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Headache Classification Committee of the International Headache Society (IHS)

The International Classification of Headache Disorders, 3rd edition (beta version)

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Preface

After two very successful editions of the *International Classification of Headache Disorders* (ICHD), a third is now close to being final. The members of the Classification Committee have all worked hard for three years in order to accomplish this beta version. Most members have chaired the work on a specific chapter of the classification, assisted by a number of other experts. For this edition, there has been a substantial body of evidence available for the classification work, in contrast to our previous editions, which were mostly based on the opinions of experts. We have tried to be conservative, making changes only where there was good published evidence to support change or where the need for change was intuitively obvious.

This is the first time that we have published a beta version ahead of the final version. The main reason is to synchronize ICHD-3 with the World Health Organization's next revision (11th edition) of the *International Classification of Diseases* (ICD-11). This classification is already well advanced, and we have not only secured a very good representation of headache within ICD-11 but also ensured congruence between ICD-11 and ICHD-3 beta. However, ICD-11 now enters a phase of field trials, and ICHD-3 should do the same. Such a test period will allow identification and correction of mistakes and enable a broad input from the members of the International Headache Society.

ICD-11 diagnostic codes will not be finalized until two or three years from now, but it would be a major advantage for ICHD-3 to be able to include these codes along with our own. WHO's ICD-11 codes will be used by health authorities for official diagnostic coding, and in many cases they will be employed

for reimbursement purposes; we must have them right.

We publish ICHD-3 beta immediately on the International Headache Society's website, and shortly after as an issue of *Cephalalgia*. Field-testing will continue for 2 or maybe 3 years. Small amendments are likely both to ICHD-3 and to the diagnostic codes of ICD-11, and these will be incorporated. At that time, we shall publish ICHD-3 in final form in *Cephalalgia*.

ICHD-3 beta is published only in English, but those throughout the world who wish to make their own careful translations of parts or in toto are welcome to do so subject to the conditions stated above. The final version of ICHD-3 should be translated into as many languages as possible, and these translations published, as happened to the second and first editions. As we expect ICHD-3 beta to be very similar to the final version, translation work begun now is likely to remain useful. Any changes necessitated later by the outcomes of field-testing can be made easily.

Clinicians and researchers should start using the criteria of ICHD-3 beta. There are many improvements over ICHD-II, and it would be unhelpful to continue to use ICHD-II for scientific work. We encourage readers to study ICHD-3 beta very closely, and document and comment on any inconsistencies they may find. Comments should be sent **not to me** but to the chairmen of the relevant working groups. Their names and email addresses are found in this publication and on the IHS website.

Jes Olesen
Chairman

Headache Classification Committee
International Headache Society

How to use this classification

This extensive document is not intended to be learnt by heart. Even members of the Classification Committee are unable to remember all of it. It is a document that should be consulted time and time again. In this way you will soon get to know the diagnostic criteria for 1.1 *Migraine without aura*, 1.2 *Migraine with aura*, the major subtypes of 2. *Tension-type headache*, 3.1 *Cluster headache* and a few others. The rest will remain something to look up. In clinical practice you do not need the classification for the obvious case of migraine or tension-type headache, but it is useful when the diagnosis is uncertain. For research, the classification is indispensable and every patient entered into a research project, be it a drug trial or a study of pathophysiology or biochemistry, must fulfil a set of diagnostic criteria.

1. This classification is hierarchical, and you must decide how detailed you want to make your diagnosis. This can range from the first-digit level to the fifth. First, one gets a rough idea about which group the patient belongs to. Is it, for example, 1. *Migraine* or 2. *Tension-type headache* or 3. *Trigeminal autonomic cephalalgias*? Then one obtains information allowing a more detailed diagnosis. The desired detail depends on the purpose. In general practice only the first- or second-digit diagnoses are usually applied, whereas in specialist practice and headache centres a diagnosis at the fourth- or fifth-digit level is appropriate.
2. For most purposes, patients receive a diagnosis according to the headache phenotypes that they currently present, or that they have presented within the last year. For genetic and some other uses, occurrence during the whole lifetime is used.
3. Each distinct type, subtype or subform of headache that the patient has must be separately diagnosed and coded. Thus, a severely affected patient in a headache centre may receive three diagnoses and codes: 1.1 *Migraine without aura*, 1.2 *Migraine with aura* and 8.2 *Medication-overuse headache*.
4. When a patient receives more than one diagnosis, these should be listed in the order of importance to the patient.
5. When one type of headache in a particular patient fulfils two different sets of diagnostic criteria, then all other available information should be used to decide which of the alternatives is the correct or more likely diagnosis. This could include the longitudinal headache history (how did the headache start?), the family history, the effect of drugs, menstrual relationship, age, gender and a range of other features. Fulfilment of the diagnostic criteria for 1. *Migraine*, 2. *Tension-type headache* or 3. *Trigeminal autonomic cephalalgias*, or any of their subtypes, always trumps fulfilment of criteria for the *probable* diagnostic categories of each, which are last-described in the respective groups. In other words, a patient whose headache fulfils criteria for both 1.5 *Probable migraine* and 2.1 *Infrequent episodic tension-type headache* should be coded to the latter. Nevertheless, consideration should always be given to the possibility that some headache attacks meet one set of criteria, whereas other attacks meet another set. In such cases, two diagnoses exist and both should be coded.
6. To receive a particular headache diagnosis the patient must, in many cases, experience a minimum number of attacks of (or days with) that headache. This number is specified in the diagnostic criteria for the headache type, subtype or subform. Further, the headache must fulfil a number of other requirements described within the criteria under separate letter headings: A, B, C etc. Some letter headings are monothetic: that is, they express a single requirement. Other letter headings are polythetic, requiring for example any two out of four listed characteristics.
7. The full set of diagnostic criteria is provided for some headache disorders only at the first- and second-digit levels. Diagnostic criteria at the third- and fourth-digit levels then demand, as criterion A, fulfilment of the criteria for levels one and/or two and, in criterion B and onwards, specify the further specific criteria to be fulfilled.
8. The frequency of primary headache disorders varies from attacks every one to two years to attacks daily. The severity of attacks also varies. ICHD-3 beta does not generally provide a possibility to code for frequency or severity, but recommends that frequency and severity be specified in free text.
9. *Primary or secondary headache or both*: When a new headache occurs *for the first time* in close temporal relation to another disorder that is known to cause headache, or fulfils other criteria for causation by that disorder, the new headache is coded as a secondary headache attributed to the causative disorder. This remains true even when the

