Spontaneous intracranial hypotension – May 24, 2013

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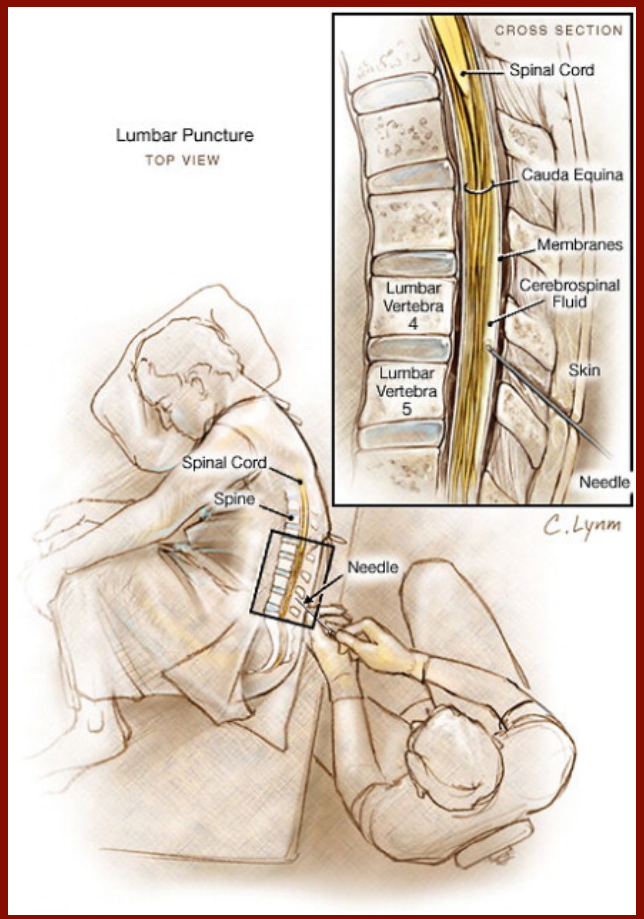
Post-dural puncture headache



Nomenclature

- Aliquorrhoea (Schaltenbrand 1936)
- Hypoliquorrhoea
- Intracranial hypotension → CSF hypovolemia
- Spontaneous spinal CSF leak

Epidemiology of spontaneous intracranial hypotension

- Prevalence: 1/50,000 (Olmsted County, MN)
- Incidence: 5/100,000/yr (LA County, CA)

How to recognize your patient

Cause of intracranial hypotension is

- **Spinal CSF leak**
- Inadequate CSF production?
- Rapid CSF absorption?

- ? CSF rhinorrhea–otorrhoea ?

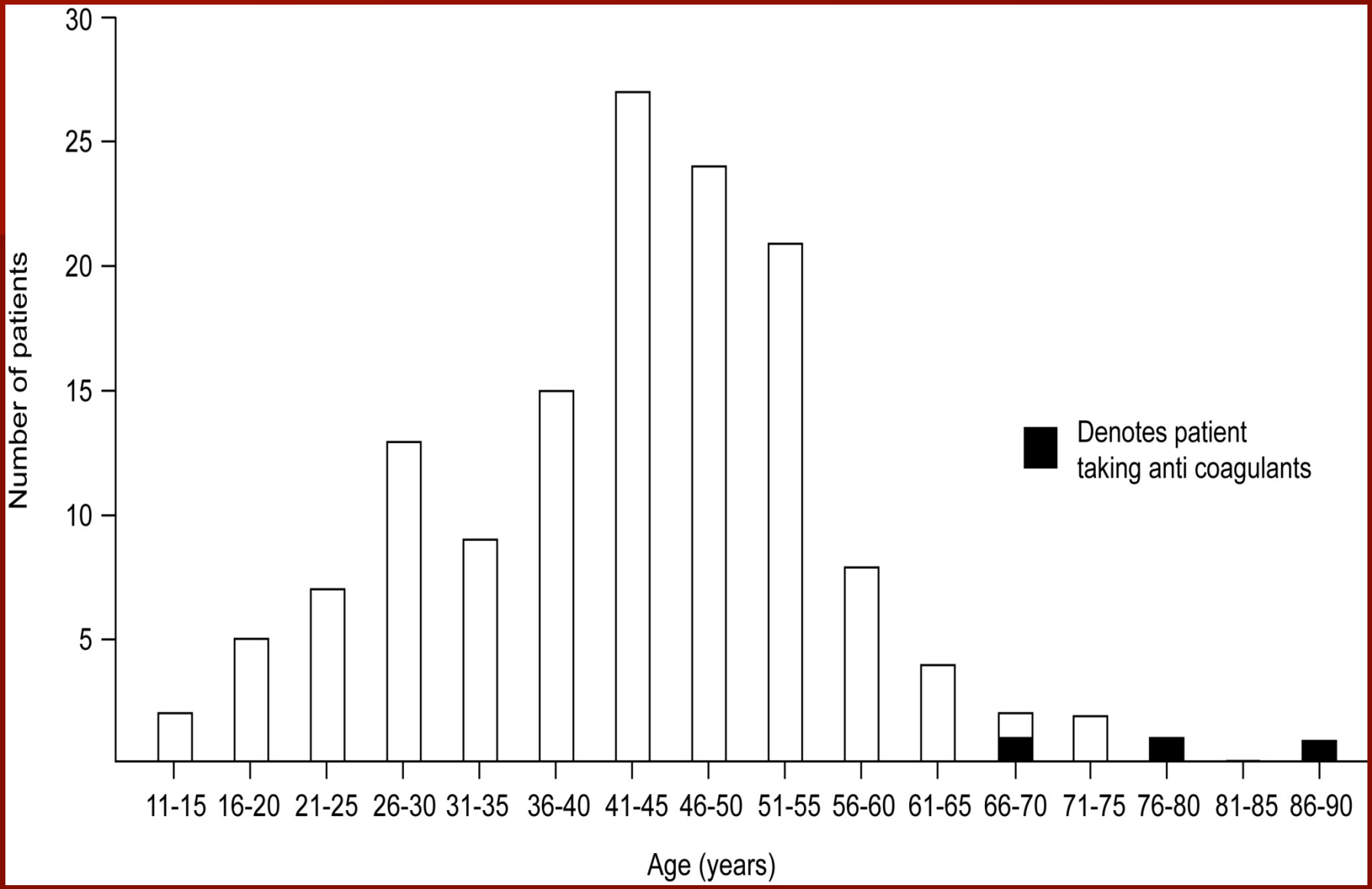
  NEVER

Spinal CSF Leak

- Cyst
- Dural hole / tear
- Bony abnormality
- Nude nerve root

Age / Sex Distribution

- **Mean age:** 43.4
- **Range:** 2 – 86
- **Sex:** 1.5 F/M ratio


30
25
20
15
10
5
Number of patients
Denotes patient
taking anti coagulants
11-15
16-20
21-25
26-30
31-35
36-40
41-45
46-50
51-55
56-60
61-65
66-70
71-75
76-80
81-85
86-90
Age (years)

N = 150

Connective Tissue Disorders

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

33/150 (22%)

Pediatric spontaneous intracranial hypotension (n=24)

- Systemic connective tissue disorders in 53%

Marfan syndrome: n = 3

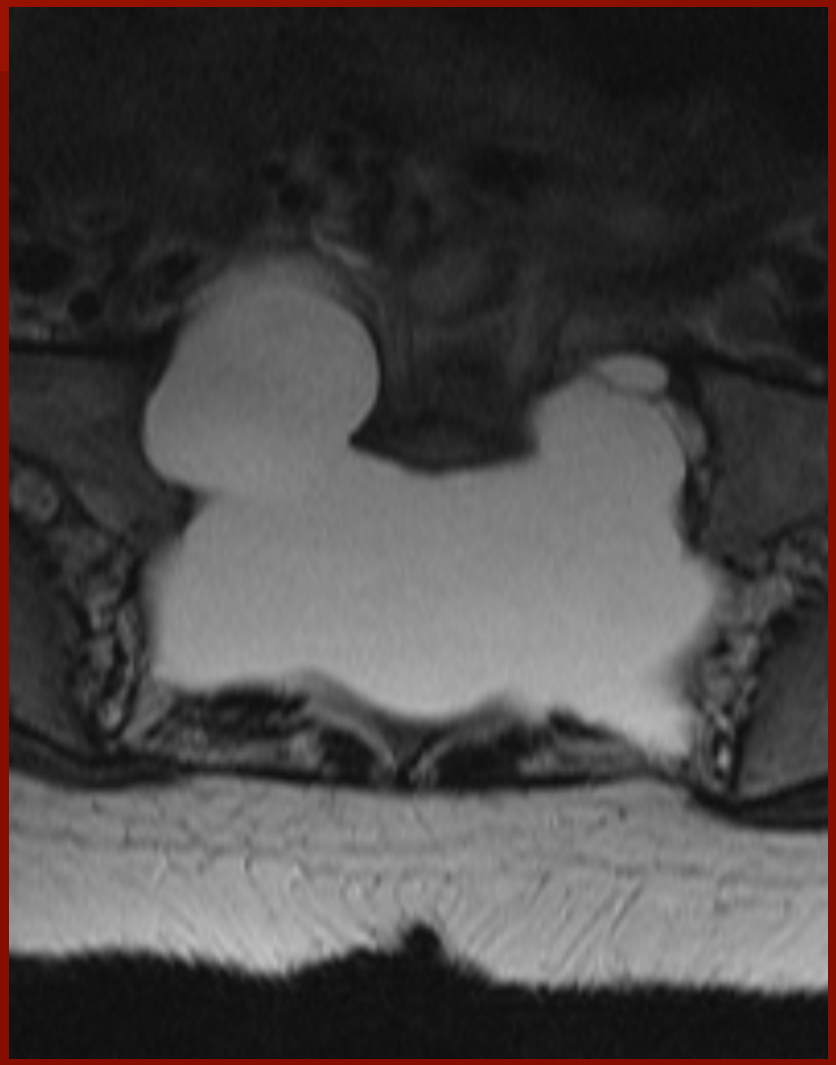
Ehlers-Danlos syndrome type III: n = 2

Undetermined: n = 8 (Congenital contractures/ bifid uvula; Marfanoid x3; Hypomelanosis of Ito; Coloboma)

