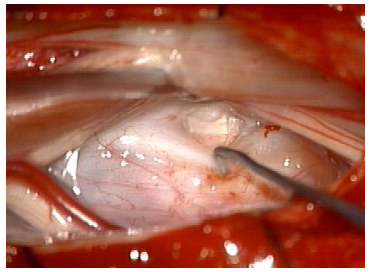
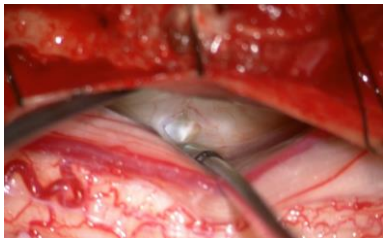
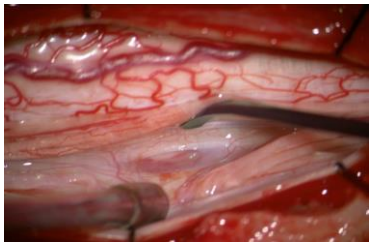
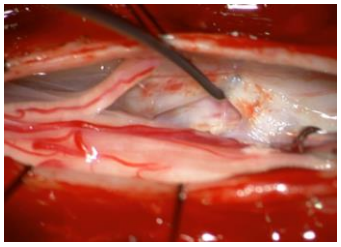
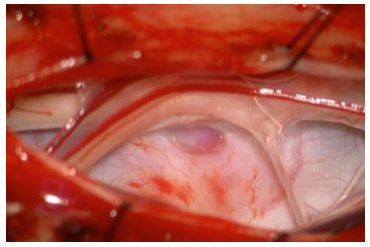
- 5% ?

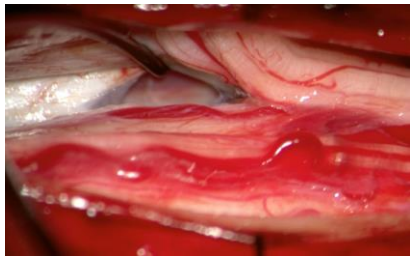
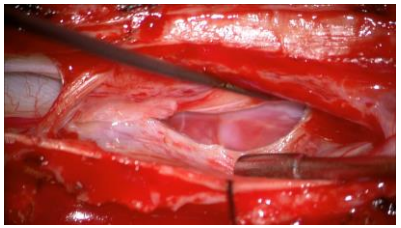
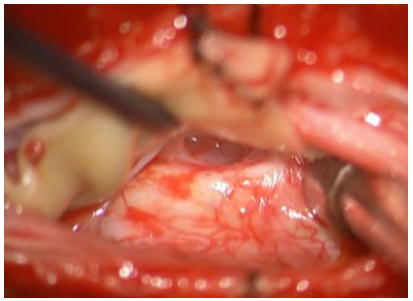
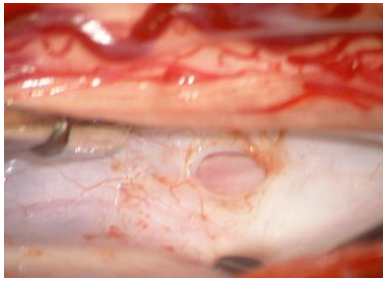
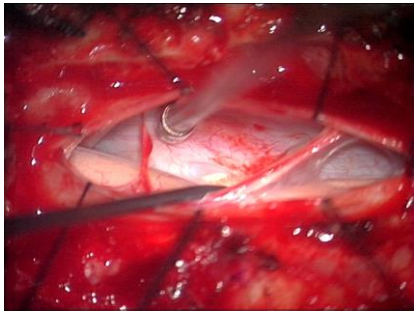
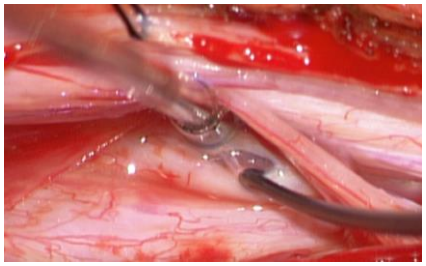
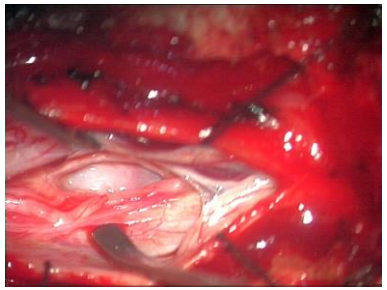
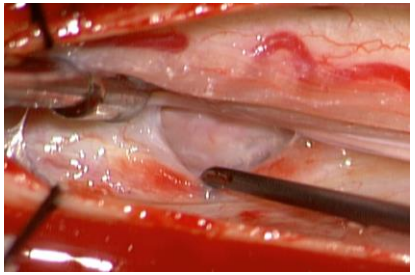
Risks of treatment

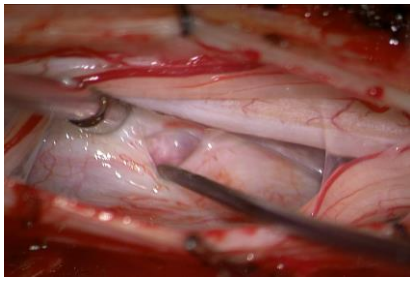
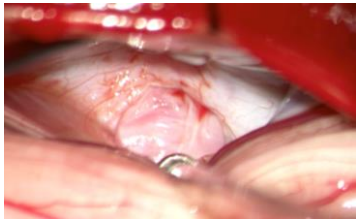
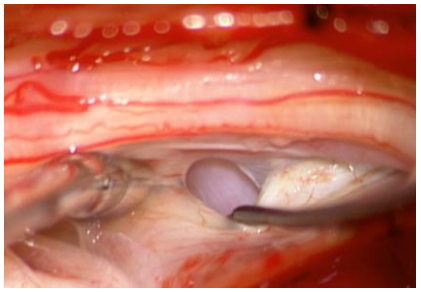
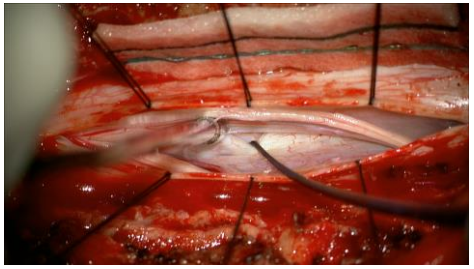
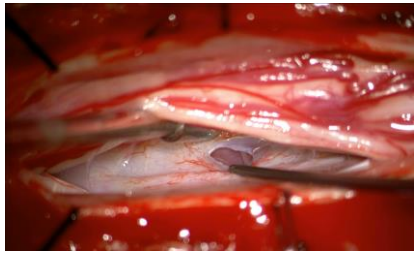
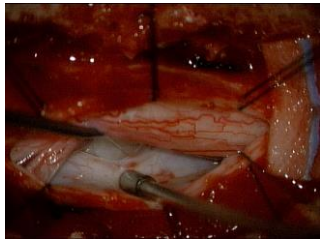
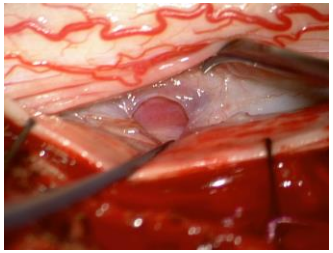
- Surgery (n=600):
  - 0.6% infection
  - 0.8% neurologic deficit
  - 2.7% pseudomeningocele
  - 0.8% presyrinx formation
  - 0.3% suicide

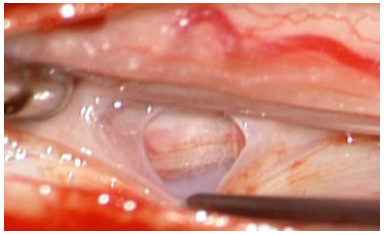
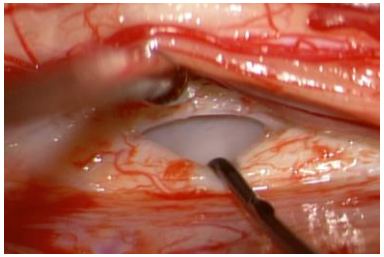
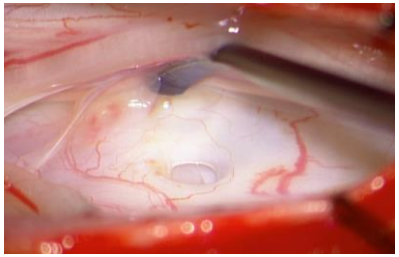
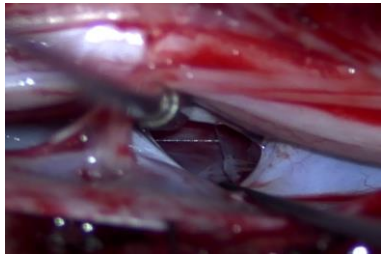
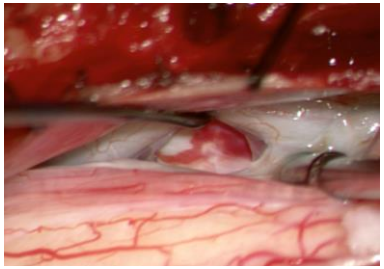
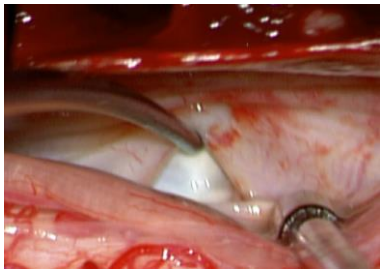
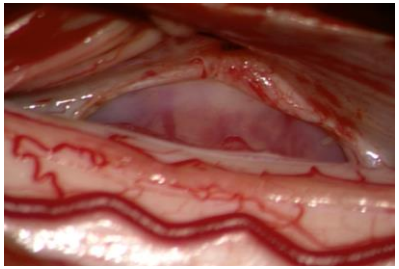
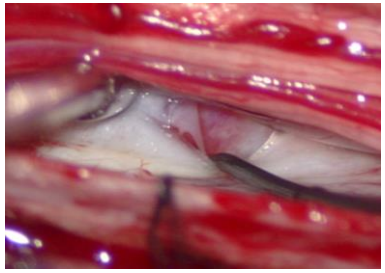
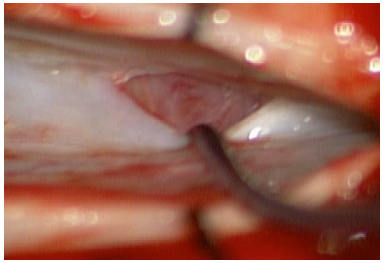
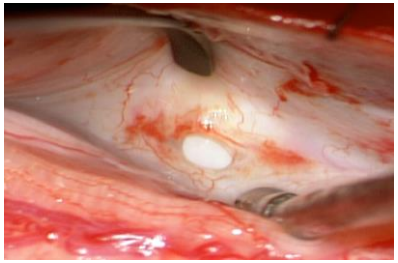
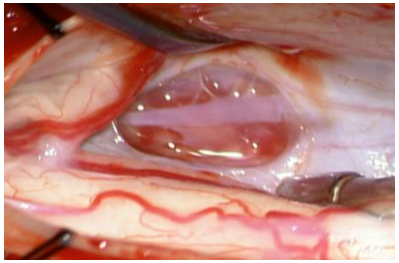
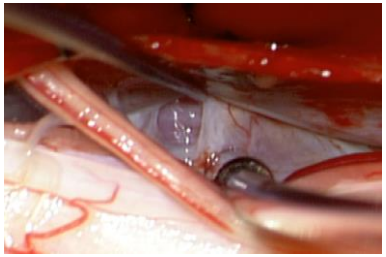
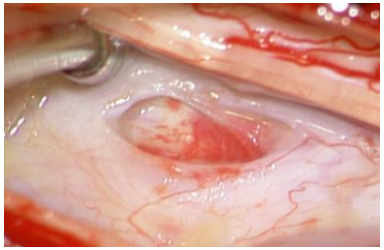
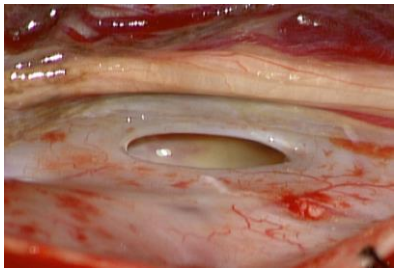
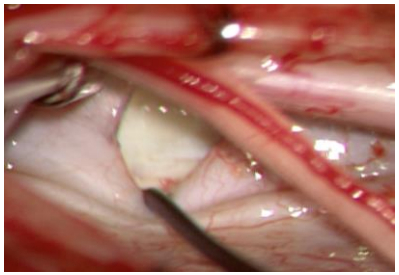
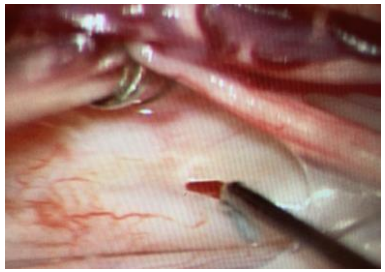