headache has the characteristics of a primary headache (migraine, tension-type headache, cluster headache or one of the other trigeminal autonomic cephalalgias). When a *pre-existing* primary headache becomes *chronic* in close temporal relation to such a causative disorder, both the primary and the secondary diagnoses should be given. When a *pre-existing* primary headache is made significantly worse (usually meaning a two-fold or greater increase in frequency and/or severity) in close temporal relation to such a causative disorder, both the primary and the secondary headache diagnoses should be given, provided that there is good evidence that the disorder can cause headache.

10. The last criterion for almost every headache disorder is 'Not better accounted for by another ICHD-3 diagnosis'. Consideration of other possible diagnoses (the *differential diagnosis*) is a routine part of the clinical diagnostic process. When a headache appears to fulfil the criteria for a particular headache disorder, this last criterion is a reminder always to consider other diagnoses that might better explain the headache. In particular this applies to assessing whether headache is secondary or primary. It may also apply to alternative causative disorders: for example, headache occurring in close temporal relation to acute ischaemic stroke may be a consequence not of the stroke but of the cause of the stroke (e.g. dissection).
11. Many patients with headache attacks fulfilling one set of explicit diagnostic criteria also have attacks that, although similar, do not quite satisfy the criteria. This can be a result of treatment, inability to recall symptoms exactly or other factors. Ask the patient to describe a typical untreated or unsuccessfully treated attack, and ascertain that there have been enough of these to establish the diagnosis. Then include the less-typical attacks when describing attack frequency.
12. When a patient is suspected of having more than one headache type or subtype, it is highly recommended that he or she fill out a diagnostic headache diary in which, for each headache episode, the important characteristics are recorded. It has been shown that such a headache diary improves diagnostic accuracy as well as allowing a more precise judgement of medication consumption. The diary helps in judging the quantity of two or more different headache types or subtypes. Finally, it teaches the patient how to distinguish between different headaches, for example between migraine without aura and episodic tension-type headache.
13. In each chapter on secondary headaches, the most well-known and well-established causes are

mentioned and criteria for these are given. However, in many chapters, for example 9. *Headache attributed to infection*, there are an almost endless number of possible causes. In order to avoid a very long list, only the most important are mentioned. In the example, rarer causes are assigned to 9.2.3 *Headache attributed to other systemic infection*. The same system is used in the other chapters on secondary headaches.

14. The diagnostic criteria for the secondary headaches no longer require remission or substantial improvement of the underlying causative disorder before the headache diagnosis can be made. The diagnostic criteria of ICHD-3 beta may be applied already on presentation or as soon after as the underlying disorder is confirmed. Criterion A is presence of the headache; criterion B is presence of the causative disorder; criterion C is the evidence of causation. In acute conditions, a close temporal relation between onset of headache and onset of the presumed causative disorder is often sufficient to establish causation, whereas less acute conditions usually require more evidence of causation. In all cases, the last criterion must be applied as a check: 'Not better accounted for by another ICHD-3 diagnosis'.
15. In a few secondary headaches, 5.2 *Persistent headache attributed to traumatic head injury* being a good example, persistent headache subforms are recognized to occur; that is, headache that was caused initially by another disorder fails to remit after that disorder has resolved. In such cases, the diagnosis changes from the acute subform (e.g. 5.1 *Acute headache attributed to traumatic head injury*) to the persistent subform (5.2 *Persistent headache attributed to traumatic head injury*) after a specified time interval (three months in this example). Evidence of causation depends on earlier fulfilment of the criteria for diagnosis of the acute subform, and persistence of the same headache. Most such diagnoses are in the Appendix because of insufficient evidence for their existence. They will not usually be applied, but are there to stimulate research into better criteria for causation.
16. The Appendix is for research. It helps clinical scientists study orphan entities for later inclusion in (or, in some cases, exclusion from) the main body of the classification. Most diagnoses and diagnostic criteria in the Appendix are either new or alternatives to criteria in the main body. Some are old entities not yet sufficiently validated; these are expected to be deleted in the next revision of ICHD if evidence is not produced.

