

Diagnostic criteria

1. Migraine

Comment: If a patient fulfils criteria for more than one type of migraine all types should receive a diagnosis. This is in contrast to tension-type headache and cluster headache where the different types at any given time are mutually exclusive. If migraine occurs for the first time in close temporal relation to one of the disorders listed in groups 5-11 code to that group. If migraine is aggravated by 100 per cent or more (headache days) in close temporal relation to one of the disorders listed in groups 5-11, this may be mentioned in parenthesis, but the patient is still coded to group 1.

The terms common migraine and classic or classical migraine have been widely confused and convey no information. Therefore, they have been replaced by “migraine without aura” and “migraine with aura”. The aura is the complex of focal neurological symptoms which initiates or accompanies an attack. Most patients will exclusively have attacks without aura. It seems that patients who have frequent attacks with aura would usually also have attacks without aura (classify 1.2 and 1.1). Premonitory symptoms occur hours to a day or two before a migraine attack (with aura or without aura). They usually consist of hyperactivity, hypoactivity, depression, craving for special foods, repetitive yawning and similar atypical symptoms. The term prodromes has been used with different meanings, most often synonymous with aura. It should therefore not be used and the same is true of the ambiguous term “warning symptoms”.

1.1 Migraine without aura

Previously used terms: Common migraine, hemicrania simplex.

Description: Idiopathic, recurring headache disorder manifesting in attacks lasting 4-72 hours. Typical characteristics of headache are unilateral location, pulsating quality, moderate or severe intensity, aggravation by routine physical activity, and association with nausea, photo- and phonophobia.

Diagnostic criteria:

- A. At least 5 attacks fulfilling B-D.
- B. Headache attacks lasting 4-72 hours* (untreated or unsuccessfully treated).

*) In children below age 15, attacks may last 2-48 hours. If the patient falls asleep and wakes up without migraine, duration of attack is until time of awakening.

- C. Headache has at least two of the following characteristics:
 - 1. Unilateral location
 - 2. Pulsating quality
 - 3. Moderate or severe intensity (inhibits or prohibits daily activities)
 - 4. Aggravation by walking stairs or similar routine physical activity
- D. During headache at least one of the following:
 - 1. Nausea and/or vomiting
 - 2. Photophobia and phonophobia
- E. At least one of the following:
 - 1. History, physical- and neurological examinations do not suggest one of the disorders listed in groups 5-11
 - 2. History and/or physical- and/or neurological examinations do suggest such disorder, but it is ruled out by appropriate investigations
 - 3. Such disorder is present, but migraine attacks do not occur for the first time in close temporal relation to the disorder

Comment: The separation of migraine without aura from episodic tension-type headache may be difficult. Therefore at least 5 attacks are required. Patients rarely seek a doctor before they have had many attacks, and this requirement therefore probably excludes very few who should be coded 1.7. The mechanisms of the attack are as yet poorly understood. Regional cerebral blood flow remains normal or is perhaps slightly increased during an attack. Changes in blood composition and platelet function initiated endogenously or by environmental influences may play a triggering role. The pathophysiological process of the attack is presumed to occur in the brain, which via the trigemino-vascular and other systems interacts with intra- and extracranial vasculature and perivascular spaces. This form of migraine accounts for most cases debilitated by migraine.

Migraine without aura may occur almost exclusively at a particular time of the menstrual cycle - so-called menstrual migraine. Generally accepted criteria for this entity are not available. It seems reasonable to demand that 90 per cent of attacks should occur between two days before menses and the last day of menses, but further epidemiological knowledge is needed.

1.2 Migraine with aura*

Previously used terms: Classic migraine, classical migraine, ophthalmic, hemiparetic, hemiplegic- or aphasic migraine, migraine accompagnée, complicated migraine.

Description: Idiopathic, recurring disorder manifesting with attacks of neurological symptoms unequivocally localizable to cerebral cortex or brain stem, usually gradually developed over 5-20 minutes and usually lasting less than 60 minutes.

*) Aura as herein used does not necessarily imply that it precedes the headache, nor does it imply any relationship with epilepsy.

Headache, nausea and/or photophobia usually follow neurological aura symptoms directly or after a free interval of less than an hour. The headache usually lasts 4-72 hours, but may be completely absent (1.2.5).

Diagnostic criteria:

- A. At least 2 attacks fulfilling B.
- B. At least 3 of the following 4 characteristics:
 - 1. One or more fully reversible aura symptoms indicating focal cerebral cortical - and/or brain stem dysfunction.
 - 2. At least one aura symptom develops gradually over more than 4 minutes or, 2 or more symptoms occur in succession.
 - 3. No aura symptom lasts more than 60 minutes. If more than one aura symptom is present, accepted duration is proportionally increased.
 - 4. Headache follows aura with a free interval of less than 60 minutes. (It may also begin before or simultaneously with the aura).
- C. At least one of the following:
 - 1. History, physical- and neurological examinations do not suggest one of the disorders listed in groups 5-11
 - 2. History and/or physical- and/or neurological examinations do suggest such disorder, but it is ruled out by appropriate investigations
 - 3. Such disorder is present, but migraine attacks do not occur for the first time in close temporal relation to the disorder

Comment: Before or simultaneously with onset of aura symptoms regional cerebral blood flow is decreased corresponding to the clinically affected area and often including an even wider area. Blood flow reduction usually starts posteriorly and spreads anteriorly. It is above or at the ischemic threshold, but not infrequently below it. After one to several hours, gradual transition into hyperemia occurs in the same region. It has been reported that hyperemia is not related to headache, which usually begins during ischemia, and may disappear during hyperemia. Cortical arteriolar vasospasm and/or spreading depression of Leao have been implied. Relationship to the headache phase and mechanisms of the headache phase are uncertain (see comment to 1.1). The cerebral blood flow changes are not fully studied in all the subforms, but for several (1.2.1, 1.2.2, 1.2.3 and 1.2.5) there seems to be only quantitative differences. Systematic studies have demonstrated that most patients with visual auras occasionally have symptoms in the extremities. Conversely patients with symptoms in the extremities virtually always also suffer visual aura symptoms. A distinction between ophthalmic migraine and hemiparesthetic/hemiparetic migraine is therefore probably artificial and is not recognized in this classification.

1.2.1 Migraine with typical aura

Previously used terms: Ophthalmic-, hemiparesthetic-, hemiparetic-, hemiplegic- or aphasic migraine, migraine accompagnée.

Description: Migraine with an aura consisting of homonymous visual disturbances, hemisensory symptoms, hemiparesis or dysphasia or combinations thereof. Gradual development, duration under one hour and complete reversibility characterize the aura which is associated with headache.

Diagnostic criteria:

- A. Fulfils criteria for 1.2 including all four criteria under B.
- B. One or more aura symptoms of the following types:
 - 1. Homonymous visual disturbance
 - 2. Unilateral paresthesias and/or numbness
 - 3. Unilateral weakness
 - 4. Aphasia or unclassifiable speech difficulty

Comment: This is the commonest form of migraine with aura, and the diagnosis is evident after a careful history alone. Visual aura is most common, usually as a fortification spectrum i.e. a star shaped figure near the point of fixation gradually spreading right or left and assuming a laterally convex shape with angulated scintillating edge leaving a variable degree of absolute or relative scotoma in its wake. In other cases it is a scotoma without positive phenomena which often is perceived as being of acute onset, but on scrutiny enlarges gradually. 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 the one side of the body and face. Numbness occurs in its wake, but numbness may also be the only symptom. Less frequent are speech disturbances, usually dysphasia but often hard to categorize, and unilateral weakness. Symptoms usually follow one another in succession beginning with visual, followed by sensory symptoms, dysphasia and weakness, but the reverse and other orders have been noted. Patients often find it hard to describe their symptoms in which case they should be instructed how to time and record their symptoms. After such prospective observation the clinical picture often becomes more clear. Common mistakes are incorrectly reported lateralization of headache, report of sudden onset when it is gradual, of monocular visual disturbances which are homonymous, as well as incorrect duration of aura.

1.2.2 Migraine with prolonged aura

Previously used terms: Complicated migraine, hemiplegic migraine.

Description: Migraine with one or more aura symptoms lasting more than 60 minutes and less than a week. Neuroimaging is normal.

Diagnostic criteria:

- A. Fulfils criteria for 1.2, but at least one symptom lasts more than 60 minutes and ≤ 7 days.
If neuroimaging reveals relevant ischemic lesion, code 1.6.2 migrainous infarction regardless of symptom duration.

Comment: Rare patients have only this form. The majority who experience prolonged aura have it rarely and intermingled with much more frequent attacks of typical aura. Prolonged acute onset aura is difficult to separate from TIA or small strokes and not sufficiently validated.

1.2.3 Familial hemiplegic migraine

Description: Migraine with aura including hemiparesis and where at least one first degree relative has identical attacks.

Diagnostic criteria:

- A. Fulfils criteria for 1.2.
- B. The aura includes some degree of hemiparesis and may be prolonged.
- C. At least one first degree relative has identical attacks.

Comment: This disorder probably has the same pathophysiology as migraine with typical aura. The reason for still keeping it separate is that families have been described where attacks are strikingly identical and sometimes long lasting. The term familial hemiplegic migraine has been abused since in most families different forms of migraine occur, and most patients with hemiplegic attacks have these intermingled with more frequent attacks of migraine without hemiparesis.

1.2.4 Basilar migraine

Previously used terms: Basilar artery migraine, Bickerstaff's migraine, syncopal migraine.

Description: Migraine with aura symptoms clearly originating from the brain stem or from both occipital lobes.

Diagnostic criteria:

- A. Fulfils criteria for 1.2.
- B. Two or more aura symptoms of the following types:
 - Visual symptoms in both the temporal and nasal fields of both eyes
 - Dysarthria
 - Vertigo
 - Tinnitus
 - Decreased hearing
 - Double vision
 - Ataxia
 - Bilateral paresthesias
 - Bilateral pareses
 - Decreased level of consciousness

Comment: Many of the symptoms listed under diagnostic criteria are subject to misinterpretation as they may occur with anxiety and hyperventilation.

Originally the term basilar artery migraine was used, but since spasm of the basilar artery may not be the mechanism of the attacks, the term basilar migraine should be preferred. Many cases have basilar attacks intermingled with attacks with typical aura. Basilar attacks are mostly seen in young adults.

1.2.5 Migraine aura without headache

Previously used terms: Migraine equivalents, acephalgic migraine.

Description: Migrainous aura unaccompanied by headache.

Diagnostic criteria:

- A. Fulfils criteria for 1.2.
- B. No headache.

Comment: It is common for migraine with aura that headache occasionally is absent. As patients get older headache may disappear completely even if auras continue. It is less common to have always suffered exclusively from migraine aura without headache. When the onset occurs after the age of forty and for other reasons the distinction between this entity and thromboembolic transient ischemic attacks may be difficult and require extensive investigation. Acute onset aura without headache is not sufficiently validated.

1.2.6 Migraine with acute onset aura

Description: Migraine with aura developing fully in less than 5 minutes.