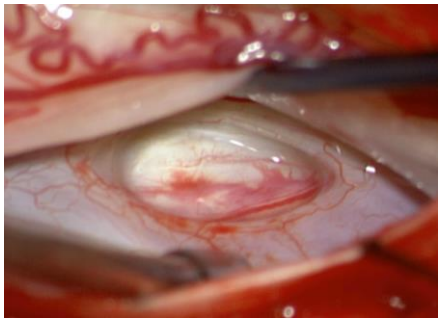
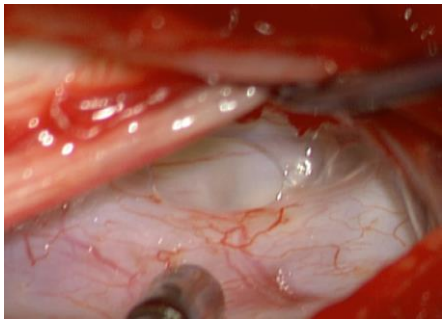
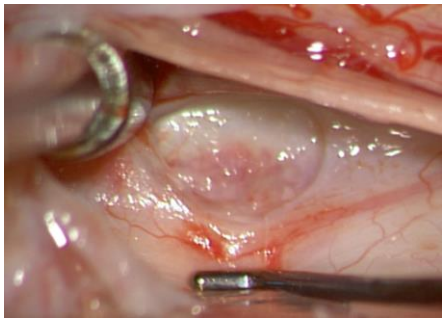
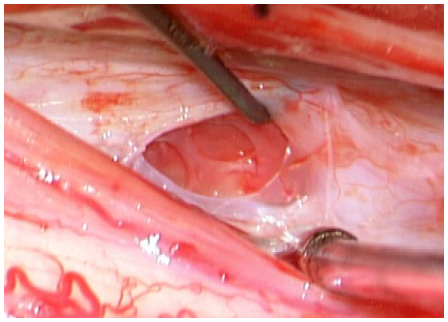
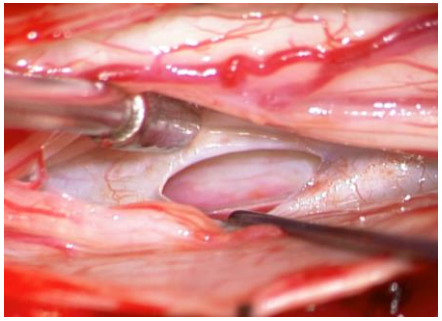
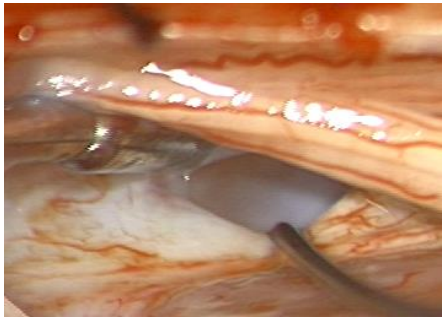
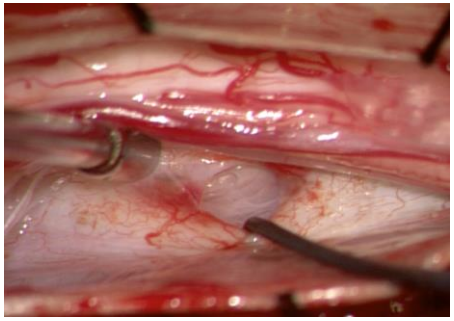
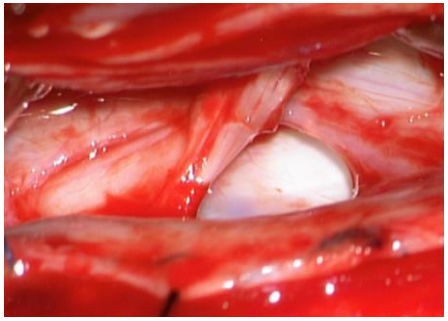
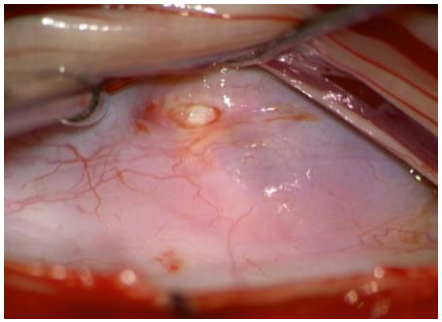
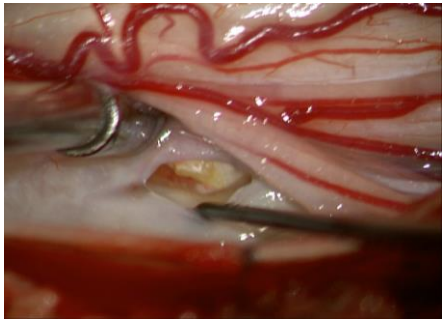
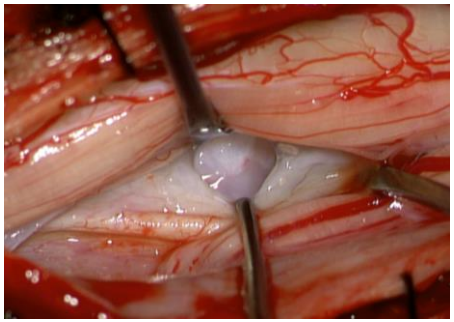
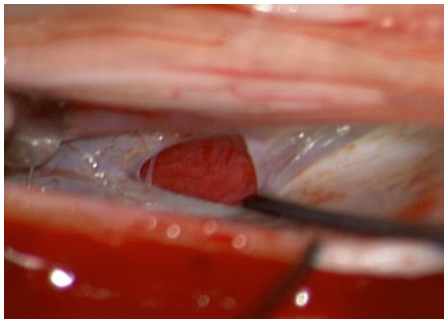
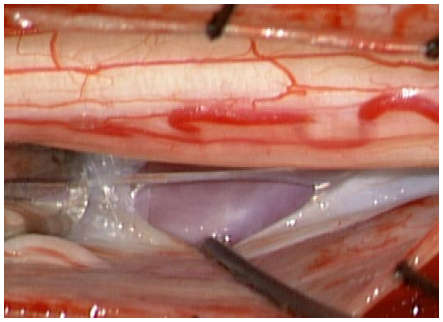
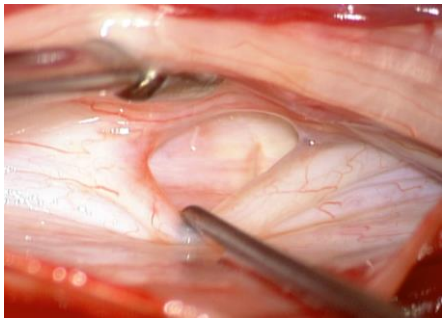
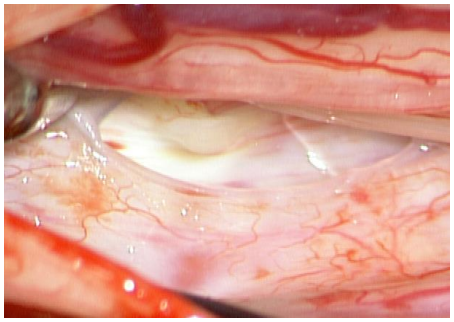
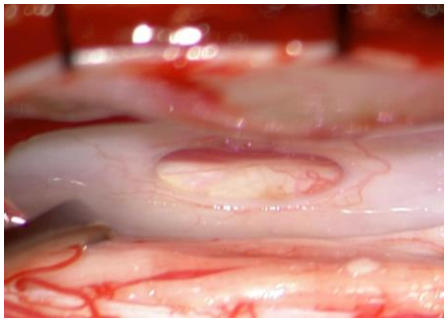