Classification

ICHD-3 code	Diagnosis
1.	Migraine
1.1	Migraine without aura
1.2	Migraine with aura
1.2.1	Migraine with typical aura
1.2.1.1	Typical aura with headache
1.2.1.2	Typical aura without headache
1.2.2	Migraine with brainstem aura
1.2.3	Hemiplegic migraine
1.2.3.1	Familial hemiplegic migraine (FHM)
1.2.3.1.1	Familial hemiplegic migraine type 1 (FHM1)
1.2.3.1.2	Familial hemiplegic migraine type 2 (FHM2)
1.2.3.1.3	Familial hemiplegic migraine type 3 (FHM3)
1.2.3.1.4	Familial hemiplegic migraine, other loci
1.2.3.2	Sporadic hemiplegic migraine
1.2.4	Retinal migraine
1.3	Chronic migraine
1.4	Complications of migraine
1.4.1	Status migrainosus
1.4.2	Persistent aura without infarction
1.4.3	Migrainous infarction
1.4.4	Migraine aura-triggered seizure
1.5	Probable migraine
1.5.1	Probable migraine without aura
1.5.2	Probable migraine with aura
1.6	Episodic syndromes that may be associated with migraine
1.6.1	Recurrent gastrointestinal disturbance
1.6.1.1	Cyclical vomiting syndrome
1.6.1.2	Abdominal migraine
1.6.2	Benign paroxysmal vertigo
1.6.3	Benign paroxysmal torticollis
2.	Tension-type headache (TTH)
2.1	Infrequent episodic tension-type headache
2.1.1	Infrequent episodic tension-type headache associated with pericranial tenderness
2.1.2	Infrequent episodic tension-type headache not associated with pericranial tenderness
2.2	Frequent episodic tension-type headache
2.2.1	Frequent episodic tension-type headache associated with pericranial tenderness
2.2.2	Frequent episodic tension-type headache not associated with pericranial tenderness
2.3	Chronic tension-type headache
2.3.1	Chronic tension-type headache associated with pericranial tenderness
2.3.2	Chronic tension-type headache not associated with pericranial tenderness
2.4	Probable tension-type headache
2.4.1	Probable infrequent episodic tension-type headache
2.4.2	Probable frequent episodic tension-type headache
2.4.3	Probable chronic tension-type headache
3.	Trigeminal autonomic cephalalgias (TACs)
3.1	Cluster headache
3.1.1	Episodic cluster headache
3.1.2	Chronic cluster headache
3.2	Paroxysmal hemicrania
3.2.1	Episodic paroxysmal hemicrania
3.2.2	Chronic paroxysmal hemicrania

- 3.3 Short-lasting unilateral neuralgiform headache attacks
 - 3.3.1 Short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing (SUNCT)
 - 3.3.1.1 Episodic SUNCT
 - 3.3.1.2 Chronic SUNCT
 - 3.3.2 Short-lasting unilateral neuralgiform headache attacks with cranial autonomic symptoms (SUNA)
 - 3.3.2.1 Episodic SUNA
 - 3.3.2.2 Chronic SUNA
- 3.4 Hemicrania continua
- 3.5 Probable trigeminal autonomic cephalgia
 - 3.5.1 Probable cluster headache
 - 3.5.2 Probable paroxysmal hemicrania
 - 3.5.3 Probable short-lasting unilateral neuralgiform headache attacks
 - 3.5.4 Probable hemicrania continua
- 4. Other primary headache disorders**
- 4.1 Primary cough headache
 - 4.1.1 Probable primary cough headache
- 4.2 Primary exercise headache
 - 4.2.1 Probable primary exercise headache
- 4.3 Primary headache associated with sexual activity
 - 4.3.1 Probable primary headache associated with sexual activity
- 4.4 Primary thunderclap headache
- 4.5 Cold-stimulus headache
 - 4.5.1 Headache attributed to external application of a cold stimulus
 - 4.5.2 Headache attributed to ingestion or inhalation of a cold stimulus
 - 4.5.3 Probable cold-stimulus headache
 - 4.5.3.1 Headache probably attributed to external application of a cold stimulus
 - 4.5.3.2 Headache probably attributed to ingestion or inhalation of a cold stimulus
- 4.6 External-pressure headache
 - 4.6.1 External-compression headache
 - 4.6.2 External-traction headache
 - 4.6.3 Probable external-pressure headache
 - 4.6.3.1 Probable external-compression headache
 - 4.6.3.2 Probable external-traction headache
- 4.7 Primary stabbing headache
 - 4.7.1 Probable primary stabbing headache
- 4.8 Nummular headache
 - 4.8.1 Probable nummular headache
- 4.9 Hypnic headache
 - 4.9.1 Probable hypnic headache
- 4.10 New daily persistent headache (NDPH)
 - 4.10.1 Probable new daily persistent headache
- 5. Headache attributed to trauma or injury to the head and/or neck**
- 5.1 Acute headache attributed to traumatic injury to the head
 - 5.1.1 Acute headache attributed to moderate or severe traumatic injury to the head
 - 5.1.2 Acute headache attributed to mild traumatic injury to the head
- 5.2 Persistent headache attributed to traumatic injury to the head
 - 5.2.1 Persistent headache attributed to moderate or severe traumatic injury to the head
 - 5.2.2 Persistent headache attributed to mild traumatic injury to the head
- 5.3 Acute headache attributed to whiplash
- 5.4 Persistent headache attributed to whiplash
- 5.5 Acute headache attributed to craniotomy
- 5.6 Persistent headache attributed to craniotomy