Diagnostic criteria:

- A. Fulfils criteria for 1.2.
- B. Neurological symptoms develop within 4 minutes.
- C. Headache lasts 4-72 hours (untreated or unsuccessfully treated).
- D. Headache has at least two of the following characteristics:
 - 1. Unilateral location
 - 2. Pulsating quality
 - 3. Moderate or severe intensity (inhibits or prohibits daily activities)
 - 4. Aggravation by walking stairs or similar routine physical activity
- E. During headache at least one of the following:
 - 1. Nausea and/or vomiting
 - 2. Photophobia and phonophobia
- F. Thromboembolic TIA and other intracranial lesion ruled out by appropriate investigations.

Comment: Inaccurate history is the most common explanation of acute onset aura. Acute onset should be confirmed by repeated close questioning and preferably by prospective observation. Presence of a typical headache phase is required and the diagnosis is supported by previous migraine attacks of other type or a strong fami-

ly history. Extensive investigations are usually necessary to rule out thromboembolic TIA.

1.3. Ophthalmoplegic migraine

Description: Repeated attacks of headache associated with paresis of one or more ocular cranial nerves in the absence of demonstrable intracranial lesion.

Diagnostic criteria:

- A. At least 2 attacks fulfilling B.
- B. Headache overlapping with paresis of one or more of cranial nerves III, IV, and VI.
- C. Parasellar lesion ruled out by appropriate investigations.

Comment: Whether ophthalmoplegic migraine in fact has anything to do with migraine is uncertain since the headache often lasts for a week or more. Association with other forms of migraine has often been noted, but a relationship to the Tolosa-Hunt syndrome has also been suggested. The condition is extremely rare.

1.4 Retinal migraine

Description: Repeated attacks of monocular scotoma or blindness lasting less than an hour and associated with headache. Ocular or structural vascular disorder must be ruled out.

Diagnostic criteria:

- A. At least 2 attacks fulfilling B-C.
- B. Fully reversible monocular scotoma or blindness lasting less than 60 minutes and confirmed by examination during attack or (after proper instruction) by patient's drawing of monocular field defect during an attack.
- C. Headache follows visual symptoms with a free interval of less than 60 minutes, but may precede them.
- D. Normal ophthalmological examination outside of attack. Embolism ruled out by appropriate investigations.

Comment: The monocular nature of the visual disturbances has been documented in only a few cases. Some cases without headache have been reported, but their migrainous nature cannot be ascertained.

1.5 Childhood periodic syndromes that may be precursors to or associated with migraine

Previously used terms: Migraine equivalents

Comment: It is not possible to propose criteria for delineation of the multiple

heterogeneous and undefined disorders comprised under the terms periodic syndromes, abdominal migraine and cyclical vomiting, and it is unlikely that any progress will be made in this uncertain area until markers are found. At the present time therefore these syndromes of childhood cannot be included in the classification despite the generally accepted view that some presentations are indeed headache free “equivalents” of migraine.

1.5.1 Benign paroxysmal vertigo of childhood

Description: This probably heterogeneous disorder is characterized by brief attacks of vertigo in otherwise healthy children.

Diagnostic criteria:

- A. Multiple, brief, sporadic episodes of disequilibrium, anxiety, and often nystagmus or vomiting.
- B. Normal neurological examination.
- C. Normal electroencephalogram.

1.5.2 Alternating hemiplegia of childhood

Description: Infantile attacks of hemiplegia involving each side alternately. Is associated with other paroxysmal phenomena and mental impairment.

Diagnostic criteria:

- A. Onset before 18 months of age.
- B. Repeated attacks of hemiplegia involving both sides of the body.
- C. Other paroxysmal phenomena, such as tonic spells, dystonic posturing, choreoathetoid movements, nystagmus or other ocular motor abnormalities, autonomic disturbances associated with the bouts of hemiplegia or occurring independently.
- D. Evidence of mental or neurological deficits.

Comment: The nature of the disorder is not clear. A relationship with migraine is suggested on clinical grounds. The possibility that the disorder is an unusual form of epilepsy cannot be ruled out.

Benign recurrent vertigo in adults has been regarded as a migraine equivalent, but is not sufficiently validated.

1.6 Complications of migraine (code for previous migraine type plus the complication)

1.6.1 Status migrainosus

Description: Attack of migraine with headache phase lasting more than 72 hours despite treatment. Headache free intervals of less than 4 hours (sleep not included) may occur.

Diagnostic criteria:

- A. The patient fulfils criteria for 1.1 or 1.2.
- B. The present attack fulfils criteria for one form of migraine except that *headache* lasts more than 72 hours whether treated or not.
- C. Headache is continuous throughout the attack or interrupted by headache free intervals lasting less than 4 hours. Interruption during sleep is disregarded.

Comment: Migraine with prolonged aura and headache lasting > 72 hours is coded 1.2.2, ophthalmoplegic migraine is coded 1.3. Status migrainosus is usually associated with prolonged drug use. See also group 8.

1.6.2 Migrainous infarction

Previously used terms: Complicated migraine

Description: One or more migrainous aura symptoms not fully reversible within 7 days and/or associated with neuroimaging confirmation of ischemic infarction.

Diagnostic criteria:

- A Patient has previously fulfilled criteria for 1.2.
- B. The present attack is typical of previous attacks, but neurological deficits are not completely reversible within 7 days and/or neuroimaging demonstrates ischemic infarction in relevant area.
- D. Other causes of infarction ruled out by appropriate investigations.

Comment: No causal relationship has been established between migraine without aura and cerebral infarction. Ischemic stroke in a migraine sufferer may be categorized as a) cerebral infarction of other cause coexisting with migraine, b) cerebral infarction of other cause presenting with symptoms resembling migraine or c) cerebral infarction occurring during the course of a typical migraine attack. Applying strict criteria, only category (c) should be coded as migrainous infarction. In so doing it is recognized that patients with stereotyped migrainous aura may suffer, although rarely, cerebral infarction possibly due to migraine mechanisms during the course of migraine-like symptoms that are not typical for that patient. Nevertheless, because of uncertainty as to mechanism, cerebral infarction under those circumstances should be designated under category b. Increased risk for stroke in migraine patients has not been found in population based studies indicating that stroke is a rare complication of migraine.

1.7 Migrainous disorder not fulfilling above criteria

Description: Headache attacks which are believed to be a form of migraine, but which do not quite meet the operational diagnostic criteria for any of the forms of migraine

Diagnostic criteria:

- A. Fulfils all criteria but one for one or more forms of migraine (specify type(s)).
- B. Does not fulfil criteria for tension-type headache.

Comment: Patients who do not have sufficient numbers of otherwise typical attacks to fulfil criteria should be coded here as should patients with sufficient numbers of attacks, which fulfil all criteria but one.

Cyclic migraine, lower half headache, facial migraine, hemicrania continua and cervical migraine are not sufficiently validated.

Coexisting migraine and tension-type headache – A comment

Previously used terms: Mixed headache, tension-vascular headache, combination headache.

Migraine and tension-type headache often coexist in the same patient. Previously the diagnosis “combination headache” has been used, but it has never been defined. Patients represent a continuum varying from those having pure migraine over those with migraine and moderate amounts of tension-type headache, those with half of each, those with preponderance of tension-type headache to those with pure tension-type headache. The concept of combination headache is therefore arbitrary, and it has been judged impossible to single out a suitable group of patients who should receive this diagnosis. Patients should instead be coded for migraine and for tension-type headache if they have both forms. Since it is a general rule that number of headache days per year should be given in brackets after each diagnosis, the evaluation of the relative importance of the two conditions is easy.

If a patient has attacks/episodes each of which fulfil criteria for migraine without aura and for episodic tension type headache, the general rule applies according to which the attacks should be coded as the type listed first in this classification, i.e. migraine without aura.

2. Tension-type headache

Previously used terms: Tension headache, muscle contraction headache, psychomyogenic headache, stress headache, ordinary headache, essential headache, idiopathic headache and psychogenic headache.

Comment: If tension-type headache occurs for the first time in close temporal relation to one of the disorders listed in groups 5-11 code to that group. If tension-type headache is aggravated by 100 per cent or more (headache days) in close temporal relation to one of the disorders listed in groups 5-11, it is still coded to group 2. The aggravating factor may be coded for using the fourth digit 9. At a given time a patient can only have one form of tension-type headache. Another type may have been present before. The subgrouping of tension-type headache into an episodic and a chronic form is introduced because patients with daily or almost daily headache constitute a large group in specialized practices and hospital clinics. Their treatment and perhaps also their pathogenic mechanisms vary to a considerable extent from the episodic form.

The subgrouping in forms with and without a muscular factor is a novel creation. For decades dispute has prevailed concerning the importance of muscle contraction mechanisms, but conclusive studies are still lacking. The classification committee believes that the diagnostic subdivision according to presence or absence of a muscular factor will stimulate research in this field. In view of the poor scientific basis for the subdivision, it should, however, be regarded as optional.

2.1. Episodic tension-type headache

Previously used terms: See above

Description: Recurrent episodes of headache lasting minutes to days. The pain is typically pressing/tightening in quality, of mild or moderate intensity, bilateral in location and does not worsen with routine physical activity. Nausea is absent, but photophobia or phonophobia may be present.

Diagnostic criteria:

- A. At least 10 previous headache episodes fulfilling criteria B-D listed below.
Number of days with such headache < 180/year (< 15/month).
- B. Headache lasting from 30 minutes to 7 days

- C. At least 2 of the following pain characteristics:
 - 1. Pressing/tightening (non-pulsating) quality
 - 2. Mild or moderate intensity (may inhibit, but does not prohibit activities)
 - 3. Bilateral location
 - 4. No aggravation by walking stairs or similar routine physical activity
- D. Both of the following:
 - 1. No nausea or vomiting (anorexia may occur)
 - 2. Photophobia and phonophobia are absent, or one but not the other is present
- E. At least one of the following:
 - 1. History, physical- and neurological examinations do not suggest one of the disorders listed in groups 5-11
 - 2. History and/or physical- and/or neurological examinations do suggest such disorder, but it is ruled out by appropriate investigations
 - 3. Such disorder is present, but tension-type headache does not occur for the first time in close temporal relation to the disorder

Comment: The exact mechanisms of tension-type headache are not known. Involuntary tightening in muscles induced mentally or physically is important as are purely psychogenic mechanisms.

2.1.1 Episodic tension-type headache associated with disorder of pericranial muscles

Previously used terms: Muscle contraction headache

Description: Episodic tension-type headache with increased levels of tenderness and/or EMG of pericranial muscles.

