

ICHD 3 – Guide to use and key definitions ASHS UCNS Board Review

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Disclosures for Dr. Morris Levin

Consultant:

Supernus, Upsher-Smith, Lilly, Amgen,
Allergan, Alder, Teva

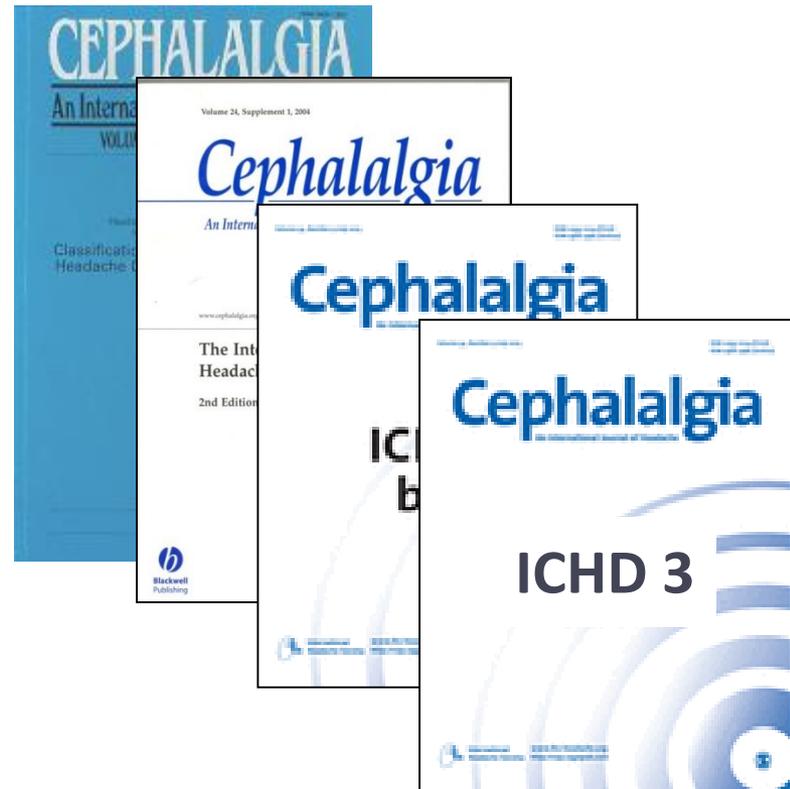
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Innovation

History

- International classification of headache disorder , 1st edition (ICHD-1) in 1988
- ICHD-2 in 2004
- ICHD-3 beta in 2013
- ICHD-3 in 2018*

*Based on 4 years of input including field testing and research aimed at increasing precision and accuracy



Painful structures in the head (neck)

- Dura
- Dural veins and arteries
- Intracran arteries
- V,VII,IX,X,C1-3
- Periosteum
- Scalp
- Scalp muscles
- Scalp vessels
- Sinuses
- Eyes
- Ears
- Teeth, TMJ
- Carotids, vertebrals
- C-spine periost
- Cerv musc

Painful structures in the head (neck)

Not painful

- *Parenchyma*
- *Pia, arachnoid*
- *Skull*
- *Ventricles*

ICHD III – Basic Organization

Part 1: Primary headaches

Part 2: Secondary headaches

Part 3: Cranial Neuralgias, etc.

The Appendix

ICHD III 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

ICHD III

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

Migraine

1.1 Migraine without aura

Diagnostic criteria:

- A. At least five attacks¹ fulfilling criteria B–D
- B. Headache attacks lasting 4–72 hours (when untreated or unsuccessfully treated)
- C. Headache has at least two of the following four characteristics:
 - 1. unilateral location
 - 2. pulsating quality
 - 3. moderate or severe pain intensity
 - 4. aggravation by or causing avoidance of routine physical activity
- D. During headache at least one of the following:
 - 1. nausea and/or vomiting
 - 2. photophobia and phonophobia

1.2 Migraine with Aura

ICHD 3 β

- A. At least two attacks fulfilling criteria B and C
- B. One or more of the following fully reversible aura symptoms:
 - 1. visual
 - 2. sensory
 - 3. speech and/or language
 - 4. motor
 - 5. brainstem
 - 6. retinal
- C. At least **2 of the following 4** characteristics:
 - 1. at least one **aura symptom spreads gradually** over ≥ 5 minutes **and/or two or more** aura 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 HA
- D. Not better accounted for by another ICHD-3 diagnosis.

New Definition of 1.2 Migraine with Aura

ICHD 3 Final

- A. At least two attacks fulfilling criteria B and C
- B. One or more of the following fully reversible aura symptoms:
 1. visual
 2. sensory
 3. speech and/or language
 4. motor
 5. brainstem
 6. retinal
- C. At least **three of the following six characteristics**:
 - 1.** at least one aura symptom spreads gradually over ≥ 5 minutes
 - 2.** two or more aura symptoms occur in succession
 3. each individual aura symptom lasts 5-60 minutes
 4. at least one aura symptom is unilateral
 - 5. at least one aura symptom is positive**
 6. the aura is accompanied, or followed within 60 minutes, by headache
- D. Not better accounted for by another ICHD-3 diagnosis.

New Definition of Migraine with Aura

Prospective testing of ICHD-3 beta diagnostic criteria for migraine with aura and migraine with typical aura in patients with transient ischemic attacks

Elena R Lebedeva^{1,2}, Natalia M Gurary³, Denis V Gilev⁴ and Jes Olesen⁵

Abstract

Introduction: The International Classification of Headache Disorders 3rd edition beta (ICHD-3 beta) gave alternative diagnostic criteria for 1.2 migraine with aura (MA) and 1.2.1 migraine with typical aura (MTA) in the appendix. The latter were presumed to better differentiate transient ischemic attacks (TIA) from MA. The aim of the present study was to field test that.

Methods: Soon after admission, a neurologist interviewed 120 consecutive patients diagnosed with TIA after MRI or CT. Semi-structured interview forms addressed all details of the TIA episode and all information necessary to apply the ICHD-3beta diagnostic criteria for 1.2, 1.2.1, A1.2 and A1.2.1.

Results: Requiring at least one identical previous attack, the main body and the appendix criteria performed almost equally well. But requiring only one attack, more than a quarter of TIA patients also fulfilled the main body criteria for 1.2. Specificity was as follows for one attack: 1.2: 0.73, A1.2: 0.91, 1.2.1: 0.88 and A1.2.1: 1.0. Sensitivity when tested against ICHD-2 criteria were 100% for the main body criteria (because they were unchanged), 96% for A1.2 and 94% for A1.2.1.

Conclusion: The appendix criteria performed much better than the main body criteria for 1.2 MA and 1.2.1 MTA when diagnosing one attack (probable MA). We recommend that the appendix criteria should replace the main body criteria in the ICHD-3.