- 6. Headache attributed to cranial or cervical vascular disorder**
- 6.1 Headache attributed to ischaemic stroke or transient ischaemic attack
 - 6.1.1 Headache attributed to ischaemic stroke (cerebral infarction)
 - 6.1.2 Headache attributed to transient ischaemic attack (TIA)
- 6.2 Headache attributed to non-traumatic intracranial haemorrhage
 - 6.2.1 Headache attributed to non-traumatic intracerebral haemorrhage
 - 6.2.2 Headache attributed to non-traumatic subarachnoid haemorrhage (SAH)
 - 6.2.3 Headache attributed to non-traumatic acute subdural haemorrhage (ASDH)
- 6.3 Headache attributed to unruptured vascular malformation
 - 6.3.1 Headache attributed to unruptured saccular aneurysm
 - 6.3.2 Headache attributed to arteriovenous malformation (AVM)
 - 6.3.3 Headache attributed to dural arteriovenous fistula (DAVF)
 - 6.3.4 Headache attributed to cavernous angioma
 - 6.3.5 Headache attributed to encephalotrigeminal or leptomeningeal angiomatosis (Sturge Weber syndrome)
- 6.4 Headache attributed to arteritis
 - 6.4.1 Headache attributed to giant cell arteritis (GCA)
 - 6.4.2 Headache attributed to primary angiitis of the central nervous system (PACNS)
 - 6.4.3 Headache attributed to secondary angiitis of the central nervous system (SACNS)
- 6.5 Headache attributed to cervical carotid or vertebral artery disorder
 - 6.5.1 Headache or facial or neck pain attributed to cervical carotid or vertebral artery dissection
 - 6.5.2 Post-endarterectomy headache
 - 6.5.3 Headache attributed to carotid or vertebral angioplasty
- 6.6 Headache attributed to cerebral venous thrombosis (CVT)
- 6.7 Headache attributed to other acute intracranial arterial disorder
 - 6.7.1 Headache attributed to an intracranial endovascular procedure
 - 6.7.2 Angiography headache
 - 6.7.3 Headache attributed to reversible cerebral vasoconstriction syndrome (RCVS)
 - 6.7.3.1 Headache probably attributed to reversible cerebral vasoconstriction syndrome (RCVS)
 - 6.7.4 Headache attributed to intracranial arterial dissection
- 6.8 Headache attributed to genetic vasculopathy
 - 6.8.1 Cerebral Autosomal Dominant Arteriopathy with Subcortical Infarcts and Leukoencephalopathy (CADASIL)
 - 6.8.2 Mitochondrial Encephalopathy, Lactic Acidosis and Stroke-like episodes (MELAS)
 - 6.8.3 Headache attributed to another genetic vasculopathy
- 6.9 Headache attributed to pituitary apoplexy
- 7. Headache attributed to non-vascular intracranial disorder**
- 7.1 Headache attributed to increased cerebrospinal fluid pressure
 - 7.1.1 Headache attributed to idiopathic intracranial hypertension (IIH)
 - 7.1.2 Headache attributed to intracranial hypertension secondary to metabolic, toxic or hormonal causes
 - 7.1.3 Headache attributed to intracranial hypertension secondary to hydrocephalus
- 7.2 Headache attributed to low cerebrospinal fluid pressure
 - 7.2.1 Post-dural puncture headache
 - 7.2.2 CSF fistula headache
 - 7.2.3 Headache attributed to spontaneous intracranial hypotension
- 7.3 Headache attributed to non-infectious inflammatory disease
 - 7.3.1 Headache attributed to neurosarcoidosis
 - 7.3.2 Headache attributed to aseptic (non-infectious) meningitis
 - 7.3.3 Headache attributed to other non-infectious inflammatory disease
 - 7.3.4 Headache attributed to lymphocytic hypophysitis
 - 7.3.5 Syndrome of transient Headache and Neurological Deficits with cerebrospinal fluid Lymphocytosis (HaNDL)

- 7.4 Headache attributed to intracranial neoplasia
 - 7.4.1 Headache attributed to intracranial neoplasm
 - 7.4.1.1 Headache attributed to colloid cyst of the third ventricle
 - 7.4.2 Headache attributed to carcinomatous meningitis
 - 7.4.3 Headache attributed to hypothalamic or pituitary hyper- or hyposecretion
- 7.5 Headache attributed to intrathecal injection
- 7.6 Headache attributed to epileptic seizure
 - 7.6.1 Hemicrania epileptica
 - 7.6.2 Post-ictal headache
- 7.7 Headache attributed to Chiari malformation type I (CM1)
- 7.8 Headache attributed to other non-vascular intracranial disorder
- 8. Headache attributed to a substance or its withdrawal**
- 8.1 Headache attributed to use of or exposure to a substance
 - 8.1.1 Nitric oxide (NO) donor-induced headache
 - 8.1.1.1 Immediate NO donor-induced headache
 - 8.1.1.2 Delayed NO donor-induced headache
 - 8.1.2 Phosphodiesterase (PDE) inhibitor-induced headache
 - 8.1.3 Carbon monoxide (CO)-induced headache
 - 8.1.4 Alcohol-induced headache
 - 8.1.4.1 Immediate alcohol-induced headache
 - 8.1.4.2 Delayed alcohol-induced headache
 - 8.1.5 Headache induced by food and/or additive
 - 8.1.5.1 Monosodium glutamate (MSG)-induced headache
 - 8.1.6 Cocaine-induced headache
 - 8.1.7 Histamine-induced headache
 - 8.1.7.1 Immediate histamine-induced headache
 - 8.1.7.2 Delayed histamine-induced headache
 - 8.1.8 Calcitonin gene-related peptide (CGRP)-induced headache
 - 8.1.8.1 Immediate CGRP-induced headache
 - 8.1.8.2 Delayed CGRP-induced headache
 - 8.1.9 Headache attributed to exogenous acute pressor agent
 - 8.1.10 Headache attributed to occasional use of non-headache medication
 - 8.1.11 Headache attributed to long-term use of non-headache medication
 - 8.1.12 Headache attributed to exogenous hormone
 - 8.1.13 Headache attributed to use of or exposure to other substance
- 8.2 Medication-overuse headache (MOH)
 - 8.2.1 Ergotamine-overuse headache
 - 8.2.2 Triptan-overuse headache
 - 8.2.3 Simple analgesic-overuse headache
 - 8.2.3.1 Paracetamol (acetaminophen)-overuse headache
 - 8.2.3.2 Acetylsalicylic acid-overuse headache
 - 8.2.3.3 Other non-steroidal anti-inflammatory drug (NSAID)-overuse headache
 - 8.2.4 Opioid-overuse headache
 - 8.2.5 Combination-analgesic-overuse headache
 - 8.2.6 Medication-overuse headache attributed to multiple drug classes not individually overused
 - 8.2.7 Medication-overuse headache attributed to unverified overuse of multiple drug classes
 - 8.2.8 Medication-overuse headache attributed to other medication
- 8.3 Headache attributed to substance withdrawal
 - 8.3.1 Caffeine-withdrawal headache
 - 8.3.2 Opioid-withdrawal headache
 - 8.3.3 Oestrogen-withdrawal headache
 - 8.3.4 Headache attributed to withdrawal from chronic use of other substance

