

Headache: classification

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Summary

Given the range of disorders that produce headache, a systematic approach to classification and diagnosis is an essential for clinical management. For the past 15 years, the first edition of the diagnostic criteria of the International Headache Society, ICHD-1 (Headache Classification Committee of the International Headache Society 1988), has been the accepted standard for headache diagnosis. The second edition of the International Classification of Headache Disorders, ICHD-2 (Headache Classification Subcommittee of the International Headache Society 2004), reflects our improved understanding of some previously included disorders as well as the development of criteria for previously excluded disorders.

Like its predecessor, the ICHD-2 separates headache into primary and secondary disorders. The four categories of primary headaches are migraine, tension-type headache, cluster headache and other trigeminal autonomic cephalalgias, and other primary headaches. There are nine categories of secondary headache. Important changes in the ICHD-2 include a restructuring of the criteria for migraine, a new subclassification of tension-type headache, introduction of the concept of trigeminal autonomic cephalalgias, and addition of previously unclassified primary headaches. Several disorders were eliminated or reclassified. Herein we present an overview of the ICHD-2, highlighting the primary headache disorders and their diagnostic criteria. We conclude by presenting an approach to headache diagnosis based on these criteria.

Introduction

Headache is one of the most common types of recurrent pain as well as one of the most frequent symptoms in neurology (Scher et al 1999). Although almost everyone gets occasional headaches, there are well-defined headache disorders that vary in incidence, prevalence and duration (Rasmussen 1995). These disorders are usually divided into two broad categories: primary headache and secondary headache disorders. In secondary disorders, headaches are attributed to another condition, such as brain tumour or head injury; for the primary disorders the headache is not due to another condition.

Given the range of disorders that can produce headache, a systematic approach to headache classification and diagnosis is an essential prelude to appropriate management. For the past 15 years, the first edition of the diagnostic criteria of the International Headache Society (ICHD-1) has been the accepted standard (Headache Classification Committee of the International Headache Society 1988). The second edition of the International Classification of Headache Disorders (ICHD-2) reflects our improved understanding of some disorders and the identification of new disorders (Headache Classification Subcommittee of the International Headache Society 2004). Like its predecessor, the ICHD-2 separates headache into primary and secondary disorders.

The ICHD-1 and ICHD-2 have established uniform terminology and consistent operational diagnostic criteria for the full range of headache disorders around the world. This has facilitated epidemiological studies and the multinational clinical trials that provide the basis for the current research and treatment guidelines (Tfelt-Hansen et al 2000). In this chapter, we will first present an overview of the ICHD-2 (Headache Classification Subcommittee of the International Headache Society 2004). We will discuss the classification of the primary headache disorders. We will review the classification of daily or near-daily primary headaches. Finally we will briefly discuss the classification of the secondary headache disorders.

The ICHD-2: an overview

Table 54.1 presents an overview of the ICHD-2. The four categories of primary headaches are 1.0, migraine; 2.0, tension-type headache; 3.0, cluster headache and other trigeminal autonomic cephalalgias; and 4.0, other primary headaches. There are nine categories of secondary headache (against eight in the ICHD-1), which are headache attributed to 5.0, head and neck trauma; 6.0, cranial or cervical vascular disorders; 7.0, nonvascular intracranial disorders; 8.0, substance or its withdrawn; 9.0, infection; 10.0, disorder of homeostasis; 11.0, disorders of cranium, neck, eyes, ears, nose, sinuses, teeth, mouth, or other facial or cranial structures; 12.0, psychiatric disorders; and 13, cranial neuralgias and central causes of facial pain. Finally, there is a fourteenth category that includes headache not classifiable elsewhere.

The ICHD-2 includes operational rules, some of which are summarized and commented on here. These rules present the general principles for headache classification and are quoted or paraphrased from the criteria (Headache Classification Subcommittee of the International Headache Society 2004).

The ICHD-2 provides a hierarchy of diagnoses with varying degrees of specificity. Headache disorders are identified with three- or sometimes four-digit codes. The first digit specifies the major diagnostic categories indicated in Table 54.1 (i.e. migraine, 1.0; tension-type headache, 2.0, etc.). The second digit indicates a disorder within the category, for example migraine without aura (1.1) (Table 54.1). Subsequent digits permit more specific diagnosis for some types of headache. For example, headaches in a patient with familial hemiplegic migraine could be coded as migraine (1.0), migraine with aura (1.2), or most precisely as familial hemiplegic migraine (1.2.4). In this example, the three-digit code provides important information about aetiology, symptom profile, and treatment.

In clinical practice, patients should receive a diagnosis for each headache type they have experienced within the last year. The criteria suggest that the 1-year period prevalence should be used in epidemiological studies, while the life prevalence should be used in genetic studies.

Table 54.1 The ICHD-2 classification			
Class	Description	Class	Description
1 Migraine	1.1 Migraine without aura 1.2 Migraine with aura 1.3 Childhood periodic syndromes that are commonly precursors of migraine 1.4 Retinal migraine 1.5 Complications of migraine 1.6 Probable migraine	8 Headache attributed to a substance or its withdrawal	8.1 Headache induced by acute substance use or exposure 8.2 Medication overuse headache 8.3 Headache as an adverse event attributed to chronic medication 8.4 Headache attributed to substance withdrawal
2 Tension-type headache	2.1 Infrequent episodic tension-type headache 2.2 Frequent episodic tension-type headache 2.3 Chronic tension-type headache 2.4 Probable tension-type headache	9 Headache attributed to infection	9.1 Headache attributed to intracranial infection 9.2 Headache attributed to systemic infection 9.3 Headache attributed to HIV/AIDS 9.4 Chronic postinfection headache
3 Cluster headache and other trigeminal autonomic cephalalgias	3.1 Cluster headache 3.2 Paroxysmal hemicrania 3.3 Short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing (SUNCT) 3.4 Probable trigeminal autonomic cephalalgia	10 Headache attributed to disorder of homeostasis	10.1 Headache attributed to hypoxia and/or hypercapnia 10.2 Dialysis headache 10.3 Headache attributed to arterial hypertension 10.4 Headache attributed to hypothyroidism 10.5 Headache attributed to fasting 10.6 Cardiac cephalalgia 10.7 Headache attributed to other disorder of homeostasis
4 Other primary headaches	4.1 Primary stabbing headache 4.2 Primary cough headache 4.3 Primary exertional headache 4.4 Primary headache associated with sexual activity 4.5 Hypnic headache 4.6 Primary thunderclap headache 4.7 Hemicrania continua 4.8 New daily-persistent headache	11 Headache or facial pain attributed to disorder of cranium, neck, eyes, ears, nose, sinuses, teeth, mouth, or other facial or cranial structures	11.1 Headache attributed to disorder of cranial bone 11.2 Headache attributed to disorder of neck 11.3 Headache attributed to disorder of eyes 11.4 Headache attributed to disorder of ears 11.5 Headache attributed to rhinosinusitis 11.6 Headache attributed to disorder of teeth, jaws, or related structures 11.7 Headache or facial pain attributed to temporomandibular joint disorder 11.8 Headache attributed to other disorder of cranium, neck, eyes, ears, nose, sinuses, teeth, mouth, or other facial or cervical structures
5 Headache attributed to head and/or neck trauma	5.1 Acute post-traumatic headache 5.2 Chronic post-traumatic headache 5.3 Acute headache attributed to whiplash injury 5.4 Chronic headache attributed to whiplash injury 5.5 Headache attributed to traumatic intracranial haematoma 5.6 Headache attributed to other head and/or neck trauma 5.7 Postcraniotomy headache	12 Headache attributed to psychiatric disorder	12.1 Headache attributed to somatization disorder 12.2 Headache attributed to psychotic disorder
6 Headache attributed to cranial or cervical vascular disorders	6.1 Headache attributed to ischaemic stroke and transient ischaemic attack 6.2 Headache attributed to non-traumatic intracranial haemorrhage 6.3 Headache attributed to unruptured vascular malformations 6.4 Headache attributed to arteritis 6.5 Carotid or vertebral artery pain 6.6 Headache attributed to cerebral venous thrombosis 6.7 Headache attributed to other intracranial vascular disorders	13 Cranial neuralgias and central causes of facial pain	13.1 Trigeminal neuralgia 13.2 Glossopharyngeal neuralgia 13.3 Nervus intermedius neuralgia 13.4 Superior laryngeal neuralgia 13.5 Nasociliary neuralgia 13.6 Supraorbital neuralgia 13.7 Other terminal branch neuralgias 13.8 Occipital neuralgia 13.9 Neck–tongue syndrome 13.10 External compression headache 13.11 Cold stimulus headache 13.12 Constant pain caused by compression, irritation or distortion of cranial nerves or upper cervical roots by structural lesions 13.13 Optic neuritis 13.14 Ocular diabetic neuropathy 13.15 Head or facial pain attributable to herpes zoster 13.16 Tolosa–Hunt syndrome 13.17 Ophthalmoplegic ‘migraine’ 13.18 Central causes of facial pain 13.19 Other cranial neuralgia or other centrally mediated facial pain
7 Headache attributed to non-vascular intracranial disorder	7.1 Headache attributed to high cerebrospinal fluid pressure 7.2 Headache attributed to low cerebrospinal fluid pressure 7.3 Headache attributed to non-infectious inflammatory disease 7.4 Headache attributed to intracranial neoplasm 7.5 Headache attributed to intrathecal injection 7.6 Headache attributed to epileptic seizure 7.7 Headache attributed to Chiari malformation type 1 7.8 Syndrome of transient headache and neurological deficits with cerebrospinal fluid lymphocytosis 7.9 Headache attributed to other non-vascular intracranial disorder	14 Other headache, cranial neuralgia, central or primary facial pain	

