First Annual Cedars-Sinai Intracranial Hypotension Symposium - October 14, 2017

Update on intracranial hypotension for patients

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Nomenclature of Spontaneous Intracranial Hypotension

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

Spontaneous intracranial hypotension
November 1991
My first patient

Memre L. DOS: 11-24-1991
22 year-old woman
6 week history of orthostatic headaches
CT brain: “normal”
CT-myelogram: single thoracic nerve root cyst
Treatment: surgery
Outcome: complete recovery, no recurrence (follow-up: 23 years)

Epidemiology of spontaneous intracranial hypotension

- Prevalence: 1/50,000 (Olmsted County, MN - 1995)
- Incidence: 5/100,000/yr (“Los Angeles”, CA - 2005)
“Epidemiology” of SAH, CAD, and SIH – Cedars-Sinai Emergency Department (2005-2010)

How to recognize your fellow CSF leaker

[Graph showing trends in SAH, CAD, and SIH]

[Images of a person lying down and a person walking]
Spontaneous Intracranial Hypotension
October 2017

 Routine practice?

Spontaneous intracranial hypotension
Cedars-Sinai Medical Center (1-1-2001 to 6-30-2017)

 Approximately 1450 patients evaluated

  977 patients met ICHD-III criteria:
  CSF leak on spinal imaging
  Brain MRI with sagging/meningeal enhancement/SDH
  Opening pressure less than 6.0

  First 107 patients evaluated: 89 (83%) met ICHD-III criteria
  Last 107 patients evaluated: 26 (24%) met ICHD-III criteria
Age / Sex Distribution

- **Mean age:** 45.4 years
- **Range:** 2 – 88 years
- **Sex:** 1.9:1 F:M ratio

Patients with spontaneous intracranial hypotension: Cedars-Sinai Medical Center (1-1-2001 to 8-31-2015)
**Connective Tissue Disorders**

- Marfan syndrome: 5 / 150
- Ehlers-Danlos syndrome III: 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
- 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%)
- Seasonal variation (peak in spring)
- Bariatric surgery
- Spider bite
Clinical Manifestations

• 90 – 100% Headache
• 50 – 90% Neck Pain
• 25 – 50%
  • Nausea / Vomiting
  • Hearing abnormalities
  • Light/noise sensitivity
• 1 – 25%
  • Diplopia (CN VI or III)
  • Cognitive decline
  • 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
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 H2O
- 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
MRI Findings - Caveats

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

Misfortunes in spontaneous intracranial hypotension
Special considerations: Subdural hematomas

Don’t let this happen to you - or your loved one!
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

Detection of CSF Leak

- MRI/MR-myelography
- Radionuclide Cisternography
- CT-Myelography/Digital Subtraction Myelography (DSM)
- Intrathecal gado-enhanced MRI
Cause of spontaneous intracranial hypotension is

- Spinal CSF leak
- Pooling of CSF
- Inadequate CSF production?
- Rapid CSF absorption?
- CSF rhinorrhea-otorrhea?

NEVER

Classification of Spontaneous Spinal CSF Leaks (n=568)

- Type 1: Dural tear
- Type 2: Arachnoid cyst
- Type 3: CSF-venous fistula
- Type 4: Indeterminate
Classification of spontaneous spinal CSF leaks (n=568)

Patient population

- 138 (24%) patients from Los Angeles County (9.8 million)
- 430 (76%) not from Los Angeles County
Type 1 CSF leak
n=151 (26.6%)

- 1 a = ventral leak 145 (96%)
- 1 b = postero-lateral leak 6 (4%)
Type 1a

Type 1b
Type 1 CSF leak

- Surgery in 125/145 (86%) patients with type 1a CSF leak
- Surgery in 5/6 (83%) patients with type 1b CSF leak

Type 2 CSF leak

n = 240 (42.3%)

- 2a = simple meningeal diverticulum  n= 218 (91%)
- 2b = complex meningeal diverticulum/dural ectasia  n=22 (9%)
Type 2a

Type 2b
Type 2 CSF leak

- Surgery in 110/218 (51%) patients with type 2a CSF leak
- Surgery in 9/22 (41%) patients with type 2b CSF leak