- 9. Headache attributed to infection**
- 9.1 Headache attributed to intracranial infection
- 9.1.1 Headache attributed to bacterial meningitis or meningoencephalitis
- 9.1.1.1 Acute headache attributed to bacterial meningitis or meningoencephalitis
- 9.1.1.2 Chronic headache attributed to bacterial meningitis or meningoencephalitis
- 9.1.1.3 Persistent headache attributed to past bacterial meningitis or meningoencephalitis
- 9.1.2 Headache attributed to viral meningitis or encephalitis
- 9.1.2.1 Headache attributed to viral meningitis
- 9.1.2.2 Headache attributed to viral encephalitis
- 9.1.3 Headache attributed to intracranial fungal or other parasitic infection
- 9.1.3.1 Acute headache attributed to intracranial fungal or other parasitic infection
- 9.1.3.2 Chronic headache attributed to intracranial fungal or other parasitic infection
- 9.1.4 Headache attributed to brain abscess
- 9.1.5 Headache attributed to subdural empyema
- 9.2 Headache attributed to systemic infection
- 9.2.1 Headache attributed to systemic bacterial infection
- 9.2.1.1 Acute headache attributed to systemic bacterial infection
- 9.2.1.2 Chronic headache attributed to systemic bacterial infection
- 9.2.2 Headache attributed to systemic viral infection
- 9.2.2.1 Acute headache attributed to systemic viral infection
- 9.2.2.2 Chronic headache attributed to systemic viral infection
- 9.2.3 Headache attributed to other systemic infection
- 9.2.3.1 Acute headache attributed to other systemic infection
- 9.2.3.2 Chronic headache attributed to other systemic infection
- 10. Headache attributed to disorder of homeostasis**
- 10.1 Headache attributed to hypoxia and/or hypercapnia
- 10.1.1 High-altitude headache
- 10.1.2 Headache attributed to aeroplane travel
- 10.1.3 Diving headache
- 10.1.4 Sleep apnoea headache
- 10.2 Dialysis headache
- 10.3 Headache attributed to arterial hypertension
- 10.3.1 Headache attributed to pheochromocytoma
- 10.3.2 Headache attributed to hypertensive crisis without hypertensive encephalopathy
- 10.3.3 Headache attributed to hypertensive encephalopathy
- 10.3.4 Headache attributed to pre-eclampsia or eclampsia
- 10.3.5 Headache attributed to autonomic dysreflexia
- 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
- 11. Headache or facial pain attributed to disorder of the cranium, neck, eyes, ears, nose, sinuses, teeth, mouth or other facial or cervical structure**
- 11.1 Headache attributed to disorder of cranial bone
- 11.2 Headache attributed to disorder of the neck
- 11.2.1 Cervicogenic headache
- 11.2.2 Headache attributed to retropharyngeal tendonitis
- 11.2.3 Headache attributed to craniocervical dystonia
- 11.3 Headache attributed to disorder of the eyes
- 11.3.1 Headache attributed to acute glaucoma
- 11.3.2 Headache attributed to refractive error
- 11.3.3 Headache attributed to heterophoria or heterotropia (latent or persistent squint)
- 11.3.4 Headache attributed to ocular inflammatory disorder
- 11.3.5 Headache attributed to trochleitis
- 11.4 Headache attributed to disorder of the ears

- 11.5 Headache attributed to disorder of the nose or paranasal sinuses
 - 11.5.1 Headache attributed to acute rhinosinusitis
 - 11.5.2 Headache attributed to chronic or recurring rhinosinusitis
- 11.6 Headache attributed to disorder of the teeth or jaw
- 11.7 Headache attributed to temporomandibular disorder (TMD)
- 11.8 Head or facial pain attributed to inflammation of the stylohyoid ligament
- 11.9 Headache or facial pain attributed to other disorder of cranium, neck, eyes, ears, nose, sinuses, teeth, mouth or other facial or cervical structure
- 12. Headache attributed to psychiatric disorder**
- 12.1 Headache attributed to somatization disorder
- 12.2 Headache attributed to psychotic disorder
- 13. Painful cranial neuropathies and other facial pains**
- 13.1 Trigeminal neuralgia
 - 13.1.1 Classical trigeminal neuralgia
 - 13.1.1.1 Classical trigeminal neuralgia, purely paroxysmal
 - 13.1.1.2 Classical trigeminal neuralgia with concomitant persistent facial pain
 - 13.1.2 Painful trigeminal neuropathy
 - 13.1.2.1 Painful trigeminal neuropathy attributed to acute Herpes zoster
 - 13.1.2.2 Post-herpetic trigeminal neuropathy
 - 13.1.2.3 Painful post-traumatic trigeminal neuropathy
 - 13.1.2.4 Painful trigeminal neuropathy attributed to multiple sclerosis (MS) plaque
 - 13.1.2.5 Painful trigeminal neuropathy attributed to space-occupying lesion
 - 13.1.2.6 Painful trigeminal neuropathy attributed to other disorder
- 13.2 Glossopharyngeal neuralgia
- 13.3 Nervus intermedius (facial nerve) neuralgia
 - 13.3.1 Classical nervus intermedius neuralgia
 - 13.3.2 Nervus intermedius neuropathy attributed to Herpes zoster
- 13.4 Occipital neuralgia
- 13.5 Optic neuritis
- 13.6 Headache attributed to ischaemic ocular motor nerve palsy
- 13.7 Tolosa-Hunt syndrome
- 13.8 Paratrigeminal oculosympathetic (Raeder's) syndrome
- 13.9 Recurrent painful ophthalmoplegic neuropathy
- 13.10 Burning mouth syndrome (BMS)
- 13.11 Persistent idiopathic facial pain (PIFP)
- 13.12 Central neuropathic pain
 - 13.12.1 Central neuropathic pain attributed to multiple sclerosis (MS)
 - 13.12.2 Central post-stroke pain (CPSP)
- 14. Other headache disorders**
- 14.1 Headache not elsewhere classified
- 14.2 Headache unspecified
- A. Appendix**
- A1. Migraine**
- A1.1 Migraine without aura
 - A1.1.1 Pure menstrual migraine without aura
 - A1.1.2 Menstrually related migraine without aura
 - A1.1.3 Non-menstrual migraine without aura
- A1.2 Migraine with aura (alternative criteria)
 - A1.2.1 Migraine with typical aura (alternative criteria)
- A1.3 Chronic migraine (alternative criteria)
 - A1.3.1 Chronic migraine with pain-free periods
 - A1.3.2 Chronic migraine with continuous pain
- A1.4 Complications of migraine
 - A1.4.5 Migraine aura status

