

# 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

# 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* +  
8.2.7 *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*