

Section Ib

Migraine and other headache syndromes

Descriptions and diagnostic criteria

General Notes

1. The criteria are labelled with letters and/or numbers to indicate their place in a hierarchy of generality and importance. General criteria, which *must* be fulfilled by all members of a group of disorders (such as the general criteria for all varieties of migraine with aura) are labelled with a capital G, plus a number. Obligatory criteria for individual disorders are distinguished by capital letters alone (A, B, C, etc.). Numbers (1, 2, 3, etc.) and lower case letters (a, b, c, etc.) are used to identify further groups and sub-groups of characteristics, only some of which are required for the diagnosis.
2. To avoid the use of 'and/or', when it is specified that *either* of two criteria is required, it is always assumed that the presence of *both* criteria also satisfies the requirement.
3. When there is a criterion that specifies frequency of attacks, life-time should be taken into account, unless it is specified differently.
4. If the patient has more than one headache disorder, all should be diagnosed in the order of importance indicated by the patient.
5. After each diagnosis the estimated number of headache days per year should be added in parentheses.
6. Patients who for the first time develop a particular form of headache in close temporal connection with the onset of one of the disorders listed in G44.3 (chronic post-traumatic headache), G44.4 (drug-induced headache), G44.8 (other specified headache syndromes) or G50–G53 (cranial neuralgia) (groups 5–11 of the IHS Classification, see Section III) are coded to these groups using additional characters to specify the etiology and additional codes to specify the type of the headache. Pre-existing migraine, tension-type headache or cluster headache aggravated in close temporal connection with one of the disorders listed in G44.3, G44.4, G44.8 or G50–G53 are still coded as migraine, tension-type headache or cluster headache (groups 1–3 of the IHS Classification). If the number of headache days increases by 100% or more, the aggravating factor may be mentioned in parentheses, but there is no separate code provided for its recording.
7. Code to the degree of specificity (number of characters) which suits your purpose.
8. If a patient has a form of headache fulfilling one set of diagnostic criteria, similar episodes which do not quite satisfy the criteria also usually occur. This can be due to treatment, lack of ability to remember symptoms exactly, and other factors. Ask the patient to describe a typical untreated attack or an unsuccessfully treated attack and ascertain that there have been enough of these attacks to establish the diagnosis. Then estimate the days per year with this type of headache adding also treated attacks and less typical attacks.
9. A major obstacle to an exact diagnosis is the reliance on patients' history to determine whether criteria are met. In less clear cases it is recommended letting the patient record attack characteristics prospectively using a headache diary before the diagnosis is made.
10. As already mentioned in the introduction, use of multiple coding is encouraged in all cases where there is a need to describe more extensively the different aspects of the patient's complaints. For instance, a depressed patient with headache would receive two codes, one for the type of headache and another one for the depression (F32) (see also list of commonly used additional categories, Section II).

DIAGNOSTIC CRITERIA

G43 MIGRAINE

Use additional external cause code (Chapter XX), if desired, to identify drug if drug-induced.

Excludes: headache NOS (R51)
atypical facial pain (G50.1)

G43.0 Migraine without aura [common migraine]

Previously used terms: hemicrania simplex.

Diagnostic criteria

- A. There have been at least five attacks fulfilling criteria B, C and D listed below.
- B. The headache attacks last between 4 and 72 h (untreated or unsuccessfully treated). Note: In children below age 15, attacks may last between 2 and 48 h. If the patient falls asleep and wakes up without migraine, the duration of the attack is until time of awakening.
- C. At least two of the following pain characteristics are present:
 1. Unilateral location.
 2. Pulsating quality.
 3. Moderate or severe intensity, inhibiting or prohibiting daily activities.
 4. Aggravation by walking stairs or similar routine physical activity.
- D. At least one of the following symptom groups is present during the attack:
 1. *Either* nausea or vomiting.
 2. *Both* photophobia and phonophobia.
- E. At least one of the following applies:
 1. History, physical and neurological examinations do not suggest one of the disorders listed in G44.3, G44.4, G44.8 or cranial neuralgia (G50–G53) (groups 5–11 of the IHS Classification, see Section III).
 2. History, physical, or neurological examinations do suggest such a disorder, but it is ruled out by appropriate investigations.
 3. Such a disorder is present, but the migraine attacks do not occur for the first time in close temporal connection with the disorder.

Comments

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% of attacks should occur between 2 days before menses and the last day of menses, but further epidemiological knowledge is needed. Menstrual migraine should be coded N94.3 (Premenstrual tension syndrome) with code G43.0 in addition.

G43.1 Migraine with aura [classical migraine]

Previously used terms: classic migraine, hemiplegic migraine, migraine accompagnée.

Diagnostic criteria

- G1. There have been at least two attacks fulfilling criterion G2 listed below.
- G2. At least three of the following characteristics are present:
 1. There are one or more fully reversible aura symptoms indicating focal cerebral cortical or brain stem dysfunction.
 2. *Either* at least one aura symptom develops gradually over more than 4 min, *or* two or more symptoms occur in succession.

3. The duration of the aura symptoms does not exceed 60 min.
4. The headache follows the aura with a free interval of less than 60 min (it may also begin before or simultaneously with the aura).

G3. At least one of the following applies:

1. History, physical and neurological examinations do not suggest one of the disorders listed in G44.3, G44.4, G44.8 or cranial neuralgia (G50–G53) (groups 5–11 of the IHS Classification, see Section III).
2. History, physical or neurological examination does suggest such a disorder, but it is ruled out by appropriate investigations.
3. Such a disorder is present, but migraine attacks do not occur for the first time in close temporal connection with the disorder.

G43.10 Migraine with typical aura

Diagnostic criteria

- A. The general criteria for G43.1 are fulfilled.
- B. No aura symptom lasts more than 60 min; if more than one aura symptom is present, the accepted duration is proportionally increased.
- C. One or more of the following types of aura symptoms are present.
 1. Homonymous visual disturbance
 2. Unilateral paraesthesias and/or numbness
 3. Unilateral weakness
 4. Aphasia or unclassifiable speech difficulty

G43.11 Migraine with prolonged aura

Diagnostic criteria

- A. The general criteria for G43.1 are fulfilled.
- B. At least one aura symptom lasts more than 60 min, but not more than 7 days.

G43.12 Migraine with acute onset aura

Diagnostic criteria

- A. The general criteria G43.1 are fulfilled.
- B. Neurological symptoms develop suddenly (in less than 4 min from the onset of the headache).
- C. The headache lasts between 4 and 72 h (untreated or unsuccessfully treated).
- D. At least two of the following pain characteristics are present:
 1. Unilateral location.
 2. Pulsating quality.
 3. Moderate or severe intensity, inhibiting or prohibiting daily activities.
 4. Aggravation by walking stairs or similar routine physical activity.
- E. At least one of the following symptom groups is present during the attack:
 1. *Either* nausea or vomiting.
 2. *Both* photophobia and phonophobia.
- F. A transient ischaemic attack (TIA) and other intracranial lesions are ruled out by appropriate investigations.