- A1.6 Episodic syndromes that may be associated with migraine
 - A1.6.4 Infantile colic
 - A1.6.5 Alternating hemiplegia of childhood
 - A1.6.6 Vestibular migraine
- A2. Tension-type headache (alternative criteria)**
- A3. Trigeminal-autonomic cephalalgias (TACs)**
- A3.6 Undifferentiated trigeminal autonomic cephalalgia
- A4. Other primary headache disorders**
- A4.11 Epicrania fugax
- A5. Headache attributed to trauma or injury to the head and/or neck**
- A5.1 Acute headache attributed to traumatic injury to the head
 - A5.1.1.1 Delayed-onset acute headache attributed to moderate or severe traumatic injury to the head
 - A5.1.2.1 Delayed-onset acute headache attributed to mild traumatic injury to the head
- A5.2 Persistent headache attributed to traumatic injury to the head
 - A5.2.1.1 Delayed-onset persistent headache attributed to moderate or severe traumatic injury to the head
 - A5.2.2.1 Delayed-onset persistent headache attributed to mild traumatic injury to the head
- A5.7 Headache attributed to radiosurgery of the brain
- A5.8 Acute headache attributed to other trauma or injury to the head and/or neck
- A5.9 Persistent headache attributed to other trauma or injury to the head and/or neck
- A6. Headache attributed to cranial or cervical vascular disorder**
- A6.10 Persistent headache attributed to past cranial or cervical vascular disorder
- A7. Headache attributed to non-vascular intracranial disorder**
- A7.6 Headache attributed to epileptic seizure
 - A7.6.3 Post-electroconvulsive therapy (ECT) headache
- A7.9 Persistent headache attributed to past non-vascular intracranial disorder
- A8. Headache attributed to a substance or its withdrawal**
- A8.4 Persistent headache attributed to past use of or exposure to a substance
- A9. Headache attributed to infection**
- A9.1 Headache attributed to intracranial infection
 - A9.1.3.3 Persistent headache attributed to past intracranial fungal or other parasitic infection
 - A9.1.6 Headache attributed to other infective space-occupying lesion
- A9.3 Headache attributed to human immunodeficiency virus (HIV) infection
- A10. Headache attributed to disorder of homeostasis**
- A10.7 Head and/or neck pain attributed to orthostatic (postural) hypotension
- A10.8 Headache attributed to other disorder of homeostasis
 - A10.8.1 Headache attributed to travel in space
 - A10.8.2 Headache attributed to other metabolic or systemic disorder
- A10.9 Persistent headache attributed to past disorder of homeostasis
- A11. Headache or facial pain attributed to disorder of the cranium, neck, eyes, ears, nose, sinuses, teeth, mouth or other facial or cervical structure**
- A11.2 Headache attributed to disorder of the neck
 - A11.2.4 Headache attributed to upper cervical radiculopathy
 - A11.2.5 Headache attributed to cervical myofascial pain
- A11.5 Headache attributed to disorder of the nose or paranasal sinuses
 - A11.5.3 Headache attributed to disorder of the nasal mucosa, turbinates or septum
- A12. Headache attributed to psychiatric disorder**
- A12.3 Headache attributed to depressive disorder
- A12.4 Headache attributed to separation anxiety disorder
- A12.5 Headache attributed to panic disorder
- A12.6 Headache attributed to specific phobia
- A12.7 Headache attributed to social anxiety disorder (social phobia)
- A12.8 Headache attributed to generalized anxiety disorder
- A12.9 Headache attributed to post-traumatic stress disorder
- A12.10 Headache attributed to acute stress disorder

Part one

The primary headaches

1. Migraine
2. Tension-type headache
3. Trigeminal autonomic cephalalgias
4. Other primary headache disorders

I. Migraine

- 1.1 Migraine without aura
- 1.2 Migraine with aura
 - 1.2.1 Migraine with typical aura
 - 1.2.1.1 Typical aura with headache
 - 1.2.1.2 Typical aura without headache
 - 1.2.2 Migraine with brainstem aura
 - 1.2.3 Hemiplegic migraine
 - 1.2.3.1 Familial hemiplegic migraine (FHM)
 - 1.2.3.1.1 Familial hemiplegic migraine type 1
 - 1.2.3.1.2 Familial hemiplegic migraine type 2
 - 1.2.3.1.3 Familial hemiplegic migraine type 3
 - 1.2.3.1.4 Familial hemiplegic migraine, other loci
 - 1.2.3.2 Sporadic hemiplegic migraine
 - 1.2.4 Retinal migraine
- 1.3 Chronic migraine
- 1.4 Complications of migraine
 - 1.4.1 Status migrainosus
 - 1.4.2 Persistent aura without infarction
 - 1.4.3 Migrainous infarction
 - 1.4.4 Migraine aura-triggered seizure
- 1.5 Probable migraine
 - 1.5.1 Probable migraine without aura
 - 1.5.2 Probable migraine with aura
- 1.6 Episodic syndromes that may be associated with migraine
 - 1.6.1 Recurrent gastrointestinal disturbance
 - 1.6.1.1 Cyclical vomiting syndrome
 - 1.6.1.2 Abdominal migraine
 - 1.6.2 Benign paroxysmal vertigo
 - 1.6.3 Benign paroxysmal torticollis

Coded elsewhere:

Migraine-like headache secondary to another disorder (*symptomatic migraine*) is coded as a secondary headache attributed to that disorder.