The ICHD-2 requires that all distinct types of headache that the patient experiences should be diagnosed. A patient with a complicated headache may receive several separate diagnoses, for example migraine without aura (1.1) plus episodic tension-type headache (2.2) plus medication overuse (8.2). Also, for patients with more than one diagnosis, diagnoses should be listed in their order of importance to the patient.

Probable diagnostic categories, such as probable migraine, probable tension-type headache, and probable cluster headache, are applied to patients missing one of the features necessary for a diagnosis. If a headache fulfils both the full criteria for one disorder and also the criteria for a probable diagnosis, the full should be coded. For example, if a patient has a headache that fulfils criteria for probable migraine (1.6) and for frequent episodic tension-type headache (2.2), the patient should be diagnosed with frequent episodic tension-type headache (2.2).

Although some headache types include frequency in their diagnostic criteria (i.e. chronic migraine and chronic tension-type headache), the ICHD-2 does not specifically code frequency or severity. Frequency and severity may be specified parenthetically at the discretion of the examiner.

If a patient experiences a new kind of headache for the first time in close temporal relation to another disorder known to cause headache and the headache is attributed to that disorder, the headache should be coded as a secondary headache. Secondary headaches are classified based on aetiology, not on symptom profile.

Patients with pre-existing primary headaches sometimes experience exacerbations in close temporal relation to a known cause of headache. In this circumstance, the known cause of headache may have worsened the pre-existing primary headache. Alternatively, it may have caused a new type of secondary headache. A secondary headache is more likely if:

1. there is a very close temporal relation to the potentially causal disorder;
2. there is a marked exacerbation of the primary headache;
3. the evidence that the causal disorder can cause headaches is strong; or
4. there is improvement or disappearance of headache after relief from the causal disorder.

Headache diaries are recommended, particularly in patients with more than one headache type, to determine the symptom profiles of each type over a period of time.

Classification of the primary headaches

The ICHD-2 divides the primary headaches into four major categories, discussed in sequence below.

Migraine (1.0)

Migraine is a chronic neurological disorder characterized by episodic attacks of headache and associated symptoms. In western countries, the condition affects 12% of the adult population (Scher et al 1999). Migraine is a heterogeneous condition that results in a range of symptom profiles both within and among different individuals (Stewart et al 1994). Migraine is divided into six major categories, the two most important of which are migraine without aura (1.1) and migraine with aura (1.2) (Table 54.2).

Class	Description
1.1	Migraine without aura
1.2	Migraine with aura
1.2.1	Typical aura with migraine headache
1.2.2	Typical aura with non-migraine headache
1.2.3	Typical aura without headache
1.2.4	Familial hemiplegic migraine
1.2.5	Sporadic hemiplegic migraine
1.2.6	Basilar-type migraine
1.3	Childhood periodic syndromes that are commonly precursors of migraine
1.3.1	Cyclical vomiting
1.3.2	Abdominal migraine
1.3.3	Benign paroxysmal vertigo of childhood
1.4	Retinal migraine
1.5	Complications of migraine
1.5.1	Chronic migraine
1.5.2	Status migrainosus
1.5.3	Persistent aura without infarction
1.5.4	Migrainous infarction
1.5.5	Migraine-triggered seizures
1.6	Probable migraine
1.6.1	Probable migraine without aura
1.6.2	Probable migraine with aura

Migraine without aura (1.1)

Migraine without aura is a clinical syndrome characterized by headache features and associated symptoms (Box 54.1; Headache Classification Subcommittee of the International Headache Society 2004). According to the ICHD-2, if a patient fulfils criteria for more than one type of migraine, each type should be diagnosed. Criteria for migraine without aura can be met by various combinations of features; no single feature is required. Because two of four pain features are required, a patient with unilateral, throbbing pain may meet the criteria, but so does a patient with bilateral pressure pain if the pain is moderate and aggravated by physical activity. Similarly, only one of two possible associated symptom combinations is required.

Box 54.1 ICHD-2 diagnostic criteria for migraine without aura (1.1)

- A. At least five attacks^a fulfilling criteria B–D
- B. Headache attacks lasting 4–72 h^b and occurring on < 15 days/month^c (untreated or unsuccessfully treated)
- C. Headache has at least two of the following characteristics
 - 1. Unilateral location^d
 - 2. Pulsating quality^e
 - 3. Moderate or severe pain intensity
 - 4. Aggravation by or causing avoidance of routine physical activity (i.e. walking or climbing stairs)
- D. During headache at least one of the following
 - 1. Nausea and/or vomiting
 - 2. Photophobia and phonophobia^f
- E. Not attributed to another disorder^g

^a*Differentiating between migraine without aura and episodic tension-type headache may be difficult, therefore at least five attacks are required. The headaches of individuals who otherwise meet criteria for migraine without aura but who have fewer than five attacks should be coded 1.6.*

^b*If the patient falls asleep during migraine and wakes up without it, duration of the attack is until time of awakening. In children, attacks may last 1–72 h. (The evidence for untreated durations < 2 h in children should be corroborated by prospective diary studies.)*

^c*If attack frequency ≥ 15 days/month and if there is no medication overuse, code 1.1 and 1.5.1 chronic migraine.*

^d*Migraine headache is often bilateral in young children; an adult pattern of unilateral pain often emerges in late adolescence or early adult life. Migraine headache is usually frontotemporal. Occipital headache, whether unilateral or bilateral, is rare in children and calls for diagnostic caution; many cases are attributable to structural lesions.*

^e*Pulsating means throbbing or varying with the heartbeat at rest or with movement.*

^f*In young children, photophobia and phonophobia may be inferred from behaviour.*

^g*History, physical and neurological examinations do not suggest one of the disorders listed in groups 5–12, or history and/or physical and/or neurological examinations do suggest such disorder but it is ruled out by appropriate investigations, or such disorder is present but migraine attacks do not occur for the first time in close temporal relation to the disorder.*

Patients with nausea but not photophobia or phonophobia fill the requirements, as do patients without nausea or vomiting but with photophobia and phonophobia.

For migraine without aura, the ICHD-2 requires at least five lifetime attacks, which last from 4 to 72 h. If the patient falls asleep during migraine and wakes up without it, the duration of the attack is timed until time of awakening. In children, attacks may last 1–72 h, and in young children photophobia and phonophobia may be inferred from behaviour. If attack frequency is ≥ 15 days/month in a patient not overusing acute medications, the ICHD-2 establishes coding 1.5.1 chronic migraine (see also *Controversies in the classification of primary chronic daily headaches of long duration*, p. 000).