To identify the neurological symptoms in G43.1 Migraine with aura, a sixth character may be used, if desired, for categories G43.10, G43.11 and G43.12, as follows:

G43.1x0 Hemianoptic and other visual migraine

Previously used term: ophthalmic migraine.

G43.1x1 Hemisensory migraine

Previously used term: hemiparaesthetic migraine.

G43.1x2 Migraine with aphasia

Previously used term: aphasic migraine.

G43.1x3 Basilar migraine

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

Diagnostic criteria

- A. The general criteria for G43.1 are fulfilled.
- B. Two or more aura symptoms of the following types are present:
 - 1. Visual symptoms in both the temporal and nasal fields of both eyes
 - 2. Dysarthria
 - 3. Vertigo
 - 4. Tinnitus
 - 5. Decreased hearing
 - 6. Double vision
 - 7. Ataxia
 - 8. Bilateral paraesthesias
 - 9. Bilateral pareses
 - 10. Decreased level of consciousness

G43.1x4 Migraine aura (all types) without headache

Previously used term: acephalgic migraine.

Diagnostic criteria

- A. The general criteria for G43.1 are fulfilled.
- B. There is no headache.

G43.1x5 Familial hemiplegic migraine

Diagnostic criteria

- A. The general criteria for G43.1 are fulfilled.
- B. The aura includes some degree of hemiparesis and may be prolonged.
- C. At least one first-degree relative has identical attacks.

G43.1x6 Multiple types of aura

G43.1x7 Other specified migraine with aura

G43.2 Status migrainosus

Diagnostic criteria

- A. Either the criteria for G43.0 or the general criteria for G43.1 are fulfilled.
- B. The attack lasts more than 72 h whether treated or not.
- C. The headache is continuous throughout the attack or interrupted by headache-free intervals lasting less than 4 h. Note: Interruption during sleep is disregarded.

G43.3 Complicated migraine

Previously used term: migrainous cerebral infarction

Diagnostic criteria

- A. The general criteria for G43.1 have been fulfilled previously.
- B. The present attack is typical of previous attacks, but neurological deficits are not completely reversible within 7 days, or neuroimaging demonstrates ischaemic infarction in the relevant area of the brain.
- C. Other causes of infarction are ruled out by appropriate investigations.

G43.8 Other migraine

G43.80 Ophthalmoplegic migraine

Diagnostic criteria

- A. There have been at least two attacks fulfilling criterion B listed below.
- B. Headache associated with paresis of at least one of cranial nerves III, IV, or VI.
- C. Parasellar lesion has been ruled out by appropriate investigations.

G43.81 Retinal (monocular) migraine

Diagnostic criteria

- A. There have been at least two attacks fulfilling criteria B and C listed below.
- B. There is a fully reversible monocular scotoma or blindness lasting less than 60 min during an attack. This must be confirmed by examination during an attack or (after proper instruction) by patient's drawing of monocular field defect.
- C. Headache follows the visual symptoms with a free interval of less than 60 min, but may precede them.
- D. Ophthalmological examination outside the attack is normal; embolism has been ruled out by appropriate investigations.

G43.82 Childhood periodic migraine syndromes

G43.820 Abdominal migraine

Includes: migraine equivalents

Comments

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.

G43.821 Benign paroxysmal vertigo of childhood

Diagnostic criteria

- A. There are multiple, brief, sporadic episodes of disequilibrium, anxiety, and often nystagmus or vomiting.
- B. Neurological examination reveals no abnormalities.
- C. The electroencephalogram is normal.

G43.822 Alternating hemiplegia of childhood

Diagnostic criteria

- A. The onset is before 18 months of age.
- B. There are repeated attacks of hemiplegia involving both sides of the body.
- C. There are other paroxysmal phenomena, such as tonic convulsions, dystonic posturing, choreoathetoid movements, nystagmus or other ocular motor abnormalities, or autonomic dysbalances associated with the bouts of hemiplegia or occurring independently.
- D. There is evidence of a mental or neurological deficit.

G43.83 Atypical migraine

Diagnostic criteria

- A. All criteria but one are fulfilled of one or more forms of migraine (specify type(s)).
- B. The criteria for tension-type headache (G44.2) are not fulfilled.

G43.9 Migraine, unspecified

This is a non-recommended residual category, to be employed when no other code from this classification can be used.

The terms "cyclic migraine", "lower half headache", "facial migraine", "hemicrania continua" and "cervical migraine" are not sufficiently validated.

G44 OTHER HEADACHE SYNDROMES

Excludes: atypical facial pain (G50.1)
glossopharyngeal neuralgia (G52.1)
headache NOS (R51)
post-lumbar puncture headache (G97.0)
trigeminal neuralgia (G50.0)
other cranial neuralgias (G52.8)

G44.0 Cluster headache syndrome

Previously used terms: Erythroprotopalgia of Bing, ciliary or migrainous neuralgia [Harris], erythromelalgia of the head, Horton's headache, histaminic cephalgia, petrosal neuralgia [Gardner], spheno-palatine, Vidian and Sluder's neuralgia, hemicrania periodica neuralgiformis.

Diagnostic criteria

- G1. There have been at least five attacks fulfilling criteria G2, G3 and G4 below.
- G2. There is severe unilateral orbital, supraorbital or temporal pain lasting between 15 and 180 min, untreated.

- G3. The headache is associated with at least one of the following symptoms, 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
- G4. The frequency of the attacks ranges from one every other day to eight per day.
- G5. At least one of the following applies:
1. History, physical and neurological examinations do not suggest one of the disorders listed in G44.3, G44.4, G44.8 or cranial neuralgia (G50-G53) (groups 5–11 of the IHS Classification, see Section III).
 2. History, physical, or neurological examination does suggest such a disorder, but it is ruled out by appropriate investigations.
 3. Such a disorder is present, but the cluster headache attacks do not occur for the first time in close temporal connection with the disorder.

G44.00 Cluster headache with periodicity undetermined

Diagnostic criteria

- A. The general criteria for G44.0 are fulfilled.
- B. The time period of 1 year needed to classify as G44.01 or G44.02 has not passed yet.

G44.01 Episodic cluster headache

Diagnostic criteria

- A. The general criteria of G44.0 are fulfilled.
- B. There have been at least two periods of headaches (cluster periods) lasting (untreated) from 7 days to 1 year, separated by remissions of at least 14 days.

G44.02 Chronic cluster headache

Diagnostic criteria

- A. The general criteria of G44.0 are fulfilled.
- B. There have been no remission phases during a period of at least 1 year, or the remissions have lasted less than 14 days.

Use an additional sixth character, if desired, to specify course:

G44.020 unremitting from onset

G44.021 evolved from episodic

G44.03 Chronic paroxysmal hemicrania

Previously used term: Sjaastad's syndrome.

