

Lifting The Burden

The Global Campaign against Headache

A collaboration between the World Health Organization, non-governmental organisations, academic institutions and individuals worldwide

Diagnostic criteria for headache disorders in primary care

Headache disorders are common. Collectively they are amongst the top 10 causes of disability worldwide.

Of the many headache disorders, only a small number are important in primary care. The purpose of this diagnostic aid is to help primary care physicians recognise and correctly diagnose these.

The International Classification of Headache Disorders

The International Classification of Headache Disorders (ICHD) published by the International Headache Society¹ is the authoritative catalogue of headache disorders, incorporating explicit diagnostic criteria for each one. There are about 200 distinct headache types or subtypes.

More information is at www.ihs-classification.org

This adaptation of ICHD is a diagnostic aid specifically for primary care, setting out the criteria for the 15 headache disorders that are important in primary care. These include the headache disorders most likely to be seen, and those that are important because they are symptomatic of another serious underlying disorder requiring treatment. They are numbered below as in the classification; WHO ICD-10 codes, used in some countries, are given in parentheses.

How the system works

This diagnostic aid should be used as a reference. Diagnoses are made by applying the criteria set out in the classification. A diagnosis is confirmed only when all criteria for that disorder are fulfilled. However, symptoms may have been modified by treatment, and this possibility should be considered in deciding whether criteria are met.

One patient may simultaneously have two or more headache disorders. Each should be separately diagnosed because each may require separate management.

The presence of more than one headache disorder can cause confusion, especially when a patient fails to distinguish between them. When this is suspected, it is recommended that he or she prospectively fills out a diagnostic headache diary, for a month or longer, recording the important characteristics of each headache episode. Diaries not only improve diagnostic accuracy but also allow precise judgement of medication consumption.

The classification distinguishes between *primary headaches*, which have no other underlying causative disorder, and *secondary headaches*, which are attributed to some other disorder.

¹ *Cephalalgia* 2004; **24** (suppl 1): 1-160 (revision published in *Cephalalgia* 2005; **25**: 460-465).

Onset in close temporal relation to another disorder known to cause headache is therefore a diagnostic criterion for all secondary headaches.

The third section of the classification covers cranial neuralgias, central and primary facial pain.

DEFINITIONS OF COMMON TERMS

Attack of headache (or pain):

Headache (or pain) that builds up, remains at a certain level for minutes to 72 hours, then wanes until it is gone completely.

Close temporal relation:

This term is used to describe the relation between an organic disorder and a secondary headache attributed to it.

Duration of attack:

Time from onset until termination of an attack of headache (or pain) meeting criteria for a particular headache type or subtype. When the patient falls asleep during an attack and wakes up relieved, duration is until time of awakening. When an attack of migraine is successfully relieved by medication but symptoms recur within 48 hours, these may represent a relapse of the same attack or a new attack (see Frequency of attacks).

Facial pain:

Pain below the orbitomeatal line, above the neck and anterior to the pinnae.

Fortification spectrum:

Angulated, arcuate and gradually enlarging visual hallucination typical of migrainous aura.

Frequency of attacks:

The rate of occurrence of attacks of headache (or pain) per time period (commonly one month). Successful relief of a migraine attack with medication may be followed by relapse within 48 hours. The IHS *Guidelines for Controlled Trials of Drugs in Migraine, 2nd edition*, recommend as a practical solution, especially in differentiating attacks recorded as diary entries over the previous month, to count as distinct attacks only those that are separated by an entire day headache-free.

Headache:

Pain located above the orbitomeatal line.

Headache days:

Number of days during an observed period of time (commonly 1 month) affected by headache for any part or the whole of the day.

Intensity of pain:

Degree of pain usually expressed in terms of its functional consequence and scored on a verbal 4-point scale: 0, no pain; 1, mild pain, does not interfere with usual activities; 2, moderate pain, inhibits but does not wholly prevent usual activities; 3, severe pain, prevents all activities. It may also be expressed on a visual analogue scale.

New headache:

Any type of headache from which the patient was not previously suffering.

Phonophobia:

Hypersensitivity to sound, usually causing avoidance.

Photophobia:

Hypersensitivity to light, usually causing avoidance.

Pressing/tightening:

Pain of a constant quality often compared to an iron band around the head.