Type 3 CSF leak

N = 14 (2.5%)
Type 3 CSF leak

Surgery in 12/14 (86%) patients with type 3 CSF leak
Type 4 CSF leak

N=163 (28.7%)

Surgery in 23/163 (14%) patients with type 4 CSF leak
Classification of spontaneous spinal CSF leaks

- Extradural CSF in 287 (50.5%) of 568 patients
  - 150 (52.3%) type 1
  - 53 (18.5%) type 2
  - 0 type 3
  - 84 (29.4%) type 4

C1-2 false localizing sign
Cervico-thoracic false localizing sign

C1-2 false localizing sign
(Oct 12, 2017)
Diagnostic evaluation

Towards a “non-invasive” diagnostic work up
i.e., no spinal tap and minimizing ionizing radiation

MRI or CT brain

MRI spine/MRMyelography
Epidural blood patching
Percutaneous fibrin glue placement
Surgery?

* When urgent treatment is required such as with coma†
Knowledge of exact site of CSF leak required

Bedrest
Oral hydration
Oral caffeine
Abdominal binder

Intrathecal infusion of saline or artificial CSF*

Epidural blood patch (repeat if necessary)

Percutaneous placement of fibrin sealant†

Surgical CSF leak repair†
Epidural blood patching

Post-dural puncture CSF leak:
10-15 cc

Spontaneous CSF leak:
7-135 cc

Percutaneous glue placement
Surgical Repair

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

Ventral leak - surgical repair
Arachnoid cysts – surgical repair

CSF–venous fistula – surgical repair
Treatment for spontaneous intracranial hypotension

Epidural blood patching: 95% initial response
80% cure rate

Percutaneous glue: 40% cure rate

Surgery - cyst + leak: 90-95% cure rate
- cyst only: 75% cure rate
- ventral/suture: 90-95% cure rate
- ventral/pre-DSM: 20% cure rate
Risks of treatment

- EBP (n=500): 0.4% paraplegia
  0.2% monoparesis
  0.8% persistent radiculitis
- Glue (n=250): 0.4% infection
  4% aseptic meningitis
- Surgery (n=500): 1% infection
  0.8% neurologic deficit
  3% pseudomeningocele

Special considerations:
Ventral (type 1a) spinal CSF leaks

- Large volume
- Rapid
- Commonly associated with calcified disc herniation
- Difficult to cure with blood patching
- Can present with delayed sequelae
- Special imaging required (DSM or dynamic CT-myelography)
- Surgically curative
Ventral spinal CSF leaks - large volume
Ventral spinal CSF leaks – Digital subtraction myelography

Delayed sequelae of ventral type 1a CSF leaks

- Superficial Siderosis
- Bibrachial Amyotrophy
- Idiopathic Ventral Spinal Cord Herniation
- ? Diffuse Intracranial Non-aneurysmal Subarachnoid Hemorrhage
Ventral spinal CSF leaks – delayed sequelae (Superficial siderosis)

Intraspinal hemorrhage – ventral spinal CSF leak
Ventral spinal CSF leak - delayed sequelae
(Bibrachial amyotrophy or ALS-like syndrome)

Bibrachial amyotrophy in SIH
Ventral spinal CSF leak –
delayed sequelae (spinal cord herniation)

Ventral spinal CSF leaks –
delayed sequelae (non-aneurysmal SAH)
Resection of transdural disc herniation and repair ventral tear
Ventral dural tear: sutures vs muscle repair
Ventral dural tear without CSF leak

Surgical solutions for the recalcitrant patient

- Lumbar dural reduction surgery
- Wearable epidural saline infusion catheter system
Brief Communication

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

Weiner I, Schievink, MD

Background and Objectives—Spontaneous intracranial hypotension has become a well-described cause of headache, particularly among young and middle-aged individuals. Treatment of the underlying spinal subarachnoid (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 laminotomy is performed, a strip of dura is excised, 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 5 years, in spite of numerous medical and surgical therapies. Significant improvement of symptoms was sustained during a 1-year period of follow-up.

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

Keywords: cerebrospinal fluid, headache, intracranial pressure, spinal cerebrospinal fluid leak, spinal dura
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

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

Wouter I. Schievink, MD,* Howard L. Rosen, MD,† and Charles Louie, 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 subdural/epidural fluid leak. However, options are limited for those patients who have exhausted these traditional treatments, especially when spinal imaging is normal. We described 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, 29 months) underwent placement of a percutaneously implanted epidural catheter attached to an external infusion pump. The Needlestick Disability Assessment Questionnaire was used to assess the severity of the symptoms.

Results: The infusion resulted in complete or near-complete symptomatic relief in 4 of 6 patients. Disability scores increased from grade 14 to grade 3. However, the clinical success was not sustained in 1 patient because of infection, and in 1 patient because of laboratory failure to provide adequate symptom control, and in 1 patient because of minimal symptom relief. Two patients required modification of medication to control pain. Neurologic examination and brain magnetic resonance imaging (MRI) findings were normal in all patients. Spinal imaging performed at the time of lumbar puncture was abnormal in all patients and showed large intradural CSF leaks. Two patients had new noncortical infarcts on follow-up MRI. CSF leak was identified in 5 of 6 patients. The presence of a spinal CSF leak could not be established in 1 of the 6 patients. All patients underwent 2 or more epidural blood patches. Two patients had undergone 1 or more surgical treatments directed at the CSF leak or lumbar spinal arterial dissection.

Patients
Six patients with spontaneous intracranial hypotension were enrolled in this trial (3 females and 3 males). The mean age of the group was 53 years (range, 49–65 years). Duration of symptoms varied from 34 to 120 months (mean, 36 months). Orthostatic symptoms occurred in all patients. Neurologic examination and brain magnetic resonance imaging (MRI) findings were normal in all patients. Spinal imaging performed at the time of lumbar puncture was abnormal in all patients and showed large intradural CSF leaks. Two patients had new noncortical infarcts on follow-up MRI. CSF leak was identified in 5 of 6 patients. The presence of a spinal CSF leak could not be established in 1 of the 6 patients. All patients underwent 2 or more epidural blood patches. Two patients had undergone 1 or more surgical treatments directed at the CSF leak or lumbar spinal arterial dissection.

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Spinal epidural infusion system

- N = 12
- 7 women and 5 men (most “without” SIH)
- Age: 40 – 79 years
- Good outcome: 10 (83%)
- Complications: Infection: 2 (17%)
  Hardware failure: 5 (42%)

Behavioral variant frontotemporal dementia-hypersomnolence syndrome due to spontaneous intracranial hypotension

- N = 31
- Insidious onset
- Sixth decade of life
- Male:Female – 4:1
- Brain sagging: 100%
- CSF leak: 3%
- Treatment: refractory; Surgery in 94%
Frontotemporal dementia in spontaneous intracranial hypotension

- Good outcome after treatment

  1 / 7 patients who had undergone Chiari decompression (14%)

  vs

  20 / 22 patients who had not undergone Chiari decompression (91%)

  $P=0.003$
bvFTD in SIH

Iatrogenic spinal CSF leak

- Lumbar puncture
- Epidural steroid injection
- Epidural analgesia
- Spine surgery
Post-dural puncture headache

Kaylee T-C

- 23 year old pregnant (31 weeks) woman

- S/P V-P and L-P shunts for idiopathic intracranial hypertension since age 11

- 2 years of orthostatic headaches and 4 shunt revisions and 11 valve adjustments.

- Diagnosed with POTS
Nick P

☎ 27 year old man from mid-west with orthostatic headache and paraplegia

☎ Visiting grandparents in Las Vegas at age 10

☎ Underwent LP to rule out meningitis
Carmen V

- 32 year old woman from Orange County with orthostatic headaches following epidural for child birth 3 years prior

- Delayed onset 1 month post-partum
Maria V

39 year old woman from Burbank with orthostatic headaches since lumbar puncture to rule out multiple sclerosis

S/P 13 epidural blood patches
Thank you

Save the Date!
Saturday October 14, 2017
Intracranial Hypotension Symposium