



**INTERNATIONAL CLASSIFICATION**  
**of**  
**HEADACHE DISORDERS**  
*2nd edition (1<sup>st</sup> revision)*  
**(ICHD-II)**

*Cephalalgia* 2004; **24** Suppl 1: 1-160);  
*Cephalalgia* 2005; **25**: 460-465

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# History

- 1st edition published as:

**Classification and diagnostic criteria for headache disorders, cranial neuralgias and facial pain.**

***Cephalalgia* 1988; 8 (Suppl 7): 1-96**

*Cephalalgia* 2004; **24** Suppl 1: 1-160);  
*Cephalalgia* 2005; **25**: 460-465

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# History

Revision anticipated after 5 years, but:

- relatively little criticism to prompt revision
- nosographic research appeared only slowly
- world-wide dissemination and translation into >20 languages took longer than expected
- 2nd edition became due after >10 years' accumulation of epidemiological and nosographic knowledge
- Revision process begun in late 1999, completed 2003
- Further minor revision to section 8.2 in 2005

# Basis

- Single classification for all purposes
- Comprehensive
- Evidence-based as far as possible
- Symptom-based for the primary headaches, aetiological for the secondary headaches
- Unambiguous
  - terms such as *sometimes, often, usually* are avoided
- Specificity weighted over sensitivity
- Separate codes for *probable* cases

# System

## Hierarchical (from 1st edition)

- major groups (1st digit)
  - types (2nd digit)
    - subtypes (3rd digit)
      - » subforms (4th digit)

## Phenomenological

- each headache present in a patient (within the last year) separately coded

# Important general rules

1. Each distinct type of headache that a patient has must be separately diagnosed and coded
  - *eg*, a severely affected patient may receive three diagnoses and codes:
    - 1.1 *Migraine without aura*,
    - 2.2 *Frequent episodic tension-type headache* and
    - 8.2 *Medication-overuse headache*

# Important general rules

2. When a patient receives more than one diagnosis these should be listed in the order of importance to the patient

# Important general rules

3. If one headache in a patient fulfils two different sets of explicit diagnostic criteria, use all other available information to decide which diagnosis is correct or more likely
  - this could include the longitudinal headache history (how did the headache start?), the family history, the effect of drugs, menstrual relationship, age, gender *etc*



# Important general rules

4. For any particular diagnosis to be given, *all* listed criteria must be fulfilled
  - *probable* diagnostic categories exist for many disorders, to be used when a single criterion is not fulfilled

# Important general rules

5. Fulfilment of explicit criteria for
  1. *Migraine*,
  2. *Tension-type headache* or
  3. *Cluster headache and other TACs*,or any of their subtypes, trumps the *probable* diagnostic categories of each
  - *eg*, a patient whose headache fulfils criteria for both 1.6 *Probable migraine* and 2.1 *Infrequent episodic tension-type headache* should be coded to the latter

# Important general rules

6. Always consider the possibility that some headache attacks in a patient meet one set of criteria whilst other attacks meet another set
  - in such cases, two diagnoses exist and both should be coded

# Important general rules

7. When a patient is suspected of having more than one headache type, a diagnostic headache diary recording the important characteristics for each headache episode
  - improves diagnostic accuracy
  - allows judgement of medication consumption
  - establishes the quantities of each of two or more different headache types or subtypes
  - teaches the patient to distinguish between different headaches

# Structure

One chapter (1-13) per major group:

- introduction
- headache types, subtypes, subforms with:
  - previously used terms
  - disorders that are related but coded elsewhere
  - short descriptions
  - **explicit diagnostic criteria**
  - notes and comments
- selected bibliography

# Structure

Final chapter (14) for:

- headache not elsewhere classified
  - headache entities still to be described
- headache unspecified
  - headaches known to be present but insufficiently described

# Structure

## Appendix for:

- research criteria for novel entities that have not been sufficiently validated
- alternative diagnostic criteria that may be preferable but for which the evidence is insufficient
- a first step in eliminating disorders included in the 1st edition for which sufficient evidence has still not been published

# Classification

## Part 1:

Primary headache disorders

## Part 2:

Secondary headache disorders

## Part 3:

Cranial neuralgias, central and primary facial pain and other headaches



# Primary or secondary headache?

## Primary:

- no other causative disorder

# Primary or secondary headache?

## Secondary

(ie, caused by another disorder):

- new headache occurring in close temporal relation to another disorder that is a known cause of headache
- coded as *attributed to* that disorder (in place of previously used term *associated with*)

# Classification

## Part 1: The primary headaches

1. Migraine
2. Tension-type headache
3. Cluster headache  
and other trigeminal autonomic cephalalgias
4. Other primary headaches

# Classification

## Part 2: The secondary headaches

5. Headache attributed to head and/or neck trauma
6. Headache attributed to cranial or cervical vascular disorder
7. Headache attributed to non-vascular intracranial disorder
8. Headache attributed to a substance or its withdrawal
9. Headache attributed to infection

# Classification

## Part 2: The secondary headaches

10. Headache attributed to disorder of homoeostasis
11. Headache or facial pain attributed to disorder of cranium, neck, eyes, ears, nose, sinuses, teeth, mouth or other facial or cranial structures
12. Headache attributed to psychiatric disorder

# Classification

## Part 3: Cranial neuralgias, central and primary facial pain and other headaches

13. Cranial neuralgias and central causes of facial pain
14. Other headache, cranial neuralgia, central or primary facial pain

# Part 1: The primary headaches

1. Migraine
2. Tension-type headache
3. Cluster headache  
and other trigeminal autonomic cephalalgias
4. Other primary headaches

# 1. Migraine

- 1.1 Migraine without aura
- 1.2 Migraine with aura
- 1.3 Childhood periodic syndromes that are commonly precursors of migraine
- 1.4 Retinal migraine
- 1.5 Complications of migraine
- 1.6 Probable migraine



# 1. Migraine

## ***Reclassification 1988-2004***

### ***1988***

- 1.1 Migraine without aura
- 1.2 Migraine with aura
- 1.3 Ophthalmoplegic migraine
- 1.4 Retinal migraine
- 1.5 Childhood periodic syndromes
- 1.6 Complications of migraine
- 1.7 Migrainous disorder

### ***2004***

- 1.1 Migraine without aura
- 1.2 Migraine with aura
- 13.17 Ophthalmoplegic 'migraine'
- 1.4 Retinal migraine
- 1.3 Childhood periodic syndromes
- 1.5 Complications of migraine
- 1.6 Probable migraine

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# 1.1 Migraine without aura

- A. At least 5 attacks fulfilling criteria B-D
- B. Headache attacks lasting 4-72 h (untreated or unsuccessfully treated)
- C. Headache has  $\geq 2$  of the following characteristics:
  1. unilateral location
  2. pulsating quality
  3. moderate or severe pain intensity
  4. aggravation by or causing avoidance of routine physical activity (*eg*, walking, climbing stairs)
- D. During headache  $\geq 1$  of the following:
  1. nausea and/or vomiting
  2. photophobia and phonophobia
- E. Not attributed to another disorder

# 1.1 Migraine without aura

## Notes

- If <5 attacks but criteria B-E otherwise met, code as 1.6.1 *Probable migraine without aura*
- When attacks occur on  $\geq 15$  d/mo for >3 mo, code as 1.1 *Migraine without aura* + 1.5.1 *Chronic migraine*
- *Pulsating* means varying with the heartbeat
- In children:
  - attacks may last 1-72 h
  - occipital headache requires caution
- In young children:
  - photophobia and/or phonophobia may be inferred from their behaviour

# 'Not attributed to another disorder'

## **Note**

For all primary headaches, this criterion means:

- History and physical/neurological examinations do not suggest any of the disorders listed in groups 5-12, or history and/or physical/ neurological examinations do suggest such disorder but it is ruled out by appropriate investigations, or such disorder is present but headache does not occur for the first time in close temporal relation to the disorder

## 1.2 Migraine with aura

- 1.2.1 Typical aura with migraine headache
- 1.2.2 Typical aura with non-migraine headache
- 1.2.3 Typical aura without headache
- 1.2.4 Familial hemiplegic migraine (FHM)
- 1.2.5 Sporadic hemiplegic migraine
- 1.2.6 Basilar-type migraine

## 1.2 Migraine with aura

- A. At least 2 attacks fulfilling criterion B
- B. Migraine aura fulfilling criteria B and C for one of the subforms 1.2.1-1.2.6
- C. Not attributed to another disorder