“(new) 1.2
criteria
performed
much better in
distinguishing
MA from TIA

ICHD is
dynamic

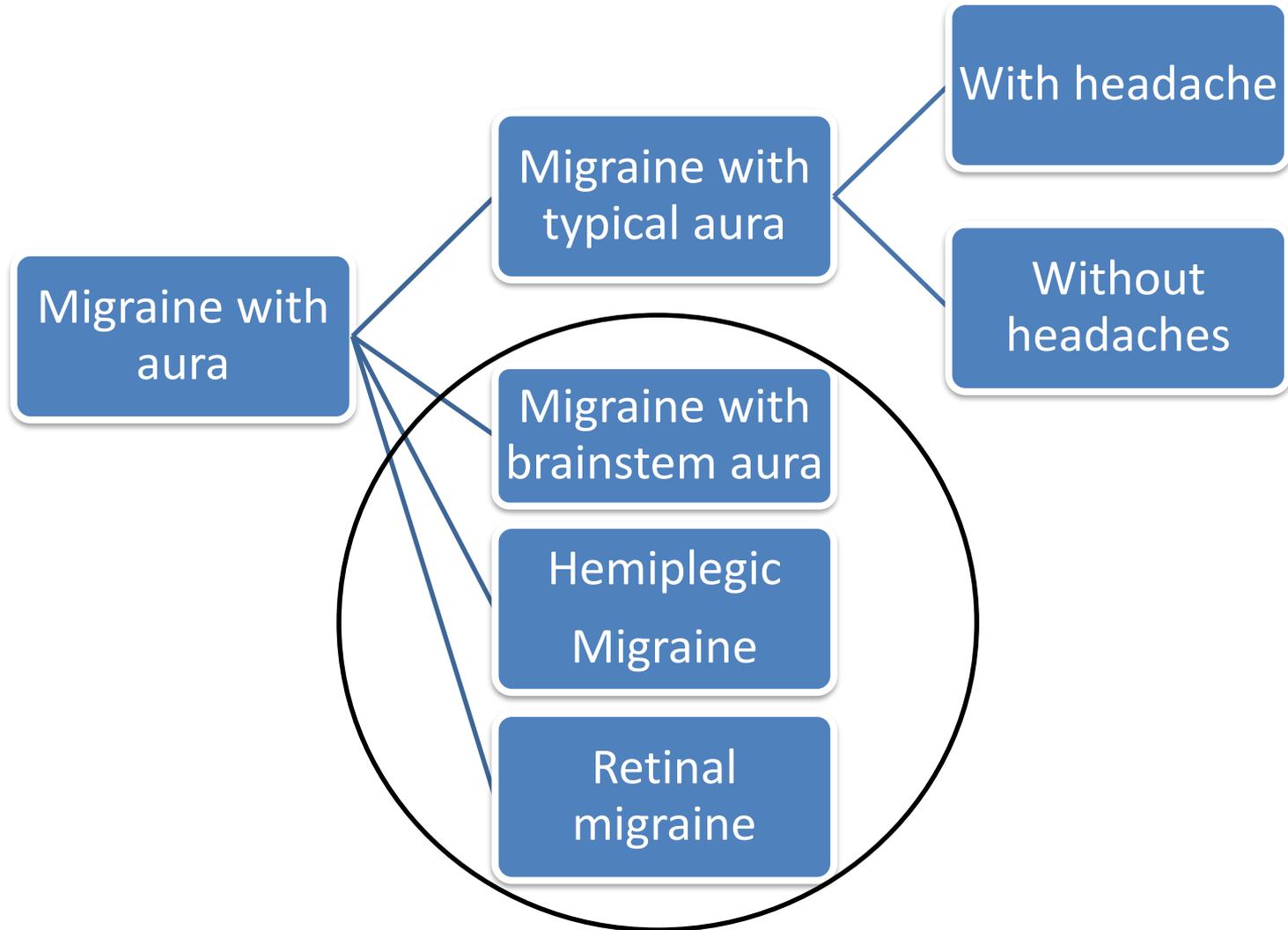
1.2 Migraine with typical aura

A. Attacks fulfilling criteria for 1.2 Migraine with aura and criterion B below

B. Aura with both of the following:

1. fully reversible visual, sensory and/or speech/language symptoms
2. no motor, brainstem or retinal symptoms.

Migraine with Aura ICHD III



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

Retinal migraine v. Binocular aura

The typical migraine visual aura is a binocular visual disturbance. Most patients describe the disturbance as monocular as it tends to occur in either the right or left visual field, which they interpret as occurring in the eye on the side of the disturbance.

It is present with eyes open or closed. Careful questioning almost always reveals that it occurs as a binocular phenomenon.

Retinal migraine v. Binocular aura

How to tell if symptoms are emanating from one eye or are homonymous?

If positive phenomena are emanating from one retina e.g. – closing that eye will NOT eliminate them

If negative phenomena are emanating from one retina, closing that eye will eliminate them.

But negative phenomena emanating from one retina may not have been noticed in the first place if that is not the dominant eye.

1.2.4 Retinal migraine

**Developmental
Neurobiology**

FORMERLY JOURNAL OF
NEUROBIOLOGY

Article

Spiral waves of spreading depression in the isolated chicken retina

N. A. Gorelova[†] and J. Bureš^{*}

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Issue



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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

Richter, F et al. Enhanced neuronal excitability in adult rat brainstem causes widespread repetitive brainstem depolarizations. *Jnl Cereb Blood Flow Metab* 2012; 32:1535-45.

Nosedá, R, & Burstein, R (2013). Migraine pathophysiology: anatomy of the trigeminovascular pathway and associated neurological symptoms, cortical spreading depression, sensitization, and modulation of pain. *PAIN* 2013, 154, S44-S53. – “descendent modulation of the brainstem trigeminovasc system” from CSD projected to brainstem via descending neuronal pathways