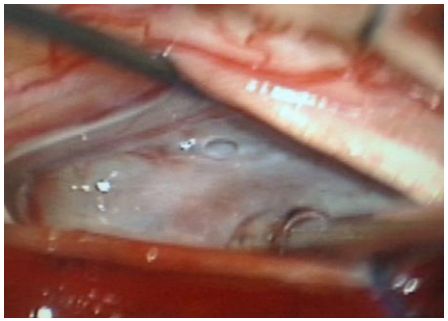
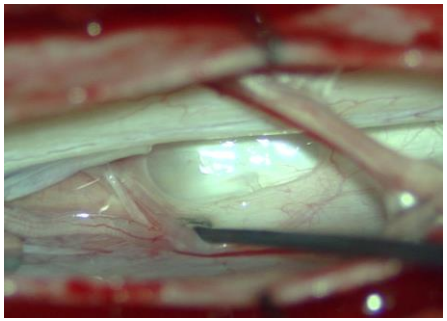
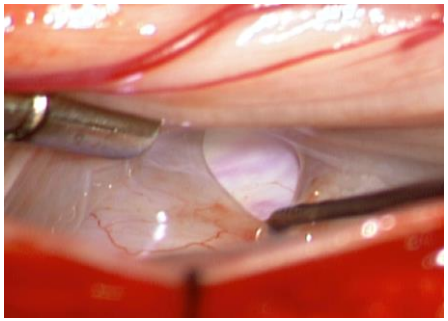
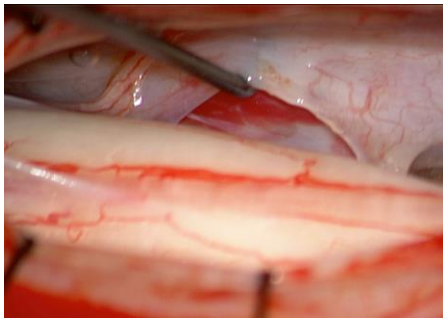
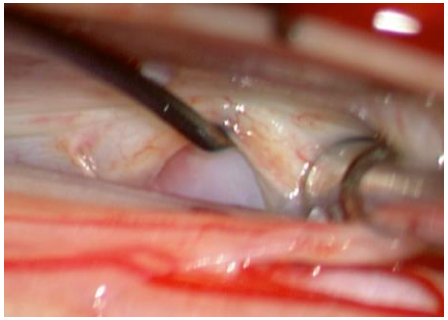
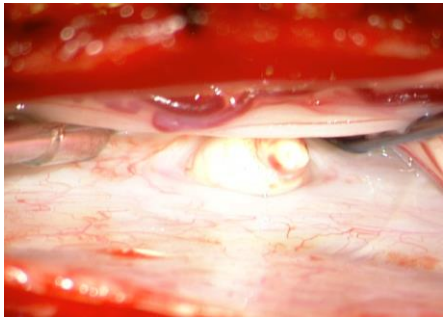
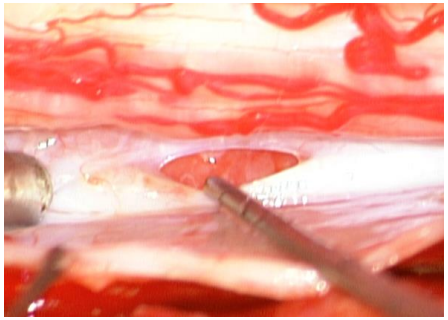
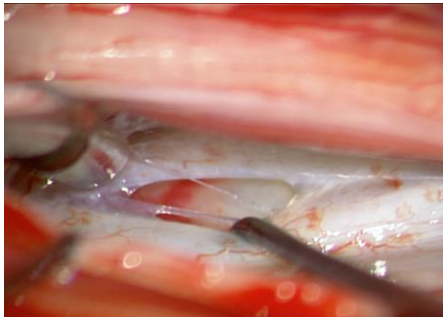
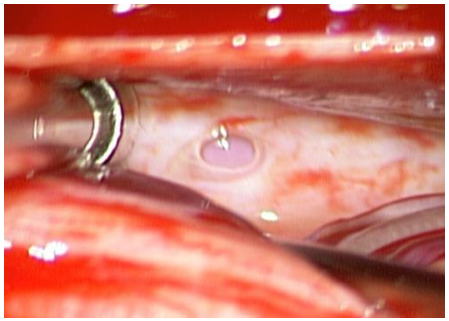
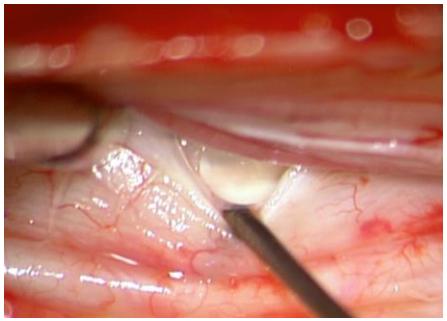
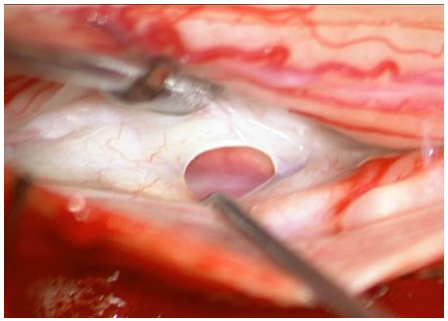
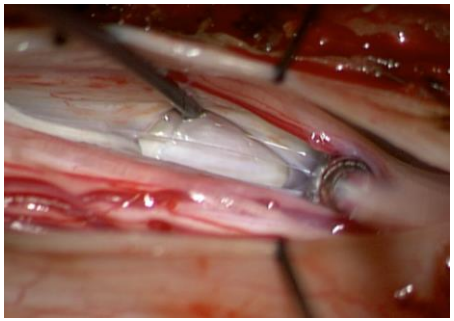
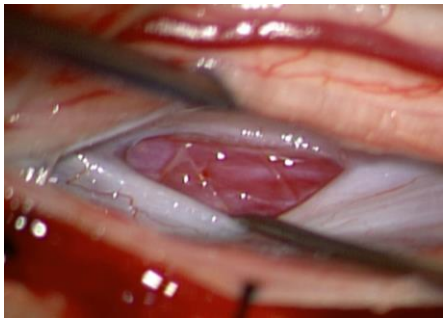
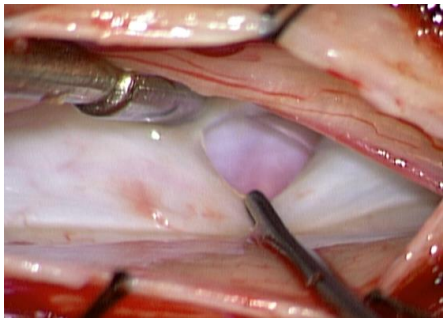
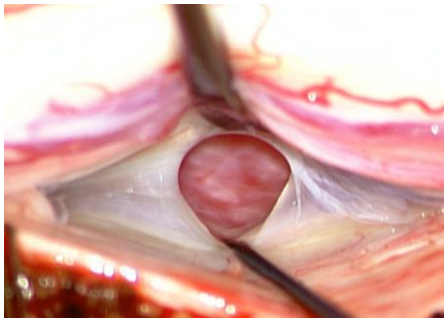
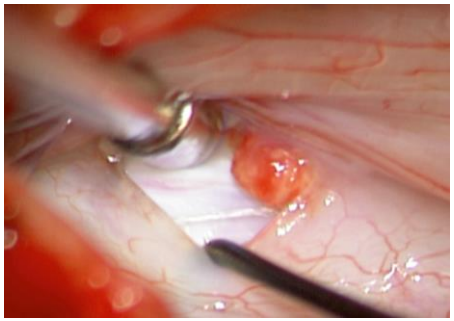

















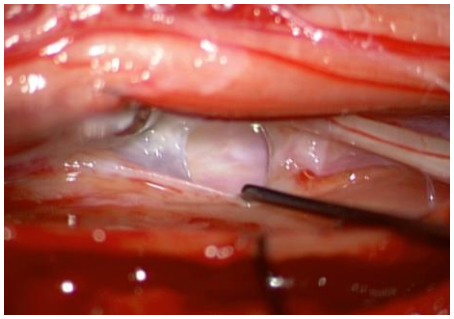
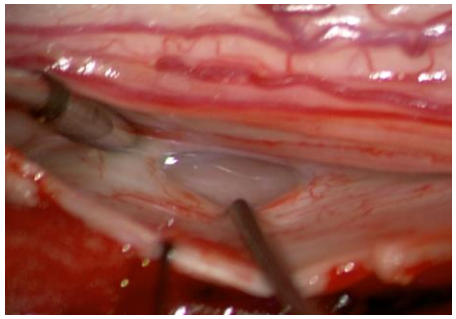
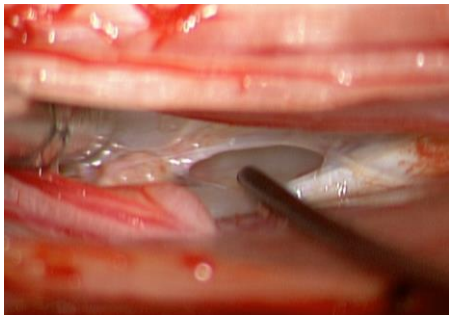
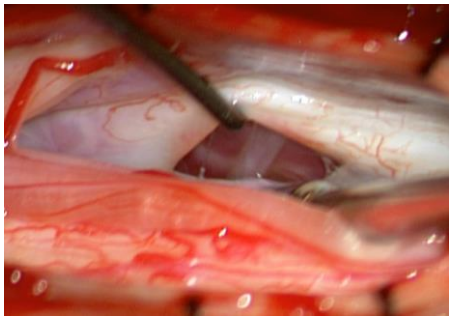
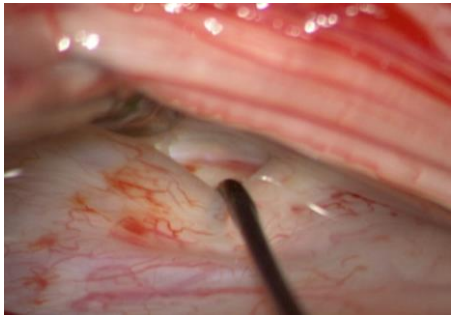
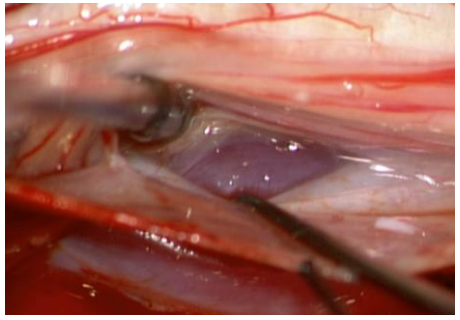
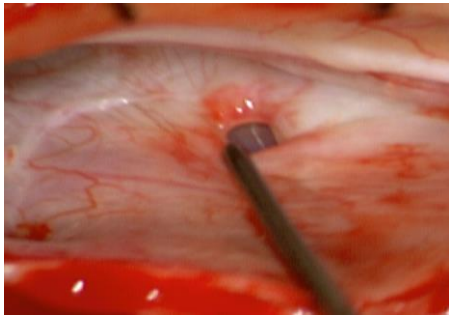
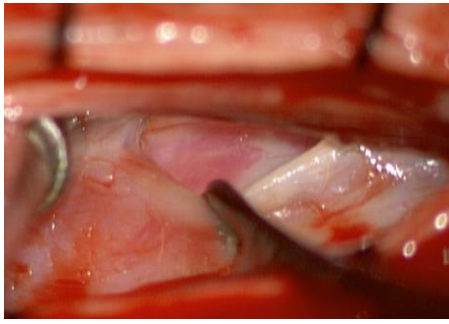
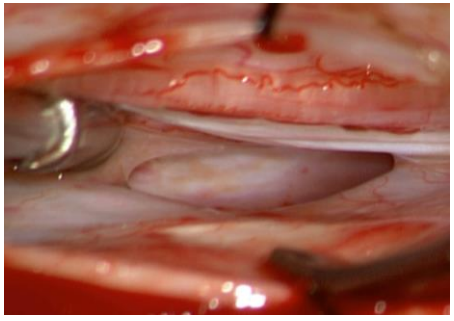
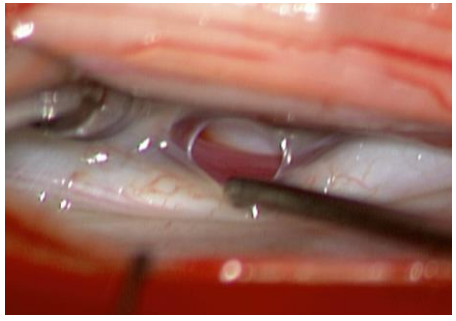
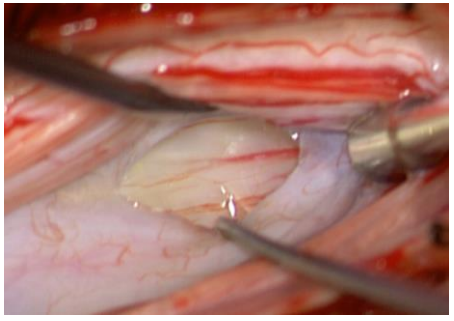
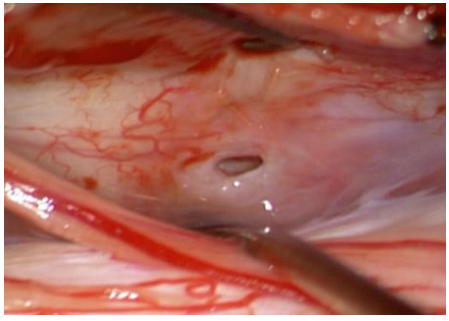
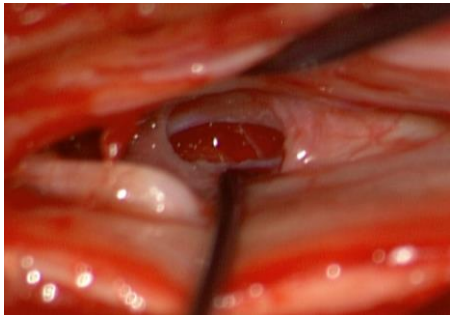
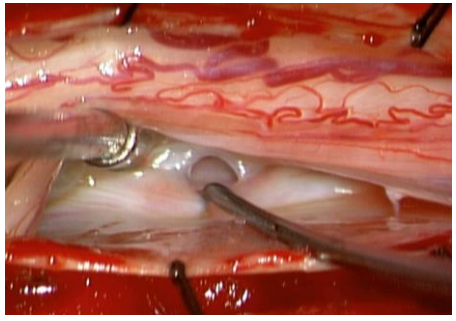
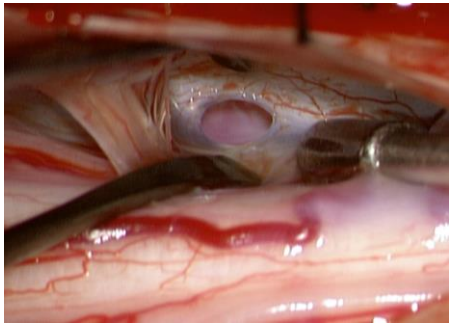
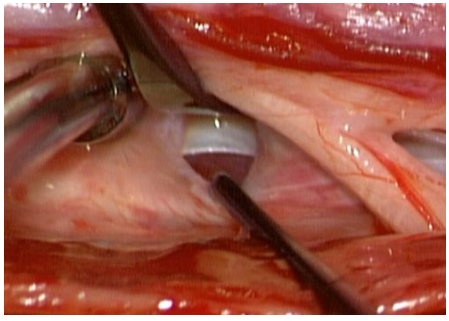