Pulsating:

Varying with the heart beat; throbbing.

Scintillation:

Visual hallucinations that are bright and fluctuate in intensity, often at approximately 8-10 cycles/second. They are typical of migraine aura.

Scotoma:

Loss of part(s) of the visual field of one or both eyes. Scotoma may be absolute (no vision) or relative (obscured or reduced vision).

PRIMARY HEADACHES

1. [G43] Migraine

Migraine is a common disabling headache disorder. It is ranked by the World Health Organization as 19th among all diseases worldwide causing disability.

Migraine has two major sub-types. 1.1 *Migraine without aura* is a clinical syndrome characterised by headache with specific features and associated symptoms. 1.2 *Migraine with aura* is primarily characterised by the focal neurological symptoms that usually precede or sometimes accompany the headache. Some patients also experience a premonitory phase, occurring hours or days before the headache, and a headache resolution phase. Premonitory and resolution symptoms include hyperactivity, hypoactivity, depression, craving for particular foods, repetitive yawning and other less typical symptoms reported by some patients.

When a patient fulfils criteria for more than one subtype of migraine, all subtypes should be diagnosed.

1.1 [G43.0] Migraine without aura

Description:

A recurrent headache disorder manifesting in attacks lasting 4-72 hours. Typical characteristics of the headache are unilateral location, pulsating quality, moderate or severe intensity, aggravation by routine physical activity and association with nausea and/or photophobia and phonophobia.

This is the commonest subtype of migraine. In women it often has a strict menstrual relationship. It is the headache disorder most prone to accelerate with frequent use of symptomatic medication, resulting in a new headache (8.2 *Medication-overuse headache*).

Diagnostic criteria:

- A. At least 5 attacks fulfilling criteria B-D
- B. Headache attacks lasting 4-72 hours (untreated or unsuccessfully treated)¹
- C. Headache has at least two of the following characteristics:
 - 1. unilateral location^{2;3}
 - 2. pulsating quality⁴
 - 3. moderate or severe pain intensity
 - 4. aggravation by or causing avoidance of routine physical activity (eg, walking or climbing stairs)
- D. During headache at least one of the following:
 - 1. nausea and/or vomiting
 - 2. photophobia and phonophobia
- E. Not attributed to another disorder⁵

Notes:

- 1. In children, attacks may last 1-72 hours.
- 2. Migraine headache is commonly bilateral in young children.

3. Migraine headache is usually frontotemporal. Occipital headache in *children*, whether unilateral or bilateral, is rare and calls for diagnostic caution; many cases are attributable to structural lesions.
4. *Pulsating* means throbbing or varying with the heartbeat.
5. History and physical and neurological examinations do not suggest any underlying and possibly causative disorder, or history and/or examination do suggest such disorder but it is ruled out by appropriate investigations, or such disorder is present but attacks do not occur for the first time in close temporal relation to it.

1.2 [G43.1] Migraine with aura

Description:

A recurrent disorder manifesting in attacks of reversible focal neurological symptoms (*aura*) that usually develop gradually over 5-20 minutes and last for less than 60 minutes. Headache with the features of migraine without aura usually follows. Less commonly, headache lacks migrainous features or is completely absent.

Two subtypes are important.

Many patients who have 1.2 *Migraine with aura* also have 1.1 *Migraine without aura*, and should receive both diagnoses.

1.2.1 [G43.10] Typical aura with migraine headache

Description:

Typical aura consisting of visual and/or sensory and/or speech symptoms. Gradual development, duration no longer than one hour, a mix of positive and negative features and complete reversibility characterise the aura which is associated with a headache fulfilling criteria for 1.1 *Migraine without aura*.

Visual aura is the most common type of aura, often presenting as a fortification spectrum, *ie*, a zigzag figure near the point of fixation that may gradually spread right or left and assume a laterally convex shape with an angulated scintillating edge leaving variable degrees of absolute or relative scotoma in its wake. In other cases, scotoma without positive phenomena may occur. Next in frequency are sensory disturbances in the form of pins-and-needles moving slowly from the point of origin and affecting a greater or smaller part of one side of the body and face. Numbness may occur in its wake, but numbness may also be the only symptom. Less frequent are speech disturbances, usually dysphasic but often hard to categorise.