Diagnostic criteria

- A. There have been at least 50 attacks fulfilling criteria B, C, D and E listed below.
- B. The attack is characterized by severe unilateral orbital, supraorbital or temporal pain always on the same side lasting between 2 and 45 min.

- C. The attack frequency is at least five a day for more than half of the time (periods with lower frequency may occur).
- D. The pain is associated with at least one of the following symptoms on the pain side:
 - 1. Conjunctival injection
 - 2. Lacrimation
 - 3. Nasal congestion
 - 4. Rhinorrhea
 - 5. Ptosis
 - 6. Eyelid edema
- E. There is absolute effectiveness of indomethacin (150 mg/day or less).
- F. At least one of the following applies:
 - 1. History, physical and neurological examinations do not suggest one of the disorders listed in G44.3, G44.4, G44.8 or cranial neuralgia (G50–G53) (groups 5–11 of the IHS Classification, see Section III).
 - 2. History, physical or neurological examination does suggest such a disorder, but it is ruled out by appropriate investigations.
 - 3. Such a disorder is present, but chronic paroxysmal hemicrania attacks do not occur for the first time in close temporal connection with the disorder.

G44.08 Other and atypical cluster headache

Diagnostic criteria

All criteria but one are fulfilled of the criteria specified for G44.00, G44.01, G44.02 or G44.03.

G44.1 Vascular headache, not elsewhere classified

Diagnostic criteria

- A. There is a headache characterized by bifronto-temporal pulsating pain.
- B. There are *neither* aura symptoms *nor* nausea or vomiting.

G44.2 Tension-type headache

Previously used terms: tension headache, psychomyogenic headache, stress headache.

Comments

In the presence of emotional conflict or psychosocial problems that are sufficient to allow the conclusion that they are the main causative influences of the headache, category F45.4 *Persistent somatoform pain disorder*, including psychogenic headache, should be used, in addition to the relevant code for the headache, either G44.21 or G44.23.

In the presence of psychological or behavioural factors that are thought to have influenced the headache, or affected the course, category F54 *Psychological and behavioural factors associated with disorders or diseases classified elsewhere* can be used in addition to the relevant code for the headache.

G44.20 Episodic tension-type headache associated with disorder of pericranial muscles

Previously used term: muscle contraction headache.

Diagnostic criteria

- A. There have been at least 10 previous headache episodes fulfilling criteria B–D listed below. The number of days with such a headache is less than 180 per year and less than 15 per month.
- B. The headache lasts from 30 min to 7 days.

- C. At least two of the following pain characteristics are present:
 - 1. Pressing or tightening (non-pulsating) quality
 - 2. Mild or moderate intensity (the pain 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 apply:
 - 1. There is *neither* nausea *nor* vomiting (anorexia may occur).
 - 2. There is no photophobia and phonophobia occurring together (but *either* the one *or* the other may be present).
- E. At least one of the following is present:
 - 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.
- F. At least one of the following applies:
 - 1. History, physical and neurological examinations do not suggest one of the disorders listed in G44.3, G44.4, G44.8 or cranial neuralgia (G50–G53) (groups 5–11 of the IHS Classification, see Section III).
 - 2. History, physical, or neurological examinations do suggest such a disorder, but it is ruled out by appropriate investigations.
 - 3. Such a disorder is present, but tension-type headache attacks do not occur for the first time in close temporal connection with the disorder.

G44.21 Episodic tension-type headache without disorder of pericranial muscles

Previously used terms: idiopathic headache, essential headache.

Diagnostic criteria

- A. There have been at least 10 previous headache episodes fulfilling criteria B–D listed below. The number of days with such a headache is less than 180 per year and less than 15 per month.
- B. The headache lasts from 30 min to 7 days.
- C. At least two of the following pain characteristics are present:
 - 1. Pressing or tightening (non-pulsating) quality
 - 2. Mild or moderate intensity (the pain 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 apply:
 - 1. There is no nausea or vomiting (anorexia may occur).
 - 2. There is no photophobia and phonophobia occurring together (but one *or* the other may be present).
- E. At least one of the following is present:
 - 1. No increased tenderness of pericranial muscles demonstrated by manual palpation or pressure algometer.
 - 2. Normal EMG level of pericranial muscles at rest or during physiological tests.

- F. At least one of the following applies:
1. History, physical and neurological examinations do not suggest one of the disorders listed in G44.3, G44.4, G44.8 or cranial neuralgia (G50–G53) (groups 5–11 of the IHS Classification, see Section III).
 2. History, physical or neurological examination does suggest such a disorder, but it is ruled out by appropriate investigations.
 3. Such a disorder is present, but tension-type headache attacks do not occur for the first time in close temporal connection with the disorder.

G44.22 Chronic tension-type headache associated with disorder of pericranial muscles

Previously used terms: chronic daily headache, chronic muscle contraction headache.

Diagnostic criteria

- A. The average headache frequency is at least 15 days per month during at least 6 months, or at least 180 days a year, fulfilling criteria B, C and D listed below.
- B. At least two of the following pain characteristics are present:
1. Pressing or tightening (non-pulsating) quality
 2. Mild or moderate intensity (the pain 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 apply:
1. There is no vomiting
 2. None or only one of the following symptoms is present:
 - a. Nausea
 - b. Photophobia
 - c. Phonophobia
- D. At least one of the following is present:
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.
- E. At least one of the following applies:
1. History, physical and neurological examinations do not suggest one of the disorders listed in G44.3, G44.4, G44.8 or cranial neuralgia (G50–G53) (groups 5–11 of the IHS Classification, see Section III).
 2. History, physical or neurological examination does suggest such a disorder, but it is ruled out by appropriate investigations.
 3. Such a disorder is present, but tension-type headache attacks do not occur for the first time in close temporal connection with the disorder.

G44.23 Chronic tension-type headache without disorder of pericranial muscles

Previously used term: chronic idiopathic headache.

Diagnostic criteria

- A. The average headache frequency is at least 15 days per month during at least 6 months, or at least 180 days a year, fulfilling criteria B, C and D listed below.
- B. At least two of the following pain characteristics are present:
1. Pressing or tightening (non-pulsating) quality
 2. Mild or moderate intensity (the pain 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 apply:
 - 1. There is no vomiting
 - 2. None or only one of the following symptoms is present:
 - a. Nausea
 - b. Photophobia
 - c. Phonophobia
- D. At least one of the following is present:
 - 1. No increased tenderness of pericranial muscles demonstrated by manual palpation or pressure algometer.
 - 2. Normal EMG level of pericranial muscles at rest or during physiological tests.
- E. At least one of the following applies:
 - 1. History, physical and neurological examinations do not suggest one of the disorders listed in G44.3, G44.4, G44.8 or cranial neuralgia (G50–G53) (groups 5–11 of the IHS Classification, see Section III).
 - 2. History, physical or neurological examination does suggest such a disorder, but it is ruled out by appropriate investigations.
 - 3. Such a disorder is present, but tension-type headache attacks do not occur for the first time in close temporal connection with the disorder.

