Classification and management of other primary headaches

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ICHD-3 beta: 4. Other primary headache disorders

- Clinically heterogeneous
  - 4.1 Primary cough headache
  - 4.2 Primary exercise headache
  - 4.3 Primary headache associated with sexual activity
  - 4.4 Primary thunderclap headache
  - 4.5 Cold-stimulus headache
  - 4.6 External pressure headache
  - 4.7 Primary stabbing headache
  - 4.8 Nummular headache
  - 4.9 Hypnic headache
  - 4.10 New daily persistent headache

Introduction

- Rare disorders – lifetime prevalence < 1.2% (Schwaiger et al., 2009)
- Dx secondary headaches!!! → careful evaluation and imaging!
- 4 categories:
  - HAs associated with physical exertion 4.1-4.4
  - HAs attributed to direct physical stimuli 4.5, 4.6
  - Epicranial HAs 4.7, 4.8
  - Miscellaneous 4.9, 4.10
- Treatment – since rare disorders → no large RCTs

4.1 Primary cough headache

- HA precipitated by coughing or other straining manoeuvre but NOT by prolonged physical exercise
- Diagnostic criteria
  - 2 HA episodes
  - Brought on by and occurring only in association with coughing, straining and/or other Valsalva manoeuvre
  - Sudden onset
  - 1 sec – 2 h

Primary cough headache

- Starts suddenly and then subsides over several seconds to a few minutes ... 2h
- Usually bilateral and posterior
- Predominantly affects patients > 40 y, M > F (3:5:1)
- 2/3 vertigo, nausea, sleep abnormalities
- NB! 40% symptomatic!!!
- Dx – Arnold-Chiari type I, basilar impression, CSF hypotension, carotid or vertebrobasilar diseases, middle cranial fossa or posterior fossa tumours, subdural haematoma, cerebral aneurysms, RCVS
- Treatment:
  - Indomethacine 50-200 mg/d, acetazolamide 125 mg tid, topiramate
  - LP 40 ml

4.2 Primary exercise headache

- Previously: primary exertional HA
- Diagnostic criteria
  - 2 episodes
  - Brought on by and occurring only during or after strenuous physical exercise
  - < 48h

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Primary exercise HA

- occurs particularly in heat or at high altitude
- precipitated by sustained physically strenuous exercise (compare – not Valsalva!)
- Mostly pulsatile
- Dx - SAH, arterial dissection, RCVS, mass lesion in the posterior fossa, sinusitis, Arnold-Chiari malformation, cardiac cephalalgia
- Treatment – indomethacine short term or 25-50 tid, propranolol 20-80 mg tid (6-8 w)
- Regular training, slow increase in sports activity, normal BMI

Primary headache associated with sexual activity

- HA precipitated by sexual activity, usually starting as a dull bilateral ache as sexual excitement increases and suddenly becoming intense at orgasm
- Diagnostic criteria
  - 2 episodes
  - only during sexual activity
  - increasing in intensity with increasing sexual excitement OR abrupt explosive intensity just before or with orgasm
  - 1 min - 24 h with severe intensity and/or up to 72 h with mild intensity

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- In majority only once in a lifetime, 44% have one or more relapses

Primary thunderclap headache

- NB!!! Dx!!! Extensive neuroimaging ± LP
- SAH, ICH, cerebral venous thrombosis, unruptured vascular malformation, aneurysm, arterial dissection, RCVS, pituitary apoplexy, meningitis, CSF hypotension
- Diagnosis of elimination!!!
- Treatment: acute treatment in the emergency settings – i/v paracetamol, opiates

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4.4 Primary thunderclap headache

- High-intensity headache with abrupt onset
- Diagnostic criteria:
  - Severe pain
  - Abrupt onset, reaching max intensity in < 1 min
  - > 5 minutes
  - No symptomatic cause!!!