Connective tissue disorders in spontaneous intracranial hypotension

- Abnormalities on EM in about 20% of patients

Dural ectasia in Marfan syndrome



Clinical Manifestations

- 90 – 100% Headache
- 50 – 90% Neck Pain
- 25 – 50%
  - Nausea / Vomiting
  - Hearing abnormalities
- 1 – 25%
  - Diplopia (CN VI or III)
  - Cognitive decline/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

Spontaneous intracranial hypotension – Asymptomatic to stroke/death

CT findings

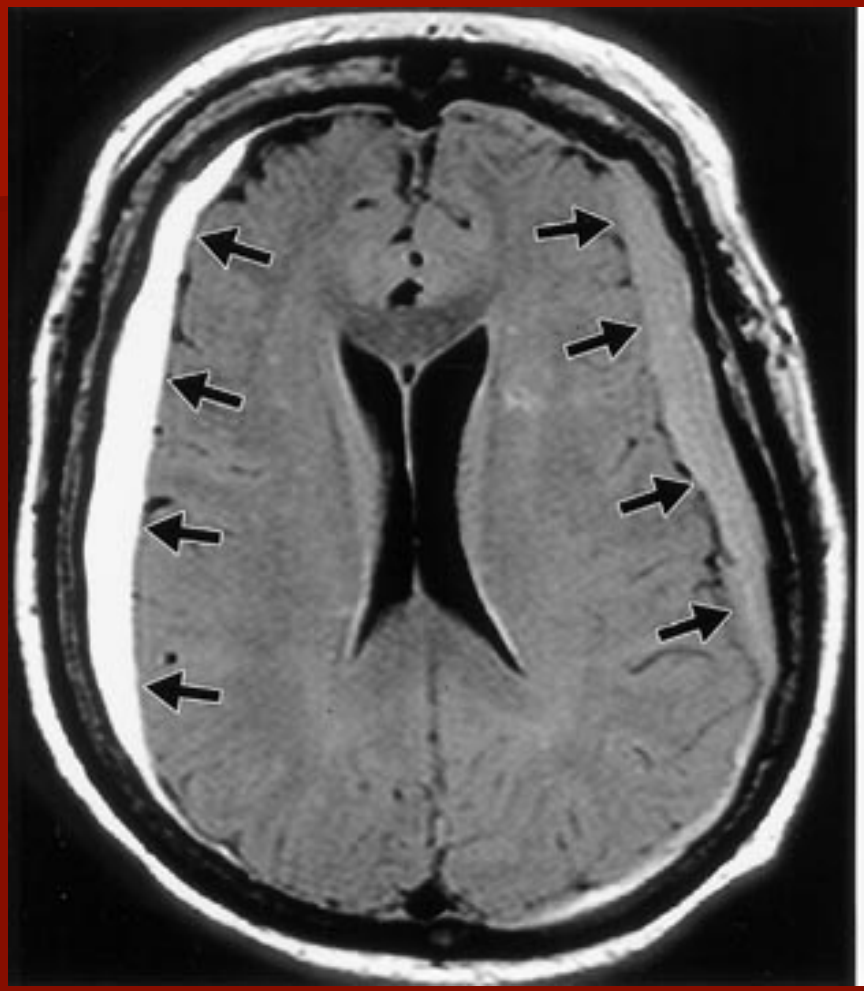
- Normal: 80+%
- Subdural fluid collections: 15-20%
- Pseudo-SAH: 5%

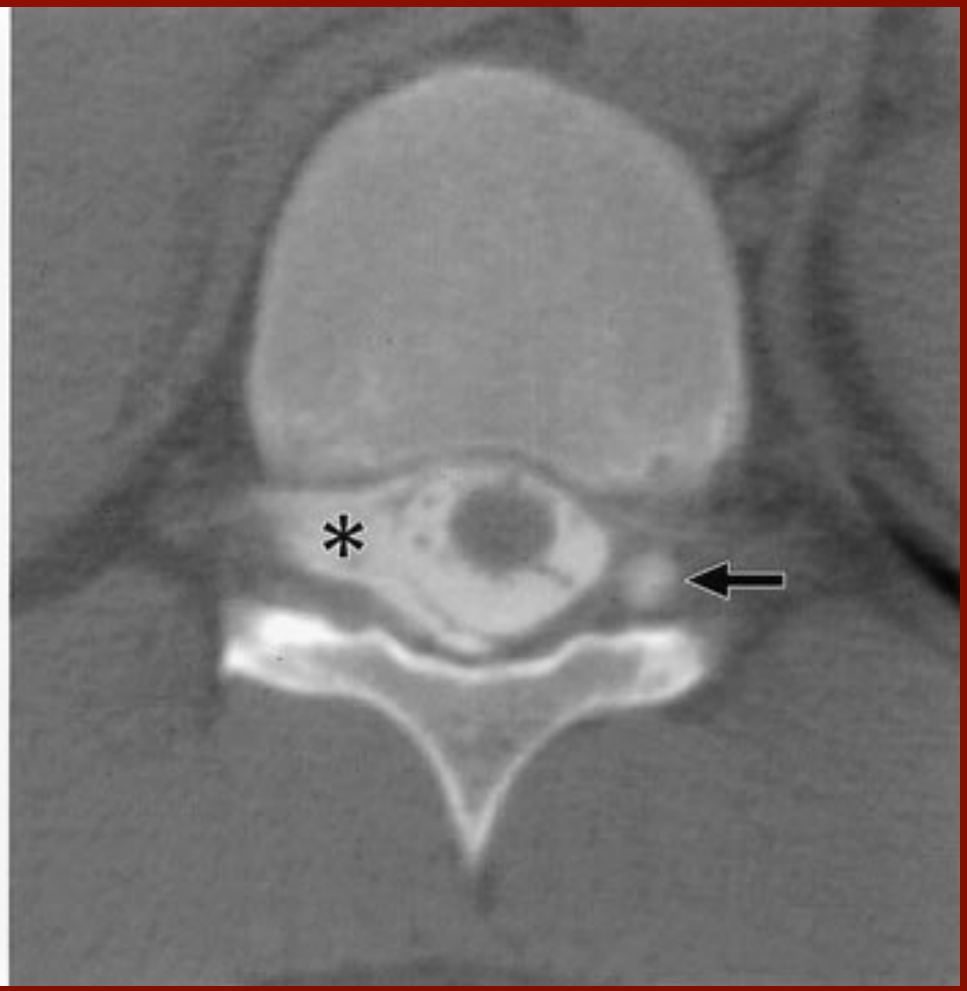
LP findings

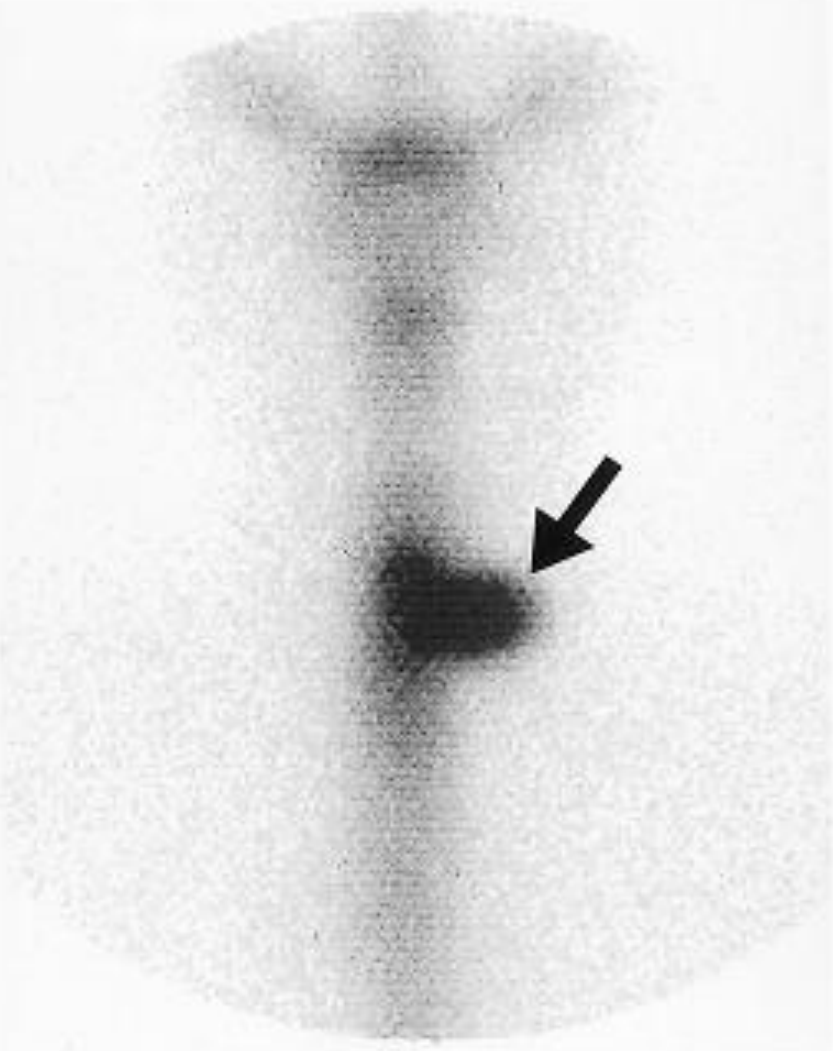
- Difficult to perform
- Traumatic tap
- Opening pressure <7 CM H_2O
- Elevated RBC, TP, and WBC