G44.28 Other specified tension-type headache

Previously used term: atypical tension-type headache.

Diagnostic criteria

- A. All criteria but one are fulfilled for one or more forms of tension-type headache (specify type(s)).
- B. The criteria for migraine without aura (G43.0) are not fulfilled.

G44.3 Chronic post-traumatic headache

G44.30 Chronic post-traumatic headache with significant head trauma or confirmatory signs of head trauma (S06)

Diagnostic criteria

- A. The severity of the head trauma is manifested by at least one of the following:
 - 1. Loss of consciousness
 - 2. Post-traumatic amnesia lasting more than 10 min
 - 3. Relevant abnormality in at least two of the following areas:
 - a. Clinical neurological examination
 - b. X-ray of skull
 - c. Neuroimaging
 - d. Evoked potentials
 - e. Spinal fluid examination
 - f. Vestibular function test
 - g. Neuropsychological testing.
- B. The onset of the headache is less than 14 days after regaining consciousness (or after the trauma, if there has been no loss of consciousness).
- C. The headache continues for more than 8 weeks after regaining consciousness (or after the trauma, if there has been no loss of consciousness).

G44.31 Chronic post-traumatic headache with minor head trauma and no confirmatory signs (S09.9)*Diagnostic criteria*

- A. There has been head trauma that does not fulfil criterion A of G44.30.
- B. The onset of the headache is less than 14 days after injury.
- C. The headache continues for more than 8 weeks after injury.

G44.4 Drug-induced headache, not elsewhere classified

Excludes: Headache associated with psychoactive substance use (G44.83)

G44.40 Headache induced by acute non-dependence-producing substance use or exposure*Diagnostic criteria*

- G1. There is a temporal connection (to be specified for each substance) between the onset of the headache and the substance intake or exposure.
- G2. The patient has taken or has been exposed to the effective minimum dose of the specific substance (to be indicated).
- G3. The headache has occurred in at least half of the exposures and at least three times.
- G4. The headache disappears when the substance is eliminated or within a specified time after elimination.

G44.400 Nitrate/nitrite-induced headache (X44)

Previously used term: hot-dog headache.

Diagnostic criteria

- A. The general criteria of G44.41 are fulfilled.
- B. The headache occurs within 1 h after absorption of nitrate/nitrite.

G44.401 Monosodium glutamate-induced headache (X44)

Previously used term: Chinese restaurant syndrome.

Diagnostic criteria

- A. The general criteria of G44.41 are fulfilled.
- B. The headache occurs within 1 h after ingestion of monosodium glutamate.
- C. The headache is associated with at least two of the following symptoms:
 - 1. Pressure in the chest
 - 2. Pressure and tightness in the face
 - 3. Burning sensation in the chest, neck or shoulders
 - 4. Flushing of face
 - 5. Dizziness
 - 6. Abdominal discomfort.

G44.402 Carbon monoxide-induced headache (X47)*Diagnostic criteria*

- A. The general criteria of G44.41 are fulfilled.

G44.408 Headache induced by other acute non-dependence-producing substance use or exposure

G44.41 Headache induced by chronic substance use or exposure*Diagnostic criteria*

- G1. The headache occurs after daily doses of a substance for at least months.
- G2. The patient has taken or has been exposed to the effective minimum dose of the specific substance (to be indicated).
- G3. The headache is chronic, i.e. occurs during at least 15 days a month.
- G4. The headache disappears within 1 month after withdrawal of the substance.

G44.410 Analgesics abuse headache (F55.2)

Excludes: Headache due to abuse of analgesics derived from opioids (F11.1 plus G44.83)

Diagnostic criteria

- A. The general criteria of G44.41 are fulfilled.
- B. One or more of the following applies:
 1. Ingestion of at least 50 g of aspirin a month or its equivalent of another mild analgesic.
 2. Ingestion of at least 100 tablets a month of analgesics, combined with barbiturates or other non-opioid compounds.

G44.411 Ergotamine-induced headache (Y52.5)*Diagnostic criteria*

- A. The general criteria of G44.41 are fulfilled.
- B. The headache is preceded by daily ergotamine intake (oral 2 mg, rectal 1 mg) for at least 3 months.
- C. The pain is diffuse, pulsating and distinguished from migraine by the absence of an attack pattern and/or the absence of associated symptoms.

G44.412 Ergotamine withdrawal headache (Y52.5)*Diagnostic criteria*

- A. The general criteria of G44.41 are fulfilled.
- B. The headache is preceded by daily ergotamine intake (oral 2 mg, rectal 1 mg) for at least 3 months.
- C. The headache occurs within 48 h after withdrawal of ergotamine.

G44.418 Headache induced by other chronic non-dependence-producing substance use or exposure*Comments*

Headache induced by birth control pills, containing estrogens (Y42.4) is not well validated. The literature on this subject is conflicting and further study is needed in this area.

Headache due to occupational exposure to toxic agents in agriculture (Z57.4) or other industries (Z57.5) should also be classified here. If desired, the substance may be specified by using one of the codes T51–T65 Toxic effect of substances chiefly non-medicinal as to source (see list of commonly used additional categories, Section II).

G44.8 Other specified headache syndromes

G44.80 Other headaches not associated with a structural lesion

G44.800 Idiopathic stabbing headache

Previously used terms: ice-pick pains, ice-pick headache, cephalgia fugax.

Diagnostic criteria

- A. The pain is 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. The pain is stabbing in nature and lasts for a fraction of a second. It occurs as single stabs or a series of stabs.
- C. It recurs at irregular intervals (hours to days).
- D. There are no structural changes at the site of the pain.

G44.801 External compression headache

Previously used term: swim-goggle headache.

Diagnostic criteria

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

G44.802 Cold stimulus headache

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

G44.8020 External application of a cold stimulus

Diagnostic criteria

- A. The headache develops during external exposure to cold.
- B. The pain is bilateral.
- C. The pain varies in intensity with the severity and duration of the cold stimulus.
- D. The headache is prevented by avoiding exposure to cold.
- E. The pain is not associated with organic cranial or intracranial disease.

G44.8021 Ingestion of a cold stimulus

Previously used term: ice-cream headache.

Diagnostic criteria

- A. The headache develops during ingestion of cold food or drink.
- B. The pain lasts for less than 5 min.

- C. The pain 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 the migraine headache (code migraine first).
- D. The headache is prevented by avoiding the rapid swallowing of cold food or drinks.
- E. The pain is not associated with organic disease.

G44.803 Benign cough headache

Diagnostic criteria

- A. There is a bilateral headache of sudden onset, lasting less than 1 min, precipitated by coughing.
- B. The pain may be prevented by avoiding coughing.
- C. Structural cerebral lesions such as posterior fossa tumour have been excluded by neuroimaging.

