Primary Headache Disorders Classification and Diagnosis

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Why do we need a headache classification system?

- It helps with research into mechanisms & treatment
- Essential for understanding headache epidemiology
- Reimbursement
- A tool for Diagnosis
- Patients want a dx
History

• International classification of headache disorder, 1st edition (ICHD-1) in 1988
• ICHD-2 in 2004
• ICHD-3 beta in 2013
• ICHD-3 in 2016
Why a beta version?

• An opportunity for broad input and detection of errors
• Research opportunities
• WHO plans field testing of ICD-11
• ICHD-3 can be field tested at the same time
• Allows final version to coincide with final WHO codes
ICHDIII – Basic Organization

Part 1: Primary headaches
Part 2: Secondary headaches
Part 3: Cranial Neuralgias, etc.
The Appendix
Part 1: Primary headaches

(no other causative disorder)

1. Migraine
2. Tension-type Headache
3. Cluster and its relatives (TACs)
4. Other primary headaches – exertional, hypnic headache, NDPH, et al
ICHHD III

Part 2: Secondary headaches
(cause by another disorder)
5. Posttraumatic
6. Vascular disease
7. Nonvasc intracranial pathology – e.g. mass, pressure
8. Substances
9. CNS infection
10. Homeostatic disorder
11. Cranial, neck, EENT, sinuses, teeth, jaw
12. Psychiatric
ICHDA III

Part 3: Cranial Neuralgias, Central and primary facial pain, other headaches

13. Neuralgias and neuropathy
14. Other Headaches (Empty for now)
ICHDI II

The Appendix:

1. Suggested criteria for possible new entities

2. Alternative diagnostic criteria for certain categories (pending evidence)

3. Some previously accepted disorders which have not been supported by evidence
Format of the ICHD

Hierarchical format allows one to decide how detailed to make the diagnosis.

E.g.:
– in primary care migraine with aura 1.2 is sufficient
– for researcher or specialist, 1.2.1 or 1.2.2 might be important (typical aura with migraine or typical aura with non-migraine)
ICHDIII

Key nomenclature

• **Chronic** – refers to high frequency for the primary headache disorders—i.e. chronic migraine occurs on more than 50% of days
• But, means non-remitting in cluster and TACs,
• And, means long duration of the problem for some secondary headaches like *chronic postinfectious headaches*. New word – **persistent** which means the same thing for PTH

• **Episodic** – less than 15 days per month for migraine and TTHA; But in cluster headache, paroxysmal hemicrania, and SUNCT it means remitting.
ICHD III

Key nomenclature

• **Probable**
  • This term replaces terms like *Migrainous headache* and is used ubiquitously to indicate generally that all but one criterion has been met for a particular diagnosis.
  • For example, if a patient has recurrent headaches which seem migrainous but fails to fulfill one of the 4 criteria for migraine
  • In ICHD III this is reserved for primary headaches only
ICHD III

Part 1: Primary headaches, chapters 1-4 (no other causative disorder)

1. Migraine
2. Tension-type Headache
3. Cluster and its relatives (TACs)
4. Other primary headaches – exertional, headaches associated with sexual activity, new daily persistent headaches, hypnic headache, etc.
1. Migraine

1.1 Migraine without aura
1.2 Migraine with aura
1.3 Chronic migraine
1.4 Complications of migraine
1.5 Probable migraine
1.6 Episodic syndromes that may be associated with migraine

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1.2 Migraine with aura

1.2.1 Migraine with typical aura
   1.2.1.1 Typical aura with HA
   1.2.1.2 Typical aura without HA

1.2.2 Migraine with brainstem aura

1.2.3 Hemiplegic migraine
   1.2.3.1 Familial hemiplegic migraine (FHM)
      1.2.3.1.1 Familial HM type 1
      1.2.3.1.2 Familial HM type 2
      1.2.3.1.3 Familial HM type 3
      1.2.3.1.4 Familial HM other loci
   1.2.3.2 Sporadic HM