General comment

Primary or secondary headache or both?

When a new headache with the characteristics of migraine occurs for the first time in close temporal relation to another disorder known to cause headache, or fulfils other criteria for causation by that disorder, the new headache is coded as a secondary headache attributed to the causative disorder. When *pre-existing* migraine becomes *chronic* in close temporal relation to such a causative disorder, both the initial migraine diagnosis and the secondary diagnosis should be given. 8.2 *Medication-overuse headache* is a particularly important example of this: both the episodic or

chronic migraine diagnosis and the diagnosis 8.2 *Medication-overuse headache* should be given when medication overuse is present. When *pre-existing* migraine is made significantly worse (usually meaning a two-fold or greater increase in frequency and/or severity) in close temporal relation to such a causative disorder, both the initial migraine diagnosis and the secondary headache diagnosis should be given, provided that there is good evidence that the disorder can cause headache.

Introduction

Migraine is a common disabling primary headache disorder. Epidemiological studies have documented its high prevalence and high socio-economic and personal impacts. In the *Global Burden of Disease Survey 2010*, it was ranked as the third most prevalent disorder and seventh-highest specific cause of disability worldwide.

Migraine has two major subtypes. 1.1 *Migraine without aura* is a clinical syndrome characterized by headache with specific features and associated symptoms. 1.2 *Migraine with aura* is primarily characterized by the transient focal neurological symptoms that usually precede or sometimes accompany the headache. Some patients also experience a premonitory phase, occurring hours or days before the headache, and a headache resolution phase. Premonitory and resolution symptoms include hyperactivity, hypoactivity, depression, cravings for particular foods, repetitive yawning, fatigue and neck stiffness and/or pain.

When a patient fulfils criteria for more than one subtype of migraine, all subtypes should be diagnosed and coded. For example, a patient who has frequent attacks with aura but also some attacks without aura should be coded as 1.2 *Migraine with aura* and 1.1 *Migraine without aura*. Attacks of either type are included in the diagnostic criteria for 1.3 *Chronic migraine*.

1.1 Migraine without aura

Previously used terms:

Common migraine; hemicrania simplex.

Description:

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

Diagnostic criteria:

- A. At least five attacks¹ fulfilling criteria B–D
- B. Headache attacks lasting 4–72 hours (untreated or unsuccessfully treated)^{2,3}
- C. Headache has at least two of the following four characteristics:
 1. unilateral location
 2. pulsating quality
 3. moderate or severe pain intensity
 4. aggravation by or causing avoidance of routine physical activity (e.g. walking or climbing stairs)
- D. During headache at least one of the following:
 1. nausea and/or vomiting
 2. photophobia and phonophobia
- E. Not better accounted for by another ICHD-3 diagnosis.

Notes:

1. One or a few migraine attacks may be difficult to distinguish from symptomatic migraine-like attacks. Furthermore, the nature of a single or a few attacks may be difficult to understand. Therefore, at least five attacks are required. Individuals who otherwise meet criteria for 1.1 *Migraine without aura* but have had fewer than five attacks, should be coded 1.5.1 *Probable migraine without aura*.
2. When the patient falls asleep during a migraine attack and wakes up without it, duration of the attack is reckoned until the time of awakening.
3. In children and adolescents (aged under 18 years), attacks may last 2–72 hours (the evidence for untreated durations of less than 2 hours in children has not been substantiated).

Comments:

Migraine headache in children and adolescents (aged under 18 years) is more often bilateral than is the case in adults; unilateral pain usually emerges in late adolescence or early adult life. Migraine headache is usually frontotemporal. Occipital headache in *children* is rare and calls for diagnostic caution. A subset of otherwise typical patients have facial location of pain, which is called ‘facial migraine’ in the literature; there is no evidence that these patients form a separate subgroup of migraine patients. In young children, photophobia and phonophobia may be inferred from their behaviour. Migraine attacks can be associated with cranial autonomic symptoms and symptoms of cutaneous allodynia.

Migraine without aura often has a menstrual relationship. ICHD-3 beta offers criteria for A1.1.1 *Pure menstrual migraine* and A1.1.2 *Menstrually related migraine*, but in the Appendix because of uncertainty over whether they should be regarded as separate entities.

Very frequent migraine attacks are now distinguished as 1.3 *Chronic migraine*. When there is associated medication overuse, both diagnoses, 1.3 *Chronic migraine* and 8.2 *Medication-overuse headache*, should be applied. 1.1 *Migraine without aura* is the disease most prone to accelerate with frequent use of symptomatic medication.

Regional cerebral blood flow imaging shows no changes suggestive of cortical spreading depression (CSD) during attacks of migraine without aura, although blood flow changes may occur in the brainstem, as may cortical changes secondary to pain activation. This contrasts with the pathognomonic spreading oligoemia of migraine with aura. Although the bulk of the literature suggests that CSD does not occur in migraine without aura, some recent studies disagree. Furthermore, it has been suggested that glial waves or other cortical phenomena may be involved in migraine without aura. The messenger molecules nitric oxide (NO), 5-hydroxytryptamine (5-HT) and calcitonin gene-related peptide (CGRP) are involved. Although the disease was previously regarded as primarily vascular, the importance of sensitization of pain pathways, and the possibility that attacks may originate in the central nervous system, have gained increasing attention over recent decades. At the same time, the circuitry of migraine pain, the trigeminovascular system, and several aspects of its neurotransmission peripherally and in the trigeminal nucleus caudalis, the central mesencephalic grey and the thalamus, have been recognized. New highly receptor-specific acute medications such as the triptans, which are 5HT_{1B/D} receptor agonists, 5-HT_{1F} receptor agonists and CGRP receptor antagonists have demonstrated efficacy in the acute treatment of attacks. Because of their high receptor-specificity, their mechanism of action provides new insight into migraine mechanisms. It is now clear that migraine without aura is a neurobiological disorder; clinical as well as basic neuroscience has advanced our knowledge of migraine mechanisms, and continues to do so.