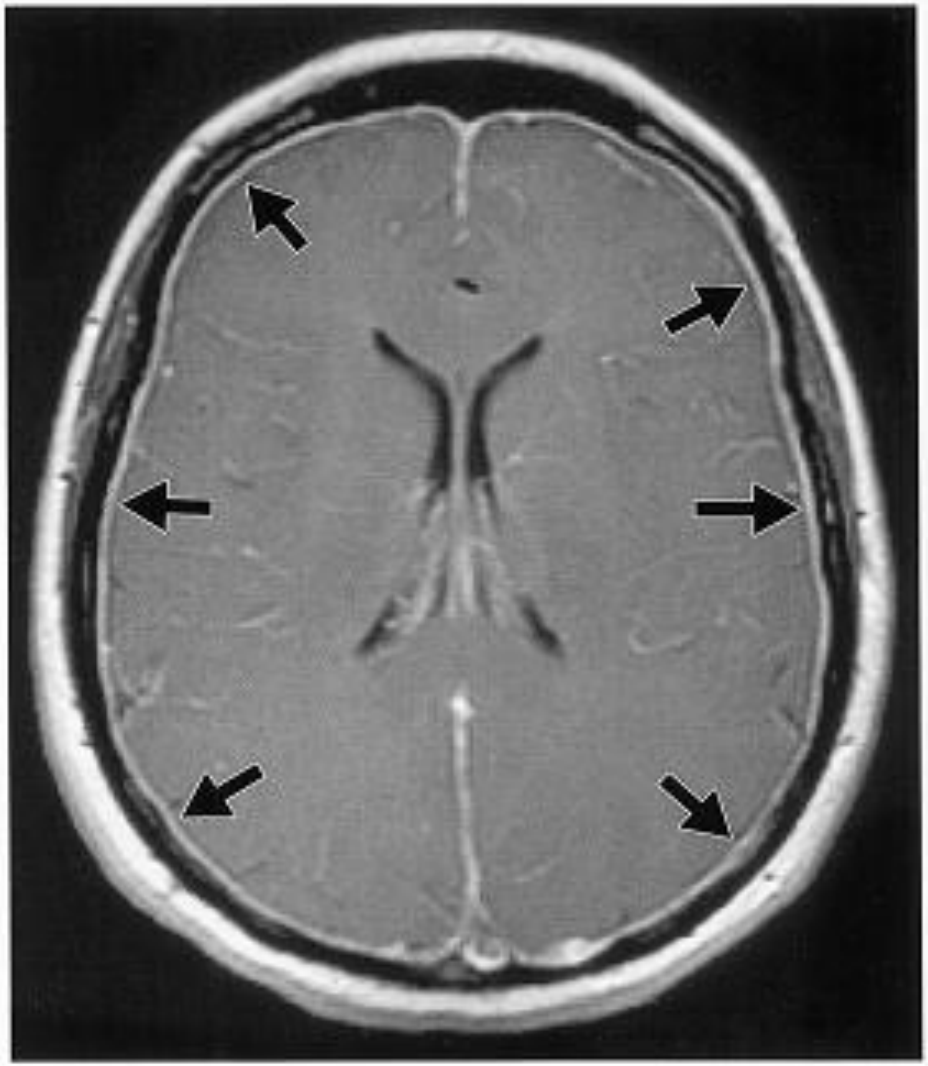
MRI findings

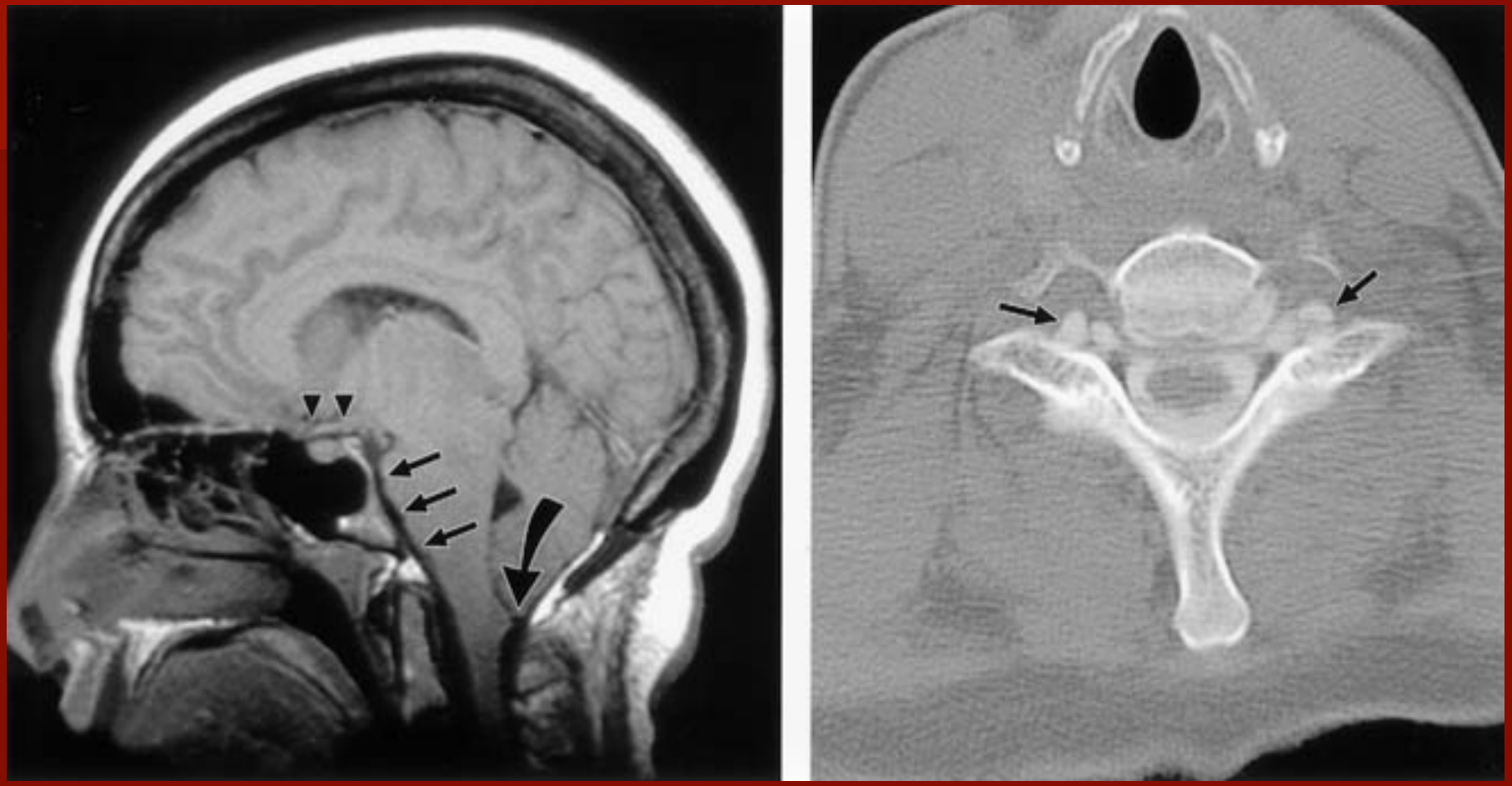
- S Subdural fluid collection
- E Enhancement of meninges
- E Engorgement of veins
- P Pituitary hyperemia
- S Sagging of brain