Migraine with aura and its subtypes (1.2)

The aura of migraine is characterized by focal neurological phenomena that usually proceed, but may accompany or occur in the absence of, the headache (Olesen et al 1990). Box 54.2 presents the features of typical aura according to the ICHD-2. Most aura symptoms develop over 5–20 min and last 20 min on average, rarely more than 60 min. Visual aura is the most common form (Jensen et al 1986). The aura often has a hemianoptic distribution in the shape of a crescent with a bright, ragged edge, which scintillates. Scotoma, photopsia or phosphenes, fortification spectra, and other visual manifestations may

Box 54.2 ICHD-2 diagnosis criteria for typical aura

- A. At least two attacks fulfilling criteria B–E
- B. Fully reversible visual and/or sensory and/or speech symptoms but no motor weakness
- C. At least two of the following
 - 1. Homonymous visual symptoms including positive features (i.e. flickering lights, spots and lines) and/or negative features (i.e. loss of vision) and/or unilateral sensory symptoms including positive features (i.e. pins and needles) and/or negative features (i.e. numbness)
 - 2. At least one symptom develops gradually over ≥ 5 min and/or different symptoms occur in succession
 - 3. Each symptom lasts ≥ 5 min and ≤ 60 min
- D. Headache that meets criteria B–D for migraine without aura (1.1) begins during the aura or follows aura within 60 min
- E. Not attributed to another disorder

occur. Visual distortions, such as metamorphopsia, micropsia or macropsia are more common in children (Silberstein & Young 1995).

Sensory symptoms are the second most common aura and occur in about one-third of patients with migraine with aura (Silberstein & Young 1995). These symptoms usually consist of numbness (a negative symptom) and tingling or paraesthesia (positive symptoms). The distribution is often cheiro-oral (face and hand) but can be hemisensory. Hemi motor weakness, dysphasia, and incoordination with other signs of brain stem dysfunctions can occur, being by far less common. Rarely, changes in level of consciousness are present (Russel & Olesen 1996).

Migraine with typical aura is the most common form of migraine with aura. The aura's duration is no less than 5 min and no longer than 60 min (and usually around 20 min) (Headache Classification Subcommittee of the International Headache Society 2004). If more than one aura symptom is present (e.g. visual and sensory symptoms), the accepted duration is proportionally increased (Box 54.2).

A number of non-migrainous headaches, including cluster headache, chronic paroxysmal hemicrania, and hemicrania continua have been described to be unusually associated with aura (Matharu & Goadsby 2001, Peres et al 2002). The ICHD-2 classifies such headaches under the code typical aura with non-migraine headache (1.2.2). In these cases, the aura occurs in close association with a headache that does not meet criteria for migraine without aura (1.1).

Some patients present otherwise typical aura not associated with headache (Lipton et al 2004). The ICHD-2 codes this phenomenon as typical aura without headache (1.2.3), a disorder that often occurs in middle-aged men (Lipton et al 1993). Within the same patient, some auras may occur without headache and others with a headache. The distinction between this entity and transient ischaemic attacks may require investigation, especially if aura begins after age 40; if negative features are predominant (i.e. hemianopia), or if the aura is prolonged or very short, other causes should be ruled out (Lipton et al 1993).

A rare form of migraine with aura, familial hemiplegic migraine (1.2.4), is an autosomal dominant disorder (Haan et al 1997). This is the first migraine syndrome to be linked to a specific genetic set of polymorphisms (Carrera et al 2001). The ICHD-2 criteria for familial hemiplegic migraine include the following (Headache Classification Subcommittee of the International Headache Society 2004):

1. the presentation fulfils criteria for migraine with aura,
2. the aura includes some degree of hemiparesis and may be prolonged, and
3. at least one first-degree relative has identical attacks.

Cerebellar ataxia may occur in 20% of familial hemiplegic migraine sufferers.

The onset of the paresis may be abrupt, resembling a stroke. Weakness usually lasts less than 1 h (Ophoff et al 1996, Staehelin-Jensen et al 1981). A person with familial hemiplegic migraine may develop migraine with aura when adult and migraine without aura later in life.

Two loci for familial hemiplegic migraine have been identified. Mutations in the *CACNA1A* gene on chromosome 19 account for about 50% of cases. This gene codes for a neuronal P/Q-type calcium channel. A second locus on chromosome 1, which codes for a sodium–potassium ATPase, has been identified. Some families do not link to either of these loci, indicating that there is at least one additional locus (De Fusco et al 2003, Ophoff et al 1996).

A new subtype of migraine with aura is presented in the ICHD-2: sporadic hemiplegic migraine (1.2.5). Patients with sporadic hemiplegic migraine have migraine with aura including motor weakness but do not have an affected first- or second-degree relative (Thomsen et al 2003).

Basilar-type migraine (1.2.6) is seen mostly in young adults. Patients with familial hemiplegic migraine have basilar-type symptoms in 60% of cases. Therefore basilar-type migraine should be diagnosed only when no motor weakness is present. A distinguishing feature of basilar-type migraine is a symptom profile that suggests posterior fossa involvement (Panayiotopoulos 1999). Symptoms are often bilateral. To be classified as basilar-type migraine according to the ICHD-2, two or more fully reversible aura symptoms of the following types must be documented: dysarthria, vertigo, tinnitus, decreased hearing, double vision, ataxia, decreased level of consciousness, simultaneous bilateral visual symptoms in both the temporal and the nasal field of both eyes, and simultaneous bilateral paraesthesias. The headache meets criteria for migraine without aura (Box 54.1; Headache Classification Subcommittee of the International Headache Society 2004).

Childhood periodic syndromes that are commonly precursors of migraine (1.3)

Childhood periodic syndromes that are commonly precursors of migraine (1.3) include the cyclic vomiting syndrome (1.3.1), abdominal migraine (1.3.2), and benign paroxysmal vertigo (1.3.3) (Hosking 1988).

Cyclic vomiting syndrome (1.3.1) was reported to affect 2–2.5% of schoolchildren in some studies (Fleischer 1999). It is characterized by recurrent episodes of unexplained nausea and vomiting. The attacks usually last a few hours and are characterized by episodic attacks of vomiting and intense nausea, stereotypical in the individual patient, in a child who is symptom-free between attacks. Vomiting occurs at least four times per hour for at least 1 h, and no signs of gastrointestinal disease can be found.

Abdominal migraine (1.3.2) is a common disorder that may affect from 8 to 12% of all schoolchildren, and is characterized by recurrent attacks of abdominal pain associated with anorexia, nausea, and sometimes vomiting (Abu-Arafeh & Russel 1995). No abnormalities can be elicited on physical or subsidiary examination. The abdominal pain has all the following characteristics:

- midline location, periumbilical or poorly localized;
- dull or just sore quality; and
- moderate or severe pain intensity.

During abdominal pain, there are at least two of the following:

- anorexia,
- nausea,

- vomiting, and
- pallor.

About 20% of schoolchildren between 5 and 15 years of age reported suffering from episodes of vertigo over a year (Drigo et al 2001). The most important cause of this presentation is benign paroxysmal vertigo (1.3.3), a disorder characterized by transient episodes of vertigo. Children with this disorder develop full migraine later in life. Criteria required for diagnoses are as follow.

- At least five attacks and duration from minutes to hours.
- Unilateral throbbing headache may be associated with some attacks.
- Multiple episodes of severe vertigo, and often nystagmus or vomiting without warning, resolving spontaneously.
- Normal neurological examination; audiometric and vestibular functions normal between attacks.
- Normal electroencephalogram.