G44.804 Benign exertional headache

Diagnostic criteria

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

G44.805 Headache associated with sexual activity

Previously used terms: benign sex headache, coital cephalalgia or cephalgia, orgasmic cephalgia.

Diagnostic criteria

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

Use additional seventh character, if desired, to specify type:

- G44.8050 dull type
- G44.8051 explosive type
- G44.8052 postural type

G44.806 Idiopathic carotidynia

Diagnostic criteria

- A. There is at least one of the following symptoms present in the area overlying the carotid artery:
 1. Tenderness
 2. Swelling
 3. Increased pulsations

- B. There is pain over the affected side of the neck, which may project to the ipsilateral side of the head.
- C. The syndrome is self-limiting of less than 2 weeks' duration.
- D. Appropriate investigations do not reveal structural abnormalities in the affected area.

G44.81 Headaches associated with other vascular disorders

Diagnostic criteria

- G1. There are symptoms indicating a vascular disorder, as specified in the subcategories.
- G2. Appropriate investigations confirm the presence of a vascular disorder, as specified in the subcategories.
- G3. There is headache as a new symptom or of a new type, occurring in close temporal connection with the onset of the vascular disorder.

G44.810 Headache associated with cerebrovascular disease

– transient ischaemic attack (TIA) (G45)

Diagnostic criteria

- A. The general criteria for G44.81 are fulfilled.
- B. The symptoms clear within 24 h.

– cerebral infarction (I63.-) (see Section 2 for appropriate subcategories)

– stroke, not specified as haemorrhage or infarction (I64)

– acute ischaemic cerebrovascular disease (G45, G46)

Diagnostic criteria

- A. The general criteria of G44.81 are fulfilled.
- B. Focal central nervous system symptoms develop within 48 h.
- C. The symptoms persist for more than 24 h.

– cerebral venous thrombosis (I63.6)

Diagnostic criteria

- A. The general criteria of G44.81 are fulfilled.
- B. At least one of the following is present:
 1. Raised intracranial pressure
 2. Focal neurological deficit
 3. Seizures
- C. The headache is located to the affected area or is diffuse.

– thalamic pain (G46.21)

Diagnostic criteria

- A. Pain and dysaesthesia 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 must apply:
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 (G35).
 3. The demonstration of a lesion in an appropriate site by computerized tomography or magnetic resonance imaging.

– **intracerebral haemorrhage (haematoma)**

- – – **non-traumatic (I61.-)** (see Section II for appropriate subcategories)
- – – **traumatic, focal (S06.3)**

– **intracranial haemorrhage (haematoma)**

- – – **non-traumatic (I62.-)** (see Section II for appropriate subcategories)
- – – **traumatic (S06.4, S06.5)** (see Section II for appropriate subcategories)

Diagnostic criteria

- A. The general criteria of G44.81 are fulfilled.
- B. Focal central nervous system symptoms develop within 24 h.

– **subarachnoid haemorrhage**

- – – **non-traumatic (I60)**
- – – **traumatic (S06.6)**

Diagnostic criteria

- A. The general criteria of G44.81 are fulfilled.
- B. There is a headache of sudden onset (within less than 60 min) if the haemorrhage is due to an aneurysm, and less than 12 h if it is due to an arteriovenous malformation.
- C. At least one of the following symptoms is present:
 1. Severe intensity of the pain.
 2. Bilateral location of the headache.
 3. Stiff neck.
 4. Increased body temperature.

– **dissection of cerebral arteries (non-ruptured) (I67.0)**

Diagnostic criteria

- A. The general criteria of G44.81 are fulfilled.
- B. At least one of the following is present:
 1. Transient ischaemic attack or cerebral infarction in the territory of the affected artery.
 2. Horner's syndrome, arterial bruit or tinnitus.
- C. There is headache and cervical pain ipsilateral to the arterial dissection.

G44.811 Headache associated with congenital malformation of the circulatory system

– **arteriovenous malformation of cerebral vessels (non-ruptured) (Q28.2)**

– **congenital cerebral aneurysm (non-ruptured) (Q28.3)**

Includes: saccular aneurysm

Diagnostic criteria

The general criteria for G44.81 are fulfilled.

G44.812 Headache associated with arteritis

– **cerebral arteritis (I67.7)**

– **arteritis in other diseases classified elsewhere (I68.-*)**

Diagnostic criteria

The general criteria for G44.81 are fulfilled.

– **giant cell arteritis (M31.6)**

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

Diagnostic criteria

A. The general criteria of G44.81 are fulfilled.

B. Two or more of the following are present:

1. Swollen and tender scalp artery (usually superficial temporal artery).
2. Elevated RBC sedimentation rate.
3. Disappearance of headache within 48 h of steroid therapy.

G44.813 Headache associated with hypertensive diseases

– **hypertension (I10)**

Previously used terms: malignant hypertension, accelerated hypertension.

Diagnostic criteria

A. There is a headache associated with grade 3 or 4 retinopathy (Keith Wagner classification).

B. The diastolic blood pressure is persistently above 120 mmHg.

C. Appropriate investigations rule out vasopressor toxins, medication or phaeochromocytoma as causative factors.

D. There is a temporal connection between the onset of the headache and the rise in blood pressure, and the headache disappears within 2 days after reduction of blood pressure. If hypertensive encephalopathy is present, the headache may persist up to 7 days after reduction of blood pressure.

– **secondary hypertension (I15.-)** (see Section II for appropriate subcategories)

Includes: acute pressor response to exogenous agent.

Diagnostic criteria

- A. There is a headache occurring with acute rise (>25%) of the diastolic blood pressure.
- B. There is evidence of an appropriate toxin or medication.
- C. The headache disappears within 24 h after normalization of the blood pressure.

– **secondary hypertension in pheochromocytoma**

– – – **malignant, unspecified site (C74.1)**

– – – **benign, unspecified site (D35.0)**

Diagnostic criteria

- A. There is a headache occurring with acute rise (>25%) of diastolic blood pressure.
- B. At least one of the following symptoms is present:
 - 1. Sweating
 - 2. Palpitations
 - 3. Anxiety
- C. There is pheochromocytoma, demonstrated by biological and imaging tests or surgery.
- D. The headache disappears within 24 h after normalization of the blood pressure.

– **pre-eclampsia (O14)**

– **mild pre-eclampsia (O13)**

– **eclampsia (O15)**

Diagnostic criteria

- A. There is a headache during pregnancy.
- B. There is a presence of edema or proteinuria and a blood pressure rise compared to the prepregnant level (not necessarily markedly increased, but at least with mean elevation of 15 mmHg or diastolic of 90 mmHg).
- C. Appropriate investigations rule out vasopressor toxins, medication or pheochromocytoma as causative factors.
- D. The headache occurs with rise in blood pressure and disappears within 7 days after blood pressure reduction or after termination of pregnancy.