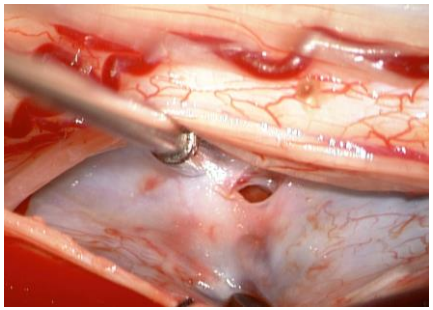
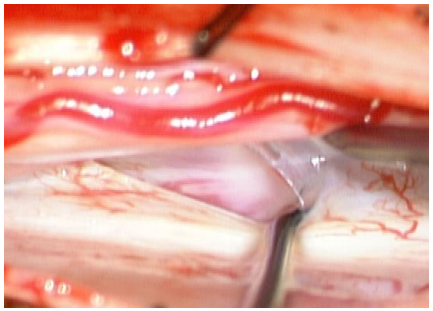
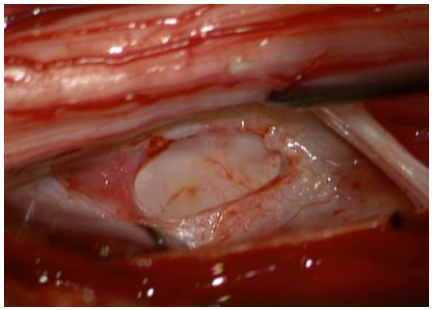
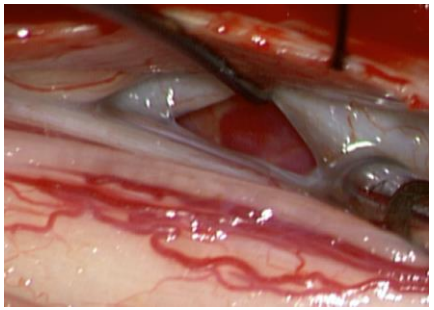
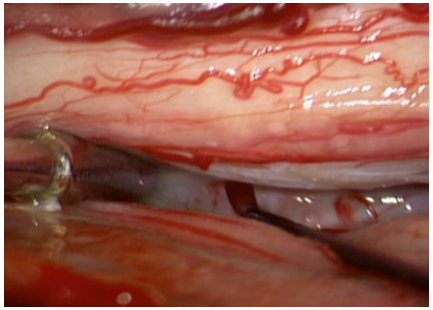
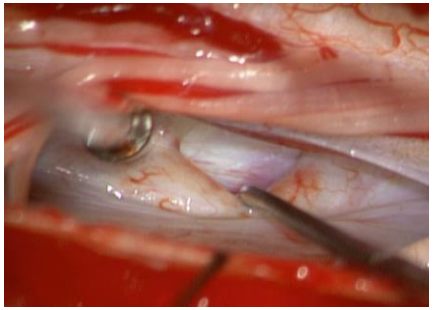
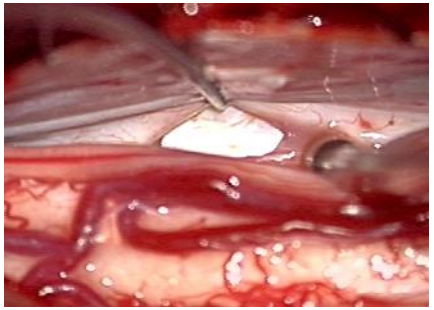
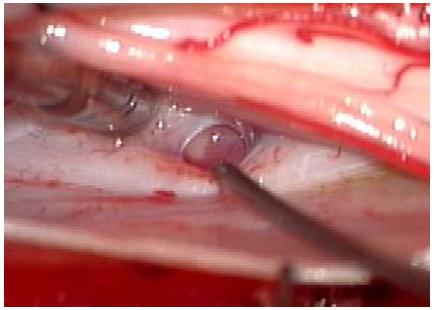
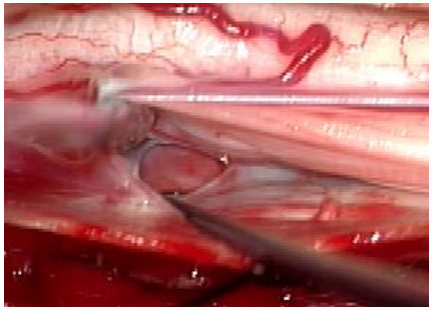
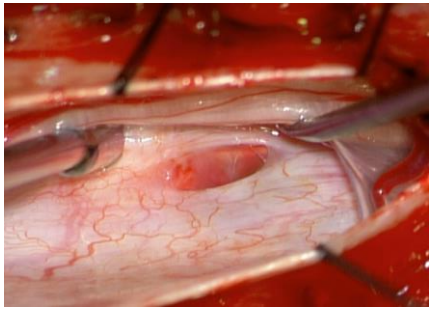
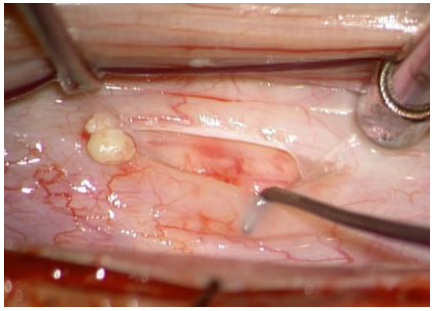
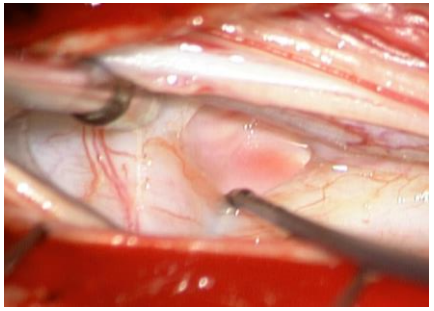
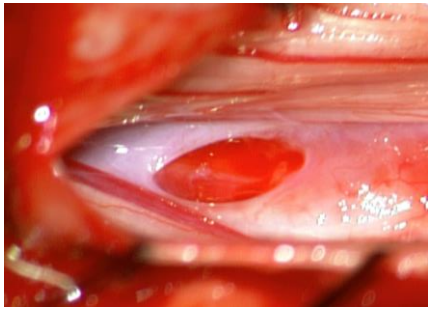
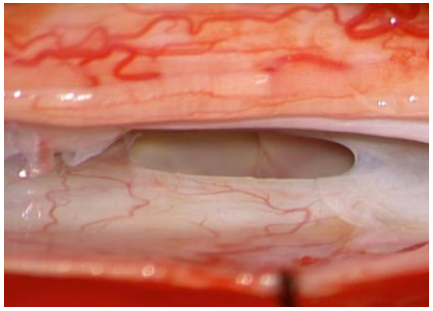
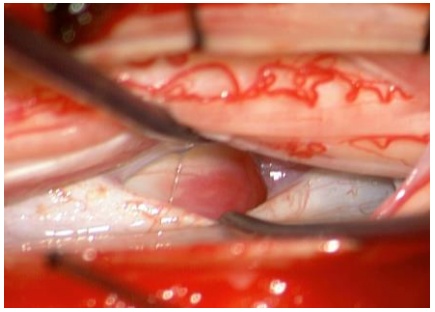
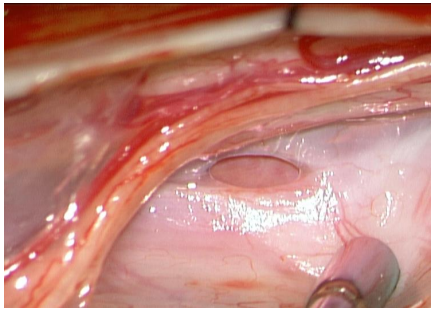








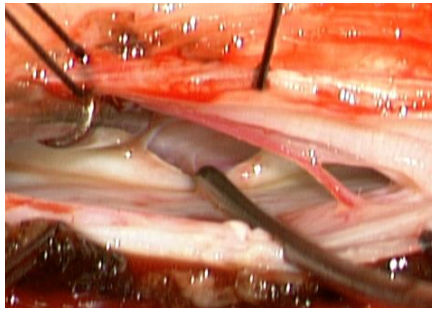
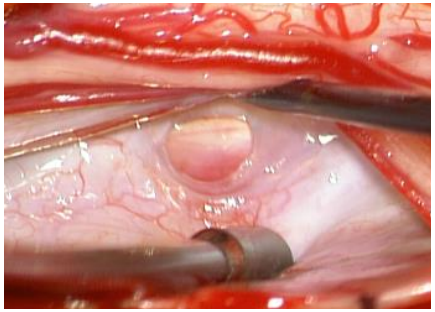
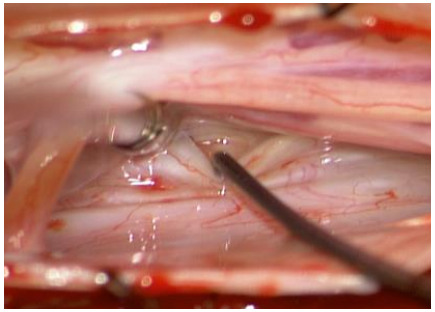
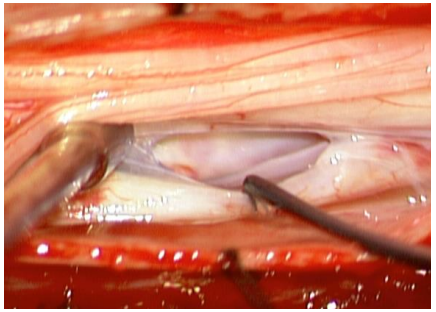
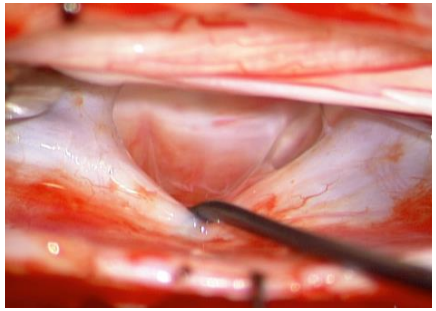
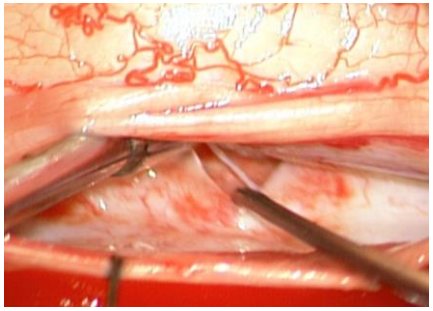
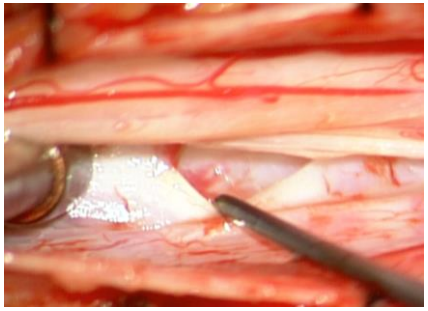
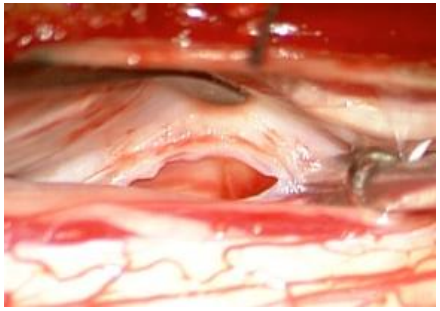
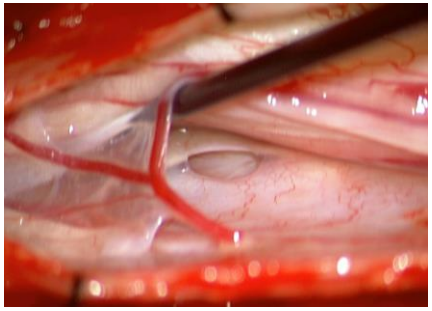
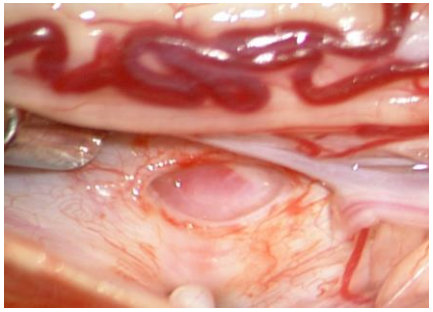
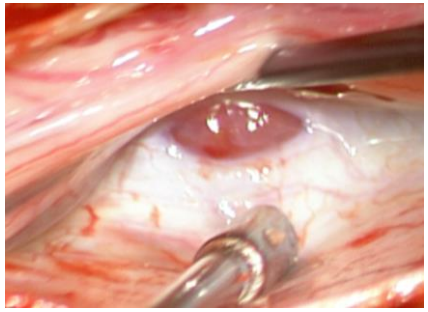

