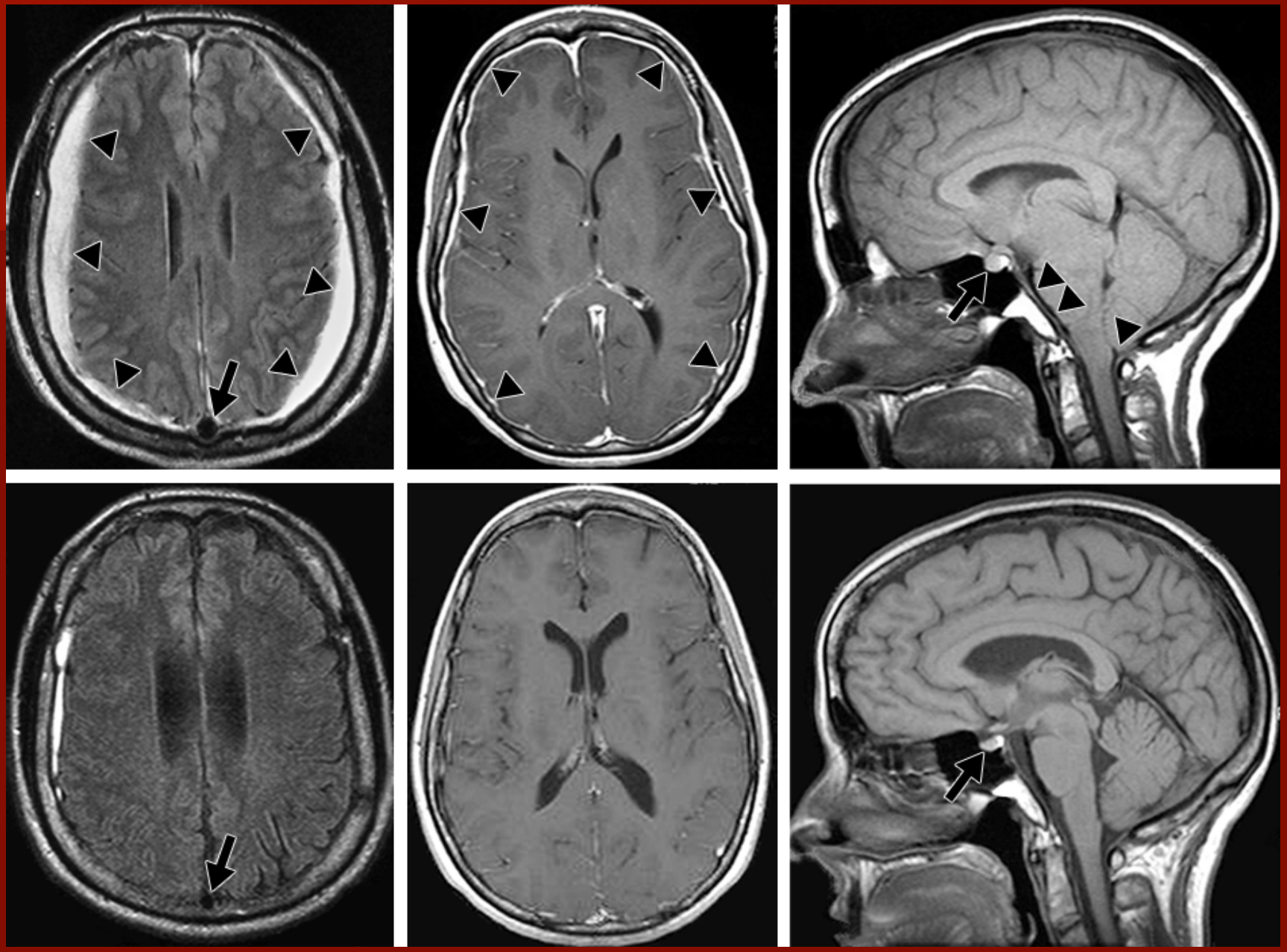






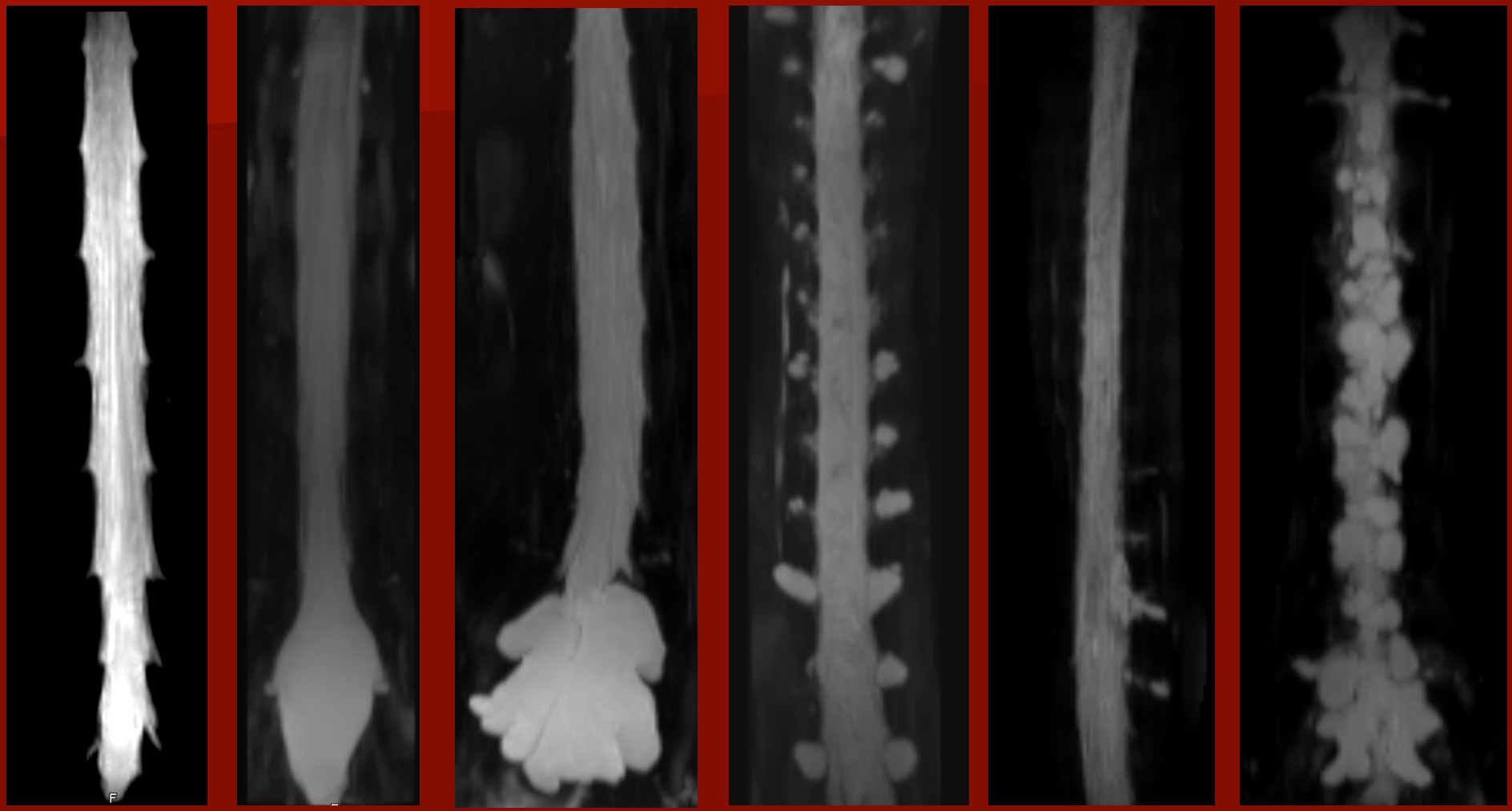




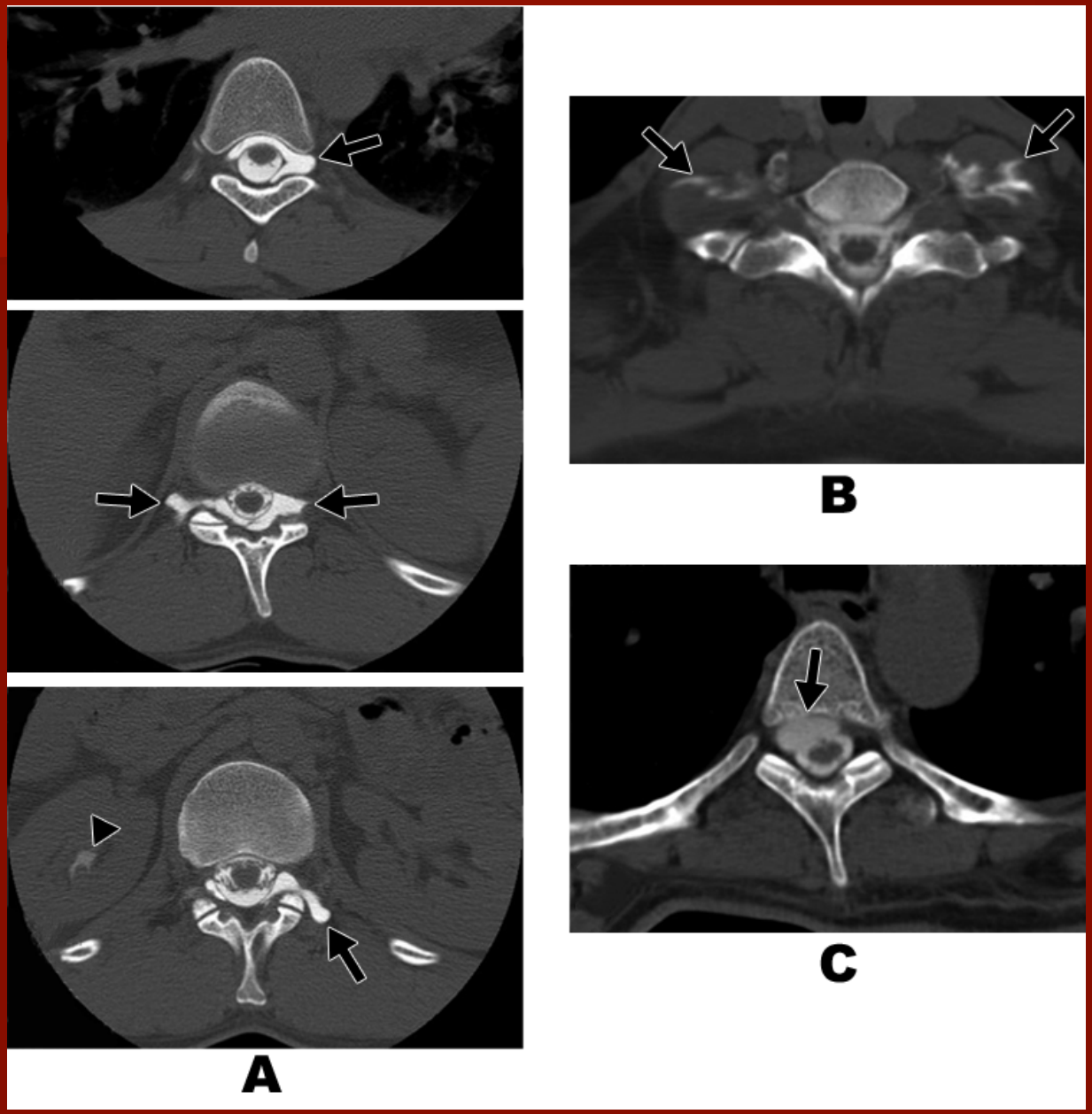


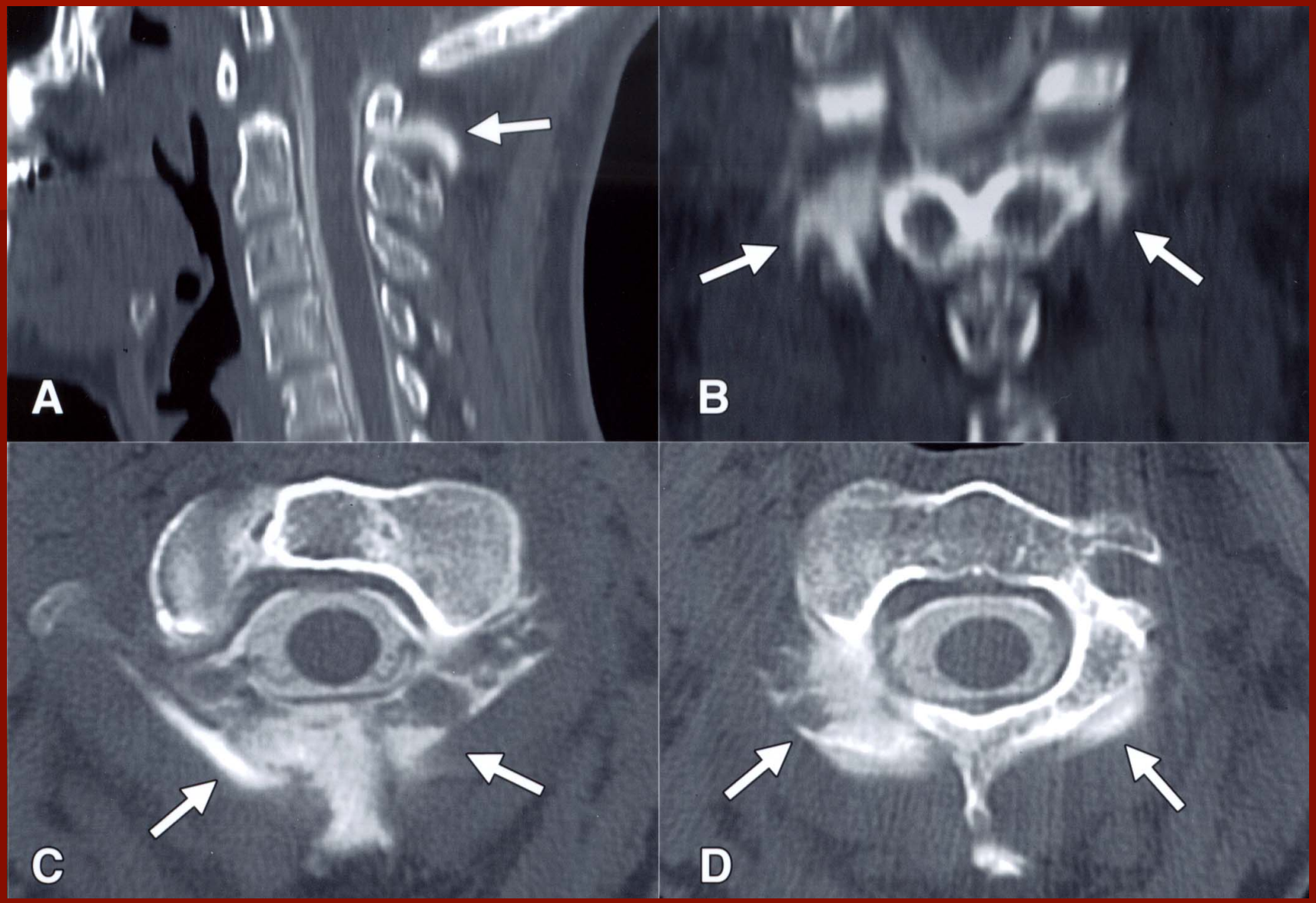
Detection of CSF Leak

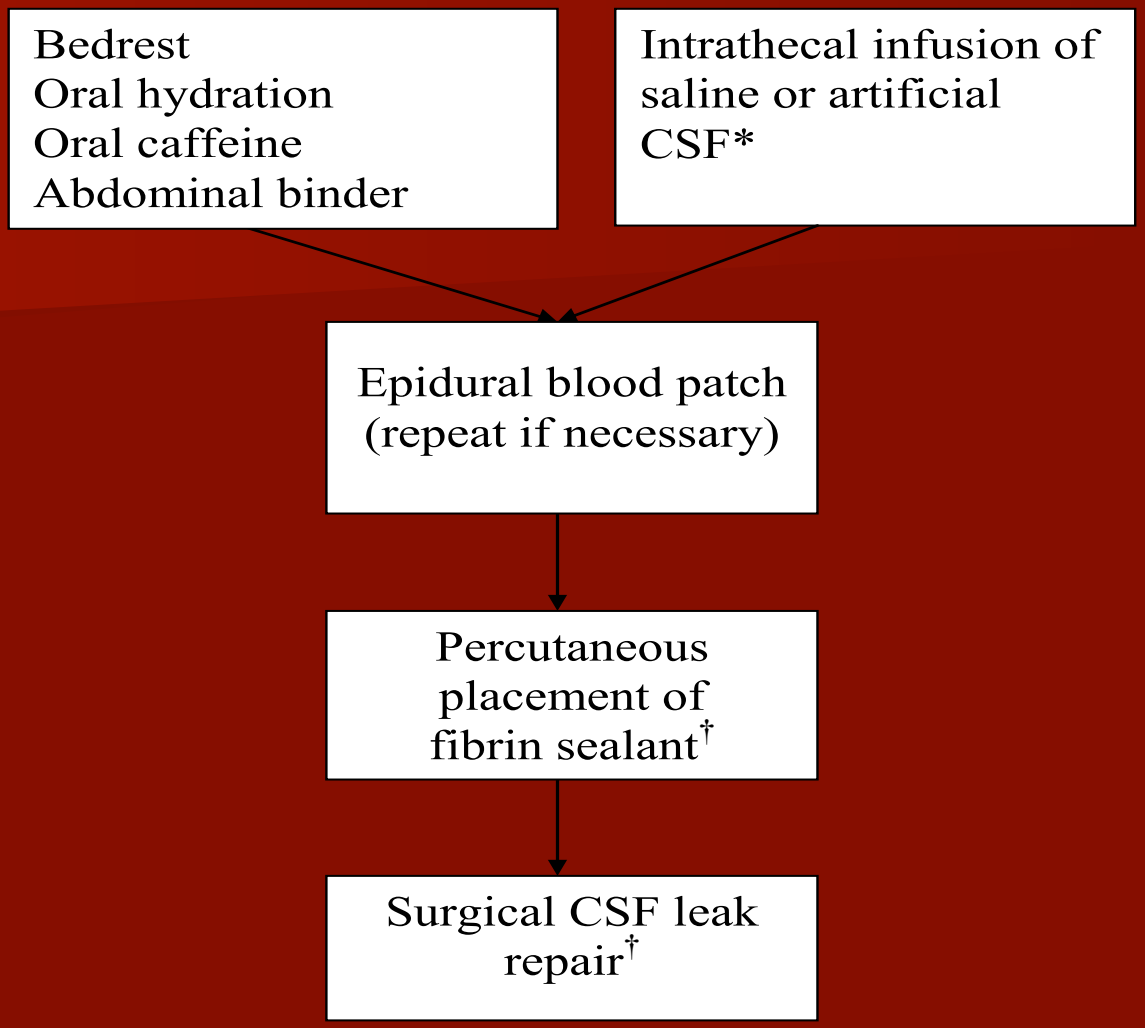
- MRI/MR-myelography
- Radionuclide Cisternography
- CT-Myelography
- Intrathecal gado-enhanced MRI