# **1.2 Migraine with aura**

## ***Subtypes new to classification***

### **1.2.1 Typical aura with migraine headache**

- most migraine auras are associated with headache fulfilling criteria for 1.1 *Migraine without aura*

### **1.2.2 Typical aura with non-migraine headache**

### **1.2.3 Typical aura without headache**

- migraine aura is sometimes associated with a headache that does not fulfil these criteria
- or occurs without headache

## 1.2.1 Typical aura with migraine headache

- A. At least 2 attacks fulfilling criteria B–D
- B. Aura consisting of  $\geq 1$  of the following, but no motor weakness:
  1. fully reversible visual symptoms including positive and/or negative features
  2. fully reversible sensory symptoms including positive and/or negative features
  3. fully reversible dysphasic speech disturbance



## 1.2.1 Typical aura with migraine headache

C. At least two of the following:

1. homonymous visual symptoms and/or unilateral sensory symptoms
2. at least one aura symptom develops gradually over  $\geq 5$  min and/or different aura symptoms occur in succession over  $\geq 5$  min
3. each symptom lasts  $\geq 5$  and  $\leq 60$  min

D. Headache fulfilling criteria B-D for 1.1 *Migraine without aura* begins during the aura or follows aura within 60 min

E. Not attributed to another disorder

## 1.2.2 Typical aura with non-migraine headache

As 1.2.1 *except*:

D. Headache that does not fulfil criteria B-D for  
1.1 *Migraine without aura* begins during the aura or  
follows aura within 60 min

## 1.2.3 Typical aura without headache

*As 1.2.1 except:*

D. Headache does not occur during aura nor follow aura  
within 60 min

## 1.2.4 Familial hemiplegic migraine (FHM)

- A. At least 2 attacks fulfilling criteria B and C
- B. Aura consisting of fully reversible motor weakness and  $\geq 1$  of:
  1. fully reversible visual symptoms including positive and/or negative features
  2. fully reversible sensory symptoms including positive and/or negative features
  3. fully reversible dysphasic speech disturbance

## 1.2.4 Familial hemiplegic migraine (FHM)

C. At least two of the following:

1. at least one aura symptom develops gradually over  $\geq 5$  min and/or different aura symptoms occur in succession over  $\geq 5$  min
2. each aura symptom lasts  $\geq 5$  min and  $< 24$  h
3. headache fulfilling criteria B-D for 1.1 *Migraine without aura* begins during the aura or follows onset of aura within 60 min

D. At least one 1st- or 2nd-degree relative fulfils these criteria

E. Not attributed to another disorder

## 1.2.6 Basilar-type migraine

*As 1.2.1 except:*

- B. Aura consisting of  $\geq 2$  of the following fully reversible symptoms, but no motor weakness:
1. dysarthria; 2. vertigo; 3. tinnitus; 4. hypacusia;
  5. diplopia; 6. visual symptoms simultaneously in both temporal and nasal fields of both eyes; 7. ataxia;
  8. decreased level of consciousness;
  9. simultaneously bilateral paraesthesias
- C. At least one of the following:
1. at least one one aura symptom develops gradually over  $\geq 5$  min and/or different aura symptoms occur in succession over  $\geq 5$  min
  2. each aura symptom lasts  $\geq 5$  and  $\leq 60$  min

## 1.2.6 Basilar-type migraine

### *Terminology change 1988-2004*

- 1.2.6 *Basilar-type migraine* was previously classified as 1.2.4 *Basilar migraine*
- Terminology has been changed because there is little evidence that the basilar artery or, necessarily, basilar-artery territory is involved

# 1.3 Childhood periodic syndromes that are commonly precursors of migraine

- 1.3.1 Cyclical vomiting
- 1.3.2 Abdominal migraine
- 1.3.3 Benign paroxysmal vertigo of childhood



## 1.3.2 Abdominal migraine

- A. At least 5 attacks fulfilling criteria B-D
- B. Attacks of abdominal pain lasting 1-72 h
- C. Abdominal pain has all of the following characteristics:
  - 1. midline location, periumbilical or poorly localised
  - 2. dull or “just sore” quality
  - 3. moderate or severe intensity
- D. During abdominal pain  $\geq 2$  of the following:
  - 1. anorexia; 2. nausea; 3. vomiting; 4. pallor
- E. Not attributed to another disorder

# 1.5 Complications of migraine

1.5.1 Chronic migraine

1.5.2 Status migrainosus

1.5.3 Persistent aura without infarction

1.5.4 Migrainous infarction

1.5.5 Migraine-triggered seizures

# 1.5 Complications of migraine

## *Reclassification 1988-2004*

**1988**

1.6.1 Status migrainosus

1.6.2 Migrainous  
infarction

**2004**

1.5.1 Chronic migraine

1.5.2 Status migrainosus

1.5.3 Persistent aura  
without infarction

1.5.4 Migrainous  
infarction

1.5.5 Migraine triggered  
seizure

## 1.5.1 Chronic migraine

### *New entrant to classification*

- A. Headache fulfilling criteria C and D for  
1.1 *Migraine without aura* on  $\geq 15$  d/mo for  $>3$  mo
- B. Not attributed to another disorder

# 1.5.1 Chronic migraine

## Notes

- When medication overuse is present, this is the most likely cause of chronic symptoms
  - code according to antecedent migraine subtype + 1.6.5 *Probable chronic migraine* + 8.2.8 *Probable MOH*
- Post-withdrawal, code as:
  - 1.5.1 *Chronic migraine* + antecedent migraine subtype if symptoms persist beyond 2 mo
  - 8.2 *Medication-overuse headache* + antecedent migraine subtype if, before 2 mo, improvement occurs and these criteria are no longer fulfilled

# 'Chronic'

## Notes

- In pain terminology, *chronic* denotes persistence over a period of more than 3 months
- In headache terminology, it retains this meaning for secondary headache disorders
- For primary headache disorders that are more usually episodic (*eg*, migraine), *chronic* is used whenever headache occurs on more days than not over more than 3 months
  - the trigeminal autonomic cephalalgias (*qv*) are an exception

# 1.6 Probable migraine

- 1.6.1 Probable migraine without aura
- 1.6.2 Probable migraine with aura
- 1.6.5 Probable chronic migraine

# 1.6 Probable migraine

## 1.6.1 Probable migraine without aura

- A. Attacks fulfilling all but one of criteria A-D for 1.1 *Migraine without aura*
- B. Not attributed to another disorder

## 1.6.2 Probable migraine with aura

- A. Attacks fulfilling all but one of criteria A-D for 1.2 *Migraine with aura*
- B. Not attributed to another disorder



# 1.6 Probable migraine

## 1.6.5 Probable chronic migraine

- A. Headache fulfilling criteria C and D for  
1.1 *Migraine without aura* on  $\geq 15$  d/mo for  $>3$  mo
- B. Not attributed to another disorder but there is, or has been within the last 2 mo, medication overuse fulfilling criterion B for any of the subforms of  
8.2 *Medication-overuse headache*

## 2. Tension-type headache

- 2.1 Infrequent episodic tension-type headache
- 2.2 Frequent episodic tension-type headache
- 2.3 Chronic tension-type headache
- 2.4 Probable tension-type headache

# Infrequent/frequent episodic TTH

## *New subdivision 1988-2004*

### Why this new subdivision?

- Infrequent TTH has very little impact on the individual and does not deserve much attention from the medical profession
- Frequent TTH sufferers can encounter considerable disability that sometimes warrants expensive drugs and prophylactic medication

## 2.1 Infrequent episodic TTH

- A. At least 10 episodes occurring on <1 d/mo (<12 d/y) and fulfilling criteria B-D
- B. Headache lasting from 30 min to 7 d
- C. Headache has  $\geq 2$  of the following 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. no nausea or vomiting (anorexia may occur)
  - 2. no more than one of photophobia or phonophobia
- E. Not attributed to another disorder

## **2.1 Infrequent episodic TTH**

### **2.1.1 Infrequent episodic tension-type headache associated with pericranial tenderness**

- A. Episodes fulfilling criteria A-E for  
*2.1 Infrequent episodic tension-type headache*
- B. Increased pericranial tenderness on manual palpation

### **2.1.2 Infrequent episodic tension-type headache not associated with pericranial tenderness**

- A. Episodes fulfilling criteria A-E for  
*2.1 Infrequent episodic tension-type headache*
- B. No increased pericranial tenderness