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

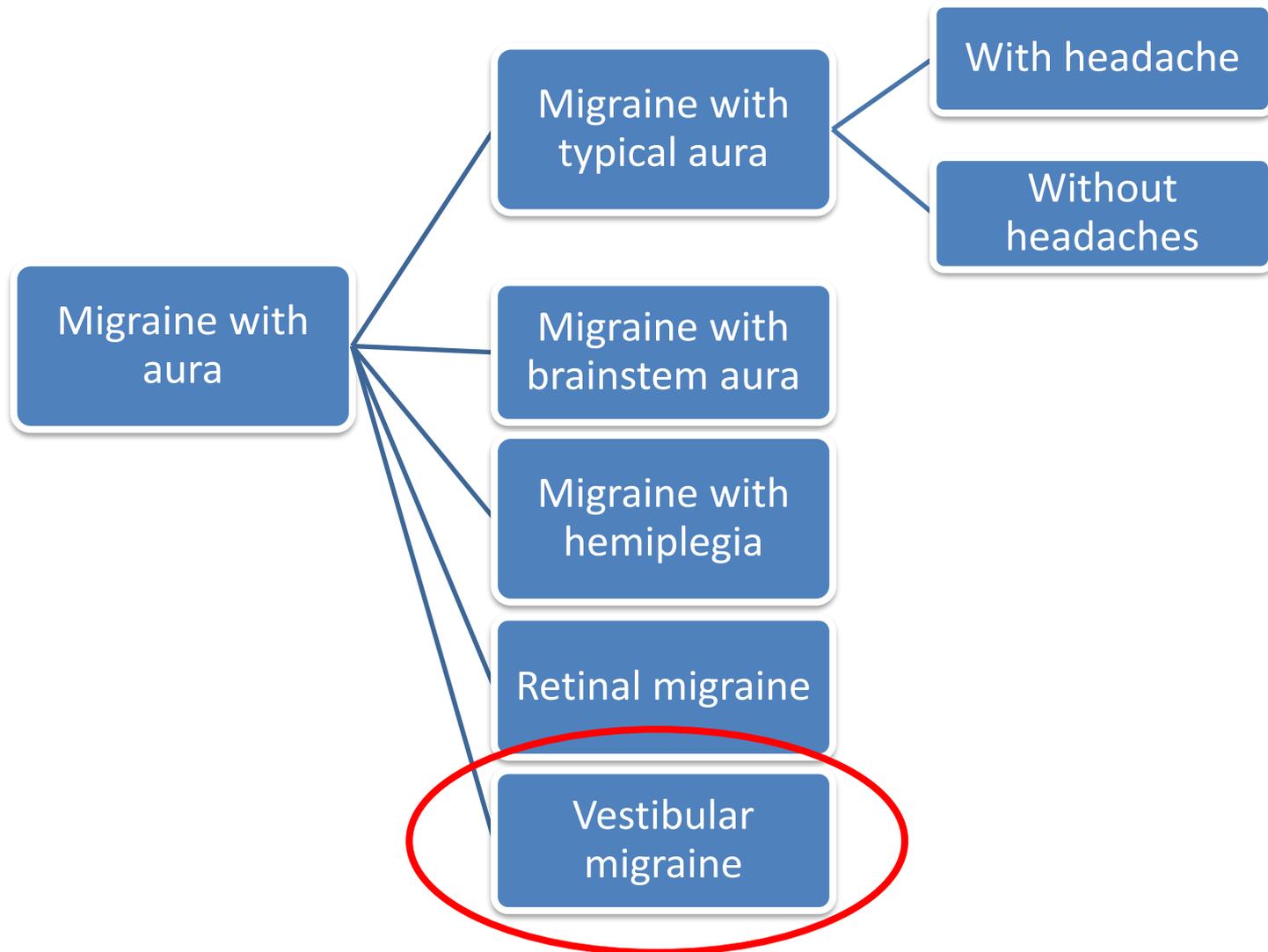
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A1.6.5 Vestibular migraine – a proposed migraine subtype

- 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 aura¹
- 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) unilateral location
 - b) pulsating quality
 - c) moderate or severe intensity
 - d) aggravation by routine activity
 2. photophobia and phonophobia
 3. visual aura

Migraine with Aura ICHD III



Menstrual migraine is not listed in the ICHD3 – considered a trigger – But:

Appendix

- A1.1.1 Pure menst mig without aura
- A1.1.2 Menstrually related mig without aura
- A1.1.3 Non-menst mig without aura

Chronic migraine defined as **migraine or strongly suspected migraine** on ≥ 15 d/mo

ICHD3 1.3 Chronic Migraine

- A. **Headache on ≥ 15 d/mo** for >3 mo
- B. In a patient who has had ≥ 5 attacks fulfilling criteria for 1.1 *Migraine without aura* or 1.2 *Migraine with aura*
- C. **On ≥ 8 d/mo** for >3 mo fulfilling any of the following:
 - 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-3 diagnosis

Imaging studies support the distinction between EM and CM

Maniyar, FH et al. 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.

Schwedt, T, et al *Headache* 2015 **Accurate subclassification** of individuals into lower and higher frequency subgroups via measurements of **cortical thickness** (and other measurements in temporal pole, anterior cingulate cortex, superior temporal lobe, entorhinal cortex, medial orbital frontal gyrus, and pars triangularis. **Threshold = 15 days per month**

1.2.3 Hemiplegic migraine

- A. At least 2 attacks fulfilling criteria B and C
- B. **Aura consisting of both of the following:**
 - 1. **fully reversible motor weakness; 2. fully reversible visual, sensory and/or speech/language symptoms**
- C. ≥ 2 of the following 4 characteristics:
 - 1. ≥ 1 aura symptom spreads gradually over ≥ 5 min, and/or ≥ 2 symptoms occur in succession
 - 2. each individual non-motor aura symptom lasts 5-60 min, and motor symptoms last < 72 h
 - 3. ≥ 1 aura symptom is unilateral
 - 4. aura accompanied or followed in < 60 min by headache
- D. Not better accounted for by another ICHD-3 diagnosis, and TIA excluded

1.2.3.1 Familial hemiplegic migraine (FHM)

- A. Fulfils criteria for 1.2.3 *Hemiplegic migraine*
- B. At least one first- or second-degree relative has had attacks fulfilling criteria for 1.2.3 *Hemiplegic migraine*

Mutations in the CACNA1A, ATP1A2, SCN1A, genes which code for ion transport channel proteins

PRRT2 gene mutation can also cause FHM – also benign familial infantile convulsions and kinesio-genic dystonia

1.2.3.1 Familial hemiplegic migraine (FHM)

- FHM1 - 1.2.3.1.1
- FHM2 - 1.2.3.1.2
- FHM3 - 1.2.3.1.3

- FHM very often presents with brainstem symptoms in addition to typical aura symptoms
- Headache almost always occurs.
- Rarely, during FHM attacks, disturbances of consciousness (including coma), confusion, fever and CSF pleocytosis can occur.

1.2.3.2 Sporadic hemiplegic migraine

- A. Fulfils criteria for 1.2.3 *Hemiplegic migraine*
- B. No first- or second-degree relative has had attacks fulfilling criteria for 1.2.3 *Hemiplegic migraine*

50% of hemiplegic migraine

Ddx for all forms of HM – TIAs, HANDL (HA, Neuro deficits and CSF lymphocytosis)