Retinal migraine (1.4)

Retinal migraine is characterized by repeated attacks of monocular scotoma or blindness lasting less than 1 h. Episodes of transient monocular visual loss may not be temporally related to the patient's headache. Patients are usually less than 44 years old and often have a history of other manifestations of migraine. It is more common than ophthalmoplegic migraine, its prevalence being around 1 in each 200 migraine sufferers (Troost & Zagami 2000). Retinal transient ischaemic attacks, as the most likely differential diagnosis, must be ruled out by appropriate investigation. The ICHD-2 criteria require at least two attacks of the following.

1. Fully reversible monocular positive visual phenomena, scotomata or blindness confirmed by examination during attack or (after proper instruction) by patient's drawing of monocular field defect during an attack.
2. Headache that meets criteria for migraine without aura (1.1), and that begins during or following the visual symptoms within 60 min.
3. Normal ophthalmological examination outside the attack.

Appropriate investigations exclude other causes of transient monocular blindness. A recent review suggests that many patients with monocular 'aura' experience retinal infarction of migrainous origin. Headache in these patients should be classified as migrainous infarction (1.5.4) (Solomon & Grosberg 2003).

Complications of migraine (1.5)

Chronic migraine (1.5.1) (see *Controversies in the classification of primary chronic daily headaches of long duration*, p. 000), according to the ICHD-2, occurs in those patients with migraine headaches at least 15 days/month for at least 3 months and with no acute medication overuse. Most cases of chronic migraine start as episodic migraine without aura (Silberstein & Lipton 2001). Therefore chronicity is regarded as a complication of episodic migraine. If medication overuse (ergotamine, a triptan, opioids, or combination medication on > 10 days/month, or simple analgesics on > 15 days/month) is present, the headaches might represent medication overuse headache (8.2), a diagnosis that can be assigned with confidence if headaches improve after medication overuse ceases. Otherwise a diagnosis of probable chronic migraine with probable medication overuse should be assigned. The classification of this disorder is discussed in the text on controversies in headache classification.

Status migrainosus (1.5.2) refers to an attack of migraine with the headache phase lasting more than 72 h despite treatment. Initially the attack resembles an unremarkable attack of migraine. Almost all attacks are accompanied by nausea and vomiting to the point of dehydration. The patient often seeks care in the emergency department (Couch & Diamond 1983). The attack is debilitating. Non-debilitating attacks lasting more than 72 h are coded 1.6 (probable migraine without aura).

Persistent aura without infarction (1.5.3) occurs when aura symptoms persist for more than 2 weeks without radiographic evidence of infarction. It is an unusual form of migraine that is being first included in the ICHD-2 (Bento & Esperanca 2000).

Migrainous infarction (1.5.4) is characterized by one or more migrainous aura symptoms not fully reversible within 7 days and/or associated with neuroimaging confirmation of ischaemic infarction. A few clues help to distinguish this part of the clinical spectrum of migraine from other causes of stroke:

1. the neurological deficit must exactly mimic the migrainous aura of previous attack;
2. stroke must occur during the course of a typical migraine attack; and
3. other cases of stroke must be excluded (Rothrock et al 1988).

Migraine and seizures are comorbid disorders (Bigal et al 2003). Headaches are common in the postictal period, but epilepsy can occur triggered by a migraine (migralepsy). The criteria for migraine-triggered seizures (1.5.5) require a seizure fulfilling diagnostic criteria for one type of epileptic attack occurs during or within 1 h of a migraine aura.

Probable migraine (1.6)

When the ICHD-1 criteria are applied to patients experiencing headaches with migrainous features, between 10 and 45% of them fail to meet the criteria for migraine (Rains et al 2001). If just one migraine criterion is missing, these patients receive the diagnosis of probable migraine (1.6, formerly migrainous headache). A recent study showed that probable migraine was as prevalent as migraine within a health plan population (Bigal et al 2004).

There are two subtypes of probable migraine: probable migraine without aura (just one criterion for migraine without aura is missing) and probable migraine with aura (one criterion for migraine with aura is missing).

Tension-type headache (2.0)

Tension-type headache is the most prevalent form of primary headache, with 1-year period prevalences ranging from 31 to 74% (Rasmussen et al 1991, Scher et al 1999). For the individual sufferer, tension-type headache is less disabling than migraine or cluster headache, but from a social perspective it is important because of its high prevalence (Schwartz et al 1998). The ICHD-1 distinguished two forms of tension-type headache: the episodic form (< 15 attacks per month) and the chronic form (> 15 attacks per month) (Headache Classification Committee of the International Headache Society 1988). The ICHD-2 distinguishes three forms of tension-type headache: infrequent episodic (2.1, < 1 attack per month), frequent episodic (2.2, 1–14 attacks per month), and chronic (2.3, > 15 attacks per month) (Headache Classification Subcommittee of the International Headache Society 2004). Each of these forms can be further

Box 54.3 ICHD-2 diagnostic criteria for tension-type headache

- A. At least 10 episodes fulfilling criteria B–E; headache < 1 day/month (episodic infrequent), 1–14 days/month (episodic frequent), or ≥ 15 days/month (chronic)
 - B. Headache lasting from 30 min to 7 days
 - C. At least two of the following pain characteristics
 1. Pressing or 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 may be present
 - E. Not attributed to another disorder
- 2.X.1.^a Associated with pericranial tenderness
 - A. Fulfils criteria for 2.X
 - B. Increased tenderness on pericranial manual palpation
 - 2.X.2.^a Not associated with pericranial tenderness
 - A. Fulfils criteria for 2.X
 - B. Not associated with increased pericranial tenderness

^aX replaced by the corresponding digit of infrequent episodic (1), frequent episodic (2), or chronic (3).

classified, using the third digit, into forms associated or not associated with pericranial tenderness.

The diagnostic criteria for tension-type headache are presented in Box 54.3. The criteria were designed to distinguish between tension-type headache and migraine. In contrast to migraine, the main pain features of tension-type headache are bilateral location, non-throbbing quality, mild to moderate intensity, and lack of aggravation by physical activity. The pain is not associated with nausea, although just one of photo- or phonophobia does not exclude the diagnosis.

Pericranial tenderness recorded by manual palpation is a significant abnormal finding in some patients with tension-type headache. When present, it is associated with the intensity and frequency of headache and may be exacerbated during headache attacks. The clinical diagnostic value of electromyography and pressure algometry is limited (Jensen & Fuglsang-Frederiksen 1994).

Like chronic migraine (1.5.1), chronic tension-type headache (2.3) is not diagnosed in patients using acute medication for more than 10 days per month (see *Controversies in the classification of primary chronic daily headaches of long duration*, p. 000). Those cases often meet criteria for medication overuse headache (8.2). Cases that phenotypically resemble chronic tension-type headache but begin de novo should be classified as new daily persistent headache (4.8).

It may be difficult to differentiate episodic tension-type headache from migraine without aura or probable migraine without aura, particularly when associated symptoms are poorly described or if more than one headache type is present.

Probable tension-type headache (4.4)

Probable tension-type headache (4.4) is diagnosed if the headache fulfils all but one of the criteria for tension-type headache and does not fulfil criteria for migraine without aura.

Cluster headache and other trigeminal autonomic cephalalgias (3.0)

As a group, the trigeminal autonomic cephalalgias are characterized by relatively short-lived attacks of unilateral pain accompanied by autonomic dysfunction ipsilateral to the pain.

Cluster headache (3.1)

Cluster headache is a form of intermittent, short-lived, excruciating, unilateral head pain associated with autonomic dysfunction. Prevalence estimates range from 0.1 to 0.4%. Men are affected more than women (ratio 4.5–6.7:1.0), and the mean age of onset is 27–31 years (Dodick et al 2000, Kudrow 1991).

Cluster headache is a severe, recurring, chronic disease. The pain of cluster headache is described variously as sharp, boring, drilling, knifelike, piercing, stabbing, but generally not throbbing like migraine. Pain usually reaches its peak intensity in 10–15 min and remains excruciatingly intense for an average of 1 h. The duration usually ranges from 15 to 180 min. During pain exacerbations, autonomic signs are usually obvious to observers and distressing to patients. During the attack, patients find it difficult to lie down, as it aggravates the pain. The pain is so excruciating that after an attack the patient remains exhausted for some time (Goadsby & Lipton 1997).

