Spontaneous Intracranial Hypotension: A Focus on Clinical Characteristics

ILC/SIH SYMPOSIUM
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FARNAZ AMOOZEGAR, MD, MSC, FRCPC
CLINICAL ASSOCIATE PROFESSOR
DEPT CLINICAL NEUROSCIENCES
UNIVERSITY OF CALGARY & HOTCHKISS BRAIN INSTITUTE
What is SIH

- SIH stands for spontaneous intracranial hypotension
- A condition where a leak occurs within the dura (the thick covering layers) at the level of the spine
- Synonomous with Spinal Cerebrospinal fluid (CSF) leak
- Different than cranial or sinonasal CSF leaks
- Although better recognized now than in the past, many health-care practitioners are still not aware of this condition
- For a number of reasons, SIH also remains difficult to diagnose and treat in many patients
Epidemiology

- No major prospective/RCT studies
- Estimated annual incidence: 5 per 100,000
- The onset of symptoms of SIH is usually in the fourth to fifth decades of life
- Peak incidence around age 40 years
- SIH affects women more than men, ratio 1.5:1
Etiology & Pathogenesis

- SIH is caused by spontaneous spinal CSF leaks
- The exact cause of the leak remains unknown in the vast majority of patients; likely multifactorial
- Underlying fragility of the spinal meninges is suspected
- A history of a traumatic event can be elicited in about one third of patients, suggesting mechanical factors as well
Etiology & Pathogenesis

- An inherited connective tissue disorder can play a role in the development of CSF leaks
- Marfan syndrome, Ehlers-Danlos syndrome (type II), and autosomal dominant polycystic kidney disease are the most common
- Familial SIH has been described
Etiology & Pathogenesis

- Some patients are found to have osseous spinal pathology
- Most commonly, degenerative disk disease with osteophytes found piercing the dura
- Occasionally, congenital bony spurs
- Dural abnormalities: dural holes or rents, meningeal diverticula, or even absence of dura
Current Understanding of SIH pathogenesis

Believed that the symptoms are related to loss of intracranial CSF volume, rather than strictly a reduction in pressure.

Many patients with SIH exhibit normal opening pressures.

The CSF volume loss is felt to occur by three major etiologies in the spine:

- Leaks secondary to dural weakness in the area of the nerve root sleeves/meningeal diverticula
- Ventral dural tears associated with degenerative disc disease/osteophytes/disc herniations
- CSF-venous fistulas

Kranz, Curr Pain Headache Reports 2017
ICHD-III Diagnostic Criteria for SIH

A. Any headache fulfilling criterion B to D

B. Low CSF pressure (<60 mm CSF) and/or evidence of CSF leakage on imaging

C. Headache has developed in temporal relation to the low CSF pressure or CSF leakage, or has led to its discovery

D. Not better accounted for by another ICHD-3 diagnosis

Cephalalgia. 33(9)629-808.
Clinical Presentation

- Prototypical & most common manifestation of SIH is an orthostatic headache
- Can occur within seconds to minutes of taking an upright position (but can be hours)
- Headache improves or resolves after lying down, usually within 30 minutes
- Headache is usually holocephalic & diffuse, but may be localized to one region of the head or asymmetric
- Headache quality can vary
Clinical Presentation

- Initial onset of headache in majority of patients is gradual or subacute
- Headache severity varies from mild to severe
- Exact cause of headache is unknown
- May be related to downward displacement of the brain due to loss of CSF buoyancy, causing traction of pain-sensitive structures
- Compensatory dilation of pain-sensitive intracranial venous structures may also play a role
Clinical Presentation

- An orthostatic headache can become less prominent or even disappear over time
- Rarely, the reverse pattern may occur
- There are also patients with no postural component to their headache
- Other headache patterns: exertional headaches, headaches at the end of the day, or even paradoxical headaches
- Think of SIH in patients with a New Daily persistent headache
Clinical Presentation

- Other relatively common symptoms of SIH include:
  - Posterior neck pain or stiffness
  - Nausea and vomiting

- Up to 25% of patients may have:
  - Change of hearing (echoing, under water)
  - Tinnitus
  - Disturbed sense of balance

- Rarely, patients can also have:
  - Visual blurring, VF defects, diplopia
  - Facial numbness or facial pain, facial weakness or spasm
  - Parkinsonism, ataxia, dementia
Evaluations

- Imaging
  - CT Brain
  - MRI
    - Brain
    - Spine
  - Radionuclide Cisternogram
  - Myelogram
    - CT
    - MR
  - Digital subtraction imaging

- Other
  - CSF
  - Profile
  - OP
CSF

- OP < 60mm H₂O
- Lymphocytic pleocytosis
  - < 50 cells
- Normal or Increased protein
  - Up to 100mg/dl
  - Rare up to 1g/dl
- Xanthochromia
Treatment

- Conservative:
  - Bed rest
  - Oral or IV hydration
  - Caffeine
- Steroids
- Epidural blood patch
- Fibrin glue injection
- Surgical repair
Epidural Blood Patch (EBP)

- Instant relief in some

**Mechanism:**
1. Volume replacement
2. Dural tamponade
3. Restriction of CSF flow within epidural space → decrease CSF absorption
4. Change in dural stiffness/resistance (long term)

- If poor response
  1. Repeat
  2. Higher volume patch (> 20 mL)
     - Need to wait 5 days in between
Recommendations for CTD

- 2013 study at Cedars-Sinai: in 50 patients diagnosed with SIH, 9 had heritable CTDs, including Marfan syndrome, Ehlers-Danlos syndrome and others.