G44.814 Post-endarterectomy headache (G97.8)*Diagnostic criteria*

- A. There has been thrombo-endarterectomy or other surgery of the extracranial carotid artery.
- B. Patent carotid artery without dissection as demonstrated by appropriate investigations.
- C. There is a headache beginning 1 to 2 days after surgery on the ipsilateral side.

G44.818 Headache associated with other specified vascular disorders

Use additional code to specify the etiology.

G44.82 Headache associated with other intracranial disorders*Diagnostic criteria*

- G1. Symptoms and/or signs of an intracranial disorder.
- G2. Confirmation by appropriate investigations.
- G3. Headache as a new symptom or of a new type occurs in a temporal connection to an intracranial disorder.

G44.820 Headache associated with disorders of cerebrospinal fluid pressure**– benign intracranial hypertension (G93.2)**

Previously used terms: high cerebrospinal fluid pressure, pseudotumour cerebri.

Diagnostic criteria

- A. The patient suffers from benign intracranial hypertension fulfilling all of the following:
 - 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 papil edema and possible VIth cranial nerve.
 - 3. Normal or low protein concentration and normal white cell count in CSF.
 - 4. No suspicion of venous sinus thrombosis, cerebral lesion or ventricular enlargement, clinically and on neuroimaging.
- B. There is a headache with an intensity and frequency related to variations of intracranial pressure with a time-lag of less than 24 h.

– high-pressure hydrocephalus (G91)**– post-traumatic hydrocephalus (G91.3)***Diagnostic criteria*

- A. The 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 h.

– **post-lumbar puncture headache (G97.0)**

Includes: post-spinal puncture headache

Diagnostic criteria

- A. There is a bilateral headache developed less than 7 days after lumbar puncture.
- B. The headache occurs or worsens less than 15 min after assuming the upright position, and disappears or improves less than 30 min after resuming the recumbent position.
- C. The headache disappears within 14 days after lumbar puncture (if duration exceeds 14 days, consider cerebrospinal fluid leak G96.0).

– **cerebrospinal fluid leak headache (G96.0)**

Diagnostic criteria

- A. Post-traumatic, post-operative 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. The headache occurs or worsens less than 15 min after assuming the upright position, and disappears or improves less than 30 min after resuming the recumbent position.
- C. Headache disappears within 14 days after effective treatment of the fistula.

G44.821 Headache associated with intracranial infection

– **brain abscess (G06.0)**

– **encephalitis (G04.9)**

– **meningitis (G03.9)**

– **subdural empyema (G06.2)**

Diagnostic criteria

The general criteria for G44.82 are fulfilled.

G44.822 Headache associated with intracranial neoplasm (C00–D48)

Diagnostic criteria

The general criteria for G44.82 are fulfilled.

G44.823 Headache associated with intracranial sarcoidosis (D86.8)

Diagnostic criteria

The general criteria for G44.82 are fulfilled.

G44.824 Headache associated with other postprocedural disorders of the nervous system**– intrathecal injections (G97.8)****– direct toxic effect of the substance injected (T80.8)***Diagnostic criteria*

- A. Headache follows intrathecal injection after less than 4 h.
- B. The headache is diffuse and present also in the recumbent position.
- C. Headache clears completely within 14 days. (If it persists consider cerebral spinal fluid leak G96.0).

– chemical meningitis (G03.8)*Diagnostic criteria*

- A. Headache follows intrathecal injection within 4–72 h.
- B. The headache is diffuse and present also in the recumbent position.
- C. There is cerebrospinal fluid pleocytosis with negative culture.

G44.828 Headache associated with other specified intracranial disorders

Use additional code for etiology.

G44.83 Headache associated with psychoactive substance use

Use additional code, if desired, to identify the substance involved:

- F10 Mental and behavioural disorder due to use of alcohol
- F11 Mental and behavioural disorder due to use of opioids
- F12 Mental and behavioural disorder due to use of cannabinoids
- F13 Mental and behavioural disorder due to use of sedatives or hypnotics
- F14 Mental and behavioural disorder due to use of cocaine
- F15 Mental and behavioural disorder due to use of other stimulants, including caffeine
- F16 Mental and behavioural disorder due to use of hallucinogens
- F17 Mental and behavioural disorder due to use of tobacco
- F18 Mental and behavioural disorder due to use of volatile solvents
- F19 Mental and behavioural disorder due to multiple drug use and use of other psychoactive substances.

This last category should be used when two or more substances are known to be involved, but it is impossible to assess which substance is contributing most to the disorders. It should also be used when the exact identity of some or even all the substances being used is uncertain or unknown, since many multiple drug users themselves often do not know the details of what they are taking.

Includes: misuse of drugs NOS

A fourth character may be used to indicate the associated condition (F1x.0–F1x.9):

- F1x.0 Acute intoxication
- F1x.1 Harmful use
- F1x.2 Dependence syndrome
- F1x.3 Withdrawal state
- F1x.4 Withdrawal state with delirium
- F1x.5 Psychotic disorder
- F1x.6 Amnestic syndrome

- F1x.7 Residual and late-onset psychotic disorder
- F1x.8 Other mental and behavioural disorder
- F1x.9 Unspecified mental and behavioural disorder

Alcohol-induced headache (F10.0)

Diagnostic criteria

Headache occurs within 3 h after ingestion of alcohol.

Alcohol withdrawal headache (F10.3)

Includes: hangover.

Diagnostic criteria

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

Caffeine withdrawal headache (F15.3)

Diagnostic criteria

- A. The patient has consumed caffeine daily and at least 15 g a month during at least 3 months.
- B. Headache occurs within 24 h after last caffeine intake.
- C. The headache is relieved within 1 h by 100 mg of caffeine.

G44.84 Headache or facial pain associated with disorders of cranium, cranial and facial structures, cranial nerves, neck and spine

Diagnostic criteria

- A. Clinical or laboratory evidence of a 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. The headache disappears within 1 month after successful treatment or spontaneous remission of the underlying disorder.

Use additional code, if desired, to identify the associated condition(s) or cause.

G44.840 Headache associated with disorders of the cranial bone (M80–M89.8) (see Section II for appropriate categories)

Comments

Most disorders of the skull, e.g. congenital abnormalities, fractures, tumours, metastases, are not usually accompanied by headache. Exceptions of importance are osteomyelitis (M86), multiple myeloma (C90.0) and Paget's disease of the skull (M88.0).

G44.841 Headache associated with biomechanical lesions of cervical spine (M99.x1) (see Section II for appropriate fourth character)

Previously used terms: cervicogenic headache.

Diagnostic criteria

- A. There is pain localized in the neck and occipital region. The pain may project to the forehead, orbital region, temples, vertex or ears.
- B. The pain is precipitated or aggravated by special neck movements or sustained neck posture.

- C. At least one of the following is present:
 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).

G44.842 Headache associated with retropharyngeal tendinitis (M79.8)

Diagnostic criteria

- A. There is pain in the back of the neck radiating to the back of the head or to the whole head.
- B. The pain is non-pulsating, uni- or bilateral and aggravated severely by bending of the 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 anti-inflammatory drugs in recommended doses.