The diagnostic criteria for cluster headache are displayed in Box 54.4. Cluster headache is divided in two forms. The term episodic cluster headache (3.1.1) indicates that remissions occur. In this form of cluster headache, attacks occur in periods lasting from 7 days to 1 year, separated by pain-free periods lasting 1 month or more. Approximately 85% of individuals affected by cluster headache have the episodic form. Patients with chronic cluster headache (3.1.2) experience daily or near-daily headaches for more than 1 year without remission or with remissions that last less than a month. The chronic form of the disease can evolve from the episodic form (secondary chronic form), or it may develop de novo as primary chronic cluster headache. Occasionally, patients begin with the chronic form and develop the episodic cluster. Chronic cluster headache occurs in approximately 15% of sufferers, is unremitting from onset (primary chronic cluster) in 10%, and evolves from the episodic form in 5%.

Some patients have described both cluster headache and trigeminal neuralgia, and their pain received the denomination of cluster tic

syndrome (Goadsby & Lipton 1997). According to the ICHD-2, they should receive both diagnoses.

Paroxysmal hemicranias (3.2)

Paroxysmal hemicranias (3.2) are a group of unusual disorders that have three main features:

1. short-lasting, strictly unilateral pain,
2. symptoms of parasympathetic activation ipsilateral to the pain, and
3. absolute response to indomethacin (Broske et al 1993, Sjaastad & Dale 1974).

The ICHD-2 includes two variants that are analogous to cluster headache: episodic paroxysmal hemicrania (3.2.1) and chronic paroxysmal hemicrania (3.2.2). Like cluster headache, these disorders are distinguished by the presence or absence of pain-free periods lasting 1 month or more. Criteria require at least 20 attacks with the following features.

- Attacks of severe unilateral orbital, supraorbital or temporal pain lasting 2–30 min.
- Attack frequency above five a day for more than half of the time, although periods with lower frequency may occur.
- Pain is associated with at least one autonomic symptom or sign on the pain side (the same described to cluster headache).
- Headache is stopped completely by indomethacin.

Similarly to with cluster headache, some patients with chronic paroxysmal hemicrania and trigeminal neuralgia have been described (chronic paroxysmal hemicrania tic syndrome) (Monzillo et al 2000).

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

The SUNCT syndrome (3.3) is one of the rarest primary headaches, with an impressive clinical presentation characterized by short-lasting unilateral attacks of moderately severe pain, of more intensity around the eye. Paroxysms peak in intensity within 3 s and are accompanied by several associated symptoms, the most prominent being ipsilateral conjunctival injection and lachrymation (Goadsby et al 2001). The criteria require at least 20 attacks of unilateral, orbital, supraorbital or temporal, stabbing or throbbing pain lasting from 5 to 240 s. Attack frequency ranges from 3 to 200 per day. Pain is accompanied by conjunctival injection and lachrymation.

Probable trigeminal autonomic cephalgia (3.4)

Those headache attacks that are believed to be a form of trigeminal autonomic cephalgia, but that do not quite meet the operational diagnostic criteria for any of the forms described in the related chapter, should be classified as probable trigeminal autonomic cephalgia.

Other primary headaches (4.0)

This group includes a variety of headache disorders that are not associated with a structural lesion. These headaches are heterogeneous from a clinical perspective. Some of them may mimic secondary headaches and need to be carefully evaluated by imaging or other appropriate tests. Some, such as hypnic headache, primary thunderclap headache, hemicrania continua, and new daily persistent headache, were not included in the ICHD-1.

Box 54.4 ICHD-2 diagnostic criteria for cluster headache

- A. At least five attacks fulfilling criteria B–D
- B. Severe or very severe unilateral orbital, supraorbital and/or temporal pain lasting 15–180 min untreated for more than half of the period (or time if chronic)
- C. Headache accompanied by at least one of the following symptoms or signs that have to be present on the side of the pain
 1. Conjunctival injection, lachrymation, or both
 2. Nasal congestion, rhinorrhoea, or both
 3. Eyelid oedema
 4. Forehead and facial sweating
 5. Miosis, ptosis, or both
 6. Headache associated with a sense of restlessness or agitation
- D. Frequency of attacks: from one every other day to eight per day for more than half of the period (or time if chronic)
- E. Not attributed to another disorder

Primary stabbing headache (4.1)

Formerly called jabs and jolts headache, primary stabbing headache consists of episodic and localized jabs of pain in the head that occur spontaneously in the absence of secondary causes. Pain is exclusively or predominantly felt in the distribution of the first division of the trigeminal nerve (orbit, temple and parietal area). Pain lasts for up to a few seconds and recurs at irregular intervals ranging from one to many per day. Other features, such as autonomic signs, are lacking (Dangond & Spierings 1993, Pareja et al 1996). Primary stabbing headache is more prevalent in migraine sufferers compared with in matched control subjects (Sjaastad et al 2003).

Primary cough headache (4.2)

In primary cough headache, headache is elicited by cough in the absence of an underlying disease (such as a brain mass or hind brain malformation). The headache is suddenly triggered by coughing or Valsalva manoeuvre, and similar headache does not occur without coughing or straining (Pascual et al 1996).

Diagnostic neuroimaging is mandatory to differentiate secondary and primary forms of cough headache. Special attention to the posterior fossa and base of the skull is required to exclude disorders in these locations.

Primary exertional headache (4.3)

Like primary cough headache, in primary exertional headache, headache is triggered by physical activity other than sexual activity (4.4) or Valsalva (4.2). Headache is often throbbing, lasting from 5 min to 48 h and occurring during physical exertion. The same headache does not occur without exertion. After the first occurrence of an exertional headache of sudden onset, appropriate investigations to exclude subarachnoid haemorrhage and mass lesions are required (Green 2001). Rarely, exertional headache may be a manifestation of angina, even in the absence of chest pain (Lipton et al 1997).

Primary headache associated with sexual activity (4.4)

Formerly benign sexual headache (coital headache), this headache is precipitated by sexual activity. It often begins as a dull bilateral ache as sexual excitement increases and suddenly becomes intense at orgasm (Lance 1976). The ICHD-2 distinguishes two subforms: preorgasmic (frequently bilateral and pressure headache) and orgasmic (explosive in nature) headaches.

Diagnosis requires appropriate investigation to rule out secondary causes, including subarachnoid haemorrhage and mass lesions.

Hypnic headache (4.5)

The hypnic headache syndrome is a primary headache disorder of the elderly, usually occurring in individuals over the age of 60 (Newman et al 1990). Like cluster headache, it is characterized by short-lived attacks (typically 30 min) of nocturnal head pain that awaken the patient at a consistent time each night, sometimes from rapid eye movement sleep. Unlike cluster headache, hypnic headache pain is usually bilateral, throbbing or diffuse, and lacks the unilateral orbital or periorbital knifelike intense pain as well as the autonomic features. Unilateral headache does not exclude the diagnosis (Dodick et al 1998).

Criteria require that headache has onset during and awakens patient from sleep, and does not occur at other times. Headache has at least two of the following characteristics.

1. Occurs more than 15 times per month.
2. Lasts at least 15 min after waking.
3. Onset after the age of 50.

No autonomic symptoms are present and no more than one migrainous feature including nausea, photophobia and phonophobia. This disorder is extremely responsive to lithium therapy.

Primary thunderclap headache (4.6)

Under this rubric, the ICHD-2 classifies those cases of severe headache that mimic the pain of ruptured cerebral aneurysm. Head pain is sudden and very severe, peaking in intensity often in less than 1 min. Pain lasts from 1 h to several days and may recur within the first week after onset, but does not recur regularly over subsequent weeks or months (Headache Classification Subcommittee of the International Headache Society 2004). The diagnosis can be established only when brain imaging and cerebrospinal fluid examination satisfactorily exclude subarachnoid haemorrhage. Depending on the clinical profile, magnetic resonance angiography may be warranted to exclude an expanding but unruptured aneurysm, which can produce this picture (Dodick et al 1999).