Diagnostic criteria:

- A. Fulfils criteria for 2.1.
- B. At least one of the following:
 - 1. Increased tenderness of pericranial muscles demonstrated by manual palpation or pressure algometer
 - 2. Increased EMG level of pericranial muscles at rest or during physiological tests

Comment: There is not yet sufficient evidence available regarding the limits of normality of pericranial muscle tenderness. Neither has sufficient attention been given to the methodology of pericranial palpation. Evidence concerning normal EMG levels of pericranial muscles is similarly deficient. Until evidence accumulates concerning tenderness on palpation and pericranial EMG, each investigator must judge as best he can on the basis of experience with non-headache sufferers and by comparing symmetrical sites. Estimation of tenderness by palpation is evidently subject to large bias. Reliable quantitation requires experience and

systematic approach. Then, judgement of tenderness is no more subjective than other elements of the sensory neurological examination. For research purposes blinding of the observer remains mandatory.

2.1.2 Episodic tension-type headache unassociated with disorder of pericranial muscles

Previously used terms: Idiopathic headache, essential headache, psychogenic headache

Description: Episodic tension-type headache with normal levels of tenderness and/or EMG of pericranial muscles.

Diagnostic criteria:

- A. Fulfilling criteria for 2.1.
- B. No increased tenderness of pericranial muscles. If studied, EMG of pericranial muscles shows normal levels of activity.

Comment: It is not known how often episodic tension-type headache is unassociated with pericranial muscle tenderness. That such cases exist is on the other hand well known. Mechanisms of headache are unknown in such cases, but psychogenic etiologies are suspected.

2.2 Chronic tension-type headache

Previously used terms: Chronic daily headache. See also p. 29.

Description: Headache present for at least 15 days a month during at least 6 months. The headache is usually pressing/tightening in quality, mild or moderate in severity, bilateral and does not worsen with routine physical activity. Nausea, photophobia or phonophobia may occur.

Diagnostic criteria:

- A. Average headache frequency ≥ 15 days/month (180 days/year) for ≥ 6 months fulfilling criteria B–D listed below.
- B. At least 2 of the following pain characteristics:
 - 1. Pressing/tightening quality
 - 2. Mild or moderate severity (may inhibit, but does not prohibit activities)
 - 3. Bilateral location
 - 4. No aggravation by walking stairs or similar routine physical activity
- C. Both of the following:
 - 1. No vomiting
 - 2. No more than one of the following:
Nausea, photophobia or phonophobia
- D. At least one of the following:

1. History, physical- and neurological examinations do not suggest one of the disorders listed in groups 5-11
2. History and/or physical- and/or neurological examinations do suggest such disorder, but it is ruled out by appropriate investigations
3. Such disorder is present, but tension-type headache does not occur for the first time in close temporal relation to the disorder

Comment: Sometimes migraine is gradually transformed into chronic tension-type headache, but more frequently it is the episodic tension-type headache which becomes chronic. In both instances overuse of drugs frequently plays a role in aggravating the disorder. Discontinuation of daily drug intake often results in improvement.

2.2.1 Chronic tension-type headache associated with disorder of pericranial muscles
Previously used terms: Chronic muscle contraction headache

Description: Chronic tension-type headache associated with increased levels of tenderness and/or EMG of pericranial muscles.

Diagnostic criteria:

- A. Fulfils criteria for 2.2.
- B. At least one of the following:
 1. Increased tenderness of pericranial muscles demonstrated by manual palpation or using pressure algometer
 2. Increased EMG level of pericranial muscles at rest or during physiological tests

2.2.2 Chronic tension-type headache unassociated with disorder of pericranial muscles

Previously used terms: Chronic idiopathic headache, chronic psychogenic headache

Description: Chronic tension-type headache with normal levels of tenderness and/or EMG of pericranial muscles

Diagnostic criteria:

- A. Fulfils criteria for 2.2.
- B. No increased tenderness of pericranial muscles. If studied, EMG of pericranial muscles shows normal levels of activity.

2.3. Headache of the tension-type not fulfilling above criteria

Description: Headache which is believed to be a form of tension-type headache, but which does not quite meet the operational diagnostic criteria for any of the forms of tension-type headache.

Diagnostic criteria:

- A. Fulfils all but one criterion for one or more forms of tension-type headache (specify type(s)).
- B. Does not fulfil criteria for migraine without aura.

Comment: Coded to this number are cases who have had < 10 typical episodes of tension-type headache or with many episodes which fail one of the criteria. Also patients who are not chronic, but who have episodes lasting longer than 7 days or with headache for more than 15 days a month which has not yet lasted for 6 months.

Fourth digit code number for group 2 indicates most likely causative factor(s). If third digit has not been coded, a zero should be inserted prior to the fourth digit.

- 0. No identifiable causative factor
- 1. More than one of the factors 2-9 (list in order of importance)
- 2. Oromandibular dysfunction

Previously used terms: Myofacial pain-dysfunction syndrome, temporo-mandibular-joint pain dysfunction syndrome, Costen's syndrome, cranio-mandibular dysfunction.

Diagnostic criteria:

Three or more of the following: temporo-mandibular joint noise on jaw movements, limited or jerky jaw movements, pain on jaw function, locking of jaw on opening, clenching of teeth, gnashing of teeth (bruxism), other oral parafunction (tongue, lips or cheek biting or pressing).

Comment: Tenderness of pericranial muscles was part of the previously used syndromes. This is not logical and has not been used in this classification, since tenderness may be part of a generalized muscle hyperactivity or caused by other factors not related to the function of the mandible or the temporo-mandibular joint. It was therefore necessary to create the new term "oromandibular dysfunction".

- 3. Psychosocial stress (DSM III-R criteria)

Diagnostic criteria:

Associated with psychosocial stressors rated 4-6 on a 1-6 scale (1 no stress, 2 mild, 3 moderate, 4 severe, 5 extreme, 6 catastrophic).

Comment: In rating, pay attention to the amount of change in the individual's life caused by the stressor, the degree to which the change is desired and under the individual's control, and to the number of stressors. Stressors may be grouped as fol-

lows: conjugal (marital and non-marital), parenting, other interpersonal, occupational, living circumstances, financial, legal, developmental (for children), physical disorder or injury, other.

4. Anxiety

Fulfilling DSM III-R criteria for one of the anxiety disorders

5. Depression

Fulfilling DSM III-R criteria for one of the depressive disorders

6. Headache as a delusion or an idea

Previously used terms: Psychogenic headache, conversion cephalgia

Diagnostic criteria:

Fulfilling DSM III-R criteria for somatic delusion or somatoform disorder

Comment: The previously used term psychogenic headache is now coded as 2.1.2.6 or 2.2.2.6 i.e. episodic- or chronic tension-type headache unassociated with a muscular factor but associated with somatic delusion or somatoform disorder.

7. Muscular stress

Associated with at least one of the following types of muscular stress: Unphysiological working position, long lasting tonic muscular contraction for other reasons, lack of rest and/or sleep

8. Drug overuse for tension-type headaches

Associated with one or more of the following:

Monthly weak analgesics exceeding 45 g of aspirin or equivalent

Morphinomimetic drugs more than twice a month

Monthly diazepam exceeding 300 mg or equivalent of other benzodiazepines

9. One of the disorders listed in groups 5-11 of this classification (specify). Use the fourth digit code only when preexisting tension-type headache is aggravated by 100 per cent or more (headache days) in close temporal relation to the organic disorder. If headache occurs for the first time in close temporal relation to the organic disorder code to group 5-11

3. Cluster headache and chronic paroxysmal hemicrania

Comment: If cluster headache or chronic paroxysmal hemicrania occur for the first time in close temporal relation to one of the disorders listed in groups 5-11 code to that group. If cluster headache or chronic paroxysmal hemicrania are aggravated by 100 per cent or more (headache days) in close temporal relation to one of the disorders listed in groups 5-11, this may be mentioned in parenthesis, but the patient is still coded to group 3. At a given time a patient can only have one type of cluster headache. Another type may have been present before.

Cluster headache and chronic paroxysmal hemicrania share the following characteristics: 1. the unilaterality of the pain 2. the severe intensity of the pain 3. the location of the pain 4. the accompanying autonomic phenomena 5. the temporal pattern of the attacks. Similarities also exist with regard to the course of the diseases (episodic or chronic pattern/stage) and other changes indicating autonomic involvement. A number of features, however, distinguish between the two: sex preponderance, frequency and duration of attacks, night preponderance, drug effects (both symptomatic and prophylactic).

So-called cluster variants and combined forms are not included in the present classification as they are considered not to be sufficiently validated.

Cluster-like headaches have occasionally been reported in patients presenting with evidence of cephalic (intra or extracranial) lesions of vascular or neoplastic type. The relationship of these lesions to the pathogenesis of cluster headache is at present not clear.

3.1 Cluster headache

Previously used terms: Erythroprosopalgia of Bing, ciliary or migrainous neuralgia (Harris), erythromelalgia of the head, Horton's headache, histaminic cephalalgia, petrosal neuralgia (Gardner), sphenopalatine, Vidian and Sluder's neuralgia, hemicrania periodica neuralgiformis

Description: Attacks of severe strictly unilateral pain orbitally, supraorbitally and/or temporally, lasting 15-180 minutes and occurring from once every other day to 8 times a day. Are associated with one or more of the following: conjunctival injection, lacrimation, nasal congestion, rhinorrhea, forehead and facial sweating, miosis, ptosis, eyelid edema. Attacks occur in series lasting for weeks or months (so-called cluster periods) separated by remission periods usually lasting months or years. About 10 per cent of the patients have chronic symptoms.

Diagnostic criteria:

- A. At least 5 attacks fulfilling B-D.
- B. Severe unilateral orbital, supraorbital and/or temporal pain lasting 15 to 180 minutes untreated.
- C. Headache is associated with at least one of the following signs which have to be present on the pain-side:
 - 1. Conjunctival injection
 - 2. Lacrimation
 - 3. Nasal congestion
 - 4. Rhinorrhea
 - 5. Forehead and facial sweating
 - 6. Miosis
 - 7. Ptosis
 - 8. Eyelid edema
- D. Frequency of attacks: from 1 every other day to 8 per day.
- E. At least one of the following:
 - 1. History, physical- and neurological examinations do not suggest one of the disorders listed in groups 5-11
 - 2. History and/or physical- and/or neurological examinations do suggest such disorder, but it is ruled out by appropriate investigations
 - 3. Such disorder is present, but cluster headache does not occur for the first time in close temporal relation to the disorder

3.1.1 Cluster headache periodicity undetermined

- A. Criteria for 3.1 fulfilled
- B. Too early to classify as 3.1.2 or 3.1.3

3.1.2 Episodic cluster headache

Description: Occurs in periods lasting 7 days to one year separated by pain free periods lasting 14 days or more

Diagnostic criteria:

- A. All the letter headings of 3.1.
- B. At least 2 periods of headaches (cluster periods) lasting (untreated patients) from 7 days to one year, separated by remissions of at least 14 days.

Comment: Cluster periods usually last between 2 weeks and 3 months.

3.1.3 Chronic cluster headache

Description: Attacks occur for more than one year without remission or with remissions lasting less than 14 days.

Diagnostic criteria:

- A. All letter headings of 3.1.

- B. Absence of remission phases for one year or more or with remissions lasting less than 14 days.