Complications of migraine

Status migrainosus

unremitting for >72 hours

pain and/or associated sx are debilitating

Persistent aura without infarction

>1 week with nl MRI

Migrainous infarction

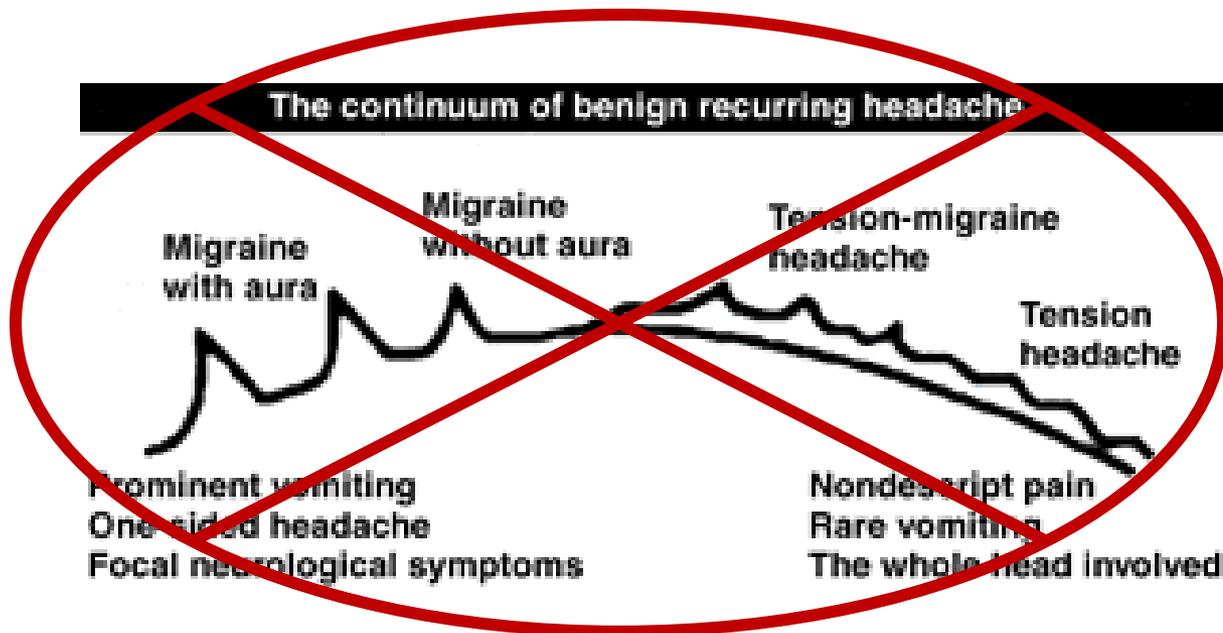
Aura sx >1 hour, MRI shows stroke in approp area

Pediatric migraine and syndromes

- Migraine in childhood – 2-72 h
- 1.6.1 Recurrent GI disturbance discomfort or nausea/vom
- Cyc vom (1.6.1.1) and Abd mig (1.6.1.2) – both include pain and nauseas; differences:
 - CV age 3-5 and inv vom 4x/h up to 10 days
 - AM age 7-10, and inv less vomiting, up to 3 d
- 1.6.3 Benign Paroxysmal Vertigo (not BPPV) – brief unprovoked attacks of vertigo, may be accomp by N/V (r/u sz, mass)
- 1.6.4 Benign Paroxysmal Torticollis – attacks of head tilt in infant or young child

Tension type headache

- Tension HA
- Muscle contraction headache
- Tension-type headache



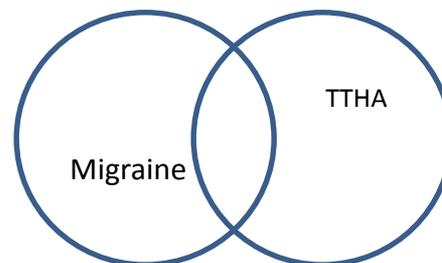
2.1 Infrequent episodic TTH

- A. At least 10 episodes of headache occurring on <1 d/mo (<12 d/y) and fulfilling criteria B-D
- B. Lasting from 30 min to 7 d
- C. ≥ 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
- D. Both of the following:
 1. no nausea or vomiting
 2. no more than one of photophobia or phonophobia
- E. Not better accounted for by another ICHD-3 diagnosis

A2. – TTH

- A. Episodes of headache fulfilling one of the tension type headache criteria
- B. Lasting from 30 min to 7 d
- C. ≥ 3 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
- D. No nausea or vomiting; No photophobia or phonophobia
- E. Not better accounted for by another ICHD-3 diagnosis

– more specific, less likely to diagnose mild migraine



2.2 Frequent episodic TTH

- A. At least 10 episodes occurring on **1-14 d/mo** for >3 mo (≥ 12 and < 180 d/y) and fulfilling criteria B-D
- B. Lasting from 30 min to 7 d
- C. ≥ 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
- D. Both of the following:
 1. no nausea or vomiting
 2. no more than one of photophobia or phonophobia
- E. Not better accounted for by another ICHD-3 diagnosis

2.3 Chronic TTH

- A. Headache occurring on ≥ 15 d/mo on average for >3 mo (≥ 180 d/y), fulfilling criteria B-D
- B. Lasting hours to days, or unremitting
- C. ≥ 2 of the following 4 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, mild nausea
 2. neither moderate or severe nausea nor vomiting
- E. Not better accounted for by another ICHD-3 diagnosis

TTHA still subdivided by presence/absence of muscle tenderness

- Infrequent (<1/mo), Freq (1-14 d/mo), CTTH (>15 d/mo)
- Each category subdivided – with or w/o pericranial tenderness.
- And still no clear evidence for etiopathology

3. Trigeminal autonomic cephalalgias (TACs)

3.1 Cluster headache

3.2 Paroxysmal hemicrania

3.3 Short-lasting unilateral neuralgiform headache.

3.4 Hemicrania continua

All are unilateral, supposedly
accompanied by restlessness and
accompanied by cranial autonomic sx.

But are they related?

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** (when untreated)

C. **Either or both** of the following:

1. ≥ 1 of the following ipsilateral symptoms or signs:

- a) conjunctival injection and/or lacrimation; b) nasal congestion and/or rhinorrhoea; c) eyelid oedema; d) forehead and facial sweating; e) forehead and facial flushing;
- f) sensation of fullness in the ear; g) miosis and/or ptosis

2. a sense of **restlessness or agitation**

D. **Frequency from 1/2 d to 8/d for > half the time when active**

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

3.1 Cluster headache

3.1.1 **Episodic** cluster headache

- A. Attacks fulfilling criteria for 3.1 *Cluster headache* and occurring in bouts (cluster periods)
- B. ≥ 2 cluster periods lasting 7 d to 1 y (when untreated) and separated by pain-free remission periods of ≥ 1 mo

3.1.2 **Chronic** cluster headache

- A. Attacks fulfilling criteria for 3.1 *Cluster headache* and criterion B below
- B. Occurring without a remission period, or with remissions lasting < 1 mo, for ≥ 1 y

3.2 Paroxysmal hemicrania

- A. At least 20 attacks fulfilling criteria B-E
- B. Severe unilateral orbital, supraorbital and/or temporal pain lasting **2-30 min**
- C. ≥ 1 of the following ipsilateral symptoms or signs:
 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
- D. Frequency **>5/d** for $>$ half the time
- E. Prevented absolutely by therapeutic doses of **indomethacin**
- F. Not better accounted for by another ICHD-3 diagnosis

3.2 Paroxysmal hemicrania

3.2.1 **Episodic** paroxysmal hemicrania

- A. Attacks fulfilling criteria for 3.2 *Paroxysmal hemicrania* and occurring in bouts
- B. ≥ 2 bouts lasting 7d to 1 y (when untreated) and separated by pain-free remission periods of ≥ 1 mo