Hemicrania continua (4.7)

Hemicrania continua is a continuous unilateral headache defined by its absolute response to indomethacin. Hemicrania continua exists in both continuous and remitting forms. The continuous variety can be subclassified into an evolutive, unremitting form that arises from the remitting form, and an unremitting form characterized by continuous headache from the onset (Sjaastad & Spierings 1984).

The ICHD-2 requires:

1. unilateral headache without side shift,
2. daily headache without pain-free periods,
3. complete response of headache to indomethacin, and
4. at least one autonomic feature in association with exacerbations of pain on the affected side.

Although one of the essential features of hemicrania continua is unilateral headache, some bilateral or alternating-side cases have been reported (Bordini et al 1991). The female:male ratio of typical cases is 2.8:1.

Autonomic symptoms occur during painful exacerbations but are less prominent than in cluster headache and chronic paroxysmal hemicrania. Symptoms of ocular discomfort and a feeling of sand in the eye have been described in patients with hemicrania continua. Migrainous features are common in hemicrania continua (Bordini et al 1991, Sjaastad & Spierings 1984).

The requirement for an absolute response to indomethacin is controversial. Because some patients do not tolerate indomethacin in the required doses and others do not respond, this requirement creates a group of cases phenotypically identical to hemicrania continua but that cannot be classified.

New daily persistent headache (4.8)

Daily or near-daily primary headache that begins without a history of evolution from episodic headache and then persists is traditionally classified as new daily persistent headache (Silberstein & Lipton

2001). According to the ICHD-2, new daily persistent headache pain resembles a tension-type headache, although some migrainous features are allowed. The pain is typically pressing, tightening in quality, of mild to moderate intensity, bilateral in location, and may worsen with routine physical activity. There may be photophobia, phonophobia, or mild nausea.

Criteria require that the headache is present more than 15 days/month for at least 3 months. At least two of the following pain characteristics must be present.

- Pressing or tightening (non-pulsating) quality.
- Mild or moderate intensity (may inhibit but does not prohibit activities).
- Bilateral location.
- No aggravation by walking stairs or similar routine physical activity.
- Both of the following: no more than one of photophobia, phonophobia, or mild nausea; and no moderate or severe nausea and no vomiting.
- Use of analgesics or other acute medication on no more than 10 days/month (see *Controversies in the classification of primary chronic daily headaches of long duration*, p. 000).

In practice, headaches with migraine features of new and persistent onset are often classified as new daily persistent headache but do not meet the ICHD-2 criteria (Bigal et al 2002).

The secondary headaches

Discussing the classification of the secondary headaches in depth is beyond the scope of this chapter. In brief, the classification of all secondary headaches follows the same format (Headache Classification Subcommittee of the International Headache Society 2004). The headache has at least one of the following characteristics specified.

- The secondary disorder known to be able to cause headache has been demonstrated.
- Headache occurs in close temporal relation to the secondary disorder and/or there is other evidence of a causal relationship.
- Headache is greatly reduced or disappears within 3 months (this may be shorter for some disorders) after successful treatment or spontaneous remission of the causative disorder. There are exceptions to this general rule, for example chronic post-traumatic headache does not disappear 3 months after the trauma. We will briefly discuss their classification.

Headache attributed to head and/or neck trauma (5.0)

This category includes headaches that occur for the first time in close temporal relation to a known trauma (Packard 1999). If there is remission within 3 months after the trauma, the headache should be classified as acute post-traumatic headache. Otherwise, chronic post-traumatic headache is the diagnosis. The same rule applies to acute and chronic postwhiplash injury headache. The ICHD-2 also classifies under this group those headaches secondary to intracranial hematoma and post craniotomy.

Headache attributed to cranial or cervical vascular disorder (6.0)

This category encompasses a large group of headaches that fulfil the following criteria: symptoms and/or signs of vascular disorder,

appropriate investigations indicate the vascular disorder, and the headache develops in close relationship with the vascular disorder. This group includes headache related to:

1. ischaemic stroke and transient ischaemic attacks (Gorelick et al 1986);
2. non-traumatic intracranial haemorrhage (Schuaib et al 1989);
3. unruptured vascular malformations (Bassi et al 1991);
4. arteritis, including giant cell arteritis (Solomon & Cappa 1987);
5. carotid or vertebral artery pain (including arterial dissection, postendarterectomy headache, etc.) (Ramadan et al 1991);
6. cerebral venous thrombosis (Boussier & Ross Russell 1997); and
7. other intracranial vascular disorders, including cerebral autosomal dominant arteriopathy with subcortical infarcts and leukoencephalopathy (CADASIL), mitochondrial encephalopathy, lactic acidosis and stroke-like episodes (MELAS), etc. (Chabriat et al 1995).

In many of these conditions, such as ischaemic or haemorrhagic stroke, headache may be unrecognized because of focal signs and/or disorders of consciousness. In others, such as subarachnoid haemorrhage and giant cell arteritis, headache may be the most prominent symptom and an initial warning symptom.

Headache attributed to non-vascular intracranial disorder (7.0)

This category includes an extensive and heterogeneous group of disorders (Bartleson et al 1981, Ramadan 1996, Wall & George 1991):

1. high cerebrospinal fluid pressure,
2. low cerebrospinal fluid pressure,
3. non-infectious inflammatory diseases,
4. intracranial neoplasm,
5. headache related to intrathecal injections,
6. postseizure headache,
7. Chiari malformation type 1, and
8. syndrome of transient headache and neurological deficits with cerebrospinal fluid lymphocytosis.

Headache attributed to a substance or its withdrawal (8.0)

When new headaches occur in close temporal relation to substance use or withdrawal, they are coded to this group. The ICHD-2 criteria classify in this group those headaches following acute exposure to the following (Altura et al 2000, Iversen et al 1989).

1. Nitric oxide donor substances
2. Phosphodiesterase inhibitor
3. Carbon monoxide
4. Alcohol
5. Food components and additives
6. Monosodium glutamate
7. Cocaine
8. Cannabis
9. Other acute substances

In addition, chronic medication overuse is a risk factor for the development of chronic daily headache (Diener & Tfelt-Hansen 1993, Rapoport 1988a). Using the ICHD-2, if a patient has a frequent headache associated with medication overuse and otherwise meets criteria for chronic migraine, a diagnosis of probable chronic migraine and probable medication overuse headache should be assigned.

Definite diagnosis of medication overuse requires that headaches remit or improve when the overused medication is withdrawn. Prior to withdrawal, the use of the 'probable' term exemplifies the difficulty of causal attribution (see *Controversies in the classification of primary chronic daily headaches of long duration*, p. 000).

Headache attributed to infection (9.0)

This is a very straightforward group where headache secondary to intracranial and extracranial (systemic) infections are classified (Gomez-Arada et al 1997). This group also includes headaches related to HIV/AIDS and chronic postinfectious headaches.

Headache attributed to disorder of homeostasis (10.0)

This group of headaches was formerly referred to as headaches associated with metabolic or systemic disease. They include the following headaches (Antoniazzi et al 2003, Moreau 1988).

1. Headache attributed to hypoxia and/or hypercapnia (high altitude, diving, or sleep apnea)
2. Dialysis
3. Arterial hypertension
4. Headache attributed to hypothyroidism
5. Headache attributed to fasting
6. Cardiac cephalgia
7. Headache attributed to other disturbance of homeostasis

Headache or facial pain attributed to disorder of cranium, neck, eyes, ears, nose, sinuses, teeth, mouth, or other facial or cranial structures (11.0)

This is a very heterogenous group classifying headache and facial pain due to disease of the cranium, neck, and facial structures (Sjaastad et al 2002). Cranial neuralgias are not classified under this chapter. The ICHD-2 includes criteria for cervicogenic headache, which are shown in Box 54.5.

Headache attributed to psychiatric disorder (12.0)

This group provides a link to classify those extremely rare headaches that are causally attributable to a psychiatric disorder. The headache may be attributed to a somatization disorder or to a psychotic disorder. This should be distinguished from psychiatric comorbidities where a headache disorder (e.g. migraine) and a psychiatric disorder

(e.g. depression) occur together in the same person (Guidetti et al 1998).

Cranial neuralgias and central causes of facial pain (13.0)

Finally, the last chapter of the ICHD-2 codes the cranial neuralgias and facial pain, including the following (Rushton et al 1981, Terrence & Jensen 2000).

1. Trigeminal neuralgia
2. Glossopharyngeal neuralgia
3. Nervus intermedius neuralgia
4. Superior laryngeal neuralgia
5. Nasociliary neuralgia (Charlin)
6. Supraorbital neuralgia
7. Other terminal branch neuralgias
8. Occipital neuralgia
9. Neck-tongue syndrome
10. External compression headache
11. Cold stimulus headache
12. Constant pain caused by compression, irritation or distortion of cranial nerves or upper cervical roots by structural lesions
13. Optic neuritis
14. Ocular diabetic neuropathy
15. Herpes zoster
16. Tolosa-Hunt syndrome
17. Ophthalmoplegic migraine
18. Central causes of facial pain

Criteria for trigeminal neuralgia, the prototype of a cranial neuralgia, are summarized in Box 54.6.

Controversies in the classification of primary chronic daily headaches of long duration

Primary chronic daily headache is defined as a group of primary headaches that occur more than 15 days/month (or 180 days/year), with a duration of at least 4 h/day (Silberstein & Lipton 2001). Chronic daily headache is one of the more frequent headache presentations to headache speciality care centres (Bigal et al 2002, Diamond 2000). Chronic daily headache has a 1-year prevalence of 4–5% in the general population (Scher et al 1998).

Several studies reported difficulties using the 1988 International Headache Society system to classify chronic daily headache (Bigal et al 2002, Sanin et al 1994). As a consequence, a few proposals for this classification have emerged. Of these proposals, the Silberstein

Box 54.5 ICHD-2 classification of cervicogenic headache

- A. Pain perceived in one or more regions of the head or face, referred to the head from a source in the neck
- B. Clinical, laboratory and/or imaging evidence of a lesion within the cervical spine or in the soft tissues of the neck known to be, or generally accepted as, a valid cause of headache
- C. Evidence that the pain can be attributed to the neck disorder, based on at least one of the following criteria
 1. Demonstration of clinical signs that implicate a source of pain in the neck
 2. Abolition of headache following diagnostic blockade of a cervical structure or its nerve supply using placebo controls or other controls

Box 54.6 ICHD-2 classification of trigeminal neuralgia

- A. Paroxysmal attacks affecting one or more divisions of the trigeminal nerve, lasting from a fraction of a second to 2 min
- B. The pain has at least one of the following characteristics: intense, sharp, superficial, stabbing, and precipitated from trigger areas or by trigger factors
- C. There is no clinically evident neurological deficit
- D. Attacks are stereotyped in the individual patient
- E. Not attributed to another disorder
- F. Headache lasts < 3 months after successful treatment of the causative disorder

Table 54.3 Classification of chronic daily headache^a according the Silberstein and Lipton Criteria^b

Class	Description
1.8	Chronic migraine
1.8.1	With overuse
1.8.2	Without overuse
2.2	Chronic tension-type headache
2.2.1	With overuse
2.2.2	Without overuse
4.7	New daily persistent headache
4.7.1	With overuse
4.7.2	Without overuse
4.8	Hemicrania continua
4.8.1	With overuse
4.8.2	Without overuse

^a Daily or near-daily headache lasting > 4 h for > 15 days/month.
^b Silberstein & Lipton 2001.

Box 54.7 Classification of transformed migraine (1.8) according the Silberstein and Lipton criteria^a

- A. Daily or almost daily (> 15 days/month) head pain for > 1 month
- B. Average headache duration of > 4 h/day (if untreated)
- C. At least one of the following
 1. History of episodic migraine meeting any International Headache Society criteria 1.1–1.6
 2. History of increasing headache frequency with decreasing severity of migrainous features over ≥ 3 months
 3. Headache at some time meets International Headache Society criteria for migraine 1.1–1.6 other than duration
- D. Does not meet criteria for new daily persistent headache (4.7) or hemicrania continua (4.8)
- E. At least one of the following
 1. There is no suggestion of one of the disorders listed in groups 5–11
 2. Such a disorder is suggested, but it is ruled out by appropriate investigation
 3. Such a disorder is present, but first migraine attacks do not occur in close temporal relation to the disorder

^a Silberstein & Lipton 2001.

headache develops in a person with a previous history of headaches. Second, one of the three following links with migraine is satisfied.

1. A prior history of migraine
2. A period of escalating headache frequency
3. Concurrent superimposed attacks of migraine that fulfil the International Headache Society criteria

New daily persistent headache is characterized by the relatively abrupt onset of an unremitting primary chronic daily headache, i.e. a patient without a previous headache syndrome develops a chronic headache that does not remit. It is the new onset of this primary daily headache that is the most important feature. The clinical features of the pain are not considered in making the diagnosis, which just requires absence of history of evolution from migraine or episodic tension-type headache (Silberstein & Lipton 2001). The S-L classification allows the diagnosis of new daily persistent headache in patients with migraine or episodic tension-type headache if these disorders do not increase in frequency to give rise to new daily persistent headache. The International Headache Society criteria consider new daily persistent headache just those cases where the headaches resemble tension-type headaches (in other words, new daily persistent headache with migrainous features or coexisting new-onset migraine do not meet criteria for this diagnosis according the ICHD-2).

As noted, the ICHD-2 includes chronic migraine (as a complication of migraine) as well as the other types of chronic daily headache. It will also address the other forms of chronic daily headache.

There are three main differences between the ICHD-2 and S-L systems. First, the ICHD-2 criteria for chronic migraine require use of acute medication for less than 10 days (or 15 days for simple analgesics) a month. If medication overuse is present, probable chronic migraine with probable medication overuse is the diagnosis.

Second, the ICHD-2 criteria for chronic migraine require that the 15 days of headache fulfil the criteria for migraine. This nicely parallels the definition of chronic tension-type headache but renders chronic migraine a relatively small group. Alternatively, in the S-L system (Box 54.7), medication overuse is defined by the use of specific amounts of medication. A diagnosis of transformed migraine with medication overuse does not imply that medication overuse caused

and Lipton (S-L) criteria have been most widely used (Silberstein & Lipton 2001).

The S-L criteria divide the primary chronic daily headache of long duration (> 4 h a day) into four main diagnoses (Table 54.3).

1. Transformed migraine
2. Chronic tension-type headache
3. New daily persistent headache
4. Hemicrania continua

The system subclassifies these main diagnoses into those with medication overuse and those without medication overuse.

Although the terms *transformed migraine* and *chronic migraine* have been used synonymously in the past, chronic migraine has a specific definition in the ICHD-2 (Headache Classification Subcommittee of the International Headache Society 2004). Transformed migraine is a headache syndrome not included in the ICHD-2. Patients with transformed migraine typically have a past history of migraine, usually migraine without aura. Patients report a process of transformation (chronification) over months or years, and as headache increases in frequency, associated symptoms become less severe and frequent. The process of transformation frequently ends in a pattern of daily or nearly daily headache that resembles chronic tension-type headache, with some attacks of full migraine superimposed (Silberstein & Lipton 2001). In clinic-based studies, transformation is most often associated with overuse of acute medication (Bigal et al 2002, Diamond 2000). In population studies, about 30% of patients with transformed migraine overuse medications (Scher et al 1998).

The S-L criteria classify transformed migraine in two situations (Box 54.7; Silberstein & Lipton 2001). First, a primary chronic daily

the transformed migraine. Many patients with headache classified as transformed migraine in the S-L criteria receive three diagnoses in the International Headache Society system: migraine, tension-type headache, and medication overuse (Bigal et al 2002).

Third, new daily persistent headache with more than 15 days of headache with migraine features cannot be classified as new daily persistent headache.