1.2.4 Retinal migraine
Migraine with typical aura

A. At least two attacks fulfilling criteria B and C
B. Aura consisting of visual, sensory and/or speech/language symptoms, each fully reversible, but no motor, brainstem or retinal symptoms
C. At least two of the following four characteristics:
   1. at least one aura symptom spreads gradually over 5 minutes, and/or two or more symptoms occur in succession
   2. each individual aura symptom lasts 5-60 minutes
   3. at least one aura symptom is unilateral
   4. the aura is accompanied, or followed within 60 minutes, by headache

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Hemiplegic Migraine

- Mutations in CACNA1A gene on chromosome 19p13 in half of the families
- ATP1A2 gene on chromosome 1 in a few families
- A few families could not be linked to either chromosome 19 or chromosome 1 and therefore at least a third gene most be involved
- In sporadic HM, genes have not been identified
1.2.4 Retinal migraine

A. At least 2 attacks fulfilling criteria B and C
B. Aura of fully reversible monocular positive and/or negative visual phenomena confirmed during an attack by either or both of the following:
   1. clinical visual field examination
   2. patient’s drawing of a monocular field defect
C. ≥2 of the following 3 characteristics:
   1. aura spreads gradually over ≥5 min
   2. aura symptoms last 5-60 min
   3. aura accompanied or followed in <60 min by headache
Migraine with Aura ICHD III

- Migraine with aura
  - Migraine with typical aura
  - Migraine with brainstem aura
  - Migraine with hemiplegia
  - Retinal migraine

With headache
Without headaches
Case 1 – Weird Aura

• 29 y/o multimillionaire software company CEO describes frequent severe holocranial headaches preceded or accompanied by vertigo & brain fog
• Nausea, phono and photosensitivity, neck pain are common accompaniments
• Exam, including Hallpike maneuver, and head MRI with gad all negative
1.2.2 Migraine with brainstem aura

A. At least 2 attacks fulfilling criteria B and C below, and criteria C and D for 1.2.1 Migraine with typical aura

B. Aura of fully reversible visual, sensory and/or speech/language symptoms, but not motor or retinal

C. ≥2 of the following brainstem symptoms:

1. dysarthria
2. vertigo
3. tinnitus
4. hypacusis
5. diplopia
6. ataxia
7. decr level of consc

Problems

1. Symptoms “clearly” emanating from the brainstem
2. What to call migraine accompanied only by vertigo? Or vertigo without HA?
A1.6.5 Vestibular migraine

A. At least five episodes fulfilling criteria C and D
B. A current or past history of 1.1 Migraine without aura or 1.2 Migraine with aura
C. Vestibular symptoms of moderate or severe intensity, lasting between 5 minutes and 72 hours
D. At least 50% of episodes are associated with at least one of the following three migrainous features:
   1. headache with at least two of the following four: a) unilat location b) pulsating quality c) moderate or severe intensity d) aggravation by routine activity
   2. photophobia and phonophobia
   3. visual aura

Yoon-Hee Cha. Long-Term Follow-Up of Vestibular Migraine. Neurology 2012
Migraine with Aura ICHD III

- Migraine with aura
  - Migraine with typical aura
  - Migraine with brainstem aura
  - Migraine with hemiplegia
  - Retinal migraine
  - Vestibular migraine

- With headache
- Without headaches
Chronic migraine

A. Headache (tension-type-like and/or migraine-like) on **15 days** per month for >3 months

B. Occurring in a patient who has had at least five attacks fulfilling criteria B-D for 1.1 Migraine without aura and/or criteria B and C for 1.2 Migraine with aura

C. On **8 days per month for >3 months**, fulfilling any of:
   1. criteria C and D for 1.1 Migraine without aura
   2. criteria B and C for 1.2 Migraine with aura
   3. believed by the patient to be migraine at onset and relieved by a triptan or ergot derivative

D. Not better accounted for by another ICHD-III diagnosis.
Is Chronic migraine different than intermittent migraine?

If so, where to draw the line? 15 days/mo? 20? 28?
And is daily migraine different from nearly daily migraine?
Is Chronic migraine different than intermittent migraine?