G44.843 Headache associated with disorders or diseases of the eye

– acute glaucoma (H40)

Diagnostic criteria

- A. Acute glaucoma is diagnosed by appropriate investigations.
- B. There is pain in, behind, or above the eye.

– refractive errors (H52) (see Section II for appropriate categories)

Diagnostic criteria

- A. There are uncorrected refractive errors, e.g. hypermetropia (H52.0), astigmatism (H52.2), presbyopia (H52.4), wearing of incorrect glasses (H52.6).
- B. There are mild headaches in the frontal region and in the eyes themselves.
- C. The pain is absent on awakening, and aggravated by prolonged visual tasks at the distance or angle where vision is impaired.

– Heterophoria or heterotropia (H50) (see Section II for appropriate categories)

Diagnostic criteria

- A. Heterophoria (H50.5), heterotropia (H50.4), or intermittent heterotropia (H50.3) is demonstrated.
- B. There is a mild to moderate constant headache in the frontal region.

- C. At least one of the following is present:
 1. Headache occurring or worsening during a visual task especially when becoming tired.
 2. Intermittent blurred vision or diplopia.
 3. Difficulty adjusting focus from near to distant objects or vice versa.
- D. There is relief or improvement of symptoms by closing one eye.

G44.844 Headache associated with disorders or diseases of the ear and mastoid process (H60–H95) (see Section II for appropriate categories)

G44.845 Headache associated with disorders or diseases of the respiratory system

- **acute sinusitis headache (J01)** (see Section II for appropriate fourth character, indicating the sinus involved)

Comments

Migraine and tension-type headache are often confused with true sinus headache because of similarities in location. In order to diagnose sinus headache the criteria mentioned below must be strictly fulfilled.

Diagnostic criteria

- A. There is a purulent discharge in the nasal passage either spontaneous or by suction.
- B. There are pathological findings in one or more of the following tests:
 1. X-ray examination
 2. Computerized tomography
 3. Transillumination
- C. There is a simultaneous onset of headache and sinusitis.
- D. The headache is located according to the sinus involved:
 1. In acute frontal sinusitis (J01.1) the headache is located directly over the sinus and may radiate to the vertex or behind the eyes.
 2. In acute maxillary sinusitis (J01.0) the headache is located over the antral area and may radiate to the upper teeth or to the forehead.
 3. In acute ethmoiditis (J01.2) the headache is located between and behind the eyes and may radiate to the temporal area.
 4. In acute sphenoiditis (J01.3) the headache is located in the occipital area, the vertex, the frontal region or behind the eyes.
- E. The headache disappears after treatment of the acute sinusitis.

- **other disorders of nose and nasal sinuses (J34)**

Comments

Other conditions which may cause headache are nasal passage abnormality due to septal deflection (J34.2), hypertrophic turbinates (J34.3) and atrophic sinus membrane (J34.8). These conditions are not sufficiently validated as a cause of headache. Chronic sinusitis (J32) is not validated as a cause of headache or facial pain unless relapsing into an acute stage. Post-operative chronic pain due to nerve damage should be classified in G44.846.

G44.846 Headache associated with disorders or diseases of the oral cavity (including teeth), salivary glands and jaws (K00–K14)
(see Section II for appropriate categories)

Comments

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 chronic (K05.3) or acute (K05.2) infection or traumatic irritation around a partially erupted lower wisdom tooth (embedded and impacted teeth) (K01).

– temporomandibular joint disorder (K07.6)

Excludes: headache due to functional temporomandibular joint disorders (G44.2)

Diagnostic criteria

- A. At least two of the following symptoms are present:
1. Pain of the jaw that is precipitated by movement and/or clenching of the jaw.
 2. Decreased range of movement of the jaw.
 3. Noise during movements of the temporomandibular joint.
 4. Tenderness of the capsule of the temporomandibular joint.
- B. There are 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.

G44.847 Headache associated with cranial neuralgia

– idiopathic trigeminal neuralgia (G50.00)

Previously used term: tic douloureux.

Diagnostic criteria

- A. There are paroxysmal attacks of facial or frontal pain which last a few seconds to less than 2 min.
- B. The pain has at least four of the following characteristics:
1. Distribution along one or more divisions of the trigeminal nerve.
 2. Sudden, intense, sharp, superficial, stabbing or burning quality.
 3. Severe intensity.
 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. There is no neurological deficit.
- D. Attacks are stereotyped in the individual patient.
- E. Other causes of facial pain are excluded by history, physical examination and special investigations when necessary.

– secondary trigeminal neuralgia
– – – anaesthesia dolorosa

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.

– – – **postzoster trigeminal neuralgia (B02.2 + plus G53.00*)**

Includes: post-herpetic trigeminal neuralgia

G53.000* with duration less than 6 months after onset of the infection

Diagnostic criteria

- A. Pain is followed by a herpetic eruption in the distribution of the trigeminal nerve, within 1 week of onset.
- B. The pain subsides within 6 months after onset of the rash.

G53.001* Chronic postzoster trigeminal neuralgia

Diagnostic criteria

- A. Pain is restricted to the distribution of the trigeminal nerve.
- B. The pain persists for more than 6 months after the onset of herpetic eruption.

– – – **secondary trigeminal neuralgia, not otherwise specified (G50.09)**

Includes: symptomatic trigeminal neuralgia, not otherwise specified

– – – **other trigeminal neuralgia secondary to diseases classified elsewhere (G53.80*)**

Diagnostic criteria

- A. There are paroxysmal attacks of facial or frontal pain which last from a few seconds to less than 2 min.
- B. The pain has at least four of the following characteristics:
 1. Distribution along one or more divisions of the trigeminal nerve.
 2. Sudden, intense, sharp, superficial, stabbing or burning quality.
 3. Severe intensity.
 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 may be entirely asymptomatic, or the aching may persist between paroxysms, with or without signs of sensory impairment in the distribution of the appropriate trigeminal division.
- C. A causative lesion is suggested or demonstrated by special investigations or posterior fossa exploration. (If the etiology can be specified, category G53.80* should be used).

– **atypical facial pain (G50.1)**

Previously used term: atypical odontalgia.

Diagnostic criteria

- A. The pain is present daily and persists for most or all of the day.
- B. The pain is confined at the onset to a limited facial area usually on one side of the nasolabial fold or chin. The pain may spread to the upper or lower jaws or a wider area of the face or neck.

- C. The pain 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.

– **nervus intermedius neuralgia, idiopathic (G51.80)**

Diagnostic criteria

- A. There are pain paroxysms felt in the depth of the ear, lasting for seconds or minutes, of intermittent occurrence.
- B. There is a trigger zone in the posterior wall of the auditory canal.
- C. A structural lesion is excluded.