1.2.1 *Typical aura with migraine headache* is the most common migraine syndrome associated with aura. The diagnosis is usually evident after a careful history alone though there are rare secondary mimics including carotid dissection, arteriovenous malformation and seizure.

Diagnostic criteria:

- A. At least 2 attacks fulfilling criteria B–D
- B. Aura consisting of at least one of the following, but no motor weakness:
 1. fully reversible visual symptoms including positive features (*eg*, flickering lights, spots or lines) and/or negative features (*ie*, loss of vision)
 2. fully reversible sensory symptoms including positive features (*ie*, pins-and-needles) and/or negative features (*ie*, numbness)
 3. fully reversible dysphasic speech disturbance
- C. At least two of the following:
 1. homonymous visual symptoms¹ and/or unilateral sensory symptoms
 2. at least one aura symptom develops gradually over ≥ 5 minutes and/or different aura symptoms occur in succession over ≥ 5 minutes
 3. each symptom lasts ≥ 5 and ≤ 60 minutes
- D. Headache fulfilling criteria B-D for 1.1 *Migraine without aura* begins during the aura or follows aura within 60 minutes
- E. Not attributed to another disorder²

Notes:

1. Additional loss or blurring of central vision may occur.
2. History and physical and neurological examinations do not suggest any underlying and possibly causative disorder, or history and/or examination do suggest such disorder but it is ruled out by appropriate investigations, or such disorder is present but attacks do not occur for the first time in close temporal relation to it.

1.2.3 [G43.104] Typical aura without headache*Description:*

Typical aura consisting of visual and/or sensory symptoms with or without speech symptoms. Gradual development, duration no longer than one hour, a mix of positive and negative features and complete reversibility characterise the aura, which is not associated with headache.

Patients may have 1.2.1 *Typical aura with migraine headache* and 1.2.3 *Typical aura without headache*. A small number have 1.2.3 *Typical aura without headache* exclusively. More commonly, as patients with 1.2.1 *Typical aura with migraine headache* become older, their headache loses migraine characteristics or disappears completely, even though auras continue.

In the absence of headache, distinction of aura from mimics signalling serious disease (*eg*, transient ischaemic attack) becomes much more important and may require investigation.

Diagnostic criteria:

- A. At least 2 attacks fulfilling criteria B–D
- B. Aura consisting of at least one of the following, with or without speech disturbance but no motor weakness:
 1. fully reversible visual symptoms including positive features (*eg*, flickering lights, spots or lines) and/or negative features (*ie*, loss of vision)
 2. fully reversible sensory symptoms including positive features (*ie*, pins-and-needles) and/or negative features (*ie*, numbness)
- C. At least two of the following:
 1. homonymous visual symptoms¹ and/or unilateral sensory symptoms
 2. at least one aura symptom develops gradually over ≥ 5 minutes and/or different aura symptoms occur in succession over ≥ 5 minutes
 3. each symptom lasts ≥ 5 and ≤ 60 minutes
- D. Headache does not occur during aura nor follow aura within 60 minutes
- E. Not attributed to another disorder²

Notes:

1. Additional loss or blurring of central vision may occur.
2. History and physical and neurological examinations do not suggest any underlying and possibly causative disorder, or history and/or examination do suggest such disorder but it is ruled out by appropriate investigations, or such disorder is present but attacks do not occur for the first time in close temporal relation to it.

2. [G44.2] Tension-type headache (TTH)

This is the most common type of primary headache: its lifetime prevalence in the general population ranges from 30 to 78%. Two subtypes are important.

2.2 [G44.2] Frequent episodic tension-type headache*Description:*

Frequent episodes of headache lasting minutes to days. The pain is typically bilateral, pressing or tightening in quality and of mild to moderate intensity, and it does not worsen with routine physical activity. There is no nausea but photophobia or phonophobia may be present.

2.2 *Frequent tension-type headache* often coexists with 1.1 *Migraine without aura*, in which case both diagnoses should be given. A diagnostic headache diary may be required to separate them.

Diagnostic criteria:

- A. At least 10 episodes occurring on ≥ 1 but < 15 days per month for at least 3 months (≥ 12 and < 180 days per year) and fulfilling criteria B-D
- B. Headache lasting from 30 minutes to 7 days
- C. Headache has at least two 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 such as walking or climbing stairs
- 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 disorder¹

Note:

- 1. History and physical and neurological examinations do not suggest any underlying and possibly causative disorder, or history and/or examination do suggest such disorder but it is ruled out by appropriate investigations, or such disorder is present but attacks do not occur for the first time in close temporal relation to it.