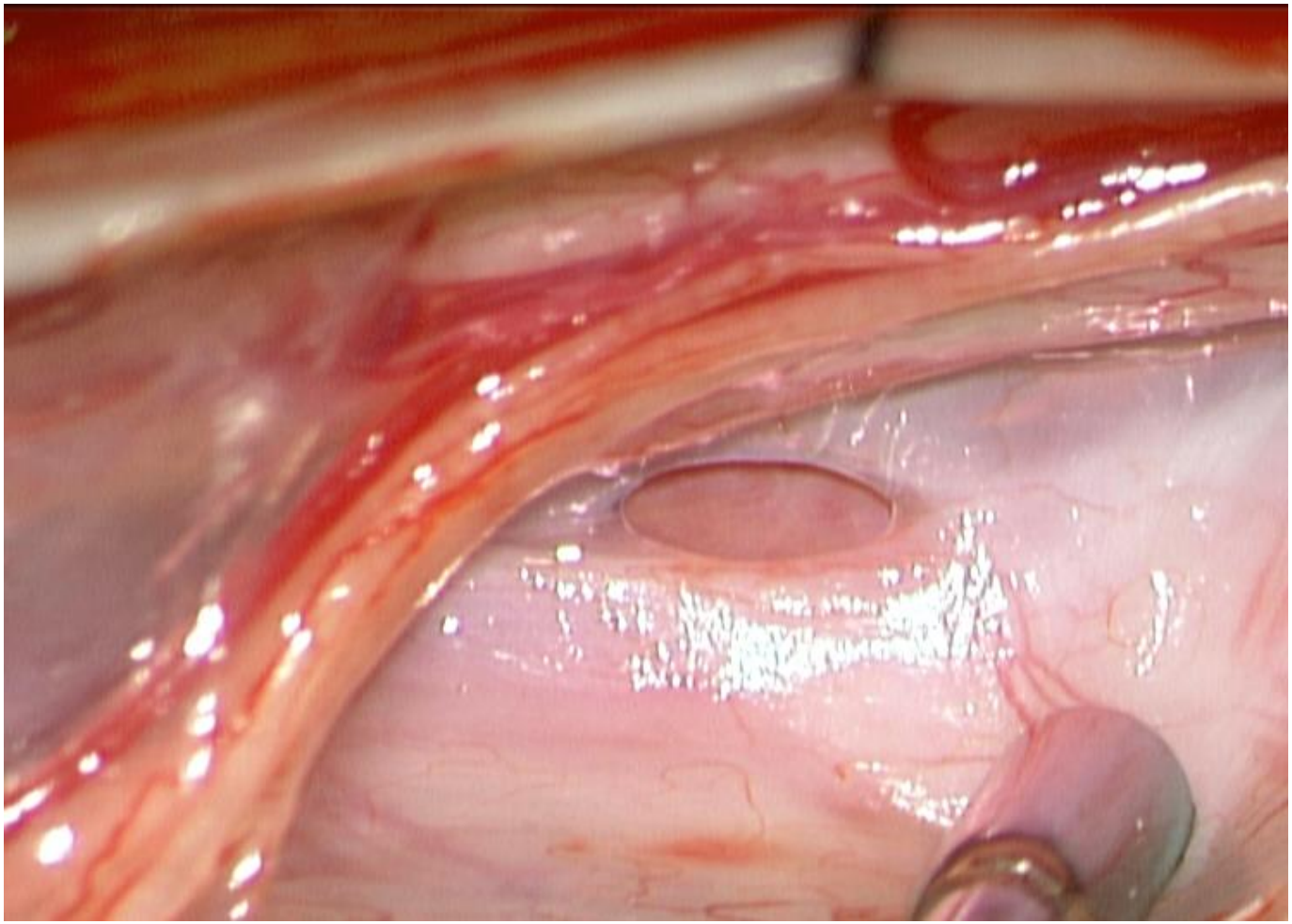

















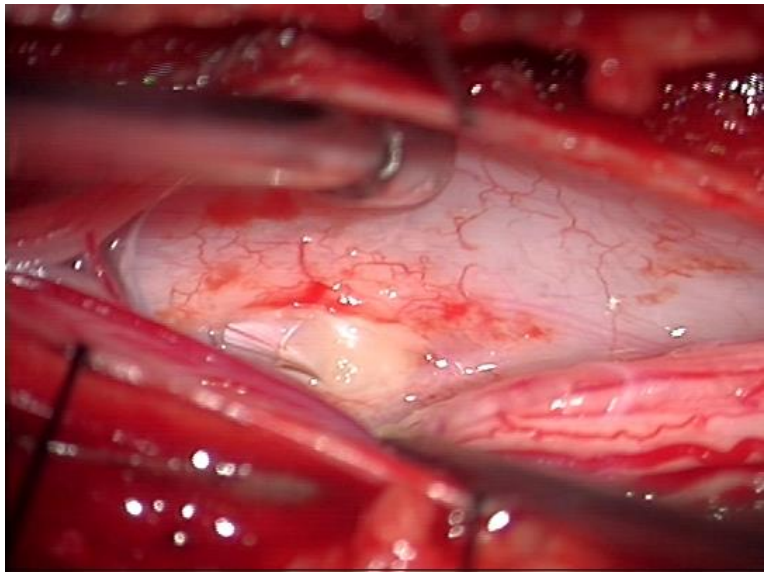
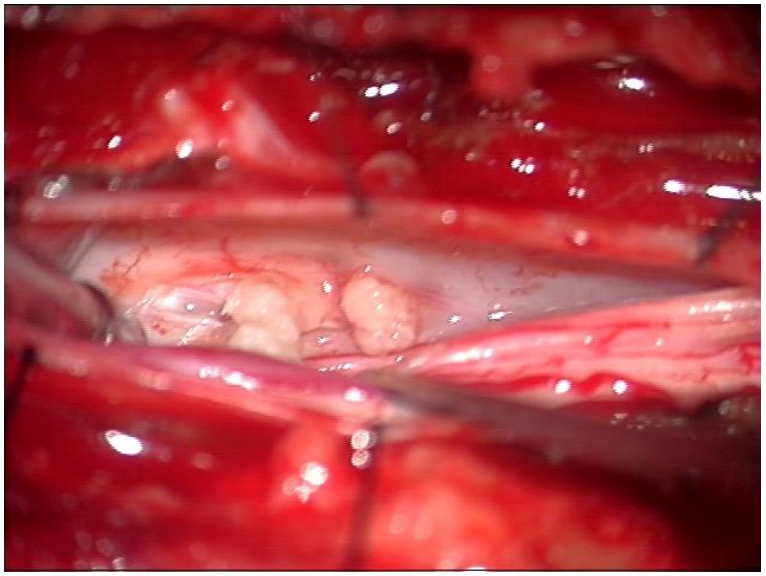
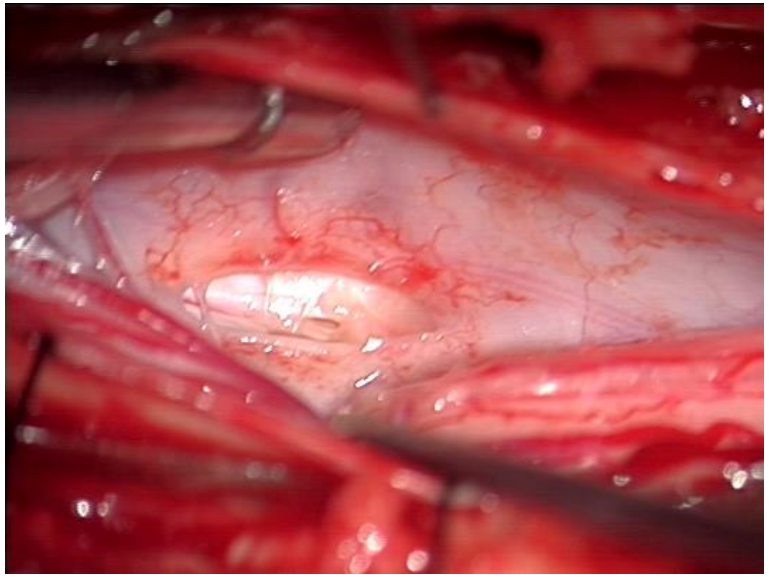
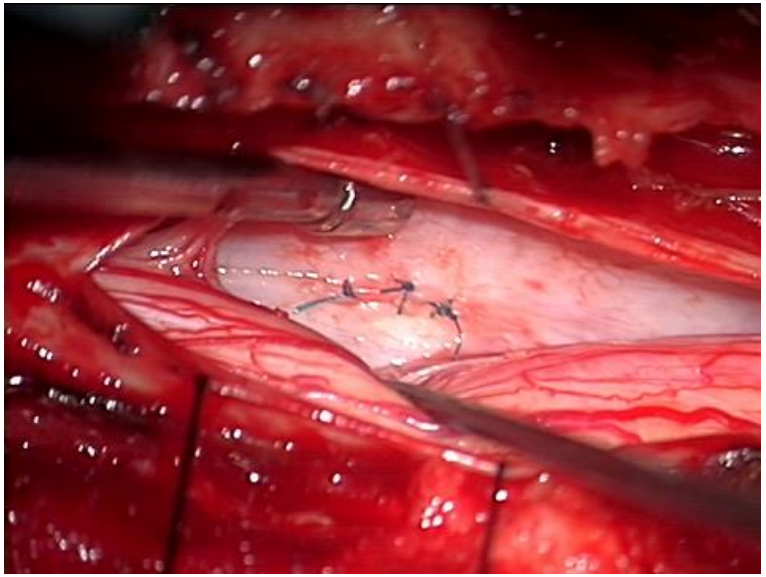

















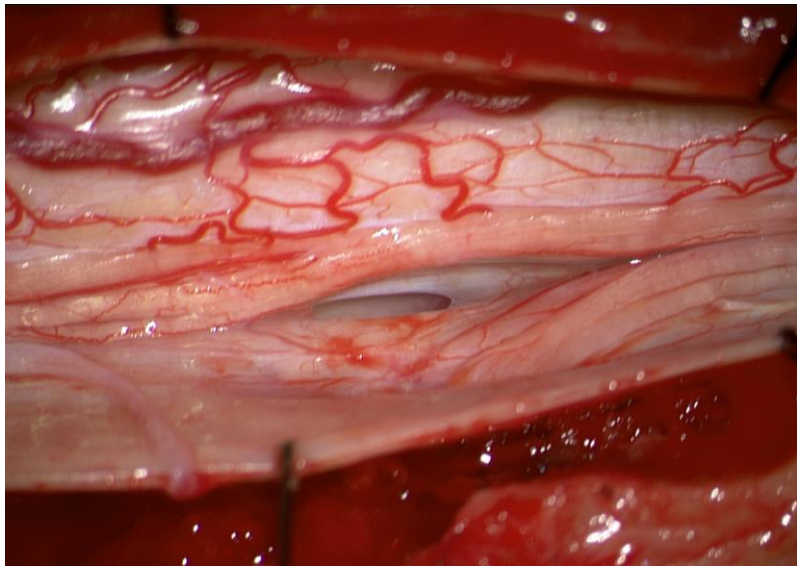
Why take pictures?