3.2.2 **Chronic** paroxysmal hemicrania

- A. Attacks fulfilling criteria for 3.2 *Paroxysmal hemicrania*
- B. Occurring without a remission period, or with remission periods lasting < 1 mo, for ≥ 1 y

3.3 Short-lasting unilateral neuralgiform headache attacks (SUN)

- A. At least 20 attacks fulfilling criteria B-D
- B. Moderate or severe unilateral head pain, with orbital, supraorbital, temporal and/or other trigeminal distribution, lasting **1-600 s and occurring as single stabs, series of stabs or in a saw-tooth pattern**
- C. ≥ 1 of the following ipsilateral cranial autonomic symptoms or signs: **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**
- D. Frequency $\geq 1/d$ for $>$ half the time when active
- E. Not better accounted for by another ICHD-3 diagnosis

3.3 Short-lasting unilateral neuralgiform headache attacks

3.3.1 Short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing (SUNCT)

3.3.2 Short lasting unilateral neuralgiform headache attacks with cranial autonomic symptoms, but with conjunctival injection or tearing or neither (SUNA)

Either can be episodic or chronic

Episodic - 7d to 1 y and separated by pain-free remission periods of ≥ 1 mo

Chronic - Occurring without a remission period, or with remissions lasting < 1 mo, and continuing for ≥ 1 y

3.4 Hemicrania continua

A. Unilateral headache fulfilling criteria B-D

B. **Present >3 mo**, with exacerbations of moderate or greater intensity

C. Either or both of the following:

1. ≥ 1 of the following ipsilateral symptoms or signs:

a) conjunctival injection and/or lacrimation; b) nasal congestion and/or rhinorrhoea; c) eyelid oedema; d) fore-head and facial sweating; e) forehead and facial flushing; f) sensation of fullness in the ear; g) miosis and/or ptosis

2. a sense of restlessness or agitation, or aggravation of pain by movement

D. Responds absolutely to therapeutic doses of **indomethacin**

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

3.4 Hemicrania continua

3.4.1 Hemicrania continua, remitting subtype

- A. Headache fulfilling criteria for 3.4 *Hemicrania continua*, and criterion B below
- B. Headache is not daily or continuous, but interrupted by remission periods of ≥ 1 d without treatment

3.4.2 Hemicrania continua, unremitting subtype

- A. Headache fulfilling criteria for 3.4 *Hemicrania continua*, and criterion B below
- B. Headache is daily and continuous for ≥ 1 y, without remission periods of ≥ 1 d

TAC's:

- Duration decreases with name length

HC

Months-Yrs

Cluster

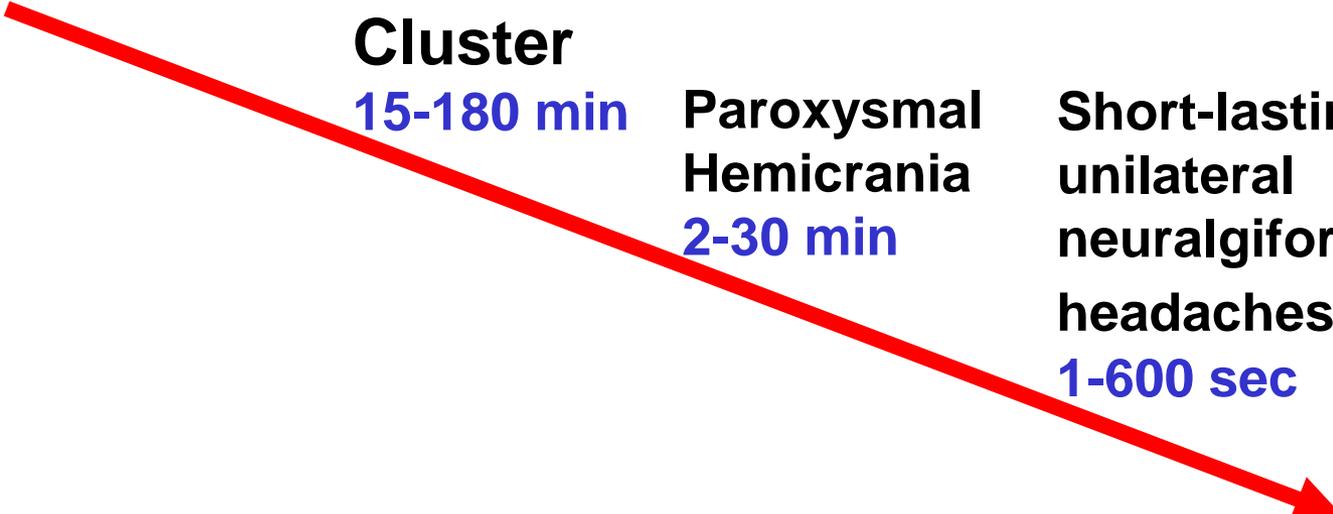
15-180 min

**Paroxysmal
Hemicrania**

2-30 min

**Short-lasting
unilateral
neuralgiform
headaches**

1-600 sec



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)

4. Other primary headache disorders

4.1 Primary cough headache – sudden with cough or Valsalva – 1 sec-2 h

4.2 Primary exercise headache – during or after sustained exercise <48 h

4.3 Primary headache associated with sexual activity - Either or both :

1. increasing in intensity with increasing sexual excitement

2. abrupt explosive intensity just before or with orgasm

4.4 Primary thunderclap headache

Diagnostic criteria:

- A. Severe head pain
- B. Abrupt onset, reaching maximum intensity in <1 minute
- C. Lasting for ≥ 5 minutes
- D. Not better accounted for by another ICHD-3 diagnosis

4.4 Primary thunderclap headache

Ddx:

Subarachnoid hemorrhage
Intracerebral hemorrhage
Pituitary Apoplexy
3rd ventric colloid cyst
Cerebral Venous Thrombosis
Arterial Dissection
CNS Vasculitis, RCVS
Intracranial hypotension
Primary Thunderclap Headache
Sex related Headache
Meningitis
Acute Sinusitis
Unruptured AVM, aneurysm

ICHD 3: “Evidence that Thunderclap headache exists as a primary condition is poor; the search for an underlying cause should be expedited and expected”



4. Other primary headache disorders

- 4.5 Cold-stimulus HA – ingest or inhale, resolves < 30 min
- 4.6 External pressure HA – pressure (hat) or traction (pony-tail) – resolves in 1 h
- 4.7 Primary stabbing HA – jabs and jolts 10sec-2 min
- 4.8 Nummular HA – 1-6 cm, usually parietal, local tenderness, allodynia
- 4.9 Hypnic HA – during sleep, USUALLY over age 50, lasting 15 min-4 h, no autonomic sx

4. Other primary headache disorders

- 4.5 Cold-stimulus HA – ingest or inhale, resolves < 30 min
- 4.6 External pressure HA – pressure (hat) or traction (pony-tail) – resolves in 1 h
- 4.7 Primary stabbing HA – jabs and jolts 10sec-2 min
- 4.8 Nummular HA – 1-6 cm, usually parietal, local tenderness, allodynia
- 4.9 Hypnic HA – during sleep, USUALLY over age 50, lasting 15 min-4 h, no autonomic sx

ICHD III 4.10 **New Daily Persistent Headache**

- A. Persistent headache
- 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.