3.1.3.1 Chronic cluster headache unremitting from onset

Previously used term: Primary chronic

Diagnostic criteria:

- A. All letter headings of 3.1.3.
- B. Absence of remission periods lasting 14 days or more from onset.

3.1.3.2 Chronic cluster headache evolved from episodic

Previously used term: Secondary chronic

Diagnostic criteria:

- A. All letter headings of 3.1.3.
- B. At least one interim remission period lasting 14 days or more within one year after onset, followed by unremitting course for at least one year.

Comment: During a cluster period and in patients with the chronic form attacks occur regularly and may be provoked by alcohol, histamine or nitroglycerine. Pain is maximal orbitally, supraorbitally and/or temporally, but may spread to other regions. Pain usually recurs on the same side of the head during an individual cluster period. During the worst attacks, the intensity of pain is excruciating. Patients are unable to lie down and typically pace the floor. Age at onset is typically 20-40 years. For unknown reasons men are afflicted 5-6 times more often than women. The mechanisms of the pain are incompletely known despite abnormalities demonstrated by studies of corneal indentation pulse, corneal temperature, forehead sweating, lacrimation and nasal secretion or by pupillometry, thermovision, extracranial and transcranial Doppler.

3.2 Chronic paroxysmal hemicrania

Previously used terms: Sjaastad's syndrome

Description: Attacks with largely the same characteristics of pain and associated symptoms and signs as cluster headache, but they are shorter lasting, more frequent, occur mostly in females, and there is absolute effectiveness of indomethacin.

Diagnostic criteria:

- A. At least 50 attacks fulfilling B-E.
- B. Attacks of severe unilateral orbital, supraorbital and/or temporal pain always on the same side lasting 2 to 45 minutes.
- C. Attack frequency above 5 a day for more than half of the time (periods with lower frequency may occur).

- D. Pain is associated with at least one of the following signs/symptoms on the pain side:
 - 1. Conjunctival injection
 - 2. Lacrimation
 - 3. Nasal congestion
 - 4. Rhinorrhea
 - 5. Ptosis
 - 6. Eyelid edema
- E. Absolute effectiveness of indomethacin (150 mg/day or less).
- F. At least one of the following:
 - 1. History, physical- and neurological examinations do not suggest one of the disorders listed in groups 5-11
 - 2. History and/or physical- and/or neurological examinations do suggest such disorder, but it is ruled out by appropriate investigations
 - 3. Such disorder is present, but chronic paroxysmal hemicrania does not occur for the first time in close temporal relation to the disorder

Comment: Most attacks last 5-20 minutes and frequency may be as high as 30 per 24 hours. Although longer lasting remissions are not seen in chronic paroxysmal hemicrania, frequency, duration and severity of the attacks may vary. Nausea and vomiting rarely accompany the attacks. There is great female predominance. Onset is usually in adulthood. The chronic stage may probably be preceded by an episodic stage similar to the pattern seen in cluster headache, but this has not yet been sufficiently validated.

3.3 Cluster headache-like disorder not fulfilling above criteria

Description: Headache attacks which are believed to be a form of cluster headache or chronic paroxysmal hemicrania, but which do not quite meet the operational diagnostic criteria for any of the forms of cluster headache or chronic paroxysmal hemicrania.

Diagnostic criteria:

- A. Fulfilling all but one of the criteria for 3.1 or 3.2.

Comment: Coded to this number are patients who do not have sufficient numbers of otherwise typical attacks as well as patients who do have enough attacks which, however, fail to fulfil one of the other criteria.

Cluster migraine and cluster tic syndromes are not sufficiently validated.

4. Miscellaneous headaches unassociated with structural lesion

4.1 Idiopathic stabbing headache

Previously used term: Ice-pick pains

Description: Transient stabs of pain in the head that occur spontaneously in the absence of organic disease of underlying structures or of the cranial nerves.

Diagnostic criteria:

- A. Pain confined to the head and exclusively or predominantly felt in the distribution of the first division of the trigeminal nerve (orbit, temple and parietal area).
- B. Pain is stabbing in nature and lasts for a fraction of a second. Occurs as single stabs or series of stabs.
- C. It recurs at irregular intervals (hours to days).
- D. Diagnosis depends upon the exclusion of structural changes at the site of pain and in the distribution of the affected cranial nerve.

Comments: Stabbing pains are more commonly experienced by people subject to migraine headache, in which case they are felt in the site habitually affected by headache in about 40 per cent of patients and tend to be more frequent at the time of headache. They commonly subside with the administration of indomethacin 25 mg orally three times daily.

4.2 External compression headache

Previously used term: Swim-goggle headache

Description: Headache resulting from continued stimulation of cutaneous nerves by the application of pressure, for example by a band around the head, a tight hat or goggles worn for the protection of eyes during swimming training.

Diagnostic criteria:

- A. Results from the application of external pressure in the forehead or the scalp.
- B. Is felt in the area subjected to pressure.
- C. Is a constant pain.
- D. Is prevented by avoiding the precipitating cause.
- E. Is not associated with organic cranial or intracranial disease.

Comment: External compression may lead to a more severe migrainous headache if the stimulus is prolonged.

4.3 Cold stimulus headache

Description: Headache resulting from the exposure of the head to low temperatures.

4.3.1 External application of a cold stimulus

Description: Generalized headache following exposure of the unprotected head to a low environmental temperature as in sub-zero weather or in diving into cold water.

Diagnostic criteria:

- A. Develops during external exposure to cold.
- B. Is bilateral.
- C. Varies in intensity with the severity and duration of the cold stimulus.
- D. Is prevented by avoiding exposure to cold.
- E. Is not associated with organic cranial- or intracranial disease.

4.3.2 Ingestion of a cold stimulus

Previously used term: Ice-cream headache

Description: Ice-cream headache is a pain produced in susceptible individuals by the passage of cold material, solid or liquid, over the palate and posterior pharyngeal wall.

Diagnostic criteria:

- A. Develops during ingestion of a cold food or drink.
- B. Lasts for less than five minutes.
- C. Is felt in the middle of the forehead, except in people subject to migraine, in which case the pain may be referred to the area habitually affected by migraine headache (code migraine first).
- D. Is prevented by avoiding the rapid swallowing of cold food or drinks.
- E. Is not associated with organic disease.

4.4 Benign cough headache

Description: Headache precipitated by coughing in the absence of any intracranial disorder.

Diagnostic criteria:

- A. Is a bilateral headache of sudden onset, lasting less than one minute, precipitated by coughing.
- B. May be prevented by avoiding coughing.

- C. May be diagnosed only after structural lesions such as posterior fossa tumour have been excluded by neuroimaging.

4.5 Benign exertional headache

Description: Headache precipitated by any form of exercise. Subvarieties, such as “weight-lifters headache” are recognised.

Diagnostic criteria:

- A. Is specifically brought on by physical exercise.
- B. Is bilateral, throbbing in nature at onset and may develop migrainous features in those patients susceptible to migraine (code for migraine first).
- C. Lasts from 5 minutes to 24 hours.
- D. Is prevented by avoiding excessive exertion, particularly in hot weather or at high altitude.
- E. Is not associated with any systemic or intracranial disorder.

Comment: Exertional headache is prevented in some patients by the ingestion of ergotamine tartrate, methysergide, propranolol or indomethacin before exercise.

4.6 Headache associated with sexual activity

Previously used terms: Benign sex headache, coital cephalalgia

Description: Headache precipitated by masturbation or coitus, usually starting as a dull bilateral ache while sexual excitement increases and suddenly becoming intense at orgasm, in the absence of any intracranial disorder.

Diagnostic criteria:

- A. Is precipitated by sexual excitement.
- B. Is bilateral at onset.
- C. Is prevented or eased by ceasing sexual activity before orgasm.
- D. Is not associated with any intracranial disorder such as aneurysm.

4.6.1 Dull type

Diagnostic criteria:

A dull ache in the head and neck that intensifies as sexual excitement increases.

4.6.2 Explosive type

Diagnostic criteria:

A sudden severe (“explosive”) headache occurring at orgasm.

4.6.3 Postural type

Diagnostic criteria:

Postural headache resembling that of low CSF pressure developing after coitus.

Fourth digit code number for groups 5-11

Types of headache

Comment: Included are as many forms of headache as can be distinguished using only headache characteristics and operational criteria. Most forms are also found elsewhere in this classification.

0. Headache is as described in the diagnostic criteria for the particular disorder
Comment: For some disorders in groups 5-11 headache characteristics are not part of the diagnostic criteria, for some they are. Fourth digit 0 applies only to the latter
1. Migraine
 Fulfilling criteria for 1.1 or 1.2 with the exception that migraine occurs for the first time in close temporal relation to one of the disorders listed in groups 5-11
2. Tension-type headache
 Fulfilling criteria for 2.1 or 2.2 with the exception that tension-type headache occurs for the first time in close temporal relation to one of the disorders listed in groups 5-11
3. Cluster headache
 Fulfilling criteria for 3.1 og 3.2 with the exception that cluster headache or chronic paroxysmal hemicrania occur for the first time in close temporal relation to one of the disorders listed in groups 5-11
4. Increased intracranial pressure type (prototype: brain tumour headache)
 - A. Crescendo time profile over 3 months or less
 - B. Moderate or severe intensity of pain
 - C. Occurs in the morning or after napping and remits or improves spontaneously after getting up
 - D. Is present at least 50 per cent of all mornings
5. Decreased intracranial pressure type (prototype: post-lumbar puncture headache)
 - A. Bilateral
 - B. Absent or mild in the recumbent position, occurs or worsens markedly in the upright position
6. Local lesion type (prototype: pain from bone metastasis)
 - A. Headache is non-pulsating and constantly present
 - B. Pain has a distinct maximum in a circumscribed area of 5 cm or less, but may irradiate to surroundings or refer to more distant areas
7. Vasodilator type (prototype: nitroglycerin, histamine and prostacyclin induced headache)

- A. Bifronto-temporal pulsating pain
- B. No aura, nausea or vomiting
- 8. Stabbing type (ice-pick type)
 - A. Stabbing head pains lasting less than a second
 - B. Occur as single stabs or series of stabs
 - C. Each stab or series of stabs occur in a small, sharply localized area.
- 9. Other type (specify)
- 10. Two or more types (specify)

5. Headache associated with head trauma

Comment: Worsening of preexisting headache is coded according to preexisting headache form. Patients who develop a new form of headache (including migraine, tension-type headache or cluster headache) in close temporal relation (specified below) to a head trauma are coded to group 5. Type of headache may be specified with the fourth digit (see p. 42). Traumatic intracranial hematoma is coded to group 6, posttraumatic hydrocephalus to group 7.

A causal relation between head trauma and headache is frequent with 5.1.1, 5.1.2 and 5.2.1 and infrequent with 5.2.2. There remains, however, a group of patients especially with acceleration/deceleration injury who do not meet criterion 5.2.1 A, but who display an abrupt decline in work performance and/or social functioning or change in personality following head trauma which indicates a possible causal relationship between headache, these symptoms and trauma. Chronic post-traumatic headache (5.2.1 and 5.2.2) is often part of the post-traumatic syndrome. The complex inter-relationship between organic and psychosocial factors in these syndromes is difficult to assess.

5.1 Acute post-traumatic headache

5.1.1 With significant head trauma and/or confirmatory signs

Diagnostic criteria:

- A. Significance of head trauma documented by at least one of the following:
 - 1. Loss of consciousness
 - 2. Posttraumatic amnesia lasting more than 10 minutes
 - 3. At least two of the following exhibit relevant abnormality: clinical neurological examination, X-ray of skull, neuroimaging, evoked potentials, spinal fluid examination, vestibular function test, neuropsychological testing
- B. Headache occurs less than 14 days after regaining consciousness (or after trauma, if there has been no loss of consciousness).
- C. Headache disappears within 8 weeks after regaining consciousness (or after trauma, if there has been no loss of consciousness).

5.1.2 With minor head trauma and no confirmatory signs

Diagnostic criteria:

- A. Head trauma that does not satisfy 5.1.1 A.

- B. Headache occurs less than 14 days after injury.
- C. Headache disappears within 8 weeks after injury.

5.2 Chronic post-traumatic headache

5.2.1 With significant head trauma and/or confirmatory signs

Diagnostic criteria:

- A. Significance of head trauma documented by at least one of the following:
 - 1. Loss of consciousness
 - 2. Posttraumatic amnesia lasting more than 10 minutes
 - 3. At least two of the following exhibit relevant abnormality: clinical neurological examination, X-ray of skull, neuroimaging, evoked potentials, spinal fluid examination, vestibular function test, neuropsychological testing
- B. Headache occurs less than 14 days after regaining consciousness (or after trauma, if there has been no loss of consciousness).
- C. Headache continues more than 8 weeks after regaining consciousness (or after trauma, if there has been no loss of consciousness).

5.2.2 With minor head trauma and no confirmatory signs

Diagnostic criteria:

- A. Head trauma that does not satisfy 5.2.1 A
- B. Headache occurs less than 14 days after injury.
- C. Headache continues more than 8 weeks after injury.

6. Headache associated with vascular disorders

Comment: Worsening of preexisting headache is coded according to preexisting headache form. Patients who develop a new form of headache (including migraine, tension-type headache and cluster headache) in close temporal relation to a vascular disorder are coded to group 6. A causal relationship is not necessarily implied, however. Type of headache may be specified with the fourth digit (see p. 42)

All headaches fulfil the following criteria, (more specific criteria are given under subforms).

Diagnostic criteria:

- A. Symptoms and/or signs of vascular disorder.
- B. Appropriate investigations indicate the vascular disorder.
- C. Headache as a new symptom or of a new type occurs in close temporal relation to onset of vascular disorder.

Comment: Headache persisting more than 1 month after successful treatment or spontaneous recovery usually has other mechanisms

6.1 Acute ischemic cerebrovascular disease

Diagnostic criteria:

- A. Focal central nervous system symptoms and/or signs developed within 48 hours.
- B. Appropriate investigations indicate acute ischemic cerebrovascular disease.

Comment: A few acute ischemic strokes take more than 48 hours to develop. Headache usually begins with the onset of acute ischemic cerebrovascular disease, but occasionally precedes the stroke by up to two weeks and rarely begins as long as two weeks after the stroke.

6.1.1 Transient ischemic attack (TIA) - Symptoms clear within 24 hours

6.1.2 Thromboembolic stroke - symptoms persist more than 24 hours

Comment: Appropriate investigations usually reveal the presence of risk factors for atherosclerosis or evidence of cardiac or cervical arterial origin for emboli. Hematologic defects such as sickle cell anemia, polycythemia vera, thrombotic thrombocytopenic purpura, and dysglobulinemia may also be found.

6.2 Intracranial hematoma

6.2.1 Intracerebral hematoma (including traumatic parenchymal hematoma)

Diagnostic criteria:

- A. Focal central nervous system symptoms and/or signs developed within 24 hours.
- B. Intracerebral hematoma diagnosed by appropriate investigations.

Comment: As for 6.1. Hematologic defects mentioned under 6.1.2 may also cause intracerebral hematoma.

6.2.2 Subdural hematoma

6.2.3 Epidural hematoma

6.3 Subarachnoid hemorrhage

Diagnostic criteria:

- A. Present or past subarachnoid bleeding demonstrated by CSF examination or CT. (If hematoma is present code 6.2).
- B. Headache of sudden onset (less than 60 minutes) if it is aneurysm, less than 12 hours if it is an arteriovenous malformation.
- C. At least one of the following:
 - 1. Severe headache intensity
 - 2. Bilateral headache location
 - 3. Stiff neck
 - 4. Increased body temperature

Comment: Although onset of headache is sudden, it may increase gradually in intensity after onset. Comatose patients may appear to develop headache gradually as they wake up.

6.4 Unruptured vascular malformation (if malformation ruptures code 6.3)

6.4.1 Arteriovenous malformation

Diagnostic criteria:

Unruptured arteriovenous malformation diagnosed by CT, MR and/or angiography.

Comment: The relationship of migraine and other headaches to this condition is poorly substantiated.

6.4.2 Saccular aneurysm

Diagnostic criteria:

Unruptured saccular aneurysm diagnosed by CT, MR and/or angiography.

Comment: The relationship of migraine and other headache to this condition is poorly substantiated (except giant aneurysms), but a major headache may be a warning of impending rupture.

Moya Moya causes headache when it ruptures resulting in intracerebral and/or subarachnoid bleeding. A relation between headache and unruptured Moya Moya is not substantiated.

6.5 Arteritis

6.5.1. Giant cell arteritis

Previously used terms: Temporal arteritis, Horton's disease.

Diagnostic criteria:

- A. One or more of the following:
 - 1. Swollen and tender scalp artery (usually superficial temporal artery)
 - 2. Elevated RBC sedimentation rate
 - 3. Disappearance of headache within 48 hours of steroid therapy
- B. Temporal artery biopsy demonstrating giant cell arteritis.

Comment: The headache usually involves one or both temporal regions, is moderate or severe, and polymyalgia rheumatica is often associated. Jaw claudication is almost pathognomonic, but uncommon. Onset is after age 50 in virtually all cases.

6.5.2 Other systemic arteritides: Headache with evidence of systemic arteritis

6.5.3 Primary intracranial arteritis: Headache with angiographic signs of arteritis or biopsy demonstrating arteritis

6.6 Carotid or vertebral artery pain

6.6.1 Carotid or vertebral dissection

Diagnostic criteria:

- A. At least one of the following:
 - 1. TIA or ischemic stroke in territory of affected artery
 - 2. Horner's syndrome, arterial bruit or tinnitus
- B. Dissection demonstrated by appropriate investigations or surgery.
- C. Headache and cervical pain ipsilateral to arterial dissection.

6.6.2 Carotidynia (idiopathic)

Diagnostic criteria:

- A. At least one of the following overlying the carotid artery:

1. Tenderness
2. Swelling
3. Increased pulsations
- B. Appropriate investigations do not reveal structural abnormality.
- C. Pain over the affected side of the neck. May project to ipsilateral side of the head.
- D. A self-limiting syndrome of less than 2 weeks duration.

Comment: Organic disease of the carotid artery (such as giant cell arteritis, atherosclerotic thrombosis, intraluminal hemorrhage, fibromuscular dysplasia, aneurysm and aneurysmal dissection) may cause a similar clinical picture. Eagle's syndrome manifested by pain in the throat and/or neck said to be caused by an elongated styloid process of the temporal bone, has not been validated as a specific entity.

6.6.3 Post endarterectomy headache

Diagnostic criteria:

- A. Thrombo-endarterectomy or other surgery of the extracranial carotid artery.
- B. Patent carotid artery without dissection as demonstrated by appropriate investigations.
- C. Headache begins within 2 days after surgery and is ipsilateral.

Comment: The headache usually disappears after days, but may persist for months.

6.7 Venous thrombosis

Diagnostic criteria:

- A. At least one of the following:
 1. Raised intracranial pressure
 2. Focal neurological deficit
 3. Seizures
- B. Venous occlusion demonstrated by appropriate investigations.
- C. Headache is located to affected area or is diffuse.

6.8 Arterial hypertension

Comment: Chronic arterial hypertension of mild or moderate degree does not cause headache (see references).

6.8.1 Acute pressor response to exogenous agent

Diagnostic criteria:

- A. Headache occurs with acute rise (> 25 per cent) of diastolic blood pressure.
- B. Evidence of appropriate toxin or medication.
- C. Headache disappears within 24 hours after normalization of blood pressure.

6.8.2 Pheochromocytoma*Diagnostic criteria:*

- A. Headache occurs with acute rise (> 25 per cent) of diastolic blood pressure.
- B. At least 1 of the following:
 - 1. Sweating
 - 2. Palpitations
 - 3. Anxiety
- C. Pheochromocytoma proved by biological *and* imaging tests or surgery.
- D. Headache disappears within 24 hours after normalization of blood pressure.

6.8.3 Malignant (accelerated) hypertension (including hypertensive encephalopathy)*Diagnostic criteria:*

- A. Headache associated with grade 3 or 4 retinopathy (Keith Wagner classification).
- B. Diastolic blood pressure persistently above 120 mm Hg.
- C. Appropriate investigations rule out vasopressor toxins, medication or pheochromocytoma as causative factors.
- D. Headache is temporally related to rise in blood pressure and disappears within 2 days after reduction of blood pressure. If hypertensive encephalopathy is present, headache may persist up to 7 days after reduction of blood pressure.

6.8.4 Pre-eclampsia and eclampsia

- A. Headache during pregnancy.
- B. Edema or proteinuria and blood pressure rise from pre-pregnant level (not necessarily markedly increased, but at least mean elevation of 15 mm Hg or diastolic of 90 mm Hg).
- C. Appropriate investigations rule out vasopressor toxins, medication or pheochromocytoma as causative factors.
- D. Headache occurs with rise in blood pressure and disappears within 7 days after blood pressure reduction or after termination of pregnancy.

6.9 Headache associated with other vascular disorder

7. Headache associated with non-vascular intracranial disorder

Comment: Worsening of preexisting headache is coded according to preexisting headache form. Patients who develop a new form of headache (including migraine, tension-type headache or cluster headache) in close temporal relation to a non-vascular intracranial disorder are coded to group 7. A causal relationship is not necessarily implied, however. Type of headache may be specified with the fourth digit (p. 42).

All headaches fulfil the following criteria (more specific criteria are given under subforms).

Diagnostic criteria:

- A. Symptoms and/or signs of intracranial disorder.
- B. Confirmation by appropriate investigations.
- C. Headache as a new symptom or of a new type occurs temporally related to intracranial disorder.

Comment: Headache persisting more than 1 month after successful treatment or spontaneous recovery usually has other mechanisms.

7.1. High cerebrospinal fluid pressure

7.1.1 Benign intracranial hypertension

Previously used terms: Pseudotumour cerebri, otitic hydrocephalus

Diagnostic criteria:

- A. Patient suffers from benign intracranial hypertension fulfilling the following criteria:
 - 1. Increased intracranial pressure (> 200 mm of water) measured by epidural or intraventricular pressure monitoring or by lumbar puncture
 - 2. Normal neurological examination except for papilledema and possible VI nerve palsy
 - 3. No mass lesion and no ventricular enlargement on neuroimaging
 - 4. Normal or low protein concentration and normal white cell count in CSF
 - 5. No clinical or neuroimaging suspicion of venous sinus thrombosis
- B. Headache intensity and frequency related to variations of intracranial pressure with a time lag of less than 24 hours.

7.1.2 High pressure hydrocephalus including posttraumatic high pressure hydrocephalus

Diagnostic criteria:

- A. Patient fulfils the following criteria for high pressure hydrocephalus:
 - 1. Ventricular enlargement on neuroimaging
 - 2. Intracranial pressure > 200 mm of water
- B. Headache occurs with increased intracranial pressure, and is improved or abolished by reduction of intracranial pressure with a time lag of less than 24 hours.

Comment: Normal pressure hydrocephalus has not been validated as a cause of headache.

7.2 Low cerebrospinal fluid pressure

7.2.1 Post-lumbar puncture headache

Diagnostic criteria:

- A. Bilateral headache developed less than 7 days after lumbar puncture.
- B. Headache occurs or worsens less than 15 minutes after assuming the upright position, and disappears or improves less than 30 minutes after resuming the recumbent position.
- C. Headache disappears within 14 days after lumbar puncture (if duration exceeds 14 days, consider 7.2.2).

7.2.2 Cerebrospinal fluid fistula headache

Diagnostic criteria:

- A. Posttraumatic, postoperative or idiopathic cerebrospinal fluid leak demonstrated by measurement of glucose concentration in leaking fluid, or by leakage of spinally injected dye or radioactive tracer.
- B. Headache characteristics as for 7.2.1.
- C. Headache disappears within 14 days after effective treatment of fistula.

7.3 Intracranial infection

(Meningitis, encephalitis, brain abscess, subdural empyema)

7.4 Intracranial sarcoidosis and other non-infectious inflammatory diseases

7.5 Headache associated with intrathecal injections

7.5.1 Direct effect (specify agent)

Diagnostic criteria:

- A. Headache follows intrathecal injection within 4 hours.
- B. Headache is diffuse and present also in the recumbent position.
- C. Headache clears completely within 14 days. (If it persists consider 7.2.2)

7.5.2 Due to chemical meningitis (specify agent)

Diagnostic criteria:

- A. Headache follows intrathecal injection within 5-72 hours.
- B. Headache is diffuse and present also in the recumbent position.
- C. Cerebrospinal fluid pleocytosis with negative culture.

7.6 Intracranial neoplasm

7.7 Headache associated with other intracranial disorder

8. Headache associated with substances or their withdrawal

Comment: Worsening of preexisting headache is coded according to preexisting headache form. Patients who develop a new form of headache (including migraine, tension-type headache or cluster headache) in close temporal relation to substance use or substance withdrawal as specified below are coded to group 8. Type of headache may be specified with the fourth digit (p. 42). Effective doses and temporal relationships have not yet been determined for most substances.

8.1 Headache induced by acute substance use or exposure

Comment: To establish that a substance really induces headache, double blind placebo controlled experiments are necessary. This has been clearly demonstrated by two studies of patients who reported headaches after dark chocolate or aspartame respectively. In both studies headache was equally frequent after placebo.

Diagnostic criteria:

- A. Occurs within a specified time after substance intake.
- B. A certain required minimum dose should be indicated.
- C. Has occurred in at least 1/2 of exposures and at least 3 times.
- D. Disappears when substance is eliminated or within a specified time thereafter.

8.1.1 Nitrate/nitrite induced headache

Previously used term: Hot dog headache

Diagnostic criteria:

Occurs within 1 hour after absorption of nitrate/nitrite

8.1.2 Monosodium glutamate induced headache

Previously used term: Chinese restaurant syndrome

Diagnostic criteria:

- A. Occurs within 1 hour after ingestion of monosodium glutamate.
- B. Is associated with at least two of the other symptoms of this syndrome:
 - 1. Pressure in chest
 - 2. Pressure and tightness in face
 - 3. Burning sensation in chest, neck or shoulders
 - 4. Flushing of face

5. Dizziness
6. Abdominal discomfort

8.1.3 Carbon monoxide induced headache

8.1.4 Alcohol induced headache

Diagnostic criteria:

Occurs within three hours after ingestion of alcohol

8.1.5 Other substances (specify)

8.2 Headache induced by chronic substance use or exposure

Diagnostic criteria:

- A. Occurs after daily doses of a substance for ≥ 3 months.
- B. A certain required minimum dose should be indicated.
- C. Headache is chronic (15 days or more a month).
- D. Headache disappears within 1 month after withdrawal of the substance.

Comment: So far headache induced by chronic use of ergotamine and analgesics has only been described when the drugs have been taken for a headache disorder, not when they have been taken for other disorders.

8.2.1 Ergotamine induced headache

Diagnostic criteria:

- A. Is preceded by daily ergotamine intake (oral ≥ 2 mg, rectal ≥ 1 mg)
- B. Is diffuse, pulsating and distinguished from migraine by absent attack pattern and/or absent associated symptoms.

Comment: The diagnosis can often only be made after withdrawal of ergotamine resulting in relief from ergotamine induced headache (but usually not from the primary headache).

8.2.2 Analgesics abuse headache

Diagnostic criteria:

One or more of the following:

1. ≥ 50 g of aspirin a month or equivalent of other mild analgesics
2. ≥ 100 tablets a month of analgesics combined with barbiturates or other non-narcotic compounds
3. One or more narcotic analgesics

Comment: This diagnosis can only be made after withdrawal of substance resulting in relief from substance induced headache (but usually not from the primary headache).

8.2.3 Other substances

8.3 Headache from substance withdrawal (acute use)

Diagnostic criteria:

- A. Follows acute use of a substance.
- B. A certain required minimum dose should be indicated.
- C. Occurs when the substance is largely or completely eliminated, but may last longer.
- D. Is relieved or improved by renewed intake of the substance.

8.3.1 Alcohol withdrawal headache (hangover)

Diagnostic criteria:

Is preceded by intake of sufficient alcohol to make the particular individual drunk.

8.3.2 Other substances

8.4 Headache from substance withdrawal (chronic use)

Diagnostic criteria:

- A. Occurs after use of a high daily dose (specified when possible under each substance) of a substance for ≥ 3 month.
- B. Occurs within hours after elimination of the substance.
- C. Is relieved by renewed intake of the substance.
- D. Headache disappears within 14 days after withdrawal of the substance.

8.4.1 Ergotamine withdrawal headache

Diagnostic criteria:

- A. Is preceded by daily ergotamine intake (oral ≥ 2 mg, rectal ≥ 1 mg)
- B. Occurs within 48 hours after withdrawal of ergotamine.

8.4.2 Caffeine withdrawal headache

Diagnostic criteria:

- A. Patient has consumed caffeine daily and ≥ 15 g a month.
- B. Occurs within 24 hours after last caffeine intake.
- C. Is relieved within 1 hour by 100 mg of caffeine.

8.4.3 Narcotics abstinence headache

8.4.4 Other substances (specify)

8.5 Headache associated with substances but with uncertain mechanism

8.5.1 Birth control pills or estrogens

Comment: The literature on this subject is conflicting. Further study is needed.

8.5.2 Other substances (specify)

9. Headache associated with non-cephalic infection

Comment: Worsening of preexisting headache is coded according to preexisting headache form. Patients who develop a new form of headache (including migraine, tension-type headache or cluster headache) in close temporal relation to a non-cephalic infection are coded to group 9. Type of headache may be specified with the fourth digit (p. 42). Headache occurring in association with *intracranial* infection is coded to group 7, cephalic infection to group 11.

All headaches fulfil the following criteria (more specific criteria are given under subforms).

Diagnostic criteria:

- A. Symptoms and/or signs of non-cephalic infection.
- B. Laboratory diagnosis of systemic or focal non-cephalic infection.
- C. Headache as a new symptom or of a new type occurs concomitantly with infection.
- D. Headache disappears less than 1 month after successful treatment or spontaneous remission of infection.

Comment: Headaches persisting after 1 month usually have other mechanisms and should be coded accordingly or to group 13.

9.1. Viral infection

Diagnostic criteria:

Clinical and laboratory (serology, microscopy or culture) diagnosis of viral infection.

9.1.1. Focal non-cephalic

Diagnostic criteria:

Infection localized to one or a few organs outside of the head.

9.1.2 Systemic

Diagnostic criteria:

Infection is systemic.

9.2 Bacterial infection

Diagnostic criteria:

Clinical and laboratory (serology, microscopy or culture) diagnosis of bacterial infection.

9.2.1 Focal non-cephalic

Diagnostic criteria:

Infection is localized to one or a few organs outside of the head.

9.2.2 Systemic (septicemia)

Diagnostic criteria:

Infection is systemic.

9.3. Headache related to other infection (specify)

10. Headache associated with metabolic disorder

Comment: Worsening of preexisting headache is coded according to preexisting headache form. Patients who develop a new form of headache (including migraine, tension-type headache or cluster headache) in close temporal relation to a metabolic disorder are coded to group 10. Type of headache may be specified with the fourth digit (p. 42).

All headaches fulfil the following criteria (more specific criteria are given under subforms).

Diagnostic criteria:

- A. Symptoms and/or signs of metabolic disorder.
- B. Confirmation by laboratory investigations when specified under the subform.
- C. Headache intensity and/or frequency related to variations in metabolic disorder with a specified time lag.
- D. Headache disappears within 7 days after normalization of metabolic state.

Comment: Headache persisting more than 7 days after the metabolic disorder has been effectively treated or has remitted spontaneously usually has other mechanisms and should be coded accordingly or to group 13.

10.1 Hypoxia

10.1.1 High altitude headache

Diagnostic criteria:

- A. Occurs within 24 hours after sudden ascent to altitudes above 3,000 m.
- B. Is associated with at least one other symptom typical of high altitude:
 - 1. Cheyne-Stokes respiration at night
 - 2. Desire to overbreathe
 - 3. Exertional dyspnea

10.1.2 Hypoxic headache (low pressure environment, pulmonary disease causing hypoxia)

Diagnostic criteria:

Occurs within 24 hours after acute onset of hypoxia with $\text{PaO}_2 \leq 70$ mm Hg or in chronic hypoxic patients with PaO_2 persistently at or below this level.

10.1.3 Sleep apnoea headache

10.2 Hypercapnia*Diagnostic criteria:*

Arterial pCO₂ increased above 50 mm Hg in the absence of hypoxia.

10.3 Mixed hypoxia and hypercapnia**10.4 Hypoglycemia***Diagnostic criteria:*

Blood glucose reduced below 2.2 mmol/l

10.5 Dialysis*Diagnostic criteria:*

- A. Onset during hemodialysis and termination within 24 hours after dialysis.
- B. Has occurred during at least half of hemodialyses and at least three times.
- C. Can be prevented by changing dialysis parameters.