2.3 [G44.2] Chronic tension-type headache

Description:

A disorder evolving from episodic tension-type headache, with daily or very frequent episodes of headache lasting minutes to days. The pain is typically bilateral, pressing or tightening in quality and of mild to moderate intensity, and it does not worsen with routine physical activity. There may be mild nausea, photophobia or phonophobia.

In many uncertain cases, because of the frequency of headache, there is overuse of acute medication for headache. The diagnosis may in that case be 8.2 *Medication-overuse headache*. Overused medication should be withdrawn. The diagnosis cannot meanwhile be certain.

Diagnostic criteria:

- A. Headache occurring on ≥ 15 days per month on average for > 3 months (≥ 180 days per year) and fulfilling criteria B-D
- B. Headache lasts hours or may be continuous
- C. Headache has at least two 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 such as walking or climbing stairs
- D. Both of the following:
 - 1. no more than one of photophobia, phonophobia or mild nausea
 - 2. neither moderate or severe nausea nor vomiting
- E. Not attributed to another disorder^{1;2}

Notes:

- 1. History and physical and neurological examinations do not suggest any underlying and possibly causative disorder, or history and/or examination do suggest such disorder but it is ruled out by appropriate investigations, or such disorder is present but attacks do not occur for the first time in close temporal relation to it.
- 2. When medication overuse is present, the diagnosis may be 8.2 *Medication-overuse headache*. This will remain uncertain until 2 months after medication has been withdrawn.

3.1 [G44.0] Cluster headache

Cluster headache is one of a group of disorders (trigeminal autonomic cephalalgias) sharing the clinical features of short-duration headache and prominent cranial parasympathetic autonomic features.

Description:

Attacks of excruciating, strictly unilateral pain which is orbital, supraorbital or temporal, lasting 15-180 minutes and occurring from once every other day to 8 times a day. Pain is associated ipsilaterally with one or more of the following: conjunctival injection, lacrimation, nasal congestion, rhinorrhoea, forehead and facial sweating, miosis, ptosis, eyelid oedema. Most patients are restless or agitated during an attack.

Attacks usually occur in series (cluster periods) lasting for weeks or months separated by remission periods. About 10-15% of patients have chronic symptoms without remissions.

Prevalence is 3-4 times higher in men than in women.

Two subtypes are important.

Diagnostic criteria:

- A. At least 5 attacks fulfilling criteria B-D
- B. Severe or very severe unilateral orbital, supraorbital and/or temporal pain lasting 15-180 minutes if untreated
- C. Headache is accompanied by at least one 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 one every other day to 8 per day
- E. Not attributed to another disorder¹

Note:

- 1. History and physical and neurological examinations do not suggest any underlying and possibly causative disorder, or history and/or examination do suggest such disorder but it is ruled out by appropriate investigations, or such disorder is present but attacks do not occur for the first time in close temporal relation to it.

3.1.1 [G44.01] Episodic cluster headache

Description:

Cluster headache attacks occurring in periods lasting 7 days to 1 year (usually 2 weeks to 3 months) separated by pain-free periods lasting 1 month or longer.

Diagnostic criteria:

- A. Attacks fulfilling criteria A-E for 3.1 *Cluster headache*
- B. At least two cluster periods lasting 7-365 days and separated by pain-free remission periods of ≥ 1 month

3.1.2 [G44.02] Chronic cluster headache

Description:

Cluster headache attacks occurring for more than 1 year without remission or with remissions lasting less than 1 month.

3.1.2 *Chronic cluster headache* may arise *de novo* or evolve from 3.1.1 *Episodic cluster headache*. Some patients may switch from chronic to episodic cluster headache.

Diagnostic criteria:

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

SECONDARY HEADACHES

A new headache occurring with another disorder recognised to be capable of causing it is always diagnosed as secondary.

Onset in close temporal relation to another disorder that is known to cause headache is therefore a diagnostic criterion for all secondary headaches. For most secondary headaches, an essential part of the evidence of a causal relationship is that the headache greatly improves or resolves after relief from the causative disorder (through treatment or spontaneous remission). However, there is usually a need to make a working ("probable") diagnosis before the disorder is treated, since this guides treatment. The diagnosis is then confirmed once the treatment results are known.