Radionuclide cisternography


A
B
C


A
B
C
D



* When urgent treatment is required such as with coma†

Knowledge of exact site of CSF leak required

Epidural blood patching

Post-dural puncture CSF leak:

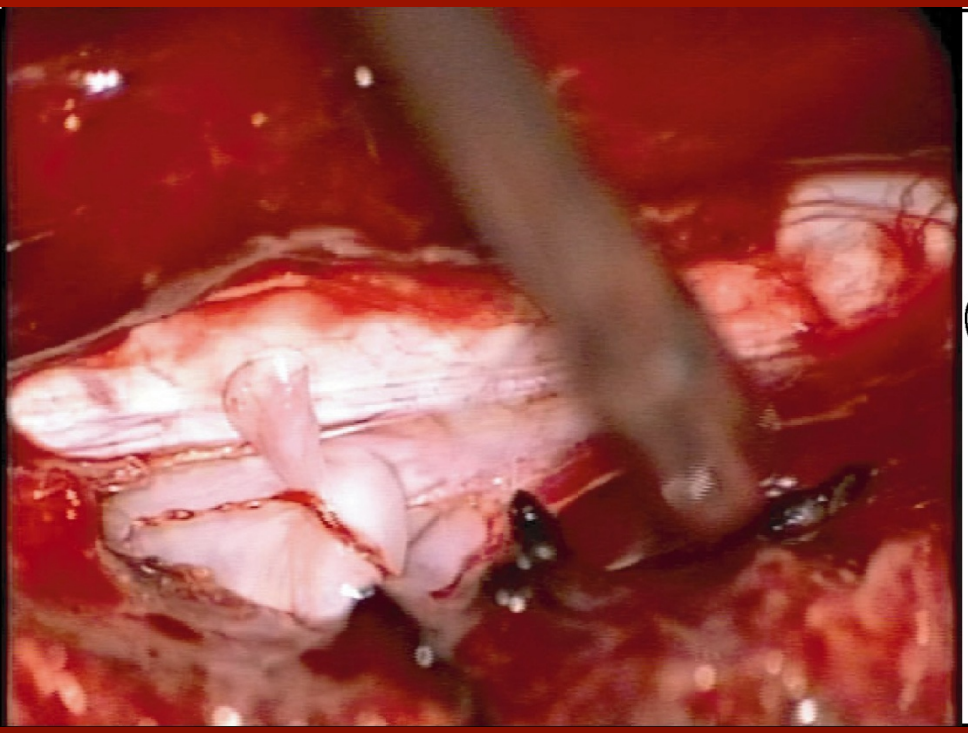
10-15 cc

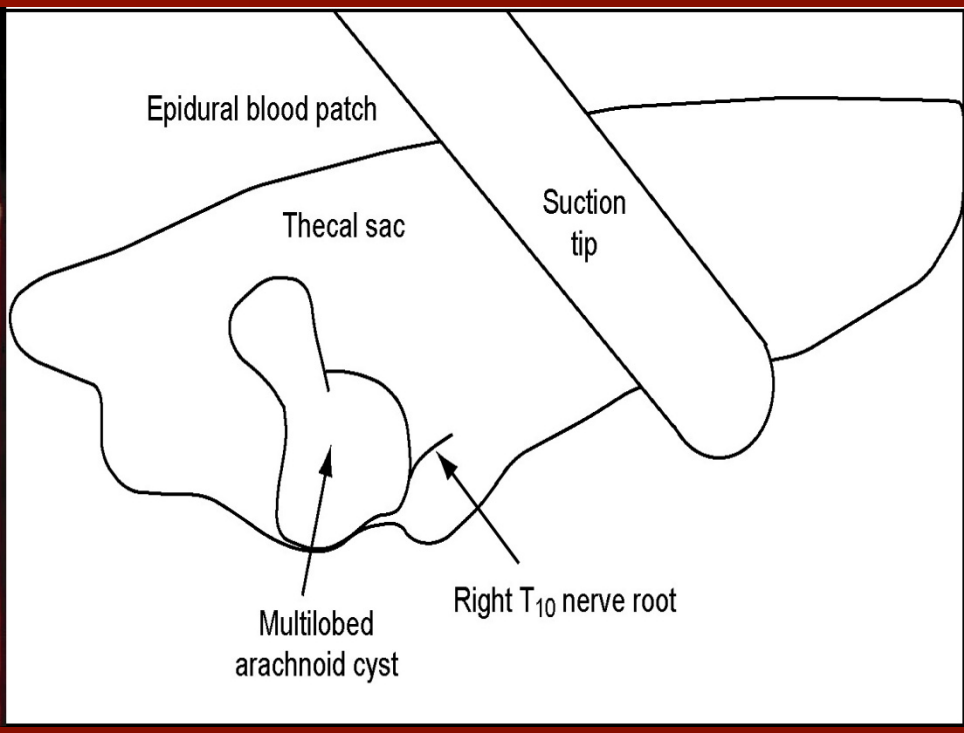
Spontaneous CSF leak:

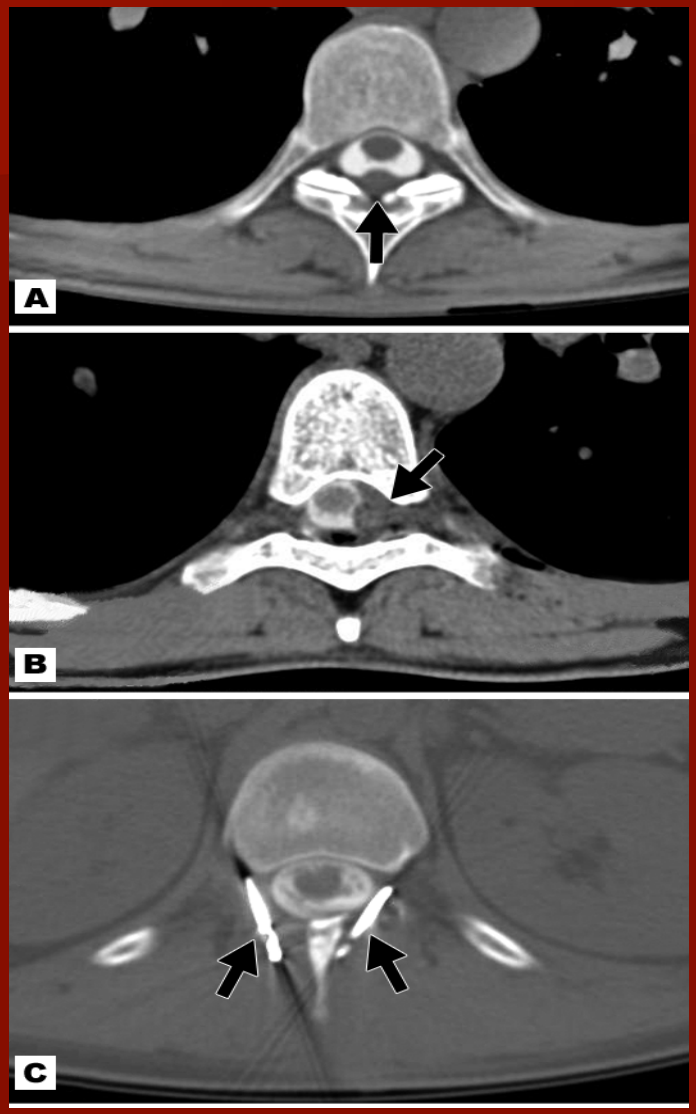
10-135 cc

Surgical Repair

- Suturing
- Ligation nerve root
- Fibrin glue
- Muscle pledgets
- Clipping of cyst




Epidural blood patch
Thecal sac
Suction tip
Multilobed arachnoid cyst
Right T_{10} nerve root

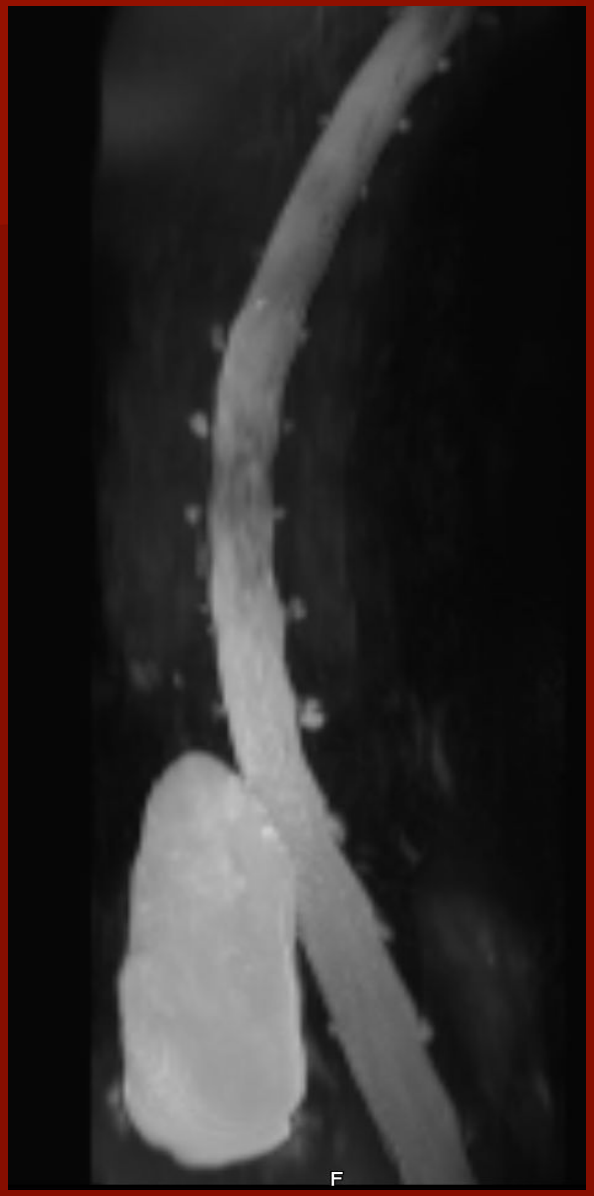

A
B
C

Treatment for spontaneous intracranial hypotension

Epidural blood patching:	95% initial response
	80% cure rate
Percutaneous glue:	40% cure rate
Surgery - cyst + leak:	90% cure rate
- cyst only:	75% cure rate
- ventral/suture:	95% cure rate
- ventral/graft:	80% cure rate
- ventral/predsm:	50% cure rate

Risks of treatment

- EBP (n=500): 0.4% paraplegia
  0.2% monoparesis
  1% persistent radiculitis
- Glue (n=100): 1% infection
  5% aseptic meningitis
- Surgery (n=200): 1% infection
  2% neurologic deficit
  4% pseudomeningocele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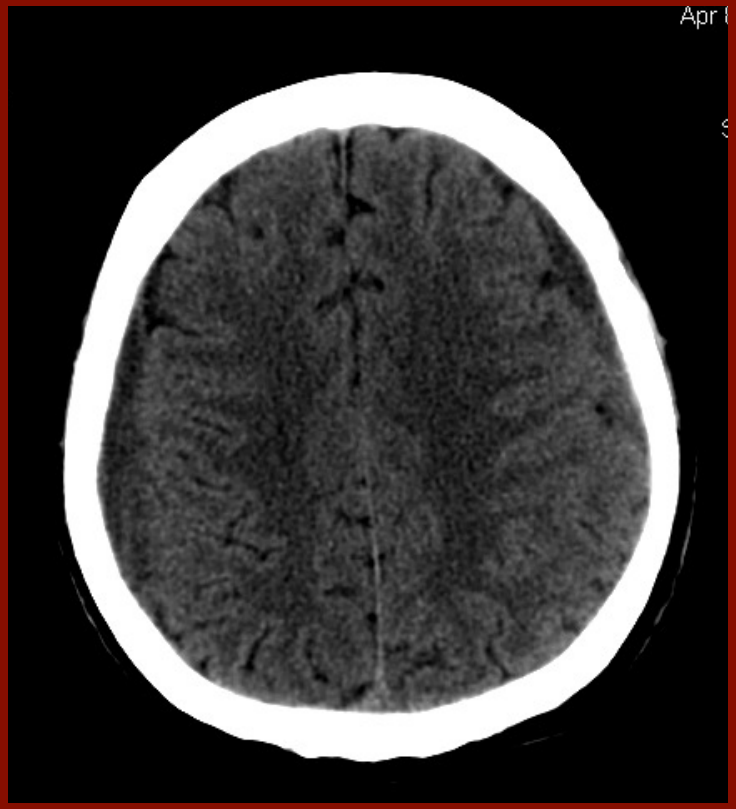
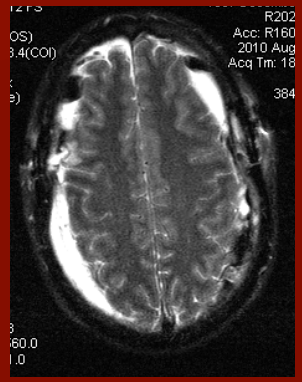
Treatment of spontaneous spinal CSF leaks

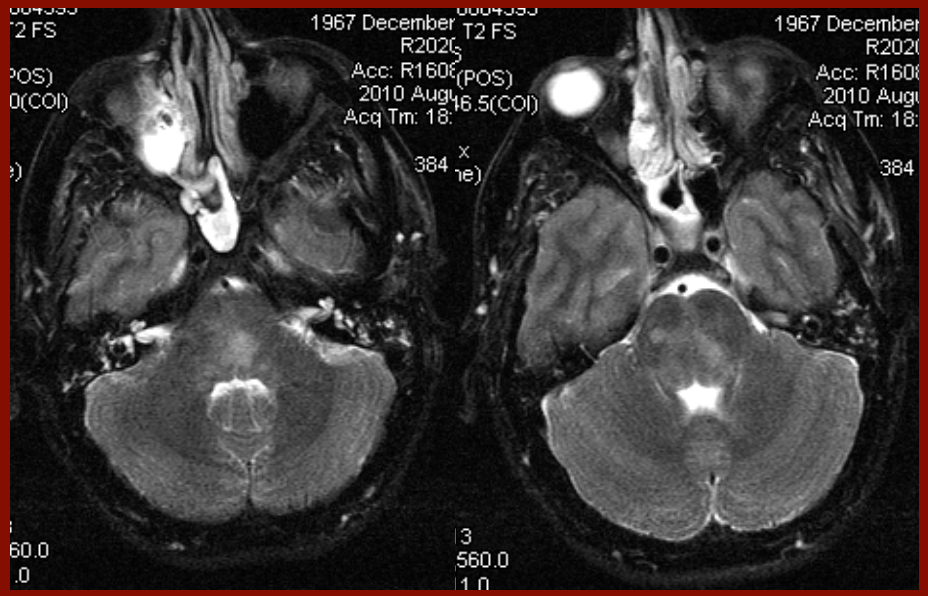
- Treatment of spontaneous spinal CSF leak has low risk and generally favorably outcome.
- Ideal patient has an abnormal brain MRI and a spinal cyst w/ active CSF leak.
- Occasionally, chronic headaches refractory to any treatment

Special considerations: Subdural hematomas



Don’t let this happen to you!





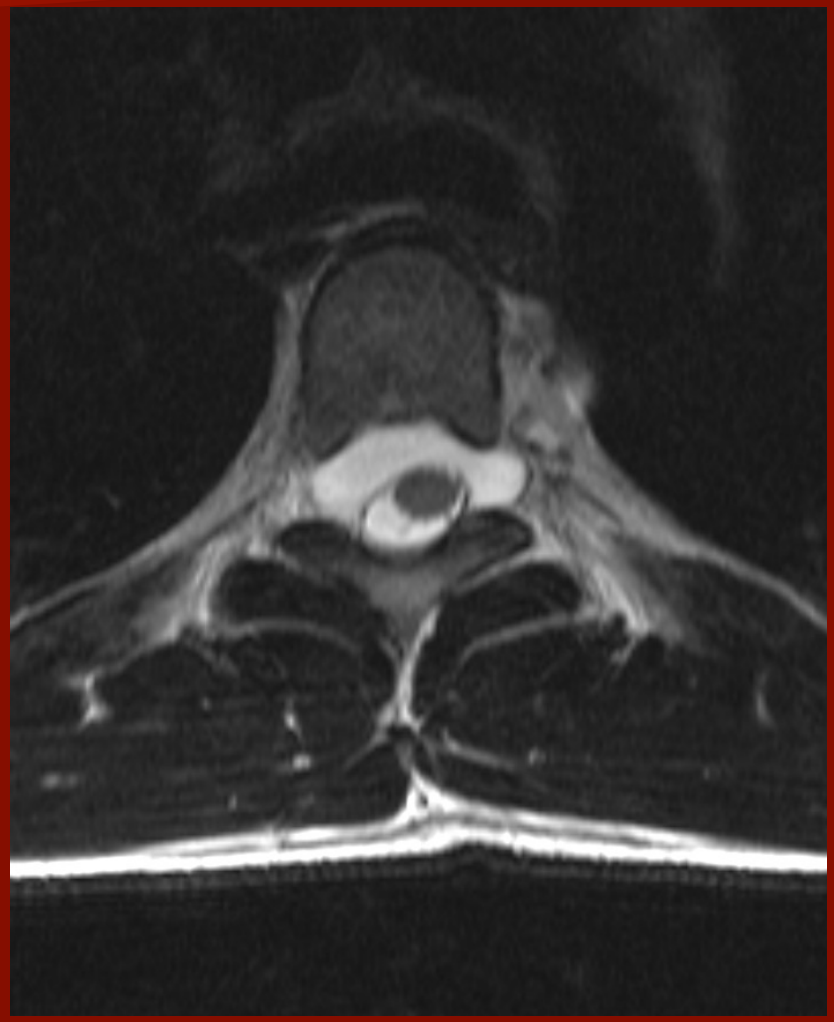
Subdural hematomas

- 128 patients 2001-2010
- Age: 14-86 years
- Sex: 63 male 65 female
- 23 craniotomy/bur hole prior to dx
- 104/105 (99%) patients successfully managed with CSF leak treatment only
- One child (1%) with craniotomy first

Special considerations: Ventral spinal CSF leaks

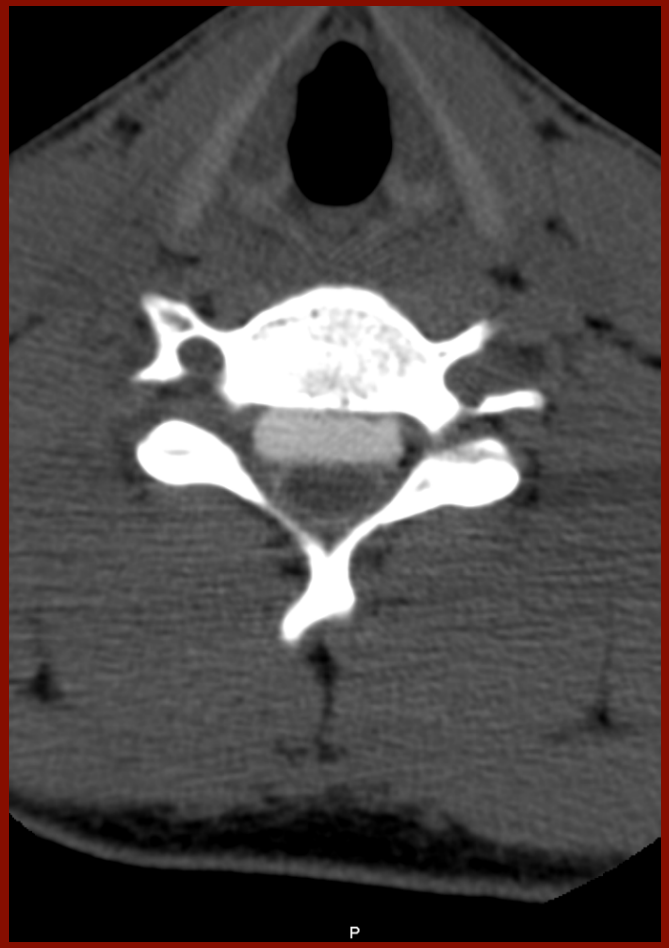
- Large volume
- Rapid
- Difficult to cure with blood patching
- Can present with delayed sequelae
- Special imaging required (DSM)
- Surgically curative

Ventral spinal CSF leaks – large volume



Ventral spinal CSF leak – delayed sequelae (ALS)

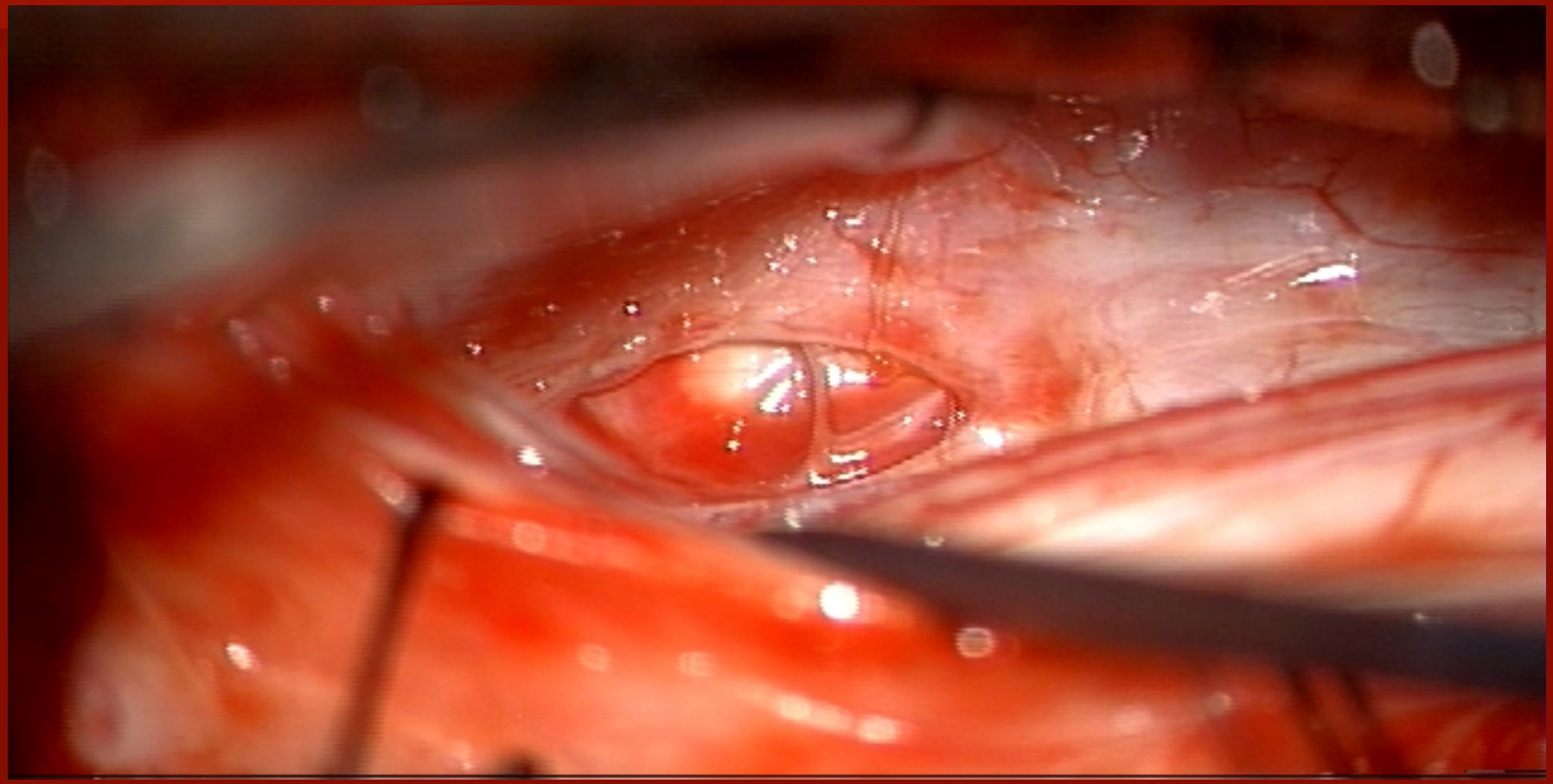
Ventral spinal CSF leaks – delayed sequelae (Myelopathy)