Ddx – exacerbation of preexisting Primary HA, Hemicrania continua, medication overuse headache

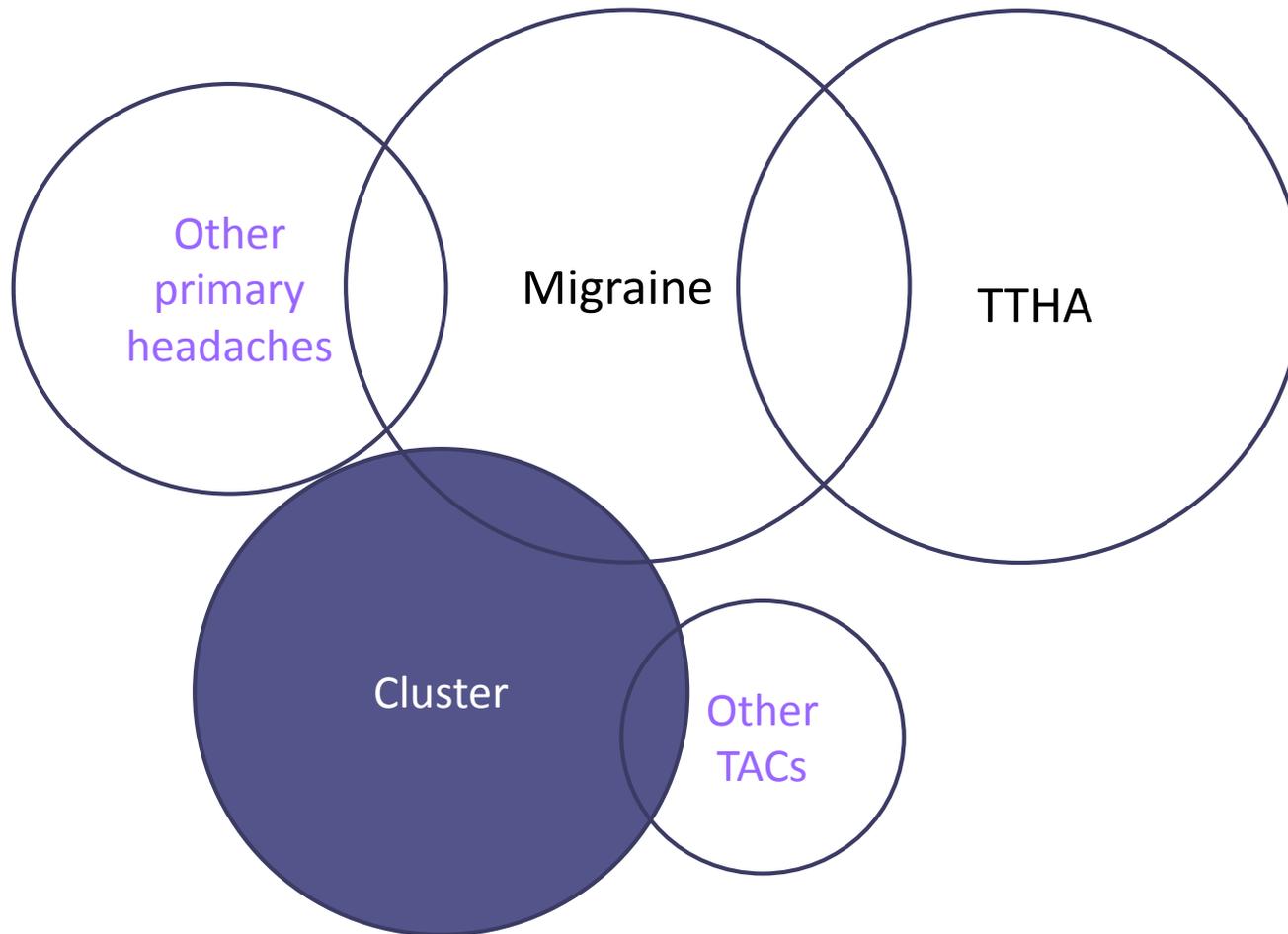
New Daily Persistent Headache

- *Description:* A daily or very frequent headache of abrupt onset. The **pain and associated features may resemble chronic migraine or chronic tension-type headache, or have elements of both.**
- A discrete entity? A mixed group?
- Is there evidence for the “self limiting subtype” and the “refractory subtype”

Lingering classification questions in migraine and primary headaches

- Is motor aura critically different from other auras
- How to diagnose/classify auras which persist beyond an hour but do not persist beyond 1 week (persistent aura 1.4.2)
- And what is the basis for long term persistent aura without stroke?
- Chapter 3 – similarities v. differences
- Is the SUNCT / SUNA distinction relevant?
- NDPH

Many primary headaches can be seen as overlapping sets?



ICHD III

Part 2: Secondary headaches *(caused 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

ICHD – Challenges with Secondary Headaches

- Morphology of HA is not necessarily diagnostic
- Location of pain is not necessarily diagnostic
- Presence of pathology is not necessarily diagnostic
- Multiple sources of pain coexist in many patients
 - relative contributions?
- Pain may not resolve when cause is gone

Lingering questions in 2° Headaches

- Post-traumatic headache require emergence within 7 days
- IIH – retained requirement for OP>250 (“...280 in obese children)
- CSF leak or hypovolemic headaches – retained requirement for OP<60 (not included – positionality)
- Are day limit differences meaningful in MOH? Are there interpatient differences? Are only migraineurs prone?
- What constitutes acceptable causes (and treatment targets) for cervicogenic headache?
- Are there persistent HA’s following resolution of viral meningitis or encephalitis (Not classified in ICHD3)
- Does HIV cause HA? (Not classified in ICHD3)

5.1 Persistent headache attributed to traumatic injury to the head

- A. Any headache fulfilling criteria C and D
- B. Traumatic injury to the head has occurred
- C. Headache is reported to have developed within 7 d after one of the following:
 1. the injury to the head
 2. regaining of consciousness following the injury
 3. discontinuation of medication(s) that impair ability to sense or report headache following the injury
- D. Headache persists for **>3 mo** after injury to the head
- E. Not better accounted for by another ICHD-3 diagnosis

5.3 Persistent headache attributed to whiplash

- A. Any headache fulfilling criteria C and D
- B. Whiplash, associated at the time with neck pain and/or headache, has occurred
- C. Headache has developed within 7 d after whiplash
- D. Headache persists for **>3** mo after whiplash
- E. Not better accounted for by another ICHD-3 diagnosis

Idiopathic Intracranial Hypertension

A. New headache, or a significant worsening of a preexisting headache, fulfilling criterion C

B. Both:

1. IIH has been diagnosed

2. CSF pressure > 250 mm (>280 mm in obese children)

C. Either or both of the following:

1. headache has developed or significantly worsened in temporal relation to the intracranial hypertension, or led to its discovery

2. headache is accompanied by either or both of the following: a) pulsatile tinnitus b) papilloedema

Intracranial Hypotension HA

- A. Any headache fulfilling criterion C
- 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 led to its discovery
- D. Not better accounted for by another ICHD-III diagnosis.