General diagnostic criteria for secondary headaches:

- A. Headache [with the following listed characteristics] fulfilling criteria C and D
- B. Another disorder known to be able to cause headache has been demonstrated
- C/D. Evidence of a causal relationship

Criteria B-D may require tests or procedures to be carried out that cannot be undertaken in primary care. In such cases, the diagnosis cannot be confirmed in primary care. The crucial role of primary care is to recognise the possibility of the diagnosis.

5.2.1 [G44.30] Chronic post-traumatic headache attributed to moderate or severe head injury

Chronic post-traumatic headache is often part of the post-traumatic syndrome, which includes symptoms such as equilibrium disturbance, poor concentration, decreased work ability, irritability, depressive mood and sleep disturbances.

Diagnostic criteria:

- 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 minutes
 2. Glasgow Coma Scale (GCS) <13
 3. post-traumatic amnesia for >48 hours
 4. imaging demonstration of a traumatic brain lesion (cerebral haematoma, intracerebral and/or subarachnoid haemorrhage, brain contusion and/or skull fracture)
- C. Headache develops within 7 days after head trauma or after regaining consciousness following head trauma
- D. Headache persists for >3 months after head trauma

6.2.2 [G44.810] Headache attributed to subarachnoid haemorrhage (SAH)

Subarachnoid haemorrhage is by far the most common cause of incapacitating headache of abrupt onset (thunderclap headache). Headache is usually the prominent initial symptom of SAH. Abrupt onset is its key feature. It is often unilateral at onset and accompanied by nausea, vomiting, disorders of consciousness and nuchal rigidity.

SAH can occur in anyone, including patients already diagnosed as having a primary headache of any type. Physicians must be alert to the occurrence of new or different headache. SAH is a neurosurgical emergency: it is fatal in 50% of cases whilst 50% of survivors are left disabled.

Diagnostic criteria:

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

6.4.1 [G44.812] Headache attributed to giant cell arteritis (GCA)

Giant cell arteritis is conspicuously associated with headache, but its characteristics are variable. Any persisting headache with recent onset in a patient over 60 years of age should suggest GCA.

GCA must be recognised. The major risk is of blindness, preventable by immediate steroid treatment. The time interval between visual loss in one eye and in the other is usually less than 1 week.

Diagnostic criteria:

- 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 days of high-dose steroid treatment

7. [G44.82] Headache attributed to non-vascular intracranial disorder

Included here are the headaches attributed to changes (increase or decrease) in intracranial pressure. There are two of importance.

7.2 [G44.820] Headache attributed to low cerebrospinal fluid pressure

There are three subtypes of this disorder, distinguished by aetiology. The key diagnostic criteria are similar for all three.

Diagnostic criteria:

- A. Headache that worsens within 15 minutes after sitting or standing and improves within 15 minutes after lying, with at least one of the following and fulfilling criteria C and D:
 - 1. neck stiffness
 - 2. tinnitus
 - 3. hypacusia
 - 4. photophobia
 - 5. nausea
- B. One of the following:
 - 1. Dural puncture has been performed
 - 2. Persistent CSF leakage (CSF fistula) has been caused by another procedure or by trauma, or low CSF pressure has developed spontaneously¹, with at least one of the following:
 - a) evidence of low CSF pressure on MRI (eg, pachymeningeal enhancement)
 - b) evidence of CSF leakage on conventional myelography, CT myelography or cisternography

- c) CSF opening pressure <60 mm H₂O in sitting position
- C. Headache develops in close temporal relation to B1² or B2
- D. Headache resolves either spontaneously within 1 week³ or after intervention to seal the spinal fluid leak

Notes:

1. A history of trivial increase in intracranial pressure (eg, on vigorous coughing) is often elicited. In other cases a sudden drop in atmospheric pressure has occurred.
2. Headache may develop up to 5 days after lumbar puncture.
3. Post-lumbar-puncture headache often resolves spontaneously within 1 week. Persistence of headache beyond a week is likely to require specialist intervention whatever the aetiology.

7.4.1 [G44.822] Headache attributed to increased intracranial pressure or hydrocephalus caused by neoplasm

A history indicating raised intracranial pressure should first suggest intracranial neoplasm.

