New daily persistent headache

What is it? Is it a diagnosis?

What is the ICHD-2 definition? How about ICHD-3?

Primary and secondary forms

Discuss some secondary causes in detail

NDPH – The original description...

Vanast et al, Headache, 1986

N = 45

Daily headache from onset, ‘no prior hist. of ha’

80% - male

1/3rd – unilateral, 1/3rd – pounding, 1/2 – nausea, up to 1/2 – photo or phono

2/3rd resolved in 2 years

Is this all one thing?

ICHD-2 definition

Diagnostic criteria:

- Headache that, within 3 days of onset, fulfills criteria B-D
- Headache is present daily, and is unremitting, for >3 months
- At least two of the following pain characteristics:
  - bilateral location
  - pressing/limiting (non-pulsating) quality
  - mild or moderate intensity
  - not aggravated by routine physical activity such as walking or climbing
- Both of the following:
  - no more than one of photophobia, phonophobia or mild nausea
  - neither moderate or severe nausea nor vomiting
- Not attributed to another disorder

In other words, like tension-type headache but daily from start

Migrainous features in NDPH

Rozen et al, Headache currents, 2010
Epidemiology

- Prevalence rates in general population studies
- Castillo et al - n = 2252, 0.1% NDPH (4.7% CDH)
- Grande et al – n = 3500, 0.03 % NDPH (30-44 years only)
- In selected CDH, 7-13% had NDPH

Pathophysiology

- Not known
- Viral infection – EBV, Herpes simplex, CMV. ? CSF raised TNF-alpha
- Stressful life event
- Head/Neck trauma
- Connective tissue - ? Low CSF pressure, ? Cervical origin of pain

Prognosis

- Self-limiting form (? Migrainous-subtype)
- Refractory form (? Featureless-subtype)
- Relapsing-remitting form

Treatment

- Takase et al, cephalalgia, 2004
  - N = 30 (out of 1760 CDH) over a 5 yr period
  - ICHD-2 defined NDPH
  - 17 males
  - Mean age – 38, mean age of onset – 35 yrs

- Meineri et al, Neurol sci, 2004
  - N = 18, retrospective
  - Amitryptiline, fluoxetine, valproic acid
  - None effective

- Evans et al, Headache 2001. n=1. effective for gabapentin, venlafaxine, topiramate, nortryptiline (modified criteria)...
- Marmura et al, Headache 2008. n = 3. mexilitine effective

Thankfully.....ICHD-3
ICHD-3 definition

• A. Persistent headache fulfilling criteria B and C
• B. Distinct and clearly remembered onset, with pain becoming continuous and unremitting within 24 hours
• C. Present for > 3 months
• D. Not better accounted for by another ICHD-3 diagnosis

NDPH - the big picture!

Spontaneous intracranial hypotension

• Orthostatic headaches first described by Georg Schaltenbrand in 1939 “aliquorrhea” – reduced CSF production
• Incidence – 5/100,000 per year (Schievink WI. JAMA. 2006;295:2286-96)
• Classical triad – orthostatic headache, pachymeningeal enhancement on imaging and low CSF pressure

Is it pressure or volume?

Is it pressure or volume?

Headache patterns in SIH

<table>
<thead>
<tr>
<th>Recent onset headache</th>
<th>Chronic headache</th>
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</thead>
<tbody>
<tr>
<td>Orthostatic headaches</td>
<td>Orthostatic headaches evolving in months to chronic lingering headaches</td>
</tr>
<tr>
<td>Exertional headaches without any orthostatic features</td>
<td>Non-orthostatic chronic daily headache</td>
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<tr>
<td>Acute thunderclap-like onset of orthostatic headaches</td>
<td>Second half of the day headaches (often with some orthostatic features)</td>
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<tr>
<td>Paradoxical orthostatic headaches</td>
<td></td>
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</tbody>
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Connective tissue disorders and SIH

- Marfans, Ehlers Danlos -2, Autosomal dominant polycystic kidney, retinal detachment, unnamed
Non-headache symptoms in SIH

- Spinal pain
- Cochleo-vestibular changes
- Diplopia
- Nausea
- Upper limb paraesthesiae
- Personality change, memory

Diagnostic criteria – ICHD-3

A. Any headache fulfilling criteria C

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

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

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

Diagnostic tests

1. Confirm the clinical suspicion of CSF leak

2. If needed, demonstrate the site of leak

Spontaneous intracranial hypotension (SIH)

- MRI, Brain with contrast

Spine MRI abnormalities in SIH

T2 vs STIR (short T1 inversion recovery) sequences

Spine MRI abnormalities in SIH

extradural extravasation of CSF

Spine MRI abnormalities in SIH
extra-arachnoid extravasation of CSF

CSF findings in SIH
- CSF pressures are less than 6 cm of water and can be unmeasurable.
- The CSF pressure can be normal in 40%
- CSF cell count can be high (upto 100 cells)
- CSF protein can be high (upto 1 gm)
- CSF glucose is never abnormal

Schievink WI. Cephalalgia. 2008 Dec;28(12):1345-56

Demonstration of CSF spinal leak
- CT myelogram
- Radionuclide cisternography
- Spinal MRI myelography (with Gd)
- Heavily T2 weighted spinal MR myelography

Demonstration of spinal CSF leak
- CT Myelogram
- Intrathecal contrast
- Early and delayed CT scans
- May miss 'fast flow' leaks – Dynamic CT maybe useful
- 'Slow flow' leaks may take several hours
- Majority of the leaks are at the cervico-thoracic junction or in the thoracic spine
- Multiple leaks are common
- Considered the most reliable test to demonstrate site of leak


Demonstration of spinal CSF leak
- Radionuclide cisternography
- Intrathecal contrast, mostly Indium based

Demonstration of spinal CSF leak

Heavily T2-weighted MR myelography

- Contrast not necessary

Management - conservative

- Most CSF leaks resolve spontaneously and the majority of patients may not require any treatment

- Treatments often tried:
  - Bed rest (with head rest lowered)
  - Hydration
  - Abdominal binder
  - Caffeine (oral or iv)
  - Steroids
  - Theophylline
  - Intrathecal saline
  - Vitamin A

Management – Epidural Blood Patch (EBP)

- Should be considered if conservative means fail

- Blind EBP at lumbar level with autologous blood – effective in 36 – 57%

- If the CSF leak is localised a targeted blood patch should be given. 87 % patients given a targeted blood patch showed clinical improvement when compared to 52 % of those given a 'blind' blood patch (p<0.05)

Complications in SIH

- Epidural blood patch related
  - Cerebral venous sinus thrombosis

- Subdural haematomas requiring intervention

- Rebound intracranial hypertension

Work-up algorithm

- Thank you for your kind attention!

- Open for questions!