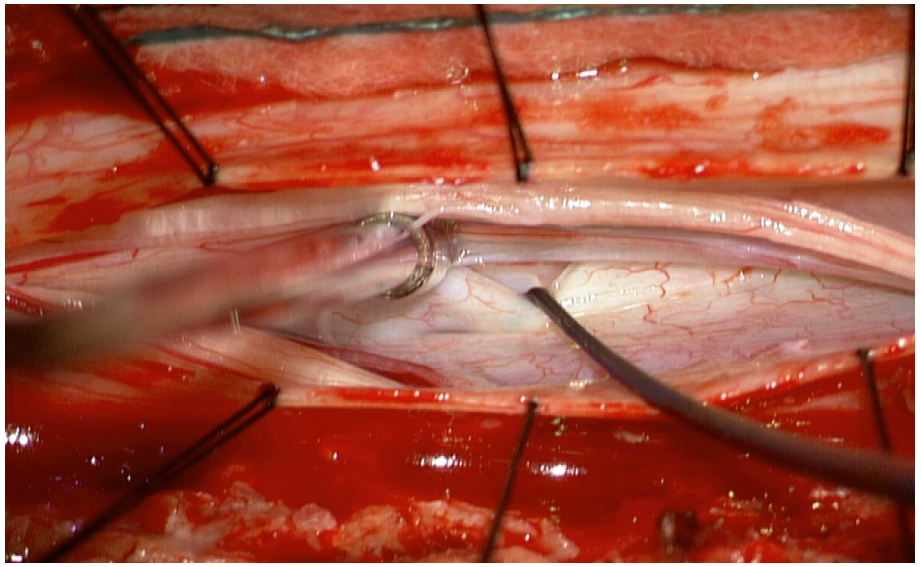


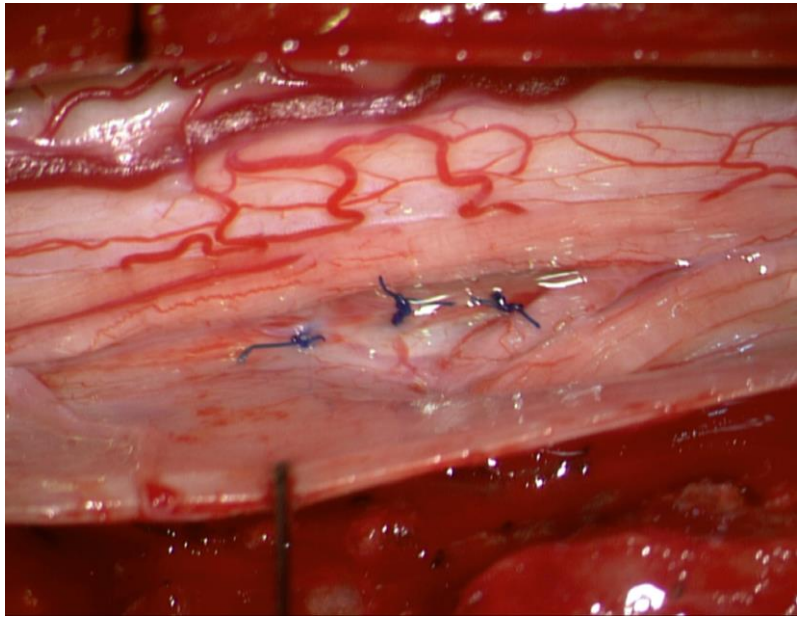
Resection of transdural disc herniation and repair ventral tear

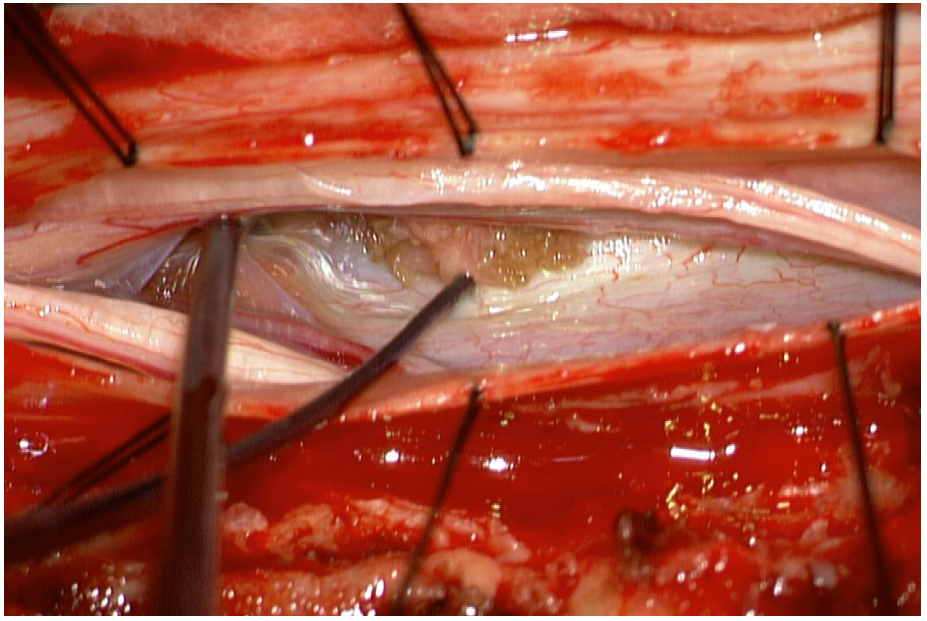





Ventral dural tear: sutures vs muscle repair

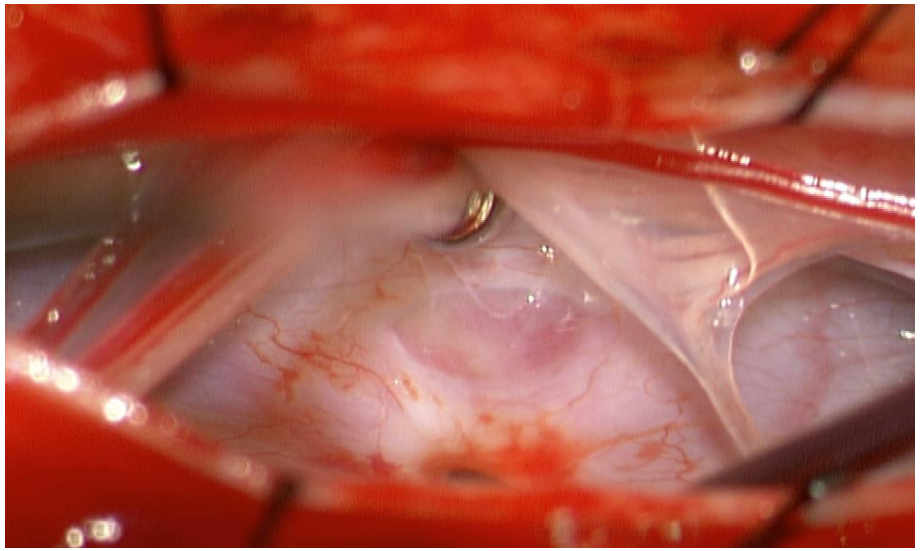


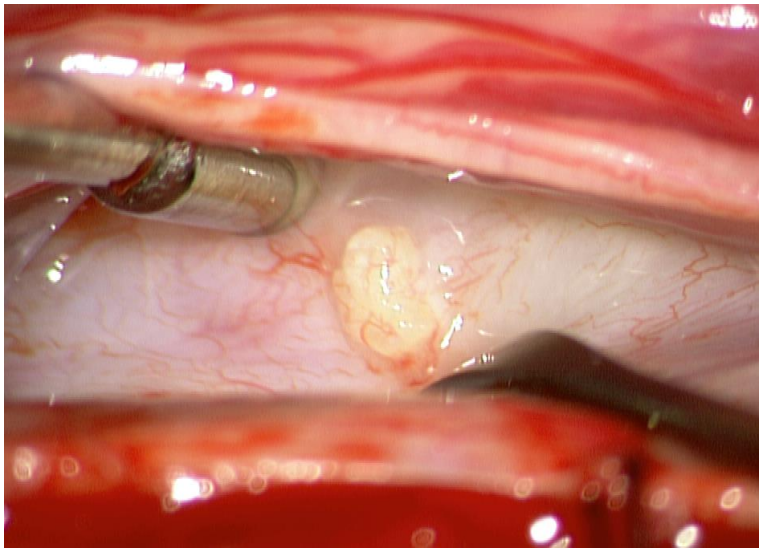


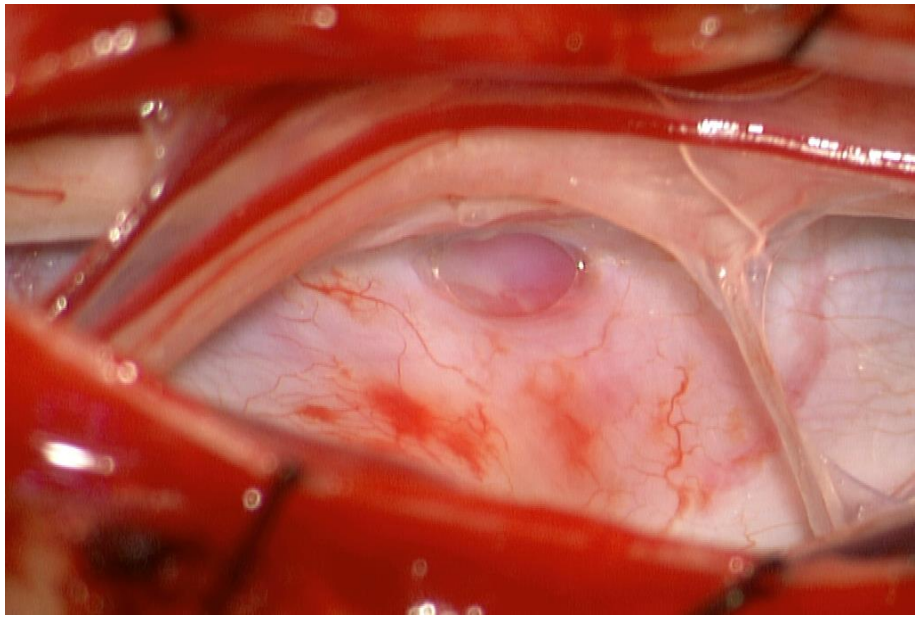


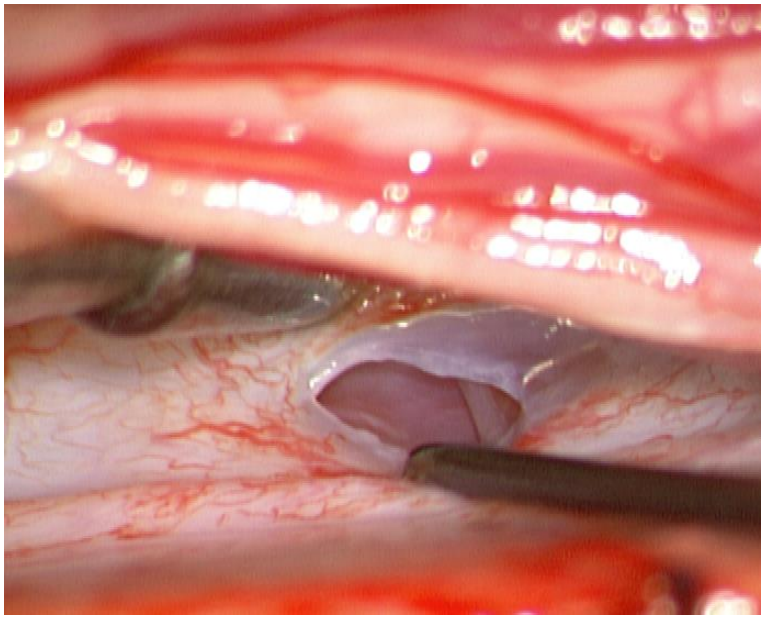


Ventral dural tear without CSF leak (Type 1a-)