7.2.1 – post LP

7.2.2 – CSF fistula

7.2.3 - SIH

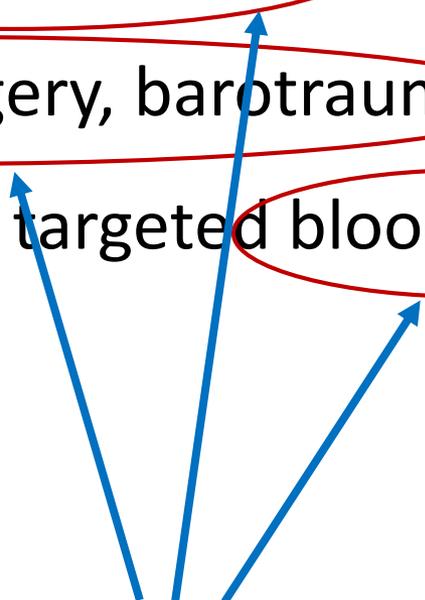
Intracranial Hypotension

HA MUCH worse upon arising

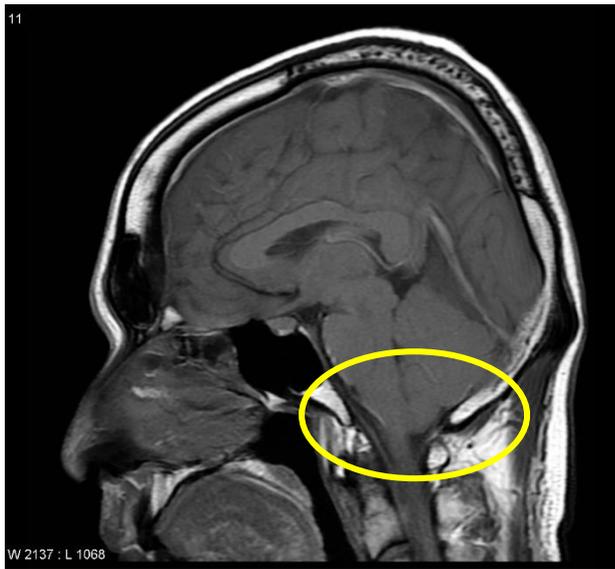
Antecedent LP, surgery, barotrauma

Goal – Responds to targeted blood patch

Not part of the definition



Imaging clues to SIH *also not in dx criteria*



Brain sag

Subdural
collections



Dural
enhancement

Drugs, substances and headaches

- Exposure to – NO donors, CO, Ethanol, Cocaine, Histamine, CGRP, pressor
- Overuse of - Ergots, Triptans, analgesics, opioids, combination meds
- Withdrawal from - caffeine, opioid, estrogen

8.2 Medication-overuse headache (MOH)

- A. Headache occurring on ≥ 15 d/mo in a patient with a pre-existing headache disorder
- B. Regular overuse for > 3 mo of one or more drugs that can be taken for acute and/or symptomatic treatment of headache
- C. Not better accounted for by another ICHD-3 diagnosis

8.2 Medication-overuse headache (MOH)

- 8.2.1 Ergotamine-overuse headache (10 d/mo use)
- 8.2.2 Triptan-overuse headache (10d/mo)
- 8.2.3 Simple analgesic-overuse headache (15d/mo)
- 8.2.4 Opioid-overuse headache (10d/mo)
- 8.2.5 Combination -analgesic-overuse headache (10d/mo)
- 8.2.6 MOH attributed to multiple drug classes not individually overused (10d/mo)
- 8.2.7 MOH headache attributed to unverified overuse of multiple drug classes (10d/mo)
- 8.2.8 MOH attributed to other medication (10d/mo)

Distinguishing CM, CTTH & MOH

- Still difficult, and using ICHD3 one could diagnose all 3 in one patient very easily –
- e.g. 23 headache per month – 8 mig, 7 thought by patient to be mig, and 8 tension type
- If patient is overusing analgesic/abortive med and fulfills CM – supposed to diagnose both and judge after med overuse has resolved

9. Infections and Headaches

- Headaches can be induced by many acute and chronic infections –
 - If a chronic infection: *Chronic HA*, e.g. “Chronic headache attrib to intracranial parasitic infection”
 - If infec has resolved but HA persists: *Persistent HA*, e.g. “Persistent HA attrib to past meningoencephalitis”
- But – no category for HA due to HIV, or persistent HA after Mono infection

Homeostasis and Headaches

10.1 Headache attributed to hypoxia and/or hypercapnia

10.1.1 High-altitude headache

10.1.2 Headache attributed to aeroplane travel

10.1.3 Diving headache

10.1.4 Sleep apnoea headache

10.2 Dialysis headache

10.3 Headache attributed to arterial hypertension

10.3.1 Headache attributed to phaeochromocytoma

10.3.2 Headache attributed to hypertensive crisis w/o hypertensive encephalopathy

10.3.3 Headache attributed to hypertensive encephalopathy

10.3.4 Headache attributed to pre-eclampsia or eclampsia

10.3.5 Headache attributed to autonomic dysreflexia

10.4 Headache attributed to hypothyroidism

10.5 Headache attributed to fasting

10.6 Cardiac cephalgia

10.6 Cardiac Cephalgia

– **classification saves lives!!!**

- Acute myocardial ischaemia has been demonstrated
- Evidence of causation -- at least 2 of the following:
 1. headache has developed in temporal relation to the onset of acute myocardial ischaemia
 2. HA worsens with ischemia and/or improves with resolution of ischemia
 3. headache has at least two of the following four characteristics:
 - a) moderate to severe intensity
 - b) accompanied by nausea
 - c) not accompanied by photophobia or phonophobia
 - d) aggravated by exertion
 4. headache is relieved by nitroglycerine or derivatives of it

11.3 Cervicogenic Headache

- Clinical and/or imaging evidence of a disorder or lesion within the cervical spine or soft tissues of the neck, known to be able to cause headache
- Evidence of causation -- at least two of the following:
 1. headache has developed in temporal relation to the onset of the cervical disorder or appearance of the lesion
 2. headache has significantly improved or resolved in parallel with improvement in or resolution of the cervical disorder or lesion
 3. cervical range of motion is reduced and headache is made significantly worse by provocative manoeuvres
 4. headache is abolished following diagnostic blockade of a cervical structure or its nerve supply

Cervicogenic Headache

Unaccepted causes in ICHD3 –

- Upper cervical radiculopathy [A11.2.4](#)
- Myofascial pain [A11.2.5](#)