Conclusion: using the criteria in clinical practice

Headache diagnosis using the ICHD-2 criteria typically proceeds in orderly fashion. First, one needs to distinguish primary and secondary headaches. The approach is to identify red flags that suggest the possibility of secondary headache (Fig. 54.1), to conduct the work-up suggested by those red flags (Table 54.4), and to diagnose specific secondary headache disorders (if present).

In the absence of secondary headache, the clinician proceeds to diagnose a specific primary headache disorder. We propose an approach to the diagnosis shown in Fig. 54.2. We first define the average monthly frequency of the headaches. We subclassify each of these groups into headache of low frequency (< 15 headache days/month) and headache of high frequency (> 15 headache days/month) based on the average headache duration a day (short duration, < 4 h; long duration, > 4 h). Low-frequency and long-duration headaches include migraine and episodic tension-type headache; high-frequency and long-duration headaches include chronic migraine, chronic tension-type headache, new daily persistent headache, and hemicrania continua.

The final step is to subdivide the headaches lasting less than 4 h (both of low and high frequency) into those triggered (or not) by cough, exercises and Valsalva. Headaches that occur less than 15 days/month, lasting less than 4 h/day, and not triggered by cough, exercises or Valsalva include episodic cluster headache, episodic paroxysmal hemicrania, and episodic tension-type headache. Headaches occurring

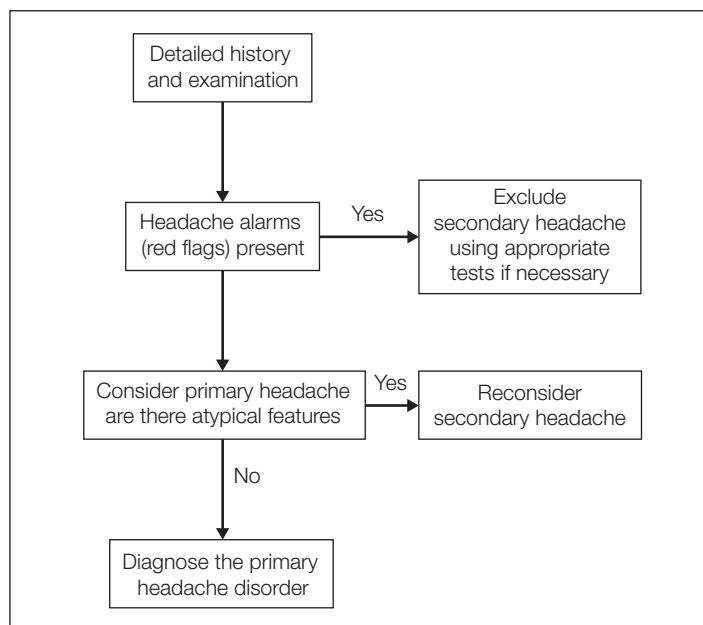


Fig. 54.1 Algorithm for headache diagnosis.

more than 15 days/month, lasting less than 4 h/day, and not triggered by cough, exercises or Valsalva include chronic cluster headache, chronic paroxysmal hemicrania, hypnic headache, and SUNCT syndrome. Headaches triggered by cough, exercises or Valsalva include primary cough headache, primary exertional headache, and primary headache associated with sexual activity.

This orderly approach should facilitate accurate diagnosis as a prelude to effective treatment.

Table 54.4 Red flags in the evaluation of headaches

Red flag	Consider	Possible investigation
Sudden onset headache	Subarachnoid haemorrhage, bleed into a mass or artery-venous malformation, mass lesion (especially posterior fossa)	Neuroimaging, lumbar puncture (after neuroimaging evaluation)
Worsening pattern headache	Mass lesion, subdural haematoma, medication overuse	Neuroimaging
Headache with systemic illness (fever, neck stiffness, cutaneous rash)	Meningitis, encephalitis, Lyme disease, systemic infection, collagen vascular disease	Neuroimaging, lumbar puncture, blood tests
Focal neurological signs other than typical visual or sensorial aura	Mass lesion, artery-venous malformation, collagen vascular disease	Neuroimaging, collagen vascular evaluation
Papilloedema	Mass lesion, pseudotumour, encephalitis, meningitis	Neuroimaging, lumbar puncture (after the neuroimaging evaluation)

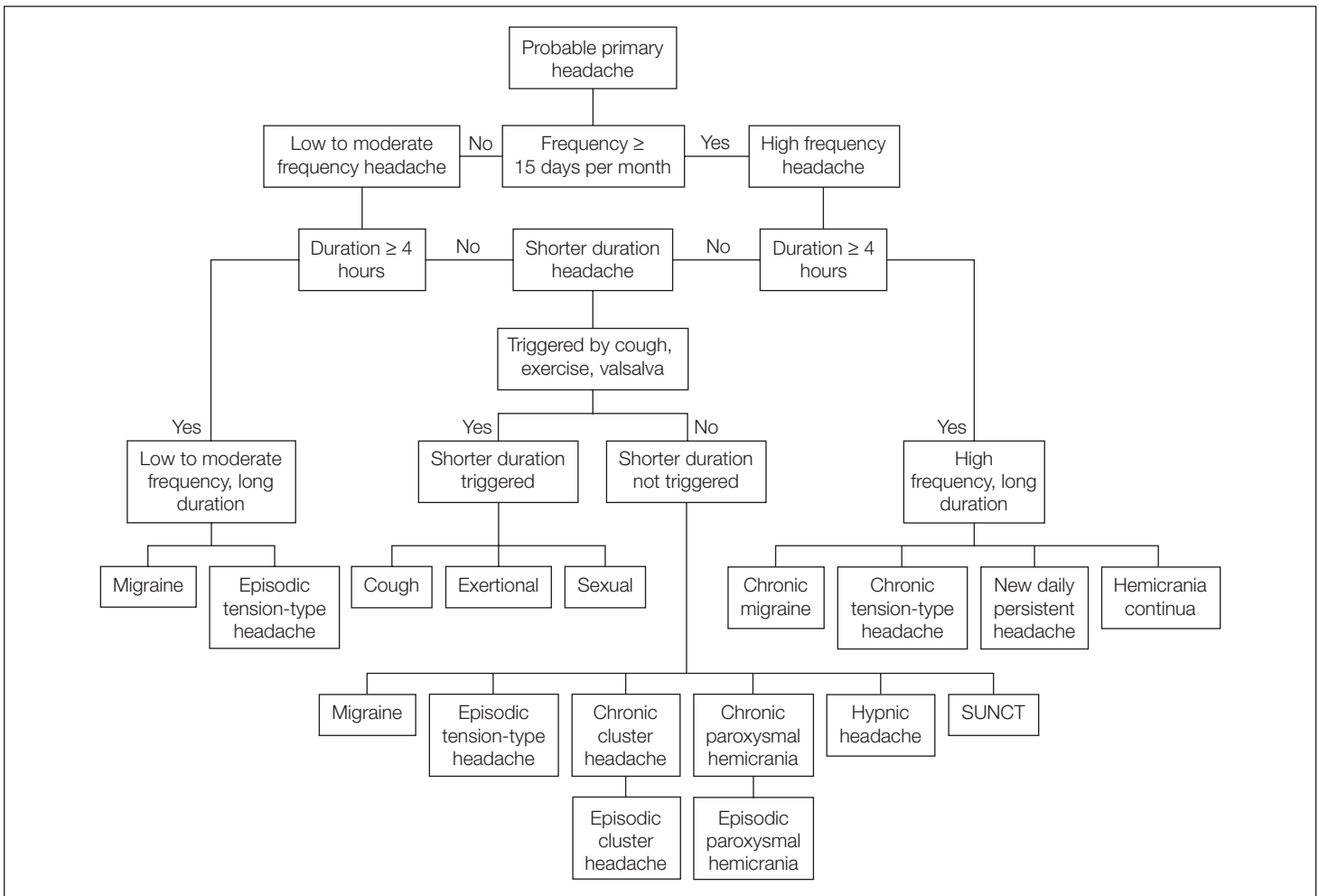


Fig. 54.2 Approach to diagnosis of primary headache disorder.

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