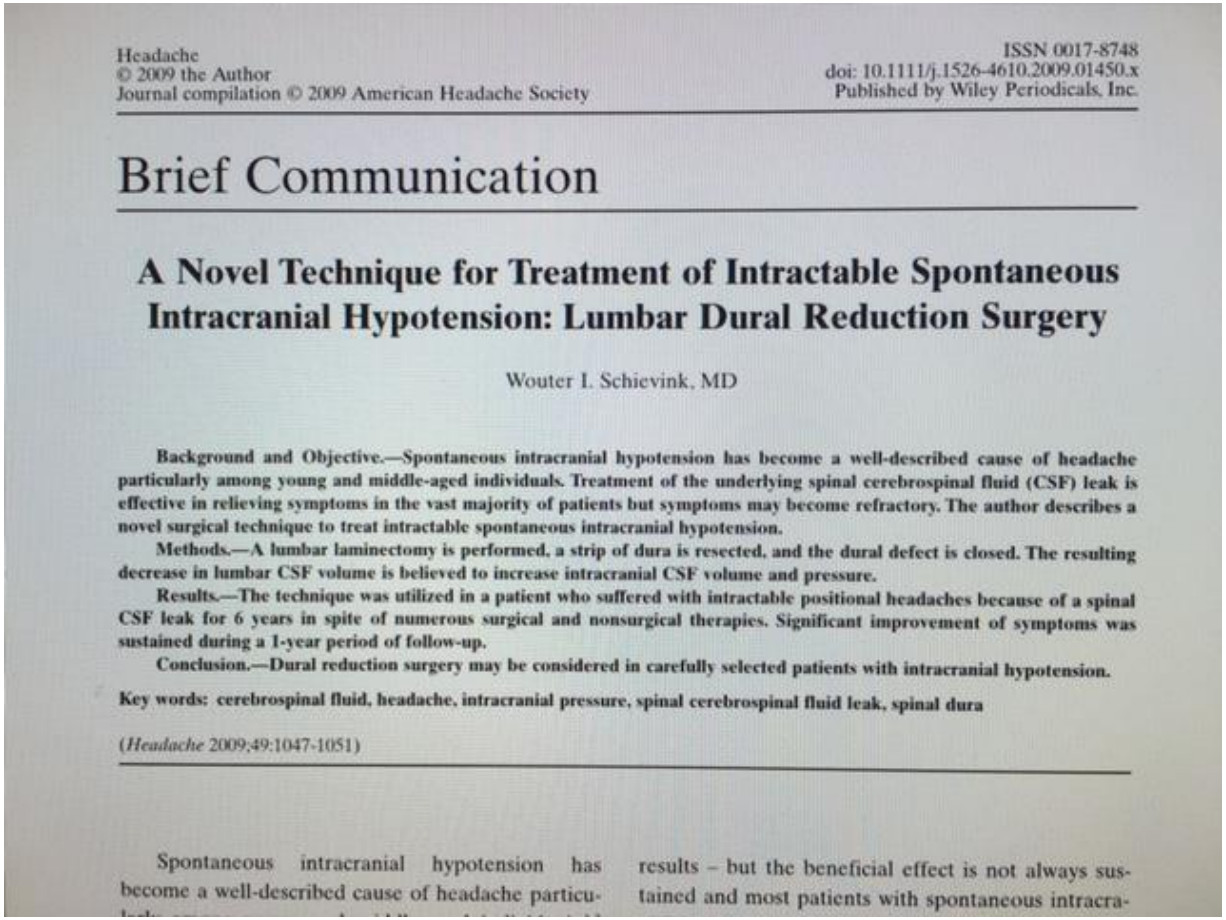




Surgical solutions for the recalcitrant patient

- Lumbar dural reduction surgery
- Wearable epidural saline infusion catheter system
- Craniotomy for brain elevation

Dural reduction surgery



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Brief Communication

A Novel Technique for Treatment of Intractable Spontaneous Intracranial Hypotension: Lumbar Dural Reduction Surgery

Wouter I. Schievink, MD

Background and Objective.—Spontaneous intracranial hypotension has become a well-described cause of headache particularly among young and middle-aged individuals. Treatment of the underlying spinal cerebrospinal fluid (CSF) leak is effective in relieving symptoms in the vast majority of patients but symptoms may become refractory. The author describes a novel surgical technique to treat intractable spontaneous intracranial hypotension.

Methods.—A lumbar laminectomy is performed, a strip of dura is resected, and the dural defect is closed. The resulting decrease in lumbar CSF volume is believed to increase intracranial CSF volume and pressure.

Results.—The technique was utilized in a patient who suffered with intractable positional headaches because of a spinal CSF leak for 6 years in spite of numerous surgical and nonsurgical therapies. Significant improvement of symptoms was sustained during a 1-year period of follow-up.

Conclusion.—Dural reduction surgery may be considered in carefully selected patients with intracranial hypotension.

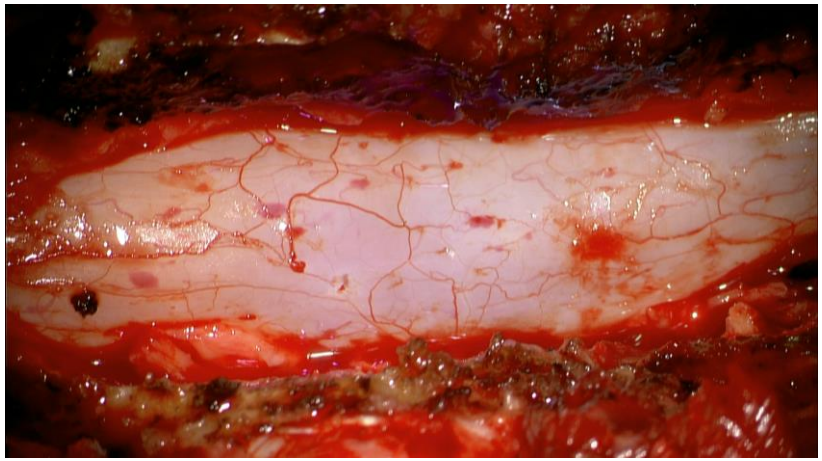
Key words: cerebrospinal fluid, headache, intracranial pressure, spinal cerebrospinal fluid leak, spinal dura

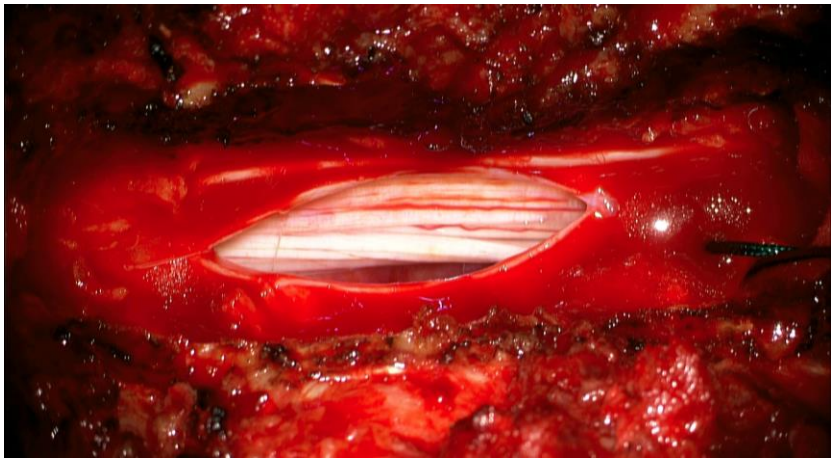
(*Headache* 2009;49:1047-1051)

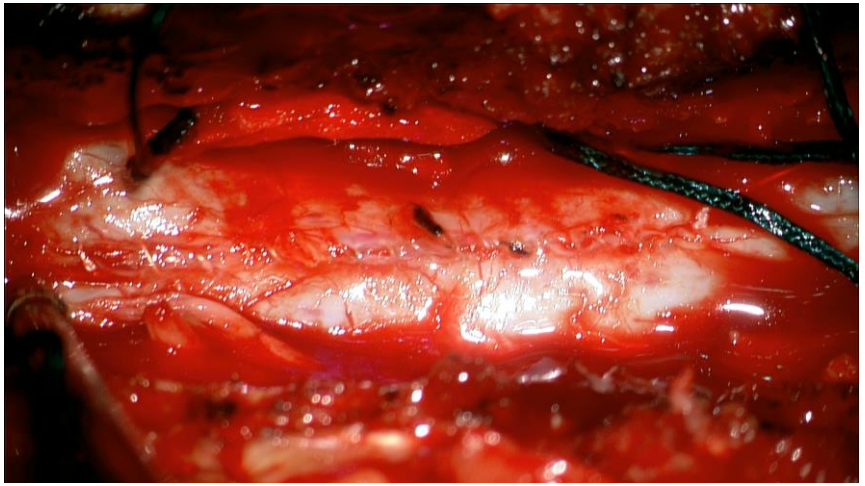
Spontaneous intracranial hypotension has become a well-described cause of headache particu-

results – but the beneficial effect is not always sustained and most patients with spontaneous intracra-

Dural reduction surgery



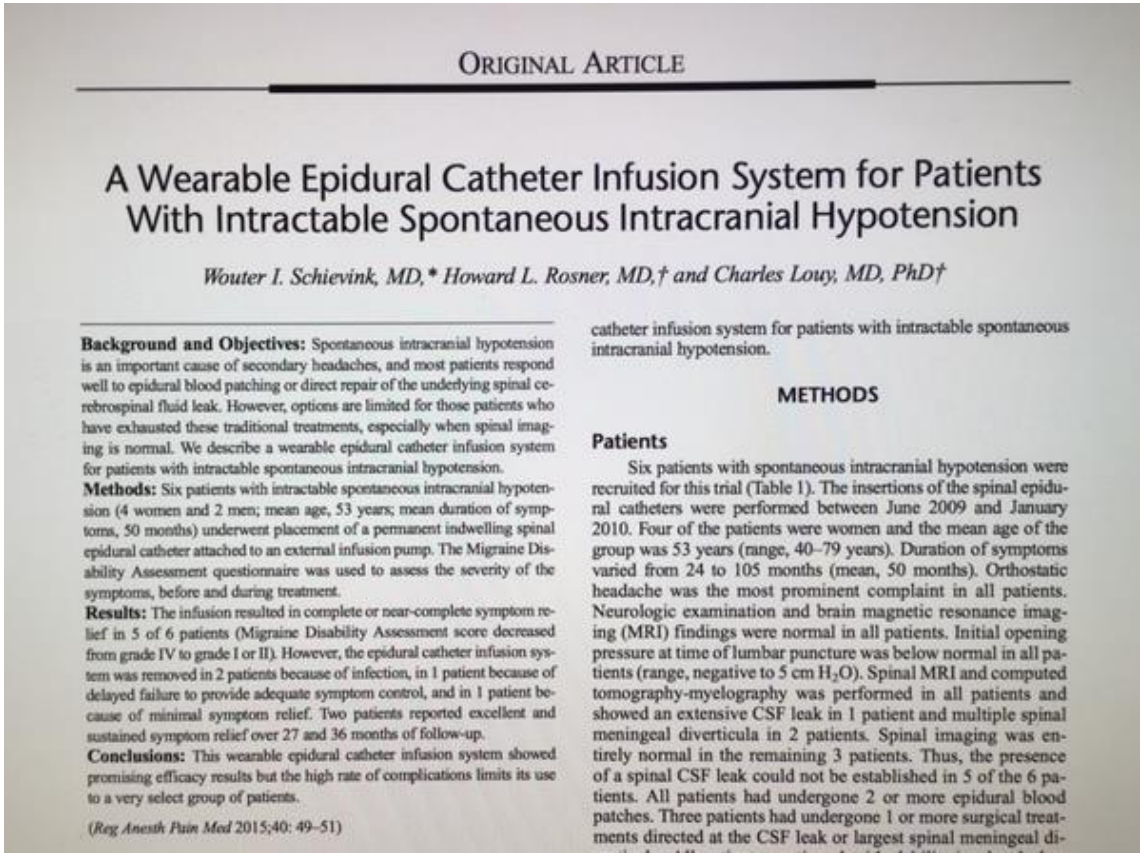




Dural reduction surgery

- N = 52
- 40 women and 12 men (most “without” SIH)
- Age: 21 – 72 years
- Good outcome: 31 (60%)
- Complications: Pseudomeningocele: 5 (10%)
  Suicide: 1 (2%)
  Infection/sepsis: 1 (2%)

Implantation of a wearable epidural spinal infusion system



ORIGINAL ARTICLE

A Wearable Epidural Catheter Infusion System for Patients With Intractable Spontaneous Intracranial Hypotension

Wouter I. Schievink, MD, Howard L. Rosner, MD,† and Charles Louy, MD, PhD†*

Background and Objectives: Spontaneous intracranial hypotension is an important cause of secondary headaches, and most patients respond well to epidural blood patching or direct repair of the underlying spinal cerebrospinal fluid leak. However, options are limited for those patients who have exhausted these traditional treatments, especially when spinal imaging is normal. We describe a wearable epidural catheter infusion system for patients with intractable spontaneous intracranial hypotension.