## 2.2 Frequent episodic TTH

*As 2.1 except:*

- A. At least 10 episodes occurring on  $\geq 1$  but  $< 15$  d/mo for  $\geq 3$  mo ( $\geq 12$  and  $< 180$  d/y) and fulfilling criteria B-D

## 2.2 Frequent episodic TTH

### 2.2.1 Frequent episodic tension-type headache associated with pericranial tenderness

- A. Episodes fulfilling criteria A-E for  
*2.2 Frequent episodic tension-type headache*
- B. Increased pericranial tenderness on manual palpation

### 2.2.2 Frequent episodic tension-type headache not associated with pericranial tenderness

- A. Episodes fulfilling criteria A-E for  
*2.2 Frequent episodic tension-type headache*
- B. No increased pericranial tenderness

## 2.3 Chronic TTH

- A. Headache occurring on  $\geq 15$  d/mo ( $\geq 180$  d/y) for  $> 3$  mo and fulfilling criteria B-D
- B. Headache lasts hours or may be continuous
- C. Headache has  $\geq 2$  of the following 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 attributed to another disorder



## 2.3 Chronic TTH

### 2.3.1 Chronic tension-type headache associated with pericranial tenderness

- A. Headache fulfilling criteria A-E for  
2.3 *Chronic tension-type headache*
- B. Increased pericranial tenderness on manual palpation

### 2.3.2 Chronic tension-type headache not associated with pericranial tenderness

- A. Episodes fulfilling criteria A-E for  
2.3 *Chronic tension-type headache*
- B. No increased pericranial tenderness

## 2.4 Probable TTH

### 2.4.1 Probable infrequent episodic TTH

- A. Episodes fulfilling all but one of criteria A-D for  
2.1 *Infrequent episodic tension-type headache*
- B. Episodes do not fulfil criteria for  
1.1 *Migraine without aura*
- C. Not attributed to another disorder

### 2.4.2 Probable frequent episodic TTH

- A. Episodes fulfilling all but one of criteria A-D for  
2.2 *Frequent episodic tension-type headache*
- B. Episodes do not fulfil criteria for  
1.1 *Migraine without aura*
- C. Not attributed to another disorder

## 2.4.3 Probable chronic TTH

*As 2.3 except:*

E. Not attributed to another disorder but there is, or has been within the last 2 mo, medication overuse fulfilling criterion B for any of the subforms of 8.2 *Medication-overuse headache*

# 3. Cluster headache and other trigeminal autonomic cephalalgias

3.1 Cluster headache

3.2 Paroxysmal hemicrania

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

3.4 Probable trigeminal autonomic cephalalgia

## 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  $\geq 1$  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

# 3.1 Cluster headache

## 3.1.1 Episodic cluster headache

- A. Attacks fulfilling criteria A-E for 3.1 *Cluster headache*
- B. At least two cluster periods lasting 7-365 d and separated by pain-free remission periods of  $\geq 1$  mo

## 3.1.2 Chronic cluster headache

- A. Attacks fulfilling criteria A-E for 3.1 *Cluster headache*
- B. Attacks recur over  $>1$  y without remission periods or with remission periods lasting  $<1$  mo

# 'Chronic'

## **Notes**

- In pain terminology, *chronic* denotes persistence over a period of more than 3 months
- For primary headache disorders that are more usually episodic, *chronic* is used whenever headache occurs on more days than not over more than 3 months
- **The trigeminal autonomic cephalalgias are an exception:**
  - **in these disorders, *chronic* is not used until the condition has been unremitting for more than 1 year**

# Episodic/chronic cluster headache

## *Reclassification 1988-2004*

**1988**

- 3.1.1 Cluster headache  
periodicity  
undetermined
- 3.1.2 Episodic cluster  
headache
- 3.1.3 Chronic cluster  
headache

**2004**

- 3.1.1 Episodic cluster  
headache
- 3.1.2 Chronic cluster  
headache

Default diagnosis until periodicity is determined or 1 y is  
3.1.1 *Episodic cluster headache*



# Episodic/chronic cluster headache

## *Definition change 1988-2004*

- The definition of remission period distinguishing  
3.1.1 *Episodic cluster headache* from  
3.1.2 *Chronic cluster headache*

is changed: duration increased from  
a minimum of 14 days to a minimum of 1 month

## 3.1.2 Chronic cluster headache

### ***Abandoned subclassification 1988-2004***

- Patients may switch from 3.1.2 *Chronic cluster headache* to 3.1.1 *Episodic cluster headache*, and *vice versa*
- Therefore the previously classified subforms  
*Chronic cluster headache unremitting from onset* and  
*Chronic cluster headache evolved from episodic*  
have been dropped

## 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  $\geq 1$  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
- 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

## **3.2 Paroxysmal hemicrania**

### ***New subdivision 1988-2004***

#### **3.2.1 Episodic paroxysmal hemicrania**

- A. Attacks fulfilling criteria A-F for 3.2 *Paroxysmal hemicrania*
- B. At least two attack periods lasting 7-365 d and separated by pain-free remission periods of  $\geq 1$  mo

#### **3.2.2 Chronic paroxysmal hemicrania**

- A. Attacks fulfilling criteria A-F for 3.2 *Paroxysmal hemicrania*
- B. Attacks recur over  $>1$  y without remission periods or with remission periods lasting  $<1$  mo

# Episodic/chronic paroxysmal hemicrania

## *New subdivision 1988-2004*

### Why this new subdivision?

- Only *chronic paroxysmal hemicrania* was previously recognised and classified
- Sufficient clinical evidence for the episodic subtype has accumulated to subdivide paroxysmal hemicranias in a manner analogous to cluster headache

### **3.3 Short-lasting Unilateral Neuralgiform headache attacks with Conjunctival injection and Tearing** ***New entrant to classification***

- A. At least 20 attacks fulfilling criteria B-D
- B. Attacks of unilateral orbital, supraorbital or temporal stabbing or pulsating pain lasting 5-240 s
- C. Pain is accompanied by ipsilateral conjunctival injection and lacrimation
- D. Attacks occur with frequency 3-200/d
- E. Not attributed to another disorder

## 3.4 Probable TAC

### 3.4.1 Probable cluster headache

### 3.4.2 Probable paroxysmal hemicrania

### 3.4.3 Probable SUNCT

- A. Attacks fulfilling all but one of the specific criteria for
  - 3.1 *Cluster headache*,
  - 3.2 *Paroxysmal hemicrania* or
  - 3.3 *SUNCT*
- B. Not attributed to another disorder

# 4. Other primary headaches

- 4.1 Primary stabbing headache
- 4.2 Primary cough headache
- 4.3 Primary exertional headache
- 4.4 Primary headache associated with sexual activity
- 4.5 Hypnic headache
- 4.6 Primary thunderclap headache
- 4.7 Hemicrania continua
- 4.8 New daily-persistent headache (NDPH)



# 4. Other primary headaches

## ***Terminology change 1988-2004***

This section was previously

4. *Miscellaneous headaches unassociated with structural lesion*

# 4.4 Primary headache associated with sexual activity

## 4.4.1 Preorgasmic headache

- A. Dull ache in the head and neck associated with awareness of neck and/or jaw muscle contraction and fulfilling criterion B
- B. Occurs during sexual activity and increases with sexual excitement
- C. Not attributed to another disorder

## 4.4.2 Orgasmic headache

- A. Sudden severe (“explosive”) headache fulfilling criterion B
- B. Occurs at orgasm
- C. Not attributed to another disorder

## 4.5 Hypnic headache

### *New entrant to classification*

- 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 times/mo
  2. lasts  $\geq$ 15 min after waking
  3. first occurs after age of 50 y
- D. No autonomic symptoms and no more than one of nausea, photophobia or phonophobia
- E. Not attributed to another disorder

## 4.6 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

## 4.7 Hemicrania continua

### *New entrant to classification*

- A. Headache for >3 mo fulfilling criteria B-D
- B. All of the following characteristics:
  - 1. unilateral pain without side-shift
  - 2. daily and continuous, without pain-free periods
  - 3. moderate intensity, with exacerbations of severe pain
- C. At least one of the following autonomic features occurs during exacerbations, ipsilateral to the pain:
  - 1. conjunctival injection and/or lacrimation
  - 2. nasal congestion and/or rhinorrhoea
  - 3. ptosis and/or miosis
- D. Complete response to therapeutic doses of indomethacin
- E. Not attributed to another disorder