– **secondary facial nerve neuralgia (G53.810*)**

Diagnostic criteria

- A. There are pain paroxysms felt in the depth of the ear, lasting for seconds or minutes, of intermittent occurrence.
- B. There is a trigger zone in the posterior wall of the auditory canal.
- C. The symptoms follow a lesion of the facial nerve.

– **idiopathic glossopharyngeal neuralgia (G52.10)**

Diagnostic criteria

- A. There are paroxysmal attacks of facial pain which last a few seconds to less than 2 min.
- B. The pain has at least four 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 quality.
 - 4. Severe intensity.
 - 5. Precipitation from trigger areas or by swallowing, chewing, talking, coughing, or yawning.
- C. There is no neurological deficit.
- D. Attacks are stereotyped in the individual patient.
- E. Other causes of pain are ruled out by history, physical and special investigations.

– **secondary glossopharyngeal neuralgia (G53.83*)**

Diagnostic criteria

- A. There are paroxysmal attacks of facial pain which last a few seconds to less than 2 min.
- B. The pain has at least four 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 quality.

4. Severe intensity.
 5. Between paroxysms the patient may be entirely asymptomatic, or the aching may persist between paroxysms, with or without signs of sensory impairment in the distribution of the glossopharyngeal or vagus nerves.
- C. A causative lesion is demonstrated by special investigations or operation.

– **postzoster glossopharyngeal neuralgia (B02.2+ plus G53.01*)**

G53.010* with duration less than 6 months after onset of the infection (B02.2+)

Diagnostic criteria

- A. Pain is followed by a herpetic eruption in the distribution of the glossopharyngeal nerve, within 1 week of onset.
- B. The pain subsides within 6 months after onset of the rash.

G53.011* Chronic postzoster glossopharyngeal neuralgia (B02.2+)

Diagnostic criteria

- A. Pain is restricted to the distribution of the glossopharyngeal nerve.
- B. The pain persists for more than 6 months after the onset of herpetic eruption.

– **superior laryngeal neuralgia (G52.20)**

Diagnostic criteria

- A. There are pain paroxysms felt in the throat, submandibular region or under the ear, lasting for minutes or hours.
- B. The 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. A structural lesion is excluded.

– **secondary vagus nerve neuralgia (g53.840*)**

Diagnostic criteria

- A. There are paroxysmal attacks of facial pain which last a few seconds to less than 2 min.
- B. The pain has at least four 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 quality.
 4. Severe intensity.
 5. Between paroxysms the patient may be entirely asymptomatic, or the aching may persist between paroxysms, with or without signs of sensory impairment in the distribution of the glossopharyngeal or vagus nerves.
- C. A causative lesion is demonstrated by special investigations or operation.

– **occipital neuralgia (G52.80)**

Diagnostic criteria

- A. Pain is felt in the distribution of greater or smaller subdivisions of occipital nerves.
- B. The pain is stabbing in quality; 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.

– **other idiopathic cranial neuralgia (G52.81)**

– **other secondary cranial nerve neuralgia (G53.8*)**

G44.848 Persistent headache of cranial nerve origin

Excludes: headache associated with cranial neuralgia (G44.847)

– **compression or distortion of cranial nerves (G53) or second or third cervical roots (G55)** (see Section II [p. 39] for the appropriate fourth, fifth and sixth character codes)

– **post-operative chronic pain due to nerve damage (G97.8)**

Diagnostic criteria

- A. There is pain in the distribution of one or more cranial nerve(s) and/or second or third cervical roots with or without projection to neighbouring areas.
- B. The presence of a relevant lesion has been demonstrated.
- C. The onset of the pain is temporally related to the onset of the cranial nerve lesion.
- D. If the lesion can be effectively treated or remits spontaneously, the pain improves or disappears.

– **optic neuritis (H46)**

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.

– **diabetic neuritis of oculomotor nerve (E10–E14 plus G53.88*)**

Diagnostic criteria

- A. Pain is felt around the eye on the affected side.
- B. The 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.

G44.85 Other specified syndromes of facial and ocular pain

G44.850 Tolosa-Hunt syndrome

Diagnostic criteria

- A. There are one or more episodes of unilateral orbital pain for an average of 8 weeks if untreated.
- B. The pain is associated 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 2 weeks.
- C. The pain is relieved within 72 h after initiation of corticosteroid therapy.
- D. Other causative lesions are excluded by neuroimaging and/or carotid angiography.

G44.851 Neck-tongue syndrome

Diagnostic criteria

- A. There is pain and numbness in the area of the distribution of the lingual nerve and second cervical root.
- B. The pain and numbness is precipitated by sudden turning of the head.

G44.88 Headache associated with other specified disorders

G44.880 Acute post-traumatic headache

- **with significant head trauma or confirmatory signs of head trauma (S06)** (see Section II for appropriate fourth character codes)

Excludes: chronic post-traumatic headache (G44.3)

Diagnostic criteria

- A. The significance of head trauma is documented by at least one of the following:
 1. Loss of consciousness.
 2. Post-traumatic amnesia lasting more than 10 min.
 3. Relevant abnormality in at least two of the following areas:
 - a. Clinical neurological examination
 - b. X-ray of skull
 - c. Neuroimaging
 - d. Evoked potentials
 - e. Spinal fluid examination
 - f. Vestibular function test
 - g. Neuro-psychological testing
- B. Headache occurs less than 14 days after consciousness has been regained (or after trauma, if there has been no loss of consciousness).
- C. The headache disappears less than 8 weeks after consciousness has been regained (or after trauma, if there has been no loss of consciousness).

– **with minor head trauma and no confirmatory signs (S09.9)**

Diagnostic criteria

- A. There has been head trauma that does not fulfil criterion A of G44.880 acute post-traumatic headache with significant head trauma or confirmatory signs of head trauma (S06).
- B. Headache occurs less than 14 days after injury.
- C. The headache disappears within 8 weeks after injury.

G44.881 Headache associated with non-cephalic infection (A00–B97)

Diagnostic criteria

- A. There are symptoms and/or signs of a non-cephalic infection.
- B. The diagnosis of a systemic or focal non-cephalic infection is confirmed by laboratory findings.
- C. Headache as a new symptom or of a new type occurs concomitantly with the infection.
- D. The headache disappears within 1 month after successful treatment or spontaneous remission of the infection.

G44.882 Headache associated with a disturbance of the metabolic state hypoxia due to

- diseases of the respiratory system (J00-J99)
- high altitude (W94)
- sleep apnoe (G47.3)
- hypercapnia, hyperventilation (R06.4)
- respiratory acidosis (E87.21)
- hypoglycaemia (E16)
- kidney dialysis (Y84.1)

Diagnostic criteria

- A. There are symptoms and/or signs of a metabolic disturbance.
- B. This is confirmed by appropriate laboratory investigations.
- C. The headache intensity and/or frequency is related to variations in the metabolic disturbance with a specified time-lag.
- D. The headache disappears within 7 days after normalization of metabolic state.

R51 Headache

Non-recommended residual category, when no other code from this classification can be used.