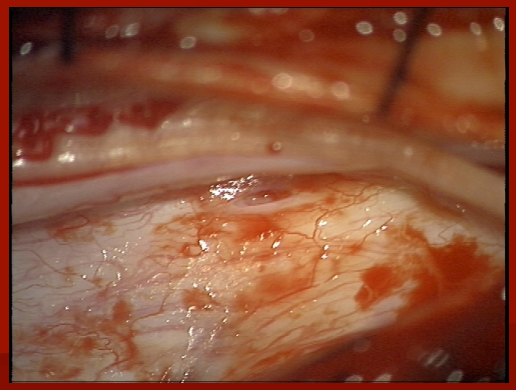
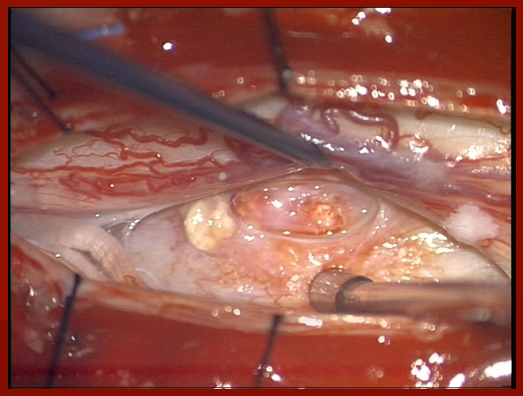
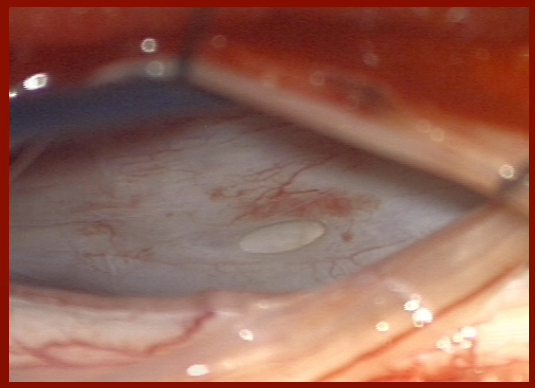
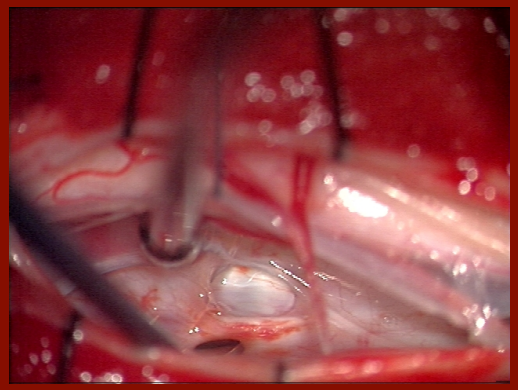
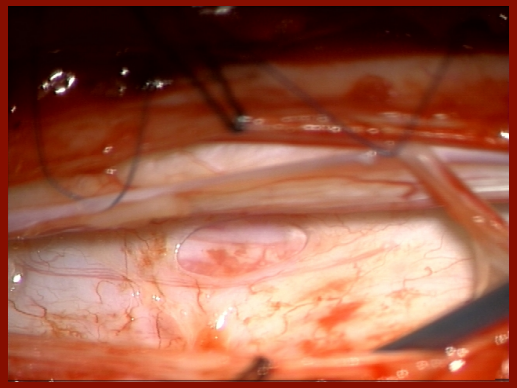
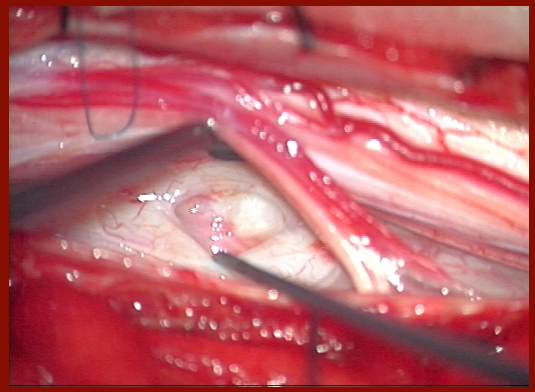
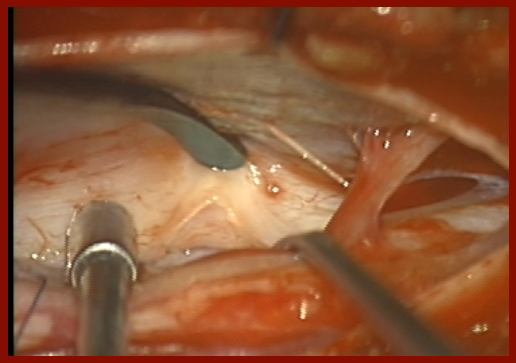
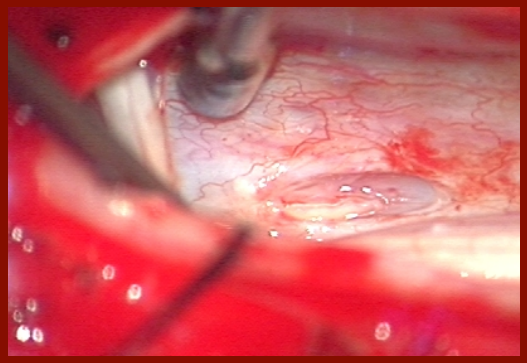
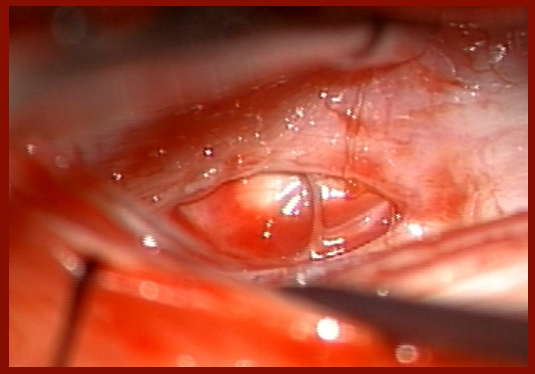


Ventral spinal CSF leaks – delayed sequelae (Superficial siderosis)

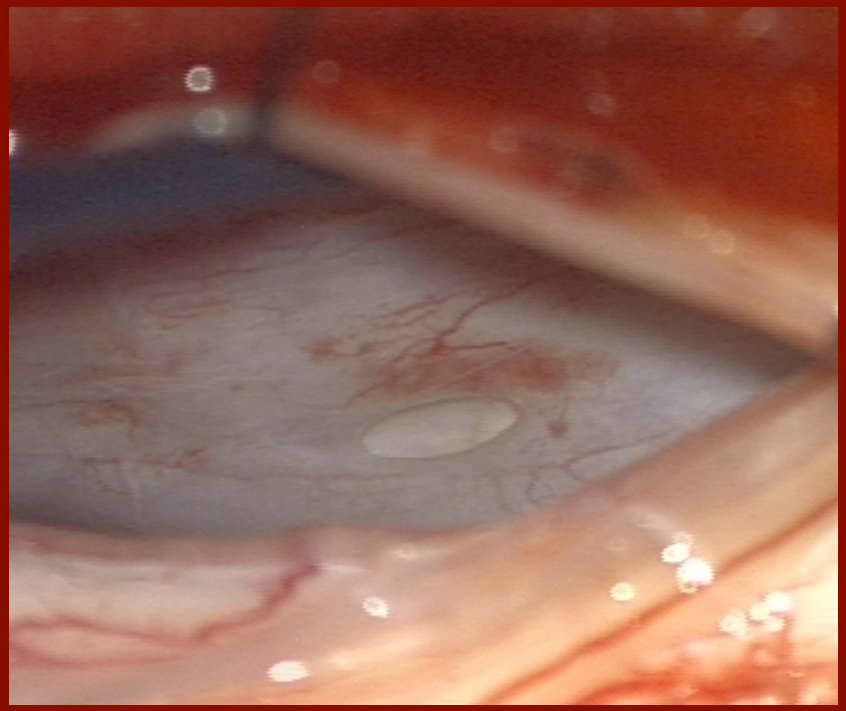
Ventral spinal CSF leaks – Digital subtraction myelography

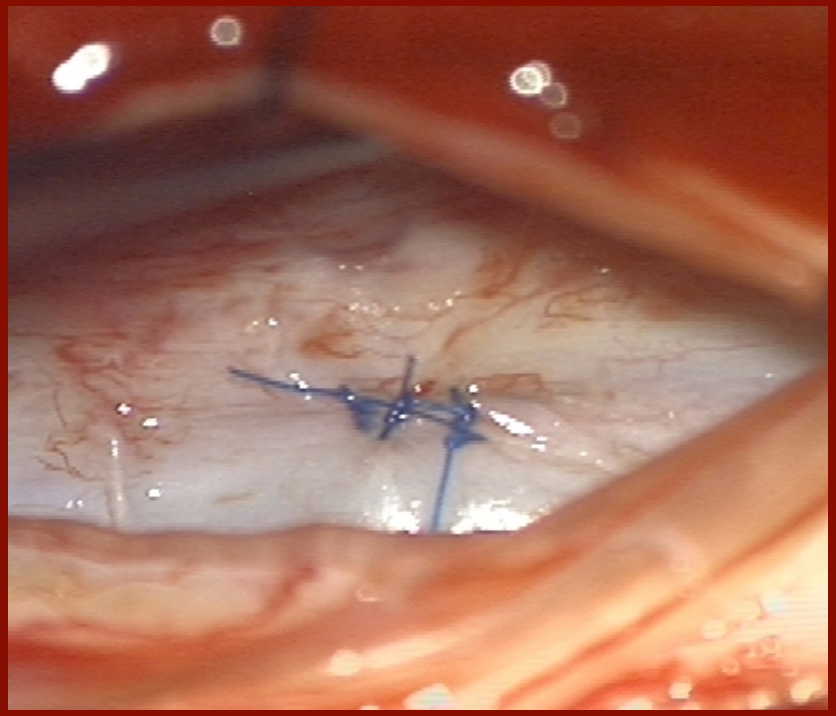
Ventral spinal CSF leak – Surgical repair



Ventral leak – surgical repair





Spontaneous intracranial hypotension - Conclusions

- Uncommon but not rare
- Headaches – Chiari – Subdural hematoma
- CSF leak in the spine – not skull base
- Epidural blood patching› glue›surgery
- Unusual manifestations: Coma – ALS – Superficial siderosis – Myelopathy – Parkinsonism – Dementia – Ataxia
- Surgery for ventral spinal CSF leaks