## 4.8 New daily-persistent headache

### *New entrant to classification*

- 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

# 4.8 New daily-persistent headache

## **Notes**

- 4.8 *New daily-persistent headache* has many similarities to 2.3 *Chronic tension-type headache*
- It is unique in that headache is daily and unremitting from, or almost from, the moment of onset
- A clear recall of such onset is necessary for the diagnosis
- If there is or has been within the last 2 mo medication overuse fulfilling criterion B for any of the subforms of 8.2 *Medication-overuse headache*, the diagnosis cannot be 4.8 *New daily-persistent headache*

## Part 2:

# The secondary headaches

5. Headache attributed to head and/or neck trauma
6. Headache attributed to cranial or cervical vascular disorder
7. Headache attributed to non-vascular intracranial disorder
8. Headache attributed to a substance or its withdrawal
9. Headache attributed to infection
10. Headache attributed to disorder of homeostasis
11. Headache or facial pain attributed to disorder of cranium, neck, eyes, ears, nose, sinuses, teeth, mouth or other facial or cranial structures
12. Headache attributed to psychiatric disorder



# Primary or secondary headache?

## Primary:

- no other causative disorder

## Secondary

(ie, caused by another disorder):

- new headache occurring in close temporal relation to another disorder that is a known cause of headache
- coded as *attributed to* that disorder

# Primary or secondary headache?

*A pre-existing* primary headache made worse in close temporal relation to another disorder:

- judgement required to code
  - *either* as the primary headache only
  - *or* as both the primary headache and a secondary headache (attributed to the other disorder)

# Primary or secondary headache?

<b>Diagnosis:</b>	<b>Primary headache only</b>	<b>Primary + secondary</b>
Temporal relation of other disorder to headache exacerbation	Loose	Close
Degree of exacerbation	Slight	Marked
Other evidence that other disorder can cause secondary headache	Weak	Strong
Other disorder eliminated	Headache unchanged	Headache returns to previous pattern

# Diagnostic criteria for secondary headaches

- A. Headache with one (or more) of the following [listed] characteristics and fulfilling criteria C and D
- B. Another disorder known to be able to cause headache has been demonstrated
- C. Headache occurs in close temporal relation to the other disorder and/or there is other evidence of a causal relationship
- D. Headache is greatly reduced or resolves within 3 mo (shorter for some disorders) after successful treatment or spontaneous remission of the causative disorder

# Important general rules

## 8. The last criterion for most secondary headaches

D. Headache is greatly reduced or resolves within [specified time] after successful treatment or spontaneous remission of the causative disorder

is part of the evidence of causation

Before treatment or spontaneous resolution, criterion D is not fulfilled; code as

*Headache probably attributed to [the disorder]*

# 5. Headache attributed to head and/or neck trauma

- 5.1 Acute post-traumatic headache
- 5.2 Chronic post-traumatic headache
- 5.3 Acute headache attributed to whiplash injury
- 5.4 Chronic headache attributed to whiplash injury
- 5.5 Headache attributed to traumatic intracranial haematoma
- 5.6 Headache attributed to other head and/or neck trauma
- 5.7 Post-craniotomy headache

## 5.1.1 Acute post-traumatic headache attributed to moderate or severe head injury

- A. Headache, no typical characteristics known, fulfilling criteria C and D
- B. Head trauma with at least one of the following:
  1. loss of consciousness for >30 min
  2. Glasgow Coma Scale (GCS) <13
  3. post-traumatic amnesia for >48 h
  4. imaging demonstration of a traumatic brain lesion
- C. Headache develops within 7 d after head trauma or after regaining consciousness following head trauma
- D. One or other of the following:
  1. headache resolves within 3 mo after head trauma
  2. headache persists but 3 mo have not yet passed

# 5.1.1 Acute post-traumatic headache attributed to moderate or severe head injury

## **Notes**

- Criterion D does not relate to evidence of causation
- Causation is established by onset in close temporal relation to trauma, whilst it is well recognised that headache after trauma often persists
- When this occurs, 5.2.1 *Chronic post-traumatic headache attributed to moderate or severe head injury* is diagnosed
- Criterion D2 allows a default diagnosis within 3 mo, before it is known whether headache will resolve or persist



## 5.2.1 Chronic post-traumatic headache attributed to moderate or severe head injury

*As 5.1.1 except:*

D. Headache persists for >3 mo after head trauma

## 5.3 Acute headache attributed to whiplash injury

- A. Headache, no typical characteristics known, fulfilling criteria C and D
- B. History of whiplash (sudden and significant acceleration/deceleration movement of the neck) associated at the time with neck pain
- C. Headache develops within 7 d after whiplash injury
- D. One or other of the following:
  - 1. headache resolves within 3 mo after whiplash injury
  - 2. headache persists but 3 mo have not yet passed since whiplash injury

# 5.3 Acute headache attributed to whiplash injury

## *Notes*

- Criterion D does not relate to evidence of causation
- Causation is established by onset in close temporal relation to whiplash, whilst it is well recognised that headache after whiplash injury may persist
- When this occurs, 5.4 *Chronic headache attributed to whiplash injury* is diagnosed
- Criterion D2 allows a default diagnosis within 3 mo, before it is known whether headache will resolve or persist

## 5.4 Chronic headache attributed to whiplash injury

*As 5.3 except:*

D. Headache persists for >3 mo after whiplash injury

# 6. Headache attributed to cranial or cervical vascular disorder

- 6.1 Headache attributed to ischaemic stroke or transient ischaemic attack
- 6.2 Headache attributed to non-traumatic intracranial haemorrhage
- 6.3 Headache attributed to unruptured vascular malformation
- 6.4 Headache attributed to arteritis
- 6.5 Carotid or vertebral artery pain
- 6.6 Headache attributed to cerebral venous thrombosis
- 6.7 Headache attributed to other intracranial vascular disorder

## 6.2 Headache attributed to non-traumatic intracranial haemorrhage

6.2.1 Headache attributed to intracerebral haemorrhage

6.2.2 Headache attributed to subarachnoid haemorrhage (SAH)

## 6.2.2 Headache attributed to subarachnoid haemorrhage

- A. Severe headache of sudden onset fulfilling criteria C and D
- B. Neuroimaging (CT or MRI T2 or flair) or CSF evidence of non-traumatic subarachnoid haemorrhage with or without other clinical signs
- C. Headache develops simultaneously with haemorrhage
- D. Headache resolves within 1 mo

## 6.3 Headache attributed to unruptured vascular malformation

6.3.1 Headache attributed to saccular aneurysm

6.3.2 Headache attributed to arteriovenous malformation (AVM)

6.3.3 Headache attributed to dural arteriovenous fistula

6.3.4 Headache attributed to cavernous angioma

6.3.5 Headache attributed to encephalotrigeminal or leptomeningeal angiomatosis (Sturge Weber syndrome)



## 6.4 Headache attributed to arteritis

- 6.4.1 Headache attributed to giant cell arteritis (GCA)
- 6.4.2 Headache attributed to primary central nervous system (CNS) angiitis
- 6.4.3 Headache attributed to secondary central nervous system (CNS) angiitis

## 6.4.1 Headache attributed to giant cell arteritis

- A. Any new persisting headache fulfilling criteria C and D
- B. At least one of the following:
  1. swollen tender scalp artery with elevated erythrocyte sedimentation rate (ESR) and/or C reactive protein (CRP)
  2. temporal artery biopsy demonstrating giant cell arteritis
- C. Headache develops in close temporal relation to other symptoms and signs of giant cell arteritis
- D. Headache resolves or greatly improves within 3 d of high-dose steroid treatment

## 6.7 Headache attributed to other intracranial vascular disorder

- 6.7.1 Cerebral Autosomal Dominant Arteriopathy with Subcortical Infarcts and Leukoencephalopathy (CADASIL)
- 6.7.2 Mitochondrial Encephalopathy, Lactic Acidosis and Stroke-like episodes (MELAS)
- 6.7.3 Headache attributed to benign angiopathy of the central nervous system
- 6.7.4 Headache attributed to pituitary apoplexy

## 6.7.1 CADASIL

- A. Attacks of migraine with aura, with or without other neurological signs
- B. Typical white matter changes on MRI T2WI
- C. Diagnostic confirmation from skin biopsy evidence or genetic testing (Notch 3 mutations)

# 7. Headache attributed to non-vascular intracranial disorder

- 7.1 Headache attributed to high cerebrospinal fluid pressure
- 7.2 Headache attributed to low cerebrospinal fluid pressure
- 7.3 Headache attributed to non-infectious inflammatory disease
- 7.4 Headache attributed to intracranial neoplasm
- 7.5 Headache attributed to intrathecal injection
- 7.6 Headache attributed to epileptic seizure
- 7.7 Headache attributed to Chiari malformation type I
- 7.8 Syndrome of transient Headache and Neurological Deficits with cerebrospinal fluid Lymphocytosis (HaNDL)
- 7.9 Headache attributed to other non-vascular intracranial disorder