- In majority only once in a lifetime, 44% have one or more relapses

4.5 Cold-stimulus headache: HA brought on by a cold stimulus applied externally to the head or ingested or inhaled

- External application of a cold stimulus
  - 2 episodes
  - Brought on by and occurring only during application of an external cold stimulus to the head
  - Resolving within 30 min after removal of the cold stimulus

- Ingestion or inhalation of a cold stimulus
  - 2 episodes of acute frontal or temporal HA
  - Brought on by and occurring immediately after a cold stimulus to the palate and/or posterior pharyngeal wall from ingestion of cold food or drink or inhalation of cold air
  - Resolving within 10 min after removal of the cold stimulus
Cold-stimulus headache

- „ice-cream headache“, „brainfreeze“
- More common in migraineurs
- Management – prevention – avoiding the known stimulus

External pressure headache

- Compression – tight band around the head, hat or helmet, goggles worn during swimming or diving, without damage to the scalp
- Traction – „ponytail headache“
- Management – removal of the causative agent

Primary stabbing headache

- 80% of stabs last 3 seconds or less (10–120 seconds)
- Attack frequency is generally low, one or a few per day
- 70% extratrigeminal regions
- 1/3 fixed location → exclude structural changes!
- NO autonomic sx!!!
- Treatment (if attack frequency high...): indomethacine 25-50 mg bid, gabapentin 400 mg bid, melatonin 3-12 mg

4.6 External pressure headache

HA resulting from sustained compression or traction on pericranial soft tissues.

External-compression headache

- 2 episodes
- Occurring within 1 h during sustained external compression of the forehead or scalp
- Maximal at the site of external compression
- Resolving within 1 h after external compression is relieved

External-traction headache

- 2 episodes
- Only during sustained external traction on the scalp
- Maximal at the traction site
- Resolving within 1 h after traction is relieved

4.7 Primary stabbing headache

- Ice-pick pains, jabs and jolts, needle-in-the-eye syndrome, ophthalmodynia periodica, short sharp short-lived head pain
- Diagnostic criteria
  - Head pain occurring spontaneously as a single stab or series of stabs
  - Each stab lasts for up to a few seconds
  - Stabs recur with irregular frequency, from one to many per day
  - NO cranial autonomic symptoms
- Treatment (if attack frequency high...): indomethacin 25-50 mg bid, gabapentin 400 mg bid, melatonin 3-12 mg

4.8 Nummular headache

- „nummus“ = „coin“ in Latin
- Diagnostic criteria:
  - Continuous or intermittent head pain
  - Felt exclusively in an area of the scalp, with all of the following 4 characteristics:
    - 1. sharply contoured
    - 2. fixed in size and shape
    - 3. round or elliptical
    - 4. 1–6 cm in diameter

8.06.2016
Nummular headache

- Usually in the parietal region
- Rarely bi- or multifocal
- 75% chronic
- Hypaesthesia, dysaesthesia, paraesthesia, allodynia and/or tenderness
- Exclude structural and dermatological lesions!
- Treatment: gabapentin 900-1800 mg/d, TCAs, botulinum toxin
- Often becomes refractory...

4.9 Hypnic headache

- Hypno = “sleep” in Greek
- “Alarm clock” headache
- Diagnostic criteria:
  - Recurrent headache attacks
  - Developing only during sleep, and causing waking
  - Occurring on 10 days per month for >3 months
  - Lasting 15 min - 4 h after waking
  - No cranial autonomic symptoms or restlessness

Hypnic headache

- Usually begins > 50 y
- 2/3 bilateral, mostly frontotemporal
- Generally tension-type like
- Dx – cluster headache, sleep apnoea, nocturnal hypertension, hypoglycaemia, medication overuse, intracranial disorders
- Treatment:
  - Caffeine, lithium 150-600 mg/die, indomethacine, melatonin, topiramate, pregabalin

4.10 New daily persistent headache

- Diagnostic criteria
  - Persistent headache
  - Distinct and clearly remembered onset, with pain becoming continuous and unremitting within 24 hours
  - Present for >3 mo

- Tension-type-like or migraine-like

NDPH

- Dx – increased CSF pressure, decreased CSF pressure, trauma, MOH

- 2 temporal profiles:
  - Self-limited form
  - Refractory, persistent form

- Treatment: aimed at matching the predominant headache phenotype
  - TTH-like – amitriptyline up to 150 mg/die
  - Migraine-like – valproic acid 600-900 mg/die, topiramate

Take home message

- Other primary headaches are rare and heterogenous
- Secondary causes must be excluded!
- Knowing about these headaches and their treatment helps your patient in a great deal... 😊
Thank you!