Imaging studies – PET, DTI - may begin to differentiate

Maniyar, FH and Goadsby, PJ. Functional imaging in Chronic Migraine. Curr Headache and Pain Reports, 2013. - Increasing frequency of migraine attacks is associated with changes in key brainstem areas, basal ganglia and various cortical areas involved in pain.

Accurate Classification of Chronic Migraine via Brain MRI
Schwedt et al Headache 2015;55:762-777

**Methods**—Study participants had MRI. Headache frequency thresholds ranging from 5-15 headache days/month were evaluated to determine the threshold allowing for the most accurate subclassification of individuals into lower and higher frequency subgroups via measurements in temporal pole, anterior cingulate cortex, superior temporal lobe, entorhinal cortex, medial orbital frontal gyrus, and pars triangularis.

**Participants** were 66 migraineurs and 54 healthy controls, 75.8% female, with an average age of 36 +/- 11 years.
• CM can be differentiated from EM with 84% accuracy
• Differences seen Temp pole, sup temp, ant cingulate, entorhinal etc
• All areas are involved in pain processing

Schwedt et al Accurate Classification of Chronic Migraine via Brain MRI. *Headache* 2015;55:762-777
Accurate Classification of Chronic Migraine via Brain MRI
Schwedt et al *Headache* 2015;55:762-777

**Conclusions** — Classifiers consisting of cortical surface area, cortical thickness, and regional volumes were highly accurate for determining if individuals have chronic migraine.

Furthermore, results provide objective support for the current use of 15 headache days/month as a threshold for dividing migraineurs into lower frequency (ie, episodic migraine) and higher frequency (ie, chronic migraine) subgroups.
Case 2 – Focal deficit

• 28 year old radiology resident with history of migraine with visual and sensory auras since childhood seen in ER with left sided numbness persistent 2 days after onset of HA.
  • Head pain has abated
  • Exam nl, MRI normal
  • Diagnosis?
  • Treatment
1.4 Complications of migraine

- 1.4.1 Status migrainosus
- 1.4.2 Persistent aura without infarction
- 1.4.3 Migrainous infarction
- 1.4.4 Migraine aura-triggered seizure
1.4 Complications of migraine

• 1.4.1 Status migrainosus
• 1.4.2 Persistent aura without infarction
• 1.4.3 Migrainous infarction
• 1.4.4 Migraine aura-triggered seizure
• None of the above
1.4 Complications of migraine

• 1.4.2 Persistent aura without infarction
  – *Migraine with aura* and typical of previous auras except that one or more aura symptoms persists for ≥1 week

• 1.4.3 Migrainous infarction
  – *Migraine with aura* and typical of previous attacks except that one or more aura symptoms persists for >60 min
  – Neuroimaging demonstrates ischaemic infarction in a relevant area
2. Tension type HA

≥2 of the following 4 characteristics:

1. bilateral location
2. pressing or tightening (non-pulsating) quality
3. mild or moderate intensity
4. not aggravated by routine physical activity

Both of the following:

1. no nausea or vomiting
2. no more than one of photophobia or phonophobia
3. TACs

Chapter 3 – Cluster Headaches and TAC

• Cluster (3.1)
• Paroxysmal hemicrania (3.2)
• SUNCT and SUNA (3.3)

• All are divided in episodic (remitting) and chronic (unremitting) forms
3.1 Cluster headache

A. At least 5 attacks fulfilling criteria B-D
B. Severe or very severe unilateral orbital, supraorbital and/or temporal pain lasting **15-180 min** if untreated
C. Headache is accompanied by ≥1 of the following:
   1. ipsilateral conjunctival injection and/or lacrimation
   2. ipsilateral nasal congestion and/or rhinorrhoea
   3. ipsilateral eyelid oedema
   4. ipsilateral forehead and facial sweating
   5. ipsilateral miosis and/or ptosis
   6. a sense of *restlessness* or agitation
D. Attacks have a frequency from 1/2 d to 8/d
E. Not attributed to another disorder
3.2 Paroxysmal hemicrania

A. At least 20 attacks fulfilling criteria B-D
B. Attacks of severe unilateral orbital, supraorbital or temporal pain lasting 2-30 min
C. Headache is accompanied by ≥1 of the following:
   1. ipsilateral conjunctival injection and/or lacrimation
   2. ipsilateral nasal congestion and/or rhinorrhea
   3. ipsilateral eyelid oedema
   4. ipsilateral forehead and facial sweating
   5. ipsilateral miosis and/or ptosis
D. Attacks have a frequency >5/d for > half of the time, although periods with lower frequency may occur
E. Attacks are prevented completely by therapeutic doses of **indomethacin**
F. Not attributed to another disorder
3.3 SUN Headaches - Short-lasting Unilateral Neuralgiform headache attacks

B. Moderate or severe unilateral head pain, with orbital, supraorbital, temporal and/or other trigeminal distribution, lasting for 1–600 sec

C. At least one of the following cranial autonomic symptoms or signs, ipsilateral to the pain:
   1. conjunctival injection and/or lacrimation
   2. nasal congestion and/or rhinorrhoea
   3. eyelid oedema
   4. forehead and facial sweating
   5. forehead and facial flushing
   6. sensation of fullness in the ear
   7. miosis and/or ptosis

SUNCT

SUNA
TAC’s:

Duration decreases with name length

Cluster
15-180 min
Paroxysmal Hemicrania
2-30 min

Short lasting unilateral neuralgiform headaches with conjunctival injection and tearing - SUNCT
1-600 sec
3.4 Hemicrania continua

Constant (>3 mo), unilateral, indomethacin-responsive headache with either

1. At least one of the following symptoms or signs ipsilateral to the headache:
   • conjunctival injection and/or lacrimation
   • nasal congestion and/or rhinorrhoea
   • eyelid oedema
   • forehead and facial sweating
   • forehead and facial flushing
   • sensation of fullness in the ear
   • miosis and/or ptosis

2. A sense of restlessness or agitation, or aggravation of the pain by movement
TAC’s:

Duration decreases with name length

- **Cluster:** 15-180 min
- **Paroxysmal Hemicrania:** 2-30 min
- Short lasting unilateral neuralgiform headaches with conjunctival injection and tearing - SUNCT: 1-600 sec
Case 3: The medicine he told me would work, didn’t

38 year old man with headaches for the last 4 years is referred for intractable migraine. Pain is strictly R-sided and is present essentially every day. He has tried everything, including triptans, beta blockers, topiramate, amitriptyline, verapamil, gabapentin and Botox.

Exam is normal except for some redness of the R eye. Dr. Ward asked if he ever had nasal drainage or tearing. Pt responded – “All the time”.

A medication was prescribed...
What is hemicrania continua which fails to respond to indomethacin

Your Options:
• It is not HC so look for a possible alternate diagnosis
• Refer the patient to someone else
• Try Depakote or Lamotrigine
What is hemicrania continua when it is bilateral?

- Bilateral Hemicrania
- Bicrania
- Transcranial
- An abomination
4. Other primary headache disorders

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 (NDPH)
Primary headache associated with sexual activity

- Brought on by & occurring only during sexual activity
- Either or both of the following:
  1. increasing in intensity with increasing sexual excitement
  2. abrupt explosive intensity around orgasm
- Lasting from 1 min to 24 hr with severe intensity and/or up to 72 hr with mild intensity
- Rare but comorbid with migraine
- Both exercise and sex induced HA can be prevented with indomethacin
Cold and Pressure

4.5 Cold-stimulus headache
   4.5.1 Headache attributed to external application of a cold stimulus
   4.5.2 Headache attributed to ingestion or inhalation of a cold stimulus

4.6 External-pressure headache
   4.6.1 External-compression headache
   4.6.2 External-traction headache
Primary stabbing headache – icepick headache

A. Head pain occurring spontaneously as a single stab or series of stabs and fulfilling criteria B-D
B. Each stab lasts for up to a few seconds
C. Stabs recur with irregular frequency, from one to many per day
D. No cranial autonomic symptoms
4.9 Hypnic headache

A. Dull headache fulfilling criteria B-D
B. Develops only during sleep, and awakens patient
C. At least two of the following characteristics:
   1. occurs >15 10 times/mo
   2. lasts ≥15 min after waking
   3. first occurs after age of 50
D. No autonomic symptoms and no more than one of nausea, photophobia or phonophobia
E. Not attributed to another disorder
Case 4 - Thunderclap HA

- 41 year old amateur runner finished a marathon (19th in a field of 350) and while cooling down, experienced a fairly abrupt headache which rapidly escalated to VERY severe holocranial pain which made race officials call 911. In the ED, VS were normal, neck was supple, neuro exam was non-focal, but pain was persistent. CT was neg, LP showed OP of 160 mm and fluid was clear with normal chem, 1 WBC and 2 RBCs.
Thunderclap HA Causes

- Intracerebral hemorrhage
- Subarachnoid hemorrhage
- Pituitary Apoplexy
- Cerebral Venous Thrombosis
- Arterial Dissection
- CNS Vasculitis, RCVS
- Intracranial hypotension
- Primary Thunderclap Headache
- Sex related Headache
- Meningitis
- Acute Sinusitis
<table>
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<th>RCVS</th>
<th>CNS Vasculitis</th>
<th>SAH</th>
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<tr>
<td>Thunderclap</td>
<td>Slower</td>
<td>Thunderclap</td>
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<td>Young female</td>
<td>Older (40-60)</td>
<td>Older (40-60) Fam hx</td>
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<td>Vasoactive drugs, pregnancy</td>
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<td>Neuro deficit less common</td>
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<td>Focal SA blood</td>
<td>CSF high prot, cells</td>
<td>Diffuse SA blood</td>
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<td>CSF nl</td>
<td>MRI abn – strokes</td>
<td>CSF xanth, blood</td>
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4.4 Primary thunderclap headache

A. Severe head pain fulfilling criteria B and C

B. Both of the following characteristics:
   1. **sudden onset**, reaching maximum intensity in <1 min
   2. lasting from 1 h to 10 d

C. **Does not recur regularly** over subsequent weeks or months

D. Not attributed to another disorder
Does primary thunderclap headache exist?

Or is it just a rapid onset migraine?
Or is it missed secondary headache that was not fatal or destructive?

“Evidence that Thunderclap headache exists as a primary condition is poor; the search for an underlying cause should be expedited and expected” (ICHD III)
4.8 New daily-persistent headache

In ICHD II -- Had to be a tension type headache

A. Headache for >3 mo fulfilling criteria B-D
B. Headache is daily and unremitting from onset or from <3 d from onset
C. At least two of the following pain characteristics:
   1. bilateral location
   2. pressing/tightening (non-pulsating) quality
   3. mild or moderate intensity
   4. not aggravated by routine physical activity
D. Both of the following:
   1. not >1 of photophobia, phonophobia or mild nausea
   2. neither moderate or severe nausea nor vomiting
E. Not attributed to another disorder

ICHDI-II. Cephalalgia 2004; 24 (Suppl 1) ©International Headache Society 2003/4
New daily persistent headache (NDPH)

A. Persistent headache fulfilling criteria B and C
B. Distinct and clearly-remembered onset, with pain becoming continuous and unremitting within 24 h
C. Present for >3 mo

“...pain and associated features may resemble chronic migraine or chronic tension-type headache, or have elements of both.”
What’s New in the Appendix?

• A1.3.1 Chronic migraine with pain free periods
• A1.3.2 Chronic migraine with continuous pain
• A1.4.5 Migraine aura status
• A1.6.1.3 Infantile Colic
• A.1.6.5 Vestibular migraine
Common errors in primary headache diagnosis I have seen

- Misdiagnose hemiplegic migraine – when migraine with typical aura
- Misdiagnose “ocular migraine” when it is really retinal or other ocular disease
- Misdiagnose sinus headache (when really migraine)
- Misdiagnose cluster
- Miss hemicrania continua and Paroxysmal Hemicrania