# 7.1 Headache attributed to high cerebrospinal fluid pressure

7.1.1 Headache attributed to idiopathic intracranial hypertension (IIH)

7.1.2 Headache attributed to intracranial hypertension secondary to metabolic, toxic or hormonal causes

7.1.3 Headache attributed to intracranial hypertension secondary to hydrocephalus

## 7.1.1 Headache attributed to IIH

- A. Progressive headache with  $\geq 1$  of the following characteristics and fulfilling criteria C and D:
  1. daily occurrence
  2. diffuse and/or constant (non-pulsating) pain
  3. aggravated by coughing or straining
- B. Intracranial hypertension (*criteria on next slide*)
- C. Headache develops in close temporal relation to increased intracranial pressure
- D. Headache improves after withdrawal of CSF to reduce pressure to 120-170 mm H<sub>2</sub>O and resolves within 72 h of persistent normalisation of intracranial pressure

# 7.1.1 Headache attributed to IIH

## B. Intracranial hypertension fulfilling the following criteria:

1. alert patient with neurological examination that either is normal or demonstrates any of the following abnormalities:
  - a) papilloedema
  - b) enlarged blind spot
  - c) visual field defect (progressive if untreated)
  - d) sixth nerve palsy
2. increased CSF pressure (>200 mm H<sub>2</sub>O [non-obese], >250 mm H<sub>2</sub>O [obese]) measured by lumbar puncture in the recumbent position or by epidural or intraventricular pressure monitoring
3. normal CSF chemistry (low CSF protein acceptable) and cellularity
4. intracranial diseases (including venous sinus thrombosis) ruled out by appropriate investigations
5. no metabolic, toxic or hormonal cause of intracranial hypertension



# 7.2 Headache attributed to low cerebrospinal fluid pressure

7.2.1 Post-dural puncture headache

7.2.2 CSF fistula headache

7.2.3 Headache attributed to spontaneous (or idiopathic) low CSF pressure

## 7.2.1 Post-dural (post-lumbar) puncture headache

- A. Headache that worsens within 15 min after sitting or standing and improves within 15 min after lying, with  $\geq 1$  of the following and fulfilling criteria C and D:
  - 1. neck stiffness; 2. tinnitus; 3. hypacusia;
  - 4. photophobia; 5. nausea
- B. Dural puncture has been performed
- C. Headache develops within 5 d after dural puncture
- D. Headache resolves either:
  - 1. spontaneously within 1 wk
  - 2. within 48 h after effective treatment of the spinal fluid leak

## 7.3 Headache attributed to non-infectious inflammatory disease

- 7.3.1 Headache attributed to neurosarcoidosis
- 7.3.2 Headache attributed to aseptic (non-infectious) meningitis
- 7.3.3 Headache attributed to other non-infectious inflammatory disease
- 7.3.4 Headache attributed to lymphocytic hypophysitis

## 7.4 Headache attributed to intracranial neoplasm

- 7.4.1 Headache attributed to increased intracranial pressure or hydrocephalus caused by neoplasm
- 7.4.2 Headache attributed directly to neoplasm
- 7.4.3 Headache attributed to carcinomatous meningitis
- 7.4.4 Headache attributed to hypothalamic or pituitary hyper- or hyposecretion

## 7.4.2 Headache attributed directly to neoplasm

- A. Headache with  $\geq 1$  of the following characteristics and fulfilling criteria C and D:
  1. progressive
  2. localised
  3. worse in the morning
  4. aggravated by coughing or bending forward
- B. Intracranial neoplasm shown by imaging
- C. Headache develops in temporal (and usually spatial) relation to the neoplasm
- D. Headache resolves within 7 d after surgical removal or volume-reduction of neoplasm or treatment with corticosteroids

# 7.6 Headache attributed to epileptic seizure

7.6.1 Hemicrania epileptica

7.6.2 Post-seizure (post-ictal) headache

## 7.6.2 Post-seizure (post-ictal) headache

- A. Headache with features of tension-type headache or, in a patient with migraine, of migraine headache and fulfilling criteria C and D
- B. The patient has had a partial or generalised epileptic seizure
- C. Headache develops within 3 h following the seizure
- D. Headache resolves within 72 h after the seizure

# 8. Headache attributed to a substance or its withdrawal

8.1 Headache induced by acute substance use or exposure

8.2 Medication-overuse headache (MOH)

8.3 Headache as an adverse event attributed to chronic medication

8.4 Headache attributed to substance withdrawal



# 8.1 Headache induced by acute substance use or exposure

- 8.1.1 Nitric oxide donor-induced headache
- 8.1.2 Phosphodiesterase inhibitor-induced headache
- 8.1.3 Carbon monoxide-induced headache
- 8.1.4 Alcohol-induced headache.
- 8.1.5 Headache induced by food components and additives
- 8.1.6 Cocaine-induced headache
- 8.1.7 Cannabis-induced headache
- 8.1.8 Histamine-induced headache
- 8.1.9 Calcitonin gene-related peptide-induced headache
- 8.1.10 Headache as an acute adverse event attributed to medication used for other indications
- 8.1.11 Headache induced by other acute substance use

## 8.1.3 Carbon monoxide (CO)-induced headache

- A. Bilateral and/or continuous headache, with quality and intensity that may be related to the severity of CO intoxication, fulfilling criteria C and D
- B. Exposure to CO
- C. Headache develops within 12 h of exposure
- D. Headache resolves within 72 h after elimination of CO

# 8.2 Medication-overuse headache

## *New entrant to classification*

- 8.2.1 Ergotamine-overuse headache
- 8.2.2 Triptan-overuse headache
- 8.2.3 Analgesic-overuse headache
- 8.2.4 Opioid-overuse headache
- 8.2.5 Combination analgesic-overuse headache
- 8.2.6 Medication-overuse headache attributed to combination of acute medications
- 8.2.7 Headache attributed to other medication overuse
- 8.2.8 Probable medication-overuse headache

# 8.2 Medication-overuse headache

## *Notes*

- The most common cause of migraine-like or mixed migraine-like and TTH-like headaches on  $\geq 15$  d/mo is overuse of symptomatic migraine drugs and/or analgesics
- Patients with migraine or TTH who develop new headache or whose migraine or TTH is made markedly worse during medication overuse should be coded for that headache + 8.2 *Medication-overuse headache*
- Diagnosis of MOH is important because patients rarely respond to preventative medications until withdrawn

## 8.2 Medication-overuse headache

- A. Headache present on  $\geq 15$  d/mo fulfilling criteria C and D
- B. Regular overuse for  $> 3$  mo of one or more drugs that can be taken for acute and/or symptomatic treatment of headache
- C. Headache has developed or markedly worsened during medication overuse
- D. Headache resolves or reverts to its previous pattern within 2 mo after discontinuation of overused medication

## 8.2.1 Ergotamine-overuse headache

- A. Headache fulfilling criteria A, C and D for 8.2  
*Medication-overuse headache*
- B. Ergotamine intake on  $\geq 10$  d/mo on a regular basis for  
>3 mo

## 8.2.2 Triptan-overuse headache

- A. Headache fulfilling criteria A, C and D for 8.2  
*Medication-overuse headache*
- B. Triptan intake (any formulation) on  $\geq 10$  d/mo on a regular basis for  $> 3$  mo

## 8.2.3 Analgesic-overuse headache

- A. Headache fulfilling criteria A, C and D for 8.2  
*Medication-overuse headache*
- B. Intake of simple analgesics on  $\geq 15$  d/mo on a regular basis for  $>3$  mo



## 8.2.3 Analgesic-overuse headache

### *Note*

- Expert opinion rather than formal evidence suggests that use on  $\geq 15$  d/mo rather than  $\geq 10$  d/mo is needed to induce analgesic-overuse headache

## 8.2.5 Combination analgesic-overuse headache

### ***Name-change in ICHD-IIR1***

- A. Headache fulfilling criteria A, C and D for 8.2  
*Medication-overuse headache*
- B. Intake of combination analgesic medications\* on  $\geq 10$  d/mo on a regular basis for  $> 3$  mo

\*Combinations typically implicated are those containing simple analgesics combined with opioids, butalbital and/or caffeine