- In 7 of these patients, SIH was the first manifestation of their illness.

- The authors suggest that patients with SIH should be screened for connective tissue disorders and vascular abnormalities.

- If there are concerns based on this, the patient can then be referred to a medical geneticist.
Recommendations for Vascular Disorders

- Vascular abnormalities occur with higher frequency in patients with SIH as compared to controls.
- One study showed intracranial aneurysms in 9% of patients, as compared to 1% of controls (J Neurosurg. 2011; 115: 113-5).
- Patients with SIH should have intracranial vascular imaging at one point during their assessment, either with CTA or MRA.

- Those with an established connective tissue disorder, such as Marfan syndrome, should be screened further with MRA of the neck, chest, abdomen and pelvis, because they are at risk of large arterial aneurysms (Schievink & Deline, Curr Pain Headache Rep (2014) 18: 457)
Clinical evaluation of CTDs in patients with SIH

Adapted with permission from Reinstein et al, EJHG (2013) 21, 386-390

General:

Height: _____________________  Weight: _____________________
Body Habitus (Marfanoid, slender, etc): _____________________
Ethnic Background: _____________________
Family History (Focused on joint, skin, eye, vascular abnormalities, sudden death): _____________________

Pregnancy, development, and neonatal history:

Delivery type (e.g. NV, CS, aids): _____________________
Presentation (cephalad): _____________________
Gestational age (term, pre-term, post-term): _____________________
☐ Pregnancy-associated maternal complications
☐ Uterine rupture/cervical insufficiency during pregnancy/delivery
☐ Delayed motor development/hypotonia

Adapted with permission from Reinstein et al, EJHG (2013) 21, 386-390
Clinical evaluation of CTDs in patients with SIH

**Signs for skin hyperextensibility and fragility:**

- Skin hyperextensibility/elasticity
- Easy bruising, ecchymoses
- Soft/thin/transparent skin
- Anal or uterine prolapse
- Surgical complications (postoperative hernias, wound dehiscence)
- Spontaneous pneumothorax
- Slow wound healing
- Widened, thin scars
- Stretch marks (unrelated to pregnancy)
Clinical evaluation of CTDs in patients with SIH

Joint hypermobility and complications:

☐ Joint dislocations
☐ Joint hypermobility Beighton score: __________________
☐ Degenerative joint disease
☐ Chronic joint and limb pain
☐ Scoliosis
☐ Flat Feet
☐ Spontaneous fractures

Cardiovascular (history and echocardiography):

☐ Aortic or other arterial aneurysm, dissection or rupture (personal or family history of): _________________________
☐ Valvular disease, including mitral valve prolapse (on Echo)
Clinical evaluation of CTDs in patients with SIH

Craniofacial:

☐ Dental history of crowding/braces
☐ High-arched palate
☐ Single/bifid uvula
☐ Dysmorphic facial features
☐ Periodontal disease and age of onset _______________________

Ophthalmologic:

☐ Personal or family history of lens dislocation
☐ Detachment of retina
☐ Other vision abnormalities: ________________________________
☐ Blue sclera
Beighton score

- A simple system to quantify joint laxity & hypermobility
- Uses a 9 point system
- A higher score indicates more laxity
- Score above 6 indicates hypermobility

Beighton's modification of the Carter and Wilkinson scoring system. Give yourself 1 point for each of the manoeuvres you can do, up to a maximum of 9 points.

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Recommendations for Cardiac Disorders

- An echocardiogram is also suggested to assess for valvular disease (such as mitral valve prolapse), and dilatation of the aortic root.

- In a 2014 study, the rate of cardiovascular abnormalities detected for the same cohort of 50 SIH patients seen at Cedars-Sinai was 9/50 or 18%.
  - 6 patients showed aortic root dilatation
  - 3 showed valvular heart disease
  - Only 2/9 patients had an underlying connective tissue disorder, indicating that cardiovascular pathology can occur even in patients with no known connective tissue disorders.

Typical SIH

If not improved after 1-2 weeks

Non-directed Lumbar EBPs, as a high a volume as patient can tolerate.

Successful

Unsuccessful

2nd non-directed lumbar EPB, as high a volume as patient can tolerate

Unsuccessful

If partial success, 3rd non-directed EPB (consider lumbar &/or lower thoracic).

1) More investigations
2) Refer to Neurology/Headache Center (if appropriate)

1) Non-invasive MRM
2) CTM (if extradural fluid on MRI, do dynamic CTM, if no extradural fluid, do conventional CTM)
3) DSM (for fast/high flow leaks, ventral leaks & CSF venous fistula)

Localized Leak

Leak not localized

Directed EPB

Unsuccessful

1) Reconsider Diagnosis
2) Conservative Measures

1st Directed EPB

Partial Success

Successful

F/U Patient PRN

Unsuccessful

3rd directed EBPs +/- Fibrin Sealant (if available)

Consider Neurosurgery
Prognosis

- Recurrence: 10% (regardless of treatment)
- Good prognostic factors
  - Classical MRI changes
  - Focal CSF leak
- Poor prognostic factors:
  - Normal initial MRI
  - Diffuse multi-level CSF leak
References

2. Schievink W. Spontaneous spinal CSF leaks and intracranial hypotension. JAMA 2006; 295(19) 2286-96