10.6 Headache related to other metabolic abnormality

Syndromes not sufficiently validated: Ischemic headache (anemia, arterial hypotension, cardiac disease), fasting without hypoglycemia, plasmapheresis induced headache

11. Headache or facial pain associated with disorder of cranium, neck, eyes, ears, nose, sinuses, teeth, mouth or other facial or cranial structures

Comment: Worsening of preexisting headache is coded according to preexisting headache form. Patients who develop a new form of headache (including migraine, tension-type headache or cluster headache) in close temporal relation to a disorder of cranium, neck, eyes, ears etc. are coded to group 11. Type of headache may be specified with the fourth digit (p. 42). Many disorders may cause headache as well as facial pain or neuralgic pain. In this section only non-neuralgic headache and facial pain are considered. All headaches fulfil the following criteria (more specific criteria are given under subforms).

Diagnostic criteria:

- A. Clinical and/or laboratory evidence of disorder in cranium, neck etc. (specify).
- B. Headache located to the affected facial or cranial structure and radiating to surroundings. Pain may or may not be referred to more distant areas of the head.
- C. Headache disappears within 1 month after successful treatment or spontaneous remission of the underlying disorder.

Comment: Headache persisting after 1 month usually has other mechanisms and should be coded accordingly or to group 13.

11.1 Cranial bone

Comment: Most disorders of the skull e.g. congenital abnormalities, fractures, tumours, metastases, are usually not accompanied by headache. Exceptions of importance are osteomyelitis, multiple myeloma and Paget's disease.

11.2 Neck

Comment: Cervicogenic headache not fulfilling 11.2.1 or 11.2.2 is not sufficiently validated.

11.2.1 Cervical spine

Diagnostic criteria:

- A. Pain localized to neck and occipital region. May project to forehead, orbital region, temples, vertex or ears.
- B. Pain is precipitated or aggravated by special neck movements or sustained neck posture.
- C. At least one of the following:

1. Resistance to or limitation of passive neck movements
 2. Changes in neck muscle contour, texture, tone or response to active and passive stretching and contraction
 3. Abnormal tenderness of neck muscles
- D. Radiological examination reveals at least one of the following:
1. Movement abnormalities in flexion/extension
 2. Abnormal posture
 3. Fractures, congenital abnormalities, bone tumours, rheumatoid arthritis or other distinct pathology (not spondylosis or osteochondrosis)

Comment: Cervical headaches are associated with movement abnormalities in cervical intervertebral segments. The disorder may be located in the joints or ligaments. The abnormal movement may occur in any component of intervertebral movement, and is manifest during either active or passive examination of the movement.

11.2.2 Retropharyngeal tendinitis

Description: Constant pain in the back of the neck and head developed to maximum within 3 weeks. Pain is aggravated by retroflexion of neck. Swelling of prevertebral soft tissue on X-ray and full recovery after treatment with non-steroidal antiinflammatory drugs are features in every case.

Diagnostic criteria:

- A. Pain in the back of the neck radiating to the back of the head or to the whole head.
- B. Pain is non-pulsating, uni- or bilateral and aggravated severely by bending head backwards.
- C. Prevertebral soft tissues in adults measure more than 7 mm at the level between C 1 and C 4 (special X-ray technique may be required).
- D. Alleviation within 2 weeks of treatment with non-steroidal antiinflammatory drugs in recommended doses.

Comment: Body temperature and erythrocyte sedimentation rate are usually elevated. Although retroflexion most consistently aggravates pain, this also usually happens with rotation and swallowing. The transverse processes of the upper 3 vertebrae are usually tender to palpation. In several cases amorphous calcific material has been aspirated from the swollen prevertebral tissues. Thin calcifications in prevertebral tissues are best seen on CT.

11.3 Eyes

11.3.1 Acute glaucoma

Diagnostic criteria:

- A. Acute glaucoma diagnosed by appropriate investigations.

- B. Pain in the eye and behind or above it.

11.3.2 Refractive errors

Diagnostic criteria:

- A. Uncorrected refractive errors e.g. hypermetropia, astigmatism, presbyopia, wearing of incorrect glasses.
- B. Mild headaches in the frontal region and in the eyes themselves.
- C. Pain absent on awakening, and aggravated by prolonged visual tasks at the distance or angle where vision is impaired.

11.3.3 Heterophoria or heterotropia (latent or manifest squint)

Diagnostic criteria:

- A. Heterophoria or heterotropia is demonstrated.
- B. Mild to moderate constant headache in the frontal region.
- C. At least one of the following:
 - 1. Headache occurs or worsens during a visual task especially when tiring
 - 2. Intermittent blurred vision or diplopia
 - 3. Difficulty adjusting focus from near to distant objects or vice versa
- D. Relief or improvement of symptoms by closing one eye.

Comment: Uncorrected refractive errors and heterophoria may cause headaches but their importance is widely overestimated.

11.4 Ears

Comment: Disorders of the middle ear may cause neuralgic pain by irritation of neural structures (see group 12).

11.5 Nose and sinuses

11.5.1 Acute sinus headache

Diagnostic criteria:

- A. Purulent discharge in the nasal passage either spontaneous or by suction.
- B. Pathological findings in one or more of the following tests:
 - 1. X-ray examination
 - 2. Computerized tomography or magnetic resonance imaging
 - 3. Transillumination
- C. Simultaneous onset of headache and sinusitis.
- D. Headache location
 - 1. In acute frontal sinusitis headache is located directly over the sinus and may radiate to the vertex or behind the eyes
 - 2. In acute maxillary sinusitis headache is located over the antral area and may radiate to the upper teeth or to the forehead
 - 3. In acute ethmoiditis headache is located between and behind the eyes and may radiate to the temporal area

4. In acute sphenoiditis headache is located in the occipital area, the vertex, the frontal region or behind the eyes
- E. Headache disappears after treatment of acute sinusitis.

11.5.2 Other diseases of nose or sinuses

Comment: Other conditions which may cause headache are nasal passage abnormality due to septal deflection, hypertrophic turbinates and atrophic sinus membrane are not sufficient validated as a cause of headache. Chronic sinusitis is not validated as a cause of headache or facial pain unless relapsing into an acute stage. Postoperative chronic pain due to nerve damage see group 12. Migraine and tension-type headache are often confused with true sinus headache because of similarity in location. In order to diagnose sinus headache the above criteria must be strictly fulfilled.

11.6 Teeth, jaws and related structures

Comment: Disorders of the teeth usually cause facial pain, and the conditions causing headache are rare. Pain from the teeth may be referred, however, and cause diffuse headache. The most common cause of headache is periodontitis or pericoronitis as the result of infection or traumatic irritation around a partially erupted lower wisdom tooth.

11.7 Temporomandibular joint disease

Diagnostic criteria:

- A. At least two of the following:
 1. Pain of the jaw precipitated by movement and/or clenching
 2. Decreased range of movement
 3. Noise during joint movements
 4. Tenderness of the joint capsule
- B. Positive X-ray and/or isotope scintigraphic findings.
- C. Pain is mild to moderate and located to the temporomandibular joint and/or radiating from there.

Comment: Pain from temporomandibular joints or from related tissues is common, but it is rarely due to definable organic disease. Moreover, when organic disease such as rheumatoid arthritis is present, significant pain or impairment of function is relatively uncommon. By far the most frequent cause of pain appearing to come from this joint is myofascial due to oromandibular dysfunction described in group 2, fourth digit number 2.

12. Cranial neuralgias, nerve trunk pain and deafferentation pain

12.1 Persistent (in contrast to tic-like) pain of cranial nerve origin

Diagnostic criteria:

- A. Pain in the distribution of one or more cranial nerve(s) and/or cervical roots 2 and 3 with or without projection to neighbouring areas.
- B. Demonstration of a relevant lesion.
- C. Onset of pain temporally related to onset of cranial nerve lesion.
- D. If lesion can be effectively treated or remits spontaneously, pain improves or disappears.

Comment: The following general criteria apply to all following diagnoses under 12.1. More specific requirements and exceptions are given under subforms.

12.1.1 Compression or distortion of cranial nerves and second or third cervical roots

Description: Headache or facial pain caused by a lesion directly compromising one or more of the afferent nerves supplying pain sensation to the head and neck (namely the trigeminal, nervus intermedius, glossopharyngeal, vagus and cervical roots 2 and 3).

Comment: Structural lesions may be space-occupying e.g. tumour, aneurysm, or contained within anatomical boundaries e.g. osteomyelitis of the cranial bones.

12.1.2 Demyelination of cranial nerves

12.1.2.1 Optic neuritis (retrobulbar neuritis)

Description: Pain behind one eye accompanied by impairment of central vision caused by demyelination of the optic nerve.

Diagnostic criteria:

- A. Pain is felt behind the affected eye.
- B. Central vision becomes impaired due to a central or paracentral scotoma.
- C. No extrinsic lesion can be demonstrated.

Comment: Pain may precede impairment of vision by hours or days, sometimes as long as four weeks.

12.1.3 Infarction of cranial nerves

12.1.3.1 Diabetic neuritis

Description: Pain around the eye and forehead associated with an oculomotor (third cranial nerve) palsy of diabetic origin.

Diagnostic criteria:

- A. Pain is felt around the eye on the affected side.
- B. Pain is of acute onset, developing over a few hours.
- C. There is a partial or complete oculomotor nerve palsy.
- D. The patient suffers from diabetes mellitus.
- E. No extrinsic lesion can be demonstrated.

Comment: In diabetic neuropathy (oculomotor nerve infarction) the pupil is usually spared.

12.1.4 Inflammation of cranial nerves

12.1.4.1 Herpes zoster

Description: Facial pain caused by acute herpes zoster.

Diagnostic criteria:

- A. Pain is followed by a herpetic eruption in the distribution of the nerve affected within one week of onset.
- B. Pain subsides within 6 months after onset of the rash.

Comment: Herpes zoster affects the trigeminal ganglion in 10-15 per cent of patients with the ophthalmic division being singled out in some 80 per cent of those patients. Herpes zoster may also involve the geniculate ganglion, causing an eruption in the external auditory meatus. The soft palate or distribution of upper cervical roots may be involved in some patients. Ophthalmic herpes may be associated with third, fourth and sixth cranial nerve palsies and geniculate herpes by facial palsy or acoustic symptoms. Zoster occurs in about 10 per cent of patients with lymphoma and 25 per cent of patients with Hodgkin's disease.

12.1.4.2 Chronic post-herpetic neuralgia

Description: Facial pain developing during the acute phase of herpes zoster and persisting more than 6 months thereafter.

Diagnostic criteria:

- A. Pain is restricted to the distribution of the affected cranial nerves or divisions thereof.
- B. Pain persists more than 6 months after the onset of herpetic eruption.