## 8.2.6 MOH attributed to combination of acute medications

***New entrant to classification in ICHD-IIR1***

- A. Headache fulfilling criteria A, C and D for 8.2  
*Medication-overuse headache*
- B. Intake of any combination of ergotamine, triptans, analgesics and/or opioids on  $\geq 10$  d/mo on a regular basis for  $> 3$  mo without overuse of any single class alone\*

\*Diagnose 8.2.1-8.2.5 if criterion B is fulfilled in respect of any single class(es) of these medications

## 8.2.8 Probable MOH

### ***Renumbered (from 8.2.7) in ICHD-IIR1***

- A. Headache fulfilling criteria A and C for 8.2  
*Medication-overuse headache*
- B. Medication overuse fulfilling criterion B for any one of the subforms 8.2.1 to 8.2.7
- C. One or other of the following:
  - 1. overused medication has not yet been withdrawn
  - 2. medication overuse has ceased within the last 2 mo but headache has not so far resolved or reverted to its previous pattern

## 8.2.8 Probable MOH

8.2.8.1 Probable ergotamine-overuse headache

8.2.8.2 Probable triptan-overuse headache

8.2.8.3 Probable analgesic-overuse headache

8.2.8.4 Probable opioid-overuse headache

8.2.8.5 Probable combination analgesic-overuse headache

8.2.8.6 Headache probably attributed to overuse of acute medication combinations

(new in ICHD-IIR1)

8.2.8.7 Headache probably attributed to other medication overuse

## 8.3 Headache as an adverse event attributed to chronic medication

### 8.3.1 Exogenous hormone-induced headache

- A. Headache or migraine fulfilling criteria C and D
- B. Regular use of exogenous hormones
- C. Headache or migraine develops or markedly worsens within 3 mo of commencing exogenous hormones
- D. Headache or migraine resolves or reverts to its previous pattern within 3 mo after total discontinuation of exogenous hormones

## 8.4 Headache attributed to substance withdrawal

- 8.4.1 Caffeine-withdrawal headache
- 8.4.2 Opioid-withdrawal headache
- 8.4.3 Oestrogen-withdrawal headache
- 8.4.4 Headache attributed to withdrawal from chronic use of other substances

## 8.4.1 Caffeine-withdrawal headache

- A. Bilateral and/or pulsating headache fulfilling criteria C and D
- B. Caffeine consumption of >200 mg/d for >2 wk, which is interrupted or delayed
- C. Headache develops within 24 h after last caffeine intake and is relieved within 1 h by 100 mg of caffeine
- D. Headache resolves within 7 d after total caffeine withdrawal



## 8.4.3 Oestrogen-withdrawal headache

- A. Headache or migraine fulfilling criteria C and D
- B. Daily use of exogenous oestrogen for  $\geq 3$  wk, which is interrupted
- C. Headache or migraine develops within 5 d after last use of oestrogen
- D. Headache or migraine resolves within 3 d

# 9. Headache attributed to infection

- 9.1 Headache attributed to intracranial infection
- 9.2 Headache attributed to systemic infection
- 9.3 Headache attributed to HIV/AIDS
- 9.4 Chronic post-infection headache

# 9.1 Headache attributed to intracranial infection

- 9.1.1 Headache attributed to bacterial meningitis
- 9.1.2 Headache attributed to lymphocytic meningitis
- 9.1.3 Headache attributed to encephalitis
- 9.1.4 Headache attributed to brain abscess
- 9.1.5 Headache attributed to subdural empyema

## 9.1.1 Headache attributed to bacterial meningitis

- A. Headache with  $\geq 1$  of the following characteristics and fulfilling criteria C and D:
  - 1. diffuse pain
  - 2. intensity increasing to severe
  - 3. associated with nausea, photophobia and/or phonophobia
- B. Evidence of bacterial meningitis from examination of CSF
- C. Headache develops during the meningitis
- D. One or other of the following:
  - 1. headache resolves within 3 mo after relief from meningitis
  - 2. headache persists but 3 mo have not yet passed since relief from meningitis

# 9.1.1 Headache attributed to bacterial meningitis

## *Notes*

- Criterion D does not relate to evidence of causation
- Causation is established by onset during diagnosed bacterial meningitis, whilst it is well recognised that this headache often persists
- When this occurs, 9.4.1 *Chronic post-bacterial meningitis headache* is diagnosed
- Criterion D2 allows a default diagnosis within 3 mo, before it is known whether headache will resolve or persist

## 9.4.1 Chronic post-bacterial meningitis headache

- A. Headache with  $\geq 1$  of the following characteristics and fulfilling criteria C and D:
  1. diffuse continuous pain
  2. associated with dizziness
  3. associated with difficulty in concentrating and/or loss of memory
- B. Evidence of previous intracranial bacterial infection from CSF examination or neuroimaging
- C. Headache is a direct continuation of  
*9.1.1 Headache attributed to bacterial meningitis*
- D. Headache persists for  $>3$  mo after resolution of infection

## 9.2 Headache attributed to systemic infection

- A. Headache with  $\geq 1$  of the following characteristics and fulfilling criteria C and D:
  1. diffuse pain
  2. intensity increasing to moderate or severe
  3. associated with fever, general malaise or other symptoms of systemic infection
- B. Evidence of systemic infection
- C. Headache develops during the systemic infection
- D. Headache resolves within 72 h after effective treatment of the infection

## 9.2 Headache attributed to systemic infection

9.2.1 Headache attributed to systemic bacterial infection

9.2.2 Headache attributed to systemic viral infection

9.2.3 Headache attributed to other systemic infection



## 9.3 Headache attributed to HIV/AIDS

- A. Headache with variable mode of onset, site and intensity fulfilling criteria C and D
- B. Confirmation of HIV infection and/or of the diagnosis of AIDS, and of the presence of HIV/AIDS-related pathophysiology likely to cause headache, by neuroimaging, CSF examination, EEG and/or laboratory investigations
- C. Headache develops in close temporal relation to the HIV/AIDS-related pathophysiology
- D. Headache resolves within 3 mo after the infection subsides

# 10. Headache attributed to disorder of homoeostasis

- 10.1 Headache attributed to hypoxia and/or hypercapnia
- 10.2 Dialysis headache
- 10.3 Headache attributed to arterial hypertension
- 10.4 Headache attributed to hypothyroidism
- 10.5 Headache attributed to fasting
- 10.6 Cardiac cephalalgia
- 10.7 Headache attributed to other disorder of homoeostasis

# 10. Headache attributed to disorder of homoeostasis

## *Terminology change 1988-2004*

- This section was previously  
*10. Headache associated with metabolic disorder*
- The new term captures more accurately their true nature
- Headaches caused by significant disturbances in arterial pressure and by myocardial ischaemia are now included in this section

# 10.1 Headache attributed to hypoxia and/or hypercapnia

- 10.1.1 High-altitude headache
- 10.1.2 Diving headache
- 10.1.3 Sleep apnoea headache

## 10.3 Headache attributed to arterial hypertension

- 10.3.1 Headache attributed to phaeochromocytoma
- 10.3.2 Headache attributed to hypertensive crisis without hypertensive encephalopathy
- 10.3.3 Headache attributed to hypertensive encephalopathy
- 10.3.4 Headache attributed to pre-eclampsia
- 10.3.5 Headache attributed to eclampsia
- 10.3.6 Headache attributed to acute pressor response to an exogenous agent

# **11. Headache or facial pain attributed to disorder of cranium, neck, eyes, ears, nose, sinuses, teeth, mouth or other facial or cranial structures**

11.1 Headache attributed to disorder of cranial bone

11.2 Headache attributed to disorder of neck

11.3 Headache attributed to disorder of eyes

11.4 Headache attributed to disorder of ears

11.5 Headache attributed to rhinosinusitis

11.6 Headache attributed to disorder of teeth, jaws or related structures

11.7 Headache or facial pain attributed to temporomandibular joint (TMJ) disorder

11.8 Headache attributed to other disorder of the above

## 11.2.1 Cervicogenic headache

- A. Pain, referred from a source in the neck and perceived in one or more regions of the head and/or face, fulfilling criteria C and D
- B. Clinical, laboratory and/or imaging evidence of a disorder or lesion within the cervical spine or soft tissues of the neck known to be, or generally accepted as, a valid cause of headache

## 11.2.1 Cervicogenic headache

- C. Evidence that the pain can be attributed to the neck disorder or lesion based on  $\geq 1$  of the following:
1. demonstration of clinical signs that implicate a source of pain in the neck
  2. abolition of headache following diagnostic blockade of a cervical structure or its nerve supply using placebo- or other adequate controls
- D. Pain resolves within 3 mo after successful treatment of the causative disorder or lesion