Psychiatric causes of Headache

12.1 – Somatization

12.2 - Psychosis

12A12. Headache attributed to psychiatric disorder

A12.3 Headache attributed to depressive disorder

A12.4 Headache attributed to separation anxiety disorder

A12.5 Headache attributed to panic disorder

A12.6 Headache attributed to specific phobia

A12.7 Headache attributed to social anxiety disorder (social phobia)

A12.8 Headache attributed to generalized anxiety disorder

A12.9 Headache attributed to post-traumatic stress disorder (PTSD)

Part 3 – Painful cranial neuropathies and other facial pain

Trigeminal nerve pain is organized differently and is the model for other neuropathies like GN and nervus intermedius neuralgia

[13.1.1 Trigeminal Neuropathy](#)

[13.1.2 Painful trigeminal neuropathy](#)

Part 3 – Painful cranial neuropathies and other facial pain - 13.1 Trigeminal Pain

13.1.1 TN - unilateral, lancinating (though may have some aching too)

13.1.1.1 Classical TN (due to vasc compression)

13.1.1.2 Secondary - e.g. mass, MS

13.1.1.3 Idiopathic (no vasc comp or lesion found)

13.1.2 Painful trigeminal neuropathy - continuous, burning and or pins and needles, sensation change or allodynia or both

13.1.2.1 Zoster

13.1.2.2 PHN

13.1.2.3 Traumatic

13.1.2.4 other

13.1.2.5 idiopathic

Part 3 – Painful cranial neuropathies etc

13.2 Glossopharyngeal neuralgia, neuropathy

13.3 Nervus intermedius neuralgia, neuropathy

13.4 Occipital neuralgia

13.5 Neck-tongue syndrome

13.6 Painful optic neuritis

13.7 Ischemic ocular motor nerve

13.8 Tolosa–Hunt syndrome

13.9 Paratrigeminal oculosympathetic (Raeder's) syndrome

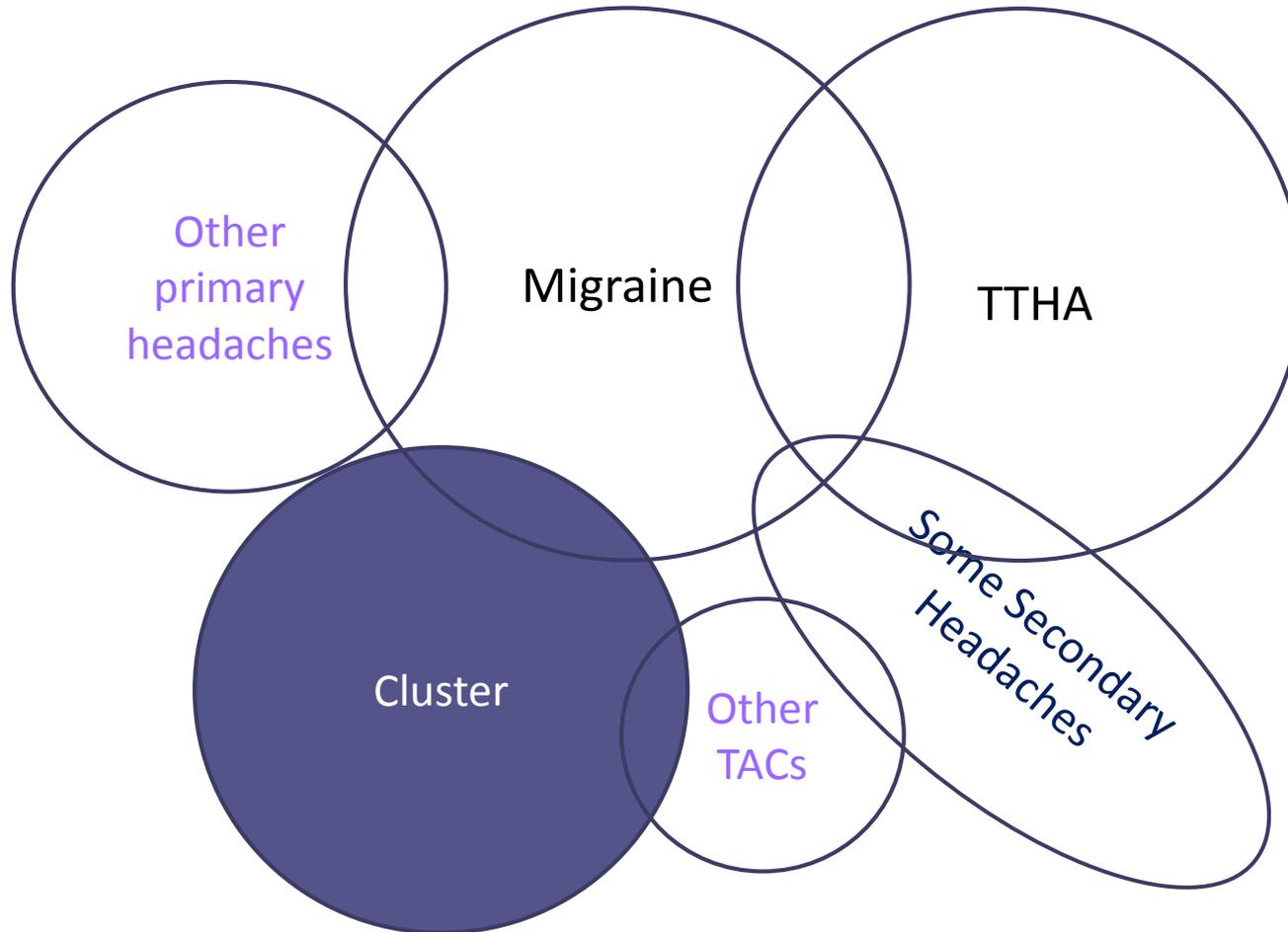
13.10 Recurrent painful ophthalmoplegic neuropathy

13.11 Burning mouth syndrome (BMS)

13.12 Persistent idiopathic facial pain (PIFP)

13.13 Central neuropathic pain

Can some primary and secondary headaches can be seen as overlapping sets?



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*
- *A 4.11 Epicrania fugax – zig-zag HA*
- *A5.1, 5.2 Delayed PTH*
- *A7.11 Post-electroconvulsive therapy (ECT) headache*
- *A7.12 Persistent post-intracranial disorder headache*
- *A 7.13 Ictal Epileptic Headache*

What's New in the Appendix?

- *A9.3 Headache attributed to HIV infection*
- *10.1.2 Headache attributed to aeroplane travel*
- *A10.9 Headache attributed to travel to space*
- *A10.10 Coat hanger pain attributed to orthostatic (postural) hypotension*
- *A11.2.4 Headache attributed to upper cervical radiculopathy*
- *A11.2.5 Headache attributed to myofascial pain in the cervical muscles*
- *A11.5.3 Headache attributed to pathology of the nasal mucosa, turbinates or septum*