Comment: Post-herpetic neuralgia is a sequel of herpes zoster more often as age advances, afflicting 50 per cent of patients over the age of 60 years. Scars resulting from the eruption, often anaesthetic, can usually be seen in the affected area.

12.1.5 Tolosa-Hunt syndrome

Description: Episodic orbital pain associated with paralysis of one or more of the

third, fourth or sixth cranial nerves which resolves spontaneously but may relapse and remit.

Diagnostic criteria:

- A. Episode or episodes of unilateral orbital pain for an average of eight weeks if untreated.
- B. Association with paralysis of one or more of the third, fourth and sixth cranial nerves which may coincide with the onset of the pain or follow it by a period of up to two weeks.
- C. Pain is relieved within 72 hours after initiation of corticosteroid therapy.
- D. Exclusion of other causative lesions by neuroimaging and (not compulsory) carotid angiography.

Comment: Some reported cases of Tolosa-Hunt syndrome had additional involvement of the trigeminal nerve (commonly the first division) or optic, facial or acoustic nerves. Sympathetic innervation of the pupil is occasionally affected. The syndrome has been caused by granulomatous material in some biopsied cases but the etiology is unknown. The sites affected are the cavernous sinus, superior orbital fissure or orbit.

Demonstration of obstruction of the superior ophthalmic vein, poor filling of the cavernous sinus and collateral venous flow can be demonstrated by orbital phlebography in more than 50 per cent of cases.

Gradenigo's syndrome and Raeder's paratrigeminal neuralgia are not specific diagnoses, but syndromes indicating a particular location of intracranial pathology.

12.1.6 Neck-tongue syndrome

Diagnostic criteria:

- A. Pain and numbness in area of distribution of the lingual nerve and second cervical root.
- B. Precipitation by sudden turning of the head.

Comment: Proprioceptive fibres from the tongue enter the central nervous system through the second cervical dorsal root via connections between lingual and hypoglossal nerves and between the latter and the second cervical root. There is clinical and operative evidence that C2 root is compromised by sudden rotation of the neck, particularly likely if subluxation of the atlanto-axial joint occurs.

12.1.7 Other causes of persistent pain of cranial nerve origin (specify)

12.2 Trigeminal neuralgia

Previously used term: Tic douloureux

12.2.1 Idiopathic trigeminal neuralgia

Description: Trigeminal neuralgia is a painful unilateral affliction of the face,

characterized by brief electric shock-like (lancinating) pains limited to the distribution of one or more divisions of the trigeminal nerve. Pain is commonly evoked by trivial stimuli including washing, shaving, smoking, talking and brushing the teeth, but may also occur spontaneously. The pain is abrupt in onset and termination and may remit for varying periods.

Diagnostic criteria:

- A. Paroxysmal attacks of facial or frontal pain which last a few seconds to less than two minutes.
- B. Pain has at least 4 of the following characteristics:
 - 1. Distribution along one or more divisions of the trigeminal nerve
 - 2. Sudden, intense, sharp, superficial, stabbing or burning in quality
 - 3. Pain intensity severe
 - 4. Precipitation from trigger areas, or by certain daily activities such as eating, talking, washing the face or cleaning the teeth
 - 5. Between paroxysms the patient is entirely asymptomatic
- C. No neurological deficit.
- D. Attacks are stereotyped in the individual patient.
- E. Exclusion of other causes of facial pain by history, physical examination and special investigations when necessary.

Comment: Pain is limited strictly to some part of the distribution of the trigeminal nerve. It usually starts in the second or third divisions, affecting the cheek or the chin, less than 5 per cent in the first division. The pain never crosses to the opposite side but it may occur bilaterally - approximately in 3-5 per cent. The pain often reflexly evokes spasm of the muscle of the face on the affected side (tic douloureux). The increasing frequency of posterior fossa exploration has demonstrated that many, possibly the majority, of patients with this condition have compression of the trigeminal root by tortuous or aberrant vessels. It is suggested that patients in whom such compression has been documented at operation and who respond favourably to surgery be transferred to category 12.2.2.

12.2.2 Symptomatic trigeminal neuralgia

12.2.2.1 Compression of trigeminal root or ganglion

Description: Pain indistinguishable from trigeminal neuralgia, caused by a demonstrable structural lesion.

Diagnostic criteria:

- A. Pain as described above for trigeminal neuralgia with or without persistence of aching between paroxysms and signs of sensory impairment in the distribution of the appropriate trigeminal division.
- B. The demonstration of a causative lesion by special investigations or posterior fossa exploration.

Comment: In patients previously considered "idiopathic", Dandy (1934) found

some source of compression in 60 per cent, most commonly an arterial branch or vein but acoustic neurinomas, cholesteatomas, osteoma, aneurysm, angiomas and adhesions were reported. Jannetta (1976) found compression by blood vessels in 88 per cent of patients, tumour or angioma in 6 per cent and multiple sclerosis in 6 per cent. Whether all trigeminal neuralgia should be regarded as "symptomatic" is still, however, uncertain.

12.2.2.2 Central lesions

Description: As for 12.2.2.1.

Diagnostic criteria:

- A. As for 12.2.2.1.
- B. The occurrence of trigeminal neuralgia during the course of clinically definite multiple sclerosis or following a brain stem infarct.

12.3 Glossopharyngeal neuralgia

12.3.1 Idiopathic glossopharyngeal neuralgia

Description: Glossopharyngeal neuralgia is a severe transient stabbing pain experienced in the ear, base of the tongue, tonsillar fossa or beneath the angle of the jaw. The pain is therefore felt in the distribution of the auricular and pharyngeal branches of the vagus nerve as well as that of the glossopharyngeal nerve. It is commonly provoked by swallowing, talking and coughing, and may remit and relapse in the fashion of trigeminal neuralgia.

Diagnostic criteria:

- A. Paroxysmal attacks of facial pain which last a few seconds to less than two minutes.
- B. Pain has at least 4 of the following characteristics:
 - 1. Unilateral location
 - 2. Distribution within the posterior part of the tongue, tonsillar fossa, pharynx, or beneath the angle of the lower jaw, or in the ear
 - 3. Sudden, sharp, stabbing or burning in quality
 - 4. Pain intensity severe
 - 5. Precipitation from trigger areas or by swallowing, chewing, talking, coughing, or yawning
- C. No neurological deficit.
- D. Attacks are stereotyped in the individual patient.
- E. Other causes of pain ruled out by history, physical and special investigations.

12.3.2 Symptomatic glossopharyngeal neuralgia

Description: as for 12.3.1 with the provision that aching pain may persist between paroxysms and sensory impairment may be found in the distribution of glossopharyngeal or vagus nerves.

Diagnostic criteria:

- A. Pain as described for 12.3.1 with or without persistence of aching between paroxysms and signs of sensory impairment in the distribution of glossopharyngeal or vagus nerves.
- B. Demonstration of a causative lesion by special investigations or operation.

12.4 Nervus intermedius neuralgia

Description: A rare disorder characterized by brief paroxysms of pain felt deeply in the auditory canal.

Diagnostic criteria:

- A. Pain paroxysms felt in the depth of the ear, lasting for seconds or minutes, of intermittent occurrence.
- B. Presence of a trigger zone in the posterior wall of the auditory canal.
- C. Exclusion of a structural lesion.

Comment: Disorders of lacrimation, salivation and taste sometimes accompany the pain. There is a common association with herpes zoster.

12.5 Superior laryngeal neuralgia

Description: A rare disorder characterized by severe pain in the lateral aspect of the throat, submandibular region and underneath the ear, precipitated by swallowing, shouting or turning the head.

Diagnostic criteria

- A. Pain paroxysms felt in the throat, submandibular region or under the ear, lasting for minutes or hours.
- B. Paroxysms are triggered by swallowing, straining the voice or head turning.
- C. Susceptibility continues for days or weeks.
- D. A trigger point is present on the lateral aspect of the throat overlying the hypothyroid membrane.
- E. Exclusion of a structural lesion.

12.6 Occipital neuralgia

Description: Occipital neuralgia is a paroxysmal jabbing pain in the distribution of the greater or lesser occipital nerves, accompanied by diminished sensation or dysaesthesiae in the affected area. It is commonly associated with tenderness over the nerve concerned.

Diagnostic criteria:

- A. Pain is felt in the distribution of greater or lesser occipital nerves.
- B. Pain is stabbing in quality although aching may persist between paroxysms.
- C. The affected nerve is tender to palpation.
- D. The condition is eased temporarily by local anaesthetic block of the appropriate nerve.

Comment: Occipital neuralgia must be distinguished from the occipital referral of pain from the atlantoaxial or upper zygapophyseal joints or from tender trigger points in neck muscles or their insertion.

Eagle's stylohyoid syndrome is not sufficiently validated.

12.7 Central causes of head and facial pain other than tic douloureux

12.7.1 Anaesthesia dolorosa

Description: Painful anaesthesia or dysaesthesia, often related to surgical trauma of the trigeminal ganglion, evoked most frequently after rhizotomy or thermocoagulation has been performed for treatment of idiopathic trigeminal neuralgia. Anaesthesia dolorosa may also follow upon trauma to the trigeminal complex, and, rarely, after vascular lesions of the central trigeminal pathways.

Diagnostic criteria:

- A. Pain or dysaesthesia is limited to the distribution of one or more divisions of the trigeminal nerve.
- B. Sensation to pinprick is diminished over the affected area.
- C. Symptoms follow a lesion of the trigeminal nerve or its central projections.

12.7.2 Thalamic pain

Description: Unilateral facial pain and dysaesthesiae attributed to a lesion of the quintothalamic pathway or thalamus. Symptoms may also involve the trunk and limbs of the affected side.

Diagnostic criteria:

- A. Pain and dysaesthesiae of one half of the face, associated with impaired sensation to pinprick, not explicable by a lesion of the trigeminal nerve.
- B. One or more of the following:
 - 1. A history of sudden onset suggesting a vascular lesion
 - 2. A remitting and relapsing history of symptoms in the face or elsewhere suggesting multiple sclerosis
 - 3. The demonstration of a lesion in an appropriate site by computerized tomography or magnetic resonance imaging

Comment: Thalamic facial pain is usually part of a hemisyn-drome, but may occur in isolation.

12.8 Facial pain not fulfilling criteria in groups 11 and 12

Previously used terms: Atypical facial pain, atypical odontalgia

Description: Persistent facial pain that does not have the characteristics of the cranial neuralgias classified above and is not associated with physical signs or a demonstrable organic cause.

Diagnostic criteria:

- A. Is present daily and persists for most or all of the day.
- B. Is confined at onset to a limited area on one side of the face. May spread to the upper or lower jaws or a wider area of the face or neck. Is deep and poorly localized.
- C. Is not associated with sensory loss or other physical signs.
- D. Laboratory investigations including X-ray of face and jaws do not demonstrate relevant abnormality.

Comment: Pain may be initiated by operation or injury to face, teeth or gums but persists without any demonstrable local cause.

13. Headache not classifiable

Diagnostic criteria:

Any type of headache which does not fulfil criteria for one of the disorders described in this book.