# 11.2.1 Cervicogenic headache

## **Notes**

- Cervical spondylosis and osteochondritis are NOT accepted as valid causes fulfilling criterion B
- When myofascial tender spots are the cause, the headache should be coded under  
*2. Tension-type headache (subform associated with pericranial tenderness)*

# 11.3 Headache attributed to disorder of eyes

- 11.3.1 Headache attributed to acute glaucoma
- 11.3.2 Headache attributed to refractive errors
- 11.3.3 Headache attributed to heterophoria or heterotropia (latent or manifest squint)
- 11.3.4 Headache attributed to ocular inflammatory disorder

# 11.3.1 Headache attributed to acute glaucoma

- A. Pain in the eye and behind or above it, fulfilling criteria C and D
- B. Raised intraocular pressure, with at least one of the following:
  1. conjunctival injection
  2. clouding of cornea
  3. visual disturbances
- C. Pain develops simultaneously with glaucoma
- D. Pain resolves within 72 h of effective treatment of glaucoma

# 11.5 Headache attributed to rhinosinusitis

- A. Frontal headache accompanied by pain in one or more regions of the face, ears or teeth and fulfilling criteria C and D
- B. Clinical, nasal endoscopic, CT and/or MRI imaging and/or laboratory evidence of acute or acute-on-chronic rhinosinusitis
- C. Headache and facial pain develop simultaneously with onset or acute exacerbation of rhinosinusitis
- D. Headache and/or facial pain resolve within 7 d after remission or successful treatment of acute or acute-on-chronic rhinosinusitis

# 11.5 Headache attributed to rhinosinusitis

## *Notes*

- 11.5 *Headache attributed to rhinosinusitis* is differentiated from “sinus headaches”, a commonly-made but non-specific diagnosis. Most such cases fulfil the criteria for 1.1 *Migraine without aura*, with headache either accompanied by prominent autonomic symptoms in the nose or triggered by nasal changes
- *Chronic sinusitis* is not a cause of headache or facial pain unless relapsing into an acute stage

## 11.7 Headache or facial pain attributed to temporomandibular joint disorder

- A. Recurrent pain in  $\geq 1$  regions of the head and/or face fulfilling criteria C and D
- B. X-ray, MRI and/or bone scintigraphy demonstrate TMJ disorder
- C. Evidence that pain can be attributed to the TMJ disorder, based on  $\geq 1$  of the following:
  1. pain is precipitated by jaw movements and/or chewing of hard or tough food
  2. reduced range of or irregular jaw opening
  3. noise from one or both TMJs during jaw movements
  4. tenderness of the joint capsule(s) of one or both TMJs
- D. Headache resolves within 3 mo, and does not recur, after successful treatment of the TMJ disorder

# 12. Headache attributed to psychiatric disorder

***New section in classification***

12.1 Headache attributed to somatisation disorder

12.2 Headache attributed to psychotic disorder

# 12. Headache attributed to psychiatric disorder

## *Notes*

- There is very limited evidence supporting psychiatric causes of headache
- The only diagnoses included are those of headache attributed to psychiatric conditions known to be symptomatically manifested by headache
- Such cases are rare
- The vast majority of headaches occurring in association with psychiatric disorders are not causally related to them but instead represent comorbidity



# 12.1 Headache attributed to somatisation disorder

- A. Headache, no typical characteristics known, fulfilling criterion C
- B. Presence of somatisation disorder fulfilling DSM-IV criteria
- C. Headache is not attributed to another cause

## 12.2 Headache attributed to psychotic disorder

- A. Headache, no typical characteristics known, fulfilling criteria C-E
- B. Delusional belief about the presence and/or aetiology of headache occurring in the context of delusional disorder, schizophrenia, major depressive episode with psychotic features, manic episode with psychotic features or other psychotic disorder fulfilling DSM-IV criteria
- C. Headache occurs only when delusional
- D. Headache resolves when delusions remit
- E. Headache is not attributed to another cause

# Part 3: Cranial neuralgias, central and primary facial pain and other headaches

13. Cranial neuralgias and central causes of facial pain
14. Other headache, cranial neuralgia, central or primary facial pain

# 13. Cranial neuralgias and central causes of facial pain

- 13.1 Trigeminal neuralgia
- 13.2 Glossopharyngeal neuralgia
- 13.3 Nervus intermedius neuralgia
- 13.4 Superior laryngeal neuralgia
- 13.5 Nasociliary neuralgia
- 13.6 Supraorbital neuralgia
- 13.7 Other terminal branch neuralgias
- 13.8 Occipital neuralgia
- 13.9 Neck-tongue syndrome
- 13.10 External compression headache
- 13.11 Cold-stimulus headache

# 13. Cranial neuralgias and central causes of facial pain

- 13.12 Constant pain caused by compression, irritation or distortion of cranial nerves or upper cervical roots by structural lesions
- 13.13 Optic neuritis
- 13.14 Ocular diabetic neuropathy
- 13.15 Head or facial pain attributed to herpes zoster
- 13.16 Tolosa-Hunt syndrome
- 13.17 Ophthalmoplegic 'migraine'
- 13.18 Central causes of facial pain
- 13.19 Other cranial neuralgia or other centrally mediated facial pain

# 13. Cranial neuralgias and central causes of facial pain

***Terminology and section number change 1988-2004***

This section was previously

*12. Cranial neuralgias, nerve trunk pain and deafferentation pain*

# 13.1 Trigeminal neuralgia



13.1.1 Classical trigeminal neuralgia

13.1.2 Symptomatic trigeminal neuralgia

# 13.1.1 Classical trigeminal neuralgia

- A. Paroxysmal attacks of pain lasting from a fraction of 1 sec to 2 min, affecting one or more divisions of the trigeminal nerve and fulfilling criteria B and C
- B. Pain has  $\geq 1$  of the following characteristics:
  - 1. intense, sharp, superficial or stabbing
  - 2. precipitated from trigger areas or by trigger factors
- C. Attacks are stereotyped in the individual patient
- D. There is no clinically evident neurological deficit
- E. Not attributed to another disorder



## 13.1.2 Symptomatic trigeminal neuralgia

*As 13.1.1 except:*

- A. Paroxysmal attacks of pain lasting from a fraction of 1 sec to 2 min, with or without persistence of aching between paroxysms, affecting one or more divisions of the trigeminal nerve and fulfilling criteria B and C
- D. (*replacing criteria D and E*)  
A causative lesion, other than vascular compression, has been demonstrated by special investigations and/or posterior fossa exploration

## 13.8 Occipital neuralgia

- A. Paroxysmal stabbing pain, with or without persistent aching between paroxysms, in the distribution(s) of the greater, lesser and/or third occipital nerves
- B. Tenderness over the affected nerve
- C. Pain is eased temporarily by local anaesthetic block of the nerve

## 13.17 Ophthalmoplegic 'migraine'

- A. At least 2 attacks fulfilling criterion B
- B. Migraine-like headache accompanied or followed within 4 d of its onset by paresis of  $\geq 1$  of the third, fourth and/or sixth cranial nerves
- C. Parasellar, orbital fissure and posterior fossa lesions ruled out by appropriate investigations

# 13.17 Ophthalmoplegic 'migraine'

## *Reclassification 1988-2004*

- 13.17 *Ophthalmoplegic 'migraine'* was previously classified as 1.3 *Ophthalmoplegic migraine*
- It is unlikely to be a variant of migraine since the headache often lasts for  $\geq 1$  wk and there is a latent period of up to 4 d from headache onset to ophthalmoplegia
- 13.17 *Ophthalmoplegic 'migraine'* may be a recurrent demyelinating neuropathy
- It is very rare

# 13.18 Central causes of facial pain

- 13.18.1 Anaesthesia dolorosa
- 13.18.2 Central post-stroke pain
- 13.18.3 Facial pain attributed to multiple sclerosis
- 13.18.4 Persistent idiopathic facial pain
- 13.18.5 Burning mouth syndrome

## 13.18.1 Anaesthesia dolorosa

- A. Persistent pain and dysaesthesia within the area of distribution of one or more divisions of the trigeminal nerve or of the occipital nerves
- B. Diminished sensation to pin-prick and sometimes other sensory loss over the affected area
- C. There is a lesion of the relevant nerve or its central connections