Diagnostic criteria:

- A. Diffuse non-pulsating headache with at least one of the following characteristics and fulfilling criteria C and D:
 1. associated with nausea and/or vomiting
 2. worsened by physical activity and/or manoeuvres known to increase intracranial pressure (such as Valsalva manoeuvre, coughing or sneezing)
 3. occurring in attack-like episodes¹
- B. Space-occupying intracranial tumour demonstrated by CT or MRI and causing hydrocephalus
- C. Headache develops and/or deteriorates in close temporal relation to the hydrocephalus
- D. Headache improves within 7 days after surgical removal or volume-reduction of tumour

Note:

1. Onset of headache can be sudden (thunderclap headache) and associated with loss of consciousness.

8.2 [G44.41 or G44.83] Medication-overuse headache (MOH)

Medication-overuse headache is an interaction between a therapeutic agent used excessively and a susceptible patient, usually one with migraine (most commonly) or tension-type headache. Such patients should be given both diagnoses: of the pre-existing headache and 8.2 *Medication-overuse headache*.

Medication overuse is defined in terms of duration and must, over that period, be both frequent and regular (eg, on 2 or more days each week).

The diagnosis of 8.2 *Medication-overuse headache* is clinically important because patients will not improve without withdrawal of the offending medication.

Diagnostic criteria:

- A. Headache present on ≥15 days/month fulfilling criteria C and D
- B. Regular overuse¹ for >3 months of one or more drugs that can be taken for acute and/or symptomatic treatment of headache²
- C. Headache has developed or markedly worsened during medication overuse
- D. Headache resolves or reverts to its previous pattern within 2 months after discontinuation of overused medication

Notes:

1. Overuse is defined as intake on ≥15 days/month for simple analgesics alone and *in all other cases* as intake on ≥10 days/month.

2. Drugs may be ergotamine, one or more triptans, simple analgesics, opioids, combination analgesics (typically containing simple analgesics plus opioids, butalbital and/or caffeine) or any combination of these.

CRANIAL NEURALGIAS, CENTRAL AND PRIMARY FACIAL PAIN

13.1.1 [G44.847] Classical trigeminal neuralgia

Description:

A disorder characterised by unilateral brief electric shock-like pains, abrupt in onset and termination, limited to the distribution of one or more divisions of the trigeminal nerve (usually the second or third). The pain often evokes spasm of the muscle of the face on the affected side (*tic douloureux*). Between paroxysms the patient is usually asymptomatic but a dull background pain may persist in long-standing cases.

The pains usually remit spontaneously for variable periods.

Diagnostic criteria:

- A. Paroxysmal attacks of unilateral pain¹ lasting from a fraction of a second to 2 minutes, affecting one or more divisions of the trigeminal nerve and fulfilling criteria B and C
- B. Pain has at least one of the following characteristics:
 1. intense, sharp, superficial or stabbing
 2. precipitated from trigger areas² or by trigger factors³
- C. Attacks are stereotyped in the individual patient
- D. There is no clinically evident neurological deficit
- E. Not attributed to another disorder⁴

Notes:

1. The pain never crosses to the opposite side but it may rarely occur bilaterally, in which case a central cause such as multiple sclerosis must be considered.
2. Small areas in the nasolabial fold and/or chin may be particularly susceptible to the precipitation of pain (trigger areas).
3. Pain frequently occurs spontaneously, but is commonly evoked by trivial stimuli (trigger factors) such as washing, shaving, smoking, talking and/or brushing the teeth.
4. History and physical and neurological examinations do not suggest any underlying and possibly causative disorder, or history and/or examination do suggest such disorder but it is ruled out by appropriate investigations, or such disorder is present but attacks do not occur for the first time in close temporal relation to it.

13.18.4 [G44.847] Persistent idiopathic facial pain

Description:

Persistent facial pain (formerly known as *Atypical facial pain*) without the characteristics of a cranial neuralgia and not attributable to another disorder (pain may be initiated by surgery or injury to the face, teeth or gums, but persists without any demonstrable local cause).

Diagnostic criteria:

- 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 face¹, 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

Note:

1. Pain at onset is commonly in the nasolabial fold or side of the chin, and may spread to the upper or lower jaw or a wider area of the face and neck.