Methods: Six patients with intractable spontaneous intracranial hypotension (4 women and 2 men; mean age, 53 years; mean duration of symptoms, 50 months) underwent placement of a permanent indwelling spinal epidural catheter attached to an external infusion pump. The Migraine Disability Assessment questionnaire was used to assess the severity of the symptoms, before and during treatment.

Results: The infusion resulted in complete or near-complete symptom relief in 5 of 6 patients (Migraine Disability Assessment score decreased from grade IV to grade I or II). However, the epidural catheter infusion system was removed in 2 patients because of infection, in 1 patient because of delayed failure to provide adequate symptom control, and in 1 patient because of minimal symptom relief. Two patients reported excellent and sustained symptom relief over 27 and 36 months of follow-up.

Conclusions: This wearable epidural catheter infusion system showed promising efficacy results but the high rate of complications limits its use to a very select group of patients.

(*Reg Anesth Pain Med* 2015;40: 49–51)

catheter infusion system for patients with intractable spontaneous intracranial hypotension.

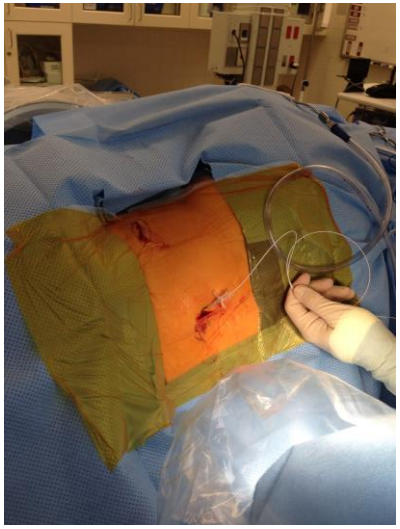
METHODS

Patients

Six patients with spontaneous intracranial hypotension were recruited for this trial (Table 1). The insertions of the spinal epidural catheters were performed between June 2009 and January 2010. Four of the patients were women and the mean age of the group was 53 years (range, 40–79 years). Duration of symptoms varied from 24 to 105 months (mean, 50 months). Orthostatic headache was the most prominent complaint in all patients. Neurologic examination and brain magnetic resonance imaging (MRI) findings were normal in all patients. Initial opening pressure at time of lumbar puncture was below normal in all patients (range, negative to 5 cm H_2O). Spinal MRI and computed tomography-myelography was performed in all patients and showed an extensive CSF leak in 1 patient and multiple spinal meningeal diverticula in 2 patients. Spinal imaging was entirely normal in the remaining 3 patients. Thus, the presence of a spinal CSF leak could not be established in 5 of the 6 patients. All patients had undergone 2 or more epidural blood patches. Three patients had undergone 1 or more surgical treatments directed at the CSF leak or largest spinal meningeal di-

A

B



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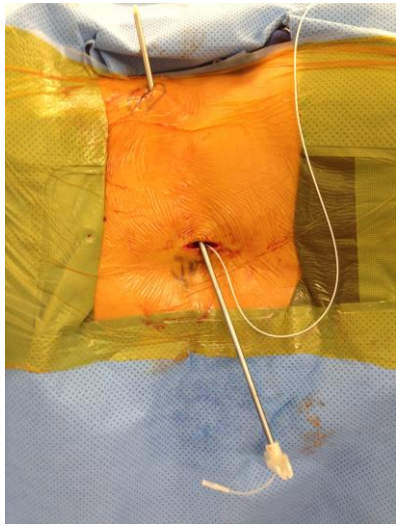

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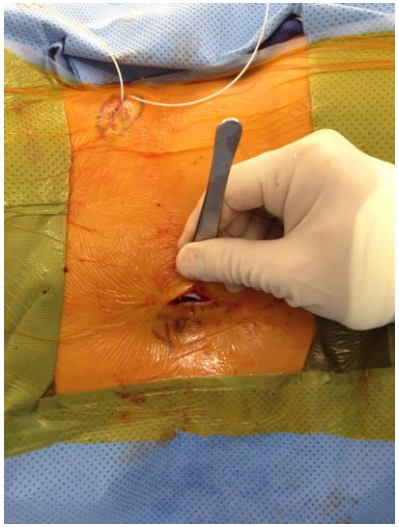

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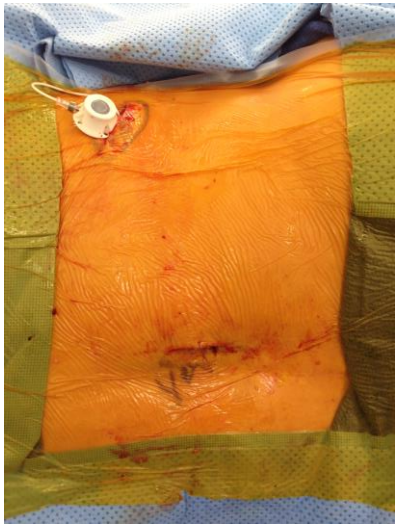


H

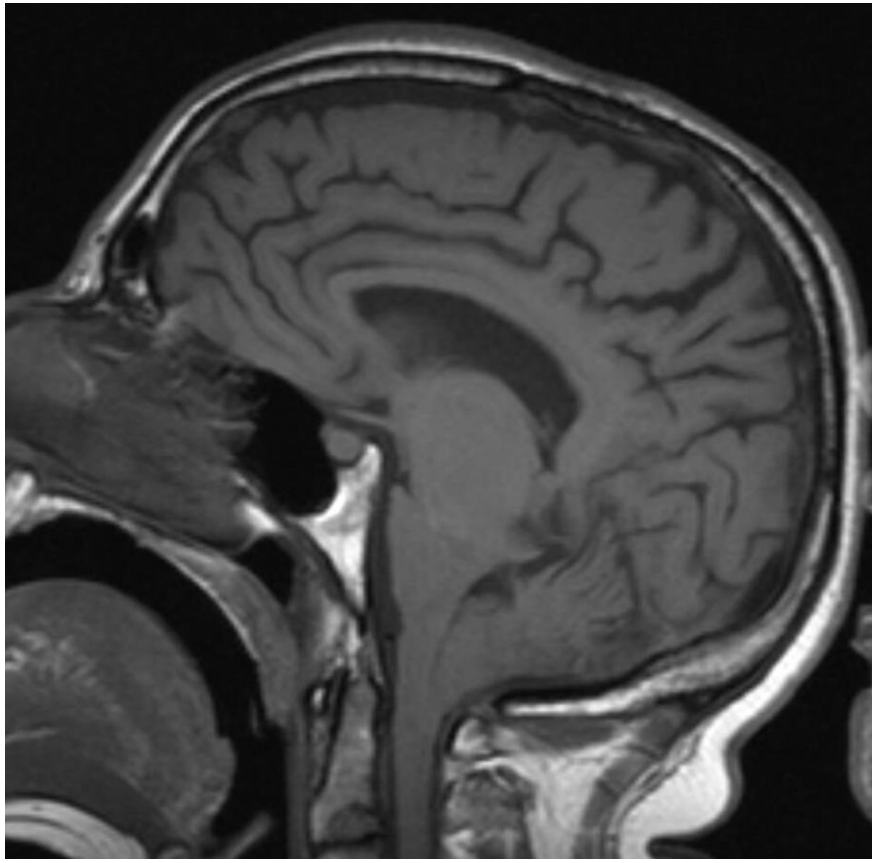
Spinal epidural infusion system

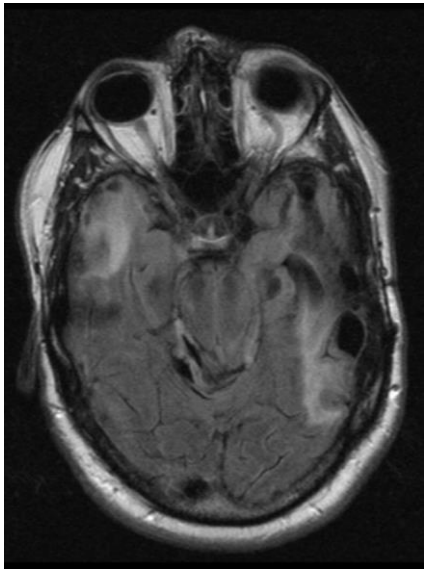
- N = 19
- 11 women and 8 men (All but 2 with SIH)
- Age: 35 – 79 years
- Good outcome: 17 (89%)

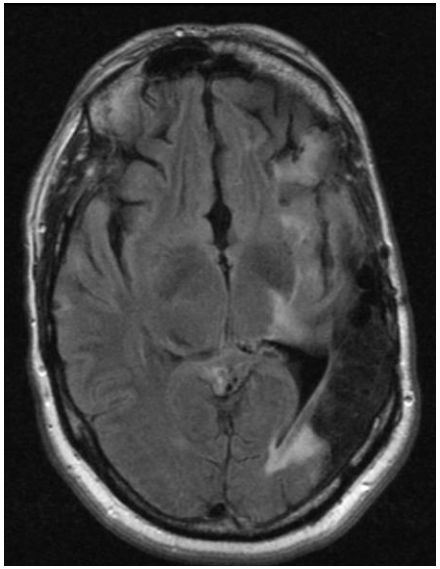
- Complications: Infection: 3 (16%)
  Hardware failure: 7 (37%)

Craniotomy for brain elevation

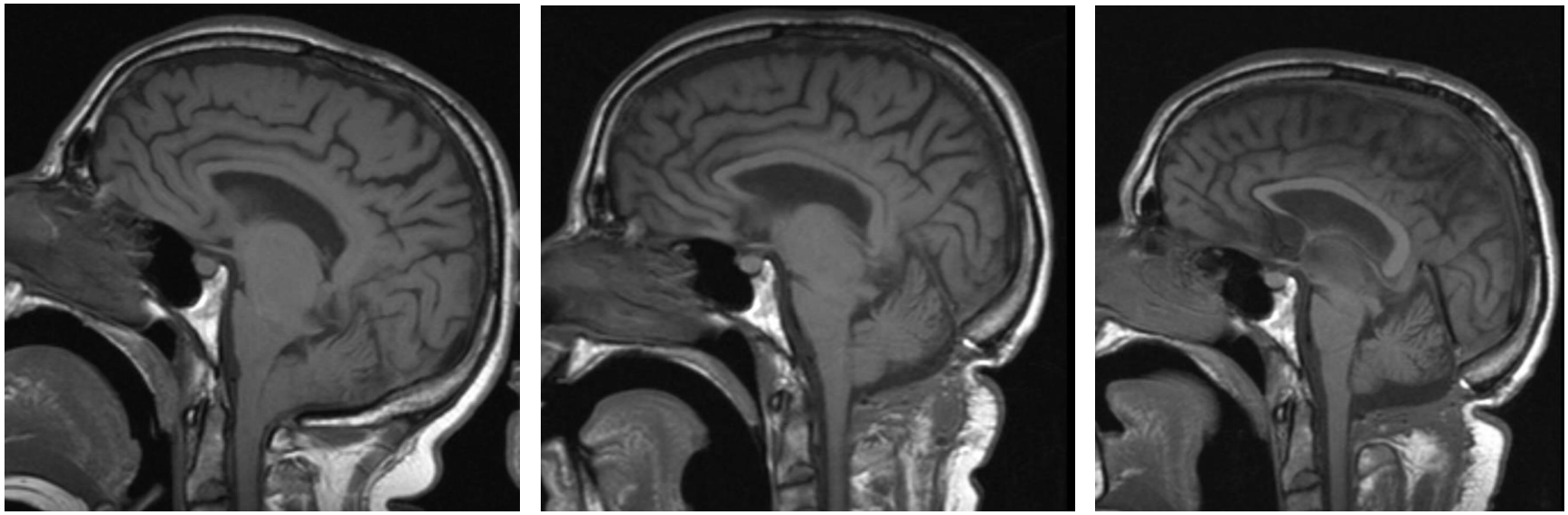
Craniotomy for brain elevation







Craniotomy for brain elevation



Thank you