## 13.18.2 Central post-stroke pain

- A. Pain and dysaesthesia in one half of the face, associated with loss of sensation to pin-prick, temperature and/or touch and fulfilling criteria C and D
- B. One or both of the following:
  1. history of sudden onset suggesting a vascular lesion (stroke)
  2. demonstration by CT or MRI of a vascular lesion in an appropriate site
- C. Pain and dysaesthesia develop within 6 mo after stroke
- D. Not explicable by a lesion of the trigeminal nerve

## 13.18.4 Persistent idiopathic facial pain

***Previously used term: Atypical facial pain***

- A. Pain in the face, present daily and persisting for all or most of the day, fulfilling criteria B and C
- B. Pain is confined at onset to a limited area on one side of the face, and is deep and poorly localised
- C. Pain is not associated with sensory loss or other physical signs
- D. Investigations including X-ray of face and jaws do not demonstrate any relevant abnormality



## 13.18.5 Burning mouth syndrome

- A. Pain in the mouth present daily and persisting for most of the day
- B. Oral mucosa is of normal appearance
- C. Local and systemic diseases have been excluded

# 14. Other headache, cranial neuralgia, central or primary facial pain

- 14.1 Headache not elsewhere classified
- 14.2 Headache unspecified

# 14. Other headache, cranial neuralgia, central or primary facial pain

## **Notes**

- There are probably headache entities still to be described; until classified, they can be coded as 14.1 *Headache not elsewhere classified*.
- When very little information is available (the patient is dead, unable to communicate or unavailable), allowing only to state that headache is or was present but not which type of headache, it is coded as 14.2 *Headache unspecified*

# 14.1 Headache not elsewhere classified

- A. Headache with characteristic features suggesting that it is a unique diagnostic entity
- B. Does not fulfil criteria for any of the headache disorders described in chapters 1-13

## 14.2 Headache unspecified

- A. Headache is or has been present
- B. Not enough information is available to classify the headache at any level of this classification

# Appendix

- Presents research criteria for a number of novel entities that have not been sufficiently validated
- Presents alternative diagnostic criteria that may be preferable but for which the evidence is insufficient
- Is a first step in eliminating disorders included in the 1st edition for which sufficient evidence has still not been published

# A1. Migraine

## **Alternative diagnostic criteria:**

A1.1 Migraine without aura

## **Proposed new subclassification:**

A1.1.1 Pure menstrual migraine without aura

A1.1.2 Menstrually-related migraine without aura

A1.1.3 Non-menstrual migraine without aura

## **Other proposed but unvalidated criteria:**

A1.2.7 Migraine aura status

A1.3.4 Alternating hemiplegia of childhood

A1.3.5 Benign paroxysmal torticollis

# A1.1 Migraine without aura

## *Alternative diagnostic criteria*

### 1.1 Migraine without aura

D. During headache  
≥**1** of the following:

1. nausea and/or vomiting
2. photophobia and phonophobia

### A1.1 Migraine without aura

D. During headache  
≥**2** of the following:

1. nausea
2. vomiting
3. photophobia
4. phonophobia
5. osmophobia



# A1.1 Migraine without aura

## **Note**

- Whilst the alternative criterion D appears easier both to understand and to apply, it is not yet sufficiently validated

# **A1.1 Migraine without aura**

## ***Proposed new subclassification\****

A1.1.1 Pure menstrual migraine without aura

A1.1.2 Menstrually-related migraine without aura

A1.1.3 Non-menstrual migraine without aura

\*This proposed subclassification is applicable only to menstruating women

# A1.1.1 Pure menstrual migraine without aura

- A. Attacks, in a menstruating woman, fulfilling criteria for 1.1 *Migraine without aura*
- B. Attacks occur exclusively on day  $1 \pm 2$  (ie, days  $-2$  to  $+3$ ) of menstruation in at least two out of three menstrual cycles and at no other times of the cycle

# A1.1.1 Pure menstrual migraine without aura

## *Notes*

- The first day of menstruation is day 1 and the preceding day is day -1; there is no day 0
- For the purposes of this classification, *menstruation* is endometrial bleeding resulting from either the normal menstrual cycle or withdrawal of exogenous progestogens (combined oral contraceptives or cyclical hormone replacement therapy)

## A1.1.2 Menstrually-related migraine without aura

- A. Attacks, in a menstruating woman, fulfilling criteria for 1.1 *Migraine without aura*
- B. Attacks occur on day  $1 \pm 2$  (ie, days  $-2$  to  $+3$ ) of menstruation in at least two out of three menstrual cycles and additionally at other times of the cycle

# A1.1.3 Non-menstrual migraine without aura

- A. Attacks, in a menstruating woman, fulfilling criteria for 1.1 *Migraine without aura*
- B. Attacks have no menstrual relationship

# A2. Tension-type headache

## *Alternative diagnostic criteria*

### 2. Tension-type headache

C. Headache has  $\geq 2$  of the following characteristics:

1. bilateral location;
2. pressing/tightening quality
3. mild or moderate intensity
4. not aggravated by routine physical activity

D. Both of the following:

1. no nausea or vomiting
2. not  $>1$  of photo- or phonophobia

### A2. Tension-type headache

C. Headache has  $\geq 3$  of the following characteristics:

D. Both of the following:

1. no nausea or vomiting
2. no photophobia or phonophobia

# A2. Tension-type headache

## *Notes*

- These alternative diagnostic criteria C and D are very specific, but have low sensitivity
- The purpose is that TTH does not become a default diagnosis



# **A3.3 Short-lasting Unilateral Neuralgiform headache attacks with cranial Autonomic symptoms (SUNA)**

## ***Proposed but unvalidated disorder***

- A. At least 20 attacks fulfilling criteria B-E
- B. Attacks of unilateral orbital, supraorbital or temporal stabbing or pulsating pain lasting from 2 sec to 10 min
- C. Pain is accompanied by one of:
  - 1. conjunctival injection and/or lacrimation
  - 2. nasal congestion and/or rhinorrhoea
  - 3. eyelid oedema
- D. Attack frequency is  $\geq 1/d$  for  $>50\%$  of the time
- E. No refractory period follows attacks triggered from trigger areas
- F. Not attributed to another disorder

# A3.3 SUNA

## *Notes*

- 3.3 *SUNCT* may be a subform of a broader problem of A3.3 *SUNA*
- This proposal requires validation
- The proposed criteria for A3.3 *SUNA* (as an alternative to 3.3 *SUNCT*) are for research purposes and need to be tested
- Cranial autonomic features should be prominent to distinguish this disorder from ophthalmic division trigeminal neuralgia

# **A9. Headache attributed to infection**

## ***Proposed but unvalidated criteria***

A9.1.6 Headache attributed to space-occupying intracranial infectious lesion or infestation

A9.1.7 Headache attributed to intracranial parasitic infestation

A9.4.2 Chronic post-non-bacterial infection headache

# A12. Headache attributed to psychiatric disorder

## *Proposed but unvalidated criteria*

A12.3 Headache attributed to major depressive disorder

A12.4 Headache attributed to panic disorder

A12.5 Headache attributed to generalised anxiety disorder

A12.6 Headache attributed to undifferentiated  
somatoform disorder

A12.7 Headache attributed to social phobia

A12.8 Headache attributed to separation anxiety disorder

A12.9 Headache attributed to post-traumatic stress  
disorder

# A12. Headache attributed to psychiatric disorder

## *Notes*

- The proposed candidate criteria sets are to facilitate research into the possible causal relationships between certain psychiatric disorders and headache
- When using them it is crucial to establish that the headache in question occurs exclusively during the course of the psychiatric disorder (*ie*, is manifest only during times when the symptoms of the psychiatric disorder are also manifest)

# A13. Cranial neuralgias and central causes of facial pain

## A13.7.1 Nummular headache

- A. Mild to moderate head pain fulfilling criteria B and C
- B. Pain is felt exclusively in a rounded or elliptical area typically 2-6 cm in diameter
- C. Pain is chronic and either continuous or interrupted by spontaneous remissions lasting weeks to months
- D. Not attributed to another disorder

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- ***The International Classification of Headache Disorders, 2<sup>nd</sup> edition (ICHD-II)***  
is published in *Cephalalgia* 2004; **24** (Suppl 1)
- ***The first revision (ICHD-IIR1)*** (with changes affecting only section 8.2) is published in *Cephalalgia* 2005; **25**: 460-465
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*Cephalalgia* 2004; **24** Suppl 1: 1-160);  
*Cephalalgia* 2005; **25**: 460-465

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