1.2 Migraine with aura*Previously used terms:*

Classic or classical migraine; ophthalmic, hemiparalytic, hemiplegic or aphasic migraine; migraine accompagnée; complicated migraine.

Description:

Recurrent attacks, lasting minutes, of unilateral fully reversible visual, sensory or other central nervous system symptoms that usually develop gradually and are usually followed by headache and associated migraine symptoms.

Diagnostic criteria:

- A. At least two attacks fulfilling criteria B and C
- B. One or more of the following fully reversible aura symptoms:
 1. visual
 2. sensory
 3. speech and/or language
 4. motor
 5. brainstem
 6. retinal
- C. At least two of the following four characteristics:
 1. at least one aura symptom spreads gradually over ≥ 5 minutes, and/or two or more symptoms occur in succession
 2. each individual aura symptom lasts 5-60 minutes¹
 3. at least one aura symptom is unilateral²
 4. the aura is accompanied, or followed within 60 minutes, by headache
- D. Not better accounted for by another ICHD-3 diagnosis, and transient ischaemic attack has been excluded.

Notes:

1. When, for example, three symptoms occur during an aura, the acceptable maximal duration is 3×60 minutes. Motor symptoms may last up to 72 hours.
2. Aphasia is always regarded as a unilateral symptom; dysarthria may or may not be.

Comments:

The aura is the complex of neurological symptoms that occurs usually before the headache of 1.2 *Migraine with aura*, but it may begin after the pain phase has commenced, or continue into the headache phase.

Visual aura is the most common type of aura, occurring in over 90% of patients with 1.2 *Migraine with aura*, at least in some attacks. It often presents as a fortification spectrum: a zigzag figure near the point of fixation that may gradually spread right or left and assume a laterally convex shape with an angulated scintillating edge, leaving absolute or variable degrees of relative scotoma in its wake. In other cases, scotoma without positive phenomena may occur; this is often perceived as being of acute onset but, on scrutiny,

usually enlarges gradually. In children and adolescents, less typical bilateral visual symptoms occur that may represent an aura. A visual aura rating scale with high specificity and sensitivity has been developed and validated.

Next in frequency are sensory disturbances, in the form of pins and needles moving slowly from the point of origin and affecting a greater or smaller part of one side of the body, face and/or tongue. Numbness may occur in its wake, but numbness may also be the only symptom.

Less frequent are speech disturbances, usually aphasic but often hard to categorize.

When the aura includes motor weakness, the disorder should be coded as 1.2.3 *Hemiplegic migraine* or one of its subforms.

Aura symptoms of these different types usually follow one another in succession, beginning with visual, then sensory, then aphasic; but the reverse and other orders have been noted. The accepted duration for most aura symptoms is 1 hour, but motor symptoms are often longer lasting.

Patients often find it hard to describe their aura symptoms, in which case they should be instructed to time and record them prospectively. The clinical picture then becomes clearer. Common mistakes are incorrect reports of lateralization, of sudden rather than gradual onset and of monocular rather than homonymous visual disturbances, as well as of duration of aura and mistaking sensory loss for weakness. After an initial consultation, use of an aura diary may clarify the diagnosis.

Many patients who have migraine attacks with aura also have attacks without aura; they should be coded as both 1.2 *Migraine with aura* and 1.1 *Migraine without aura*.

Premonitory symptoms may begin hours or a day or two before the other symptoms of a migraine attack (with or without aura). They include various combinations of fatigue, difficulty in concentrating, neck stiffness, sensitivity to light and/or sound, nausea, blurred vision, yawning and pallor. The terms 'prodrome' and 'warning symptoms' are best avoided, because they are often mistakenly used to include aura.

Migraine aura is sometimes associated with a headache that does not fulfil criteria for 1.1 *Migraine without aura*, but this is still regarded as a migraine headache because of its relation to the aura. In other cases, migraine aura may occur without headache.

Before or simultaneously with the onset of aura symptoms, regional cerebral blood flow is decreased in the cortex corresponding to the clinically affected area and often over a wider area. Blood flow reduction usually starts posteriorly and spreads anteriorly, and is usually above the ischaemic threshold. After 1 to

several hours, gradual transition into hyperaemia occurs in the same region. Cortical spreading depression of Leão is the likely underlying mechanism.

Systematic studies have demonstrated that many patients with visual aura occasionally have symptoms in the extremities and/or speech symptoms. Conversely, patients with symptoms in the extremities and/or speech or language symptoms almost always also experience visual aura symptoms at least during some attacks. A distinction between migraine with visual aura, migraine with hemiparaesthetic aura and migraine with speech and/or language aura is probably artificial, and therefore is not recognized in this classification. They are all coded as 1.2.1 *Migraine with typical aura*. Patients with aura symptoms arising from the brainstem are coded as 1.2.2 *Migraine with brainstem aura*, but they almost always have additional typical aura symptoms. Patients with 1.2.3 *Hemiplegic migraine* have motor weakness, and this is classified as a separate subform because of genetic and pathophysiological differences from migraine with typical aura. Such patients often have brainstem symptoms in addition.

The previously defined syndromes, *migraine with prolonged aura* and *migraine with acute-onset aura*, have been abandoned. The great majority of patients with such attacks have other attacks that fulfil criteria for one of the recognized subforms of 1.2 *Migraine with aura*, and should be coded to that diagnosis. The rest should be coded to 1.5.2 *Probable migraine with aura*, specifying the atypical feature (prolonged aura or acute onset aura) in parenthesis. The diagnosis is usually evident after a careful history alone, although there are rare secondary mimics including carotid dissection, arteriovenous malformation and seizure.

1.2.1 Migraine with typical aura

Description:

Migraine with aura in which aura consists of visual and/or sensory and/or speech/language symptoms, but no motor weakