Idiopathic intracranial hypotension: from diagnosis to treatment

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Idiopathic intracranial hypotension outline

Definition
Clinical presentation and incidence
Pathogenesis
Intracranial MR findings
Spinal MR findings
Spinal imaging issues
Treatment
Idiopathic intracranial hypotension
= spontaneous intracranial hypotension

underrecognized though curable condition
caused by low CSF pressure due to a spinal CSF leak
patients present with a wide variety of symptoms and signs:

• characteristic
  postural headache relieved by recumbancy
dizziness, diplopia, CN deficits (- coma, in 7 pats reported)

• characteristic imaging findings
  intracranial and spinal
### TABLE. International Classification of Headache Disorders SIH Criteria

<table>
<thead>
<tr>
<th>A. At Least One of the Following</th>
<th>B. At Least One of the Following</th>
<th>C. Must Be Fulfilled</th>
<th>D. Must Be Fulfilled</th>
</tr>
</thead>
<tbody>
<tr>
<td>Neck stiffness</td>
<td>Evidence of low CSF pressure on MRI (eg, pachymeningeal enhancement)</td>
<td>No history of dural puncture or other cause of CSF fistula</td>
<td>Headache resolves within 72 h after epidural blood patch</td>
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<tr>
<td>Tinnitus</td>
<td>Evidence of CSF leakage on conventional myelography, CT myelography, or cisternography</td>
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<tr>
<td>Hypacusia</td>
<td>CSF opening pressure &lt;60 mm H₂O in sitting position</td>
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<td>Photophobia</td>
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<tr>
<td>Nausea</td>
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</tbody>
</table>

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*Rahman, M et al. Spontaneous intracranial hypotension: dilemmas in diagnosis: Neurosurgery 2011;69:4-14*
Idiopathic intracranial hypotension incidence

- prevalence of SIH syndrome ~ 1 in 50 000
- women >> men, peak incidence ~ 40 years
- rarely associated with connective tissue disorder (Marfan, Ehlers-Danlos, polycystic kidney syndrome)
- symptoms → indistinguishable from post LP headache
- posture-related component may reduce or disappear over time when IIH is not effectively treated and becomes chronic!
spontaneous dural tear
at sites of focal structural vulnerability of spinal meninges
(often trivial trauma)
→ leads to occult cerebro-spinal fluid leak
→ decrease in CSF volume and pressure! (< 60mm)

Modified from: Cohen G. Neurosurgery 2006; 58, Suppl. ONS-238-245
Idiopathic intracranial hypotension

pathogenesis

volume of CSF & of intracranial blood vary inversely (Monroe-Kelly law)

→ descent of the brain and traction on supporting pain sensitive meningeal structures

→ compensated by hypervolemia of intradural & epidural venous plexus → venous engorgement

→ subdural fluid collections (hygromas/ hematomas) as compensatory phenomena to maintain constant intracranial fluid volume

⇒ characteristic MRI findings
Idiopathic intracranial hypotension

**cranial MR examination**

⇒ *establish diagnosis*

- descent of brain
- reduction of CSF spaces
- venous distension-dilatation
- pachymeningeal enhancement
- subdural effusion
- pituitary bulging-hyperemia
descent of the brain

tonsillar herniation, tentorial bulging, corpus callosum ↓
shallow basal cisterns, small ventricles
reduction of perioptic CSF space

Sensitivity and specificity 100% each on gross visual inspection

Rohr A et al. MR imaging of the optic nerve sheath in patients with craniospinal hypotension. AJNR 2010;31:1752-1757
Given greater capacity and compliance most of the compensation \( \Rightarrow \) through dilatation of the venous side.
venous engorgement

basilar plexus

thick linear pachymeningeal, no nodularity, infra-supratentorial
venous distension sign

Venous distension sign: sensitivity & specificity of 94% each

subdural effusions

pressure driven fluid extravasation from engorged dural veins
→ transsudation rather than leakage
→ compensate for more severe volume loss
subdural effusion

- effusions are always associated with meningeal enhancement
- abnormal meningeal enhancement in patients with SIH is not the absolute rule.

Size of pituitary appears larger during symptomatic phase - due to venous distension of adjacent intercavernous sinus?

Idiopathic intracranial hypotension

spinal MR examination

⇒ confirm cranial diagnosis

• dilatation of the anterior internal epidural venous plexus
• (meningeal enhancement ?)
• extradural fluid collections !

⇒ reveal location of CSF leak
dilatation of epidural venous plexus

- No meningeal enhancement, but epidural venous enhancement
- Width of epidural plexus ~ with size of subdural fluid collection*

extradural spinal fluid collections

T2 w TSE sequence sag/ax with fat signal suppression
Localization of CSF leak

majority of CSF leaks occur at the thoracic level rarely lower cervical or lumbar spine and are due to dural defects at arachnoid diverticula (bony spurs, disk herniation ?)

⇒ root sleeve (Tarlov) cysts
3D MR myelography

Location of CSF leakage

Myelography and CT myelography
Radionuclide cisternography
MR- Haste myelography
MR- Gd myelography
MR-Haste “myelography”

Thin MIP sagittal / VR coronal
MR-Myelo Haste (3DT2)  

MR-Myelo with ith Gd (0.5ml/4.5ml NaCl) Dixon
Intrathecal Gd injection- of label use

MR Gd myelography 0.5ml Magnevist + 4.5ml NaCl
(gadopentetate dimeglumine)

**Jinkins JR et al.** Intrathecal Gadolinium enhanced MR myelography in the evaluation of clinically suspected CSF Rhinorrhea in Humans. Early experience. Radiology 2002; 222 : 555-559

15 pats with CSF rhinorrhea; leakage confirmed in 13/15
0.5ml via LP; feasibility and safety demonstrated

**Albayram S et al.** Gadolinium enhanced MR-cisternography to evaluate dural leaks in intracranial hypotension syndrome. AJNR 2008;29:116-121

19 pats with SIH; leakage in 17 pats, leakage site in 14,
0.5ml via LP; safety, accuracy and feasibility confirmed
False localizing sign C1-2

Visualization of leakage site

8 patients investigated by CT myelography:

“CSF leak sites were directly visualized in 37% (3/8) of patients”

Kranz PG et al. CT-Guided Epidural Blood Patching of Directly Observed or Potential Leak Sites for the Targeted Treatment of Spontaneous Intracranial Hypotension
Indications for intrathecal Gd

negative MR-Haste, false localizing sign, multiplicity of sites

Multiplicity 28.6-38.5%
Brain MR to confirm the diagnosis, → conservative measures for up to two weeks.

if patient remains symptomatic,  
→≤ 3 non-directed lumbar epidural blood patches

If unsuccessful, localize the leak with  
MR myelo, radionuclide cisternography, MR Gd or CT-myelo

If leak is localized,  
directed EPBs should be considered,  
followed by fibrin sealant or neurosurgery if necessary
Idiopathic intracranial hypotension therapy options medical/surgical

- Caffeine
- Glucocorticoids / mineralocorticoids
- Theophylline
- Desmopressin acetate

Resection of diverticulum, closure of dural defect with piece of muscle or Gelfoam wrapped around root sleeve and sutures
Therapy: epidural blood patch

forms a dural tamponade and triggers an aseptic inflammation to permanently seal CSF leak.

⇒ nonselective initial epidural blood patch at the lumbar level regardless of the level of the CSF leak

• optimal volume of autologous blood is controversial, recommended: distant 15–20 ml

• cervical / upper thoracic spine 5-10ml epidural blood

• if lumbar procedure fails,

⇒ targeted selective epidural blood patch at the exact site of the CSF leak!
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Results of targeted epidural blood patch

- 12 pats with clinically and MR confirmed IIH
- CSF leak visualized in 10 pats
- single leak in 8, two sites in 2 pats
- 2 patients with suspected sites 2 each

Targeted blood patch at 16 sites (12 pats)
  retreatment in 1 patient within 4 months

⇒ clinical and imaging resolution all 12 pats
Treatment results

26 patients: multilevel leaks in 23/26 pats
cervical 12, thoracic 25, lumbar 21

Treatment in 25 pats w 20cc of blood
single level 9, multilevel 16

Clinical and/or radiological improvement
following 1 treatment (16), 2 (5), 3 (3) and 5 (1)

Treatment control: spine

F-up after 2 months
Treatment control
Idiopathic intracranial hypotension

**conclusion**

- treatable cause of headache and variable nonspecific symptoms (dizziness, cranial nerve deficits - coma !)
- MR imaging is crucial for diagnosis and localisation of CSF leak (s) (multiplicity not uncommon)
- Imaging findings reflect pathophysiology
- Treatment may be performed non-invasively under imaging guidance
- MR documentation of treatment success, failure or recurrence
Shokran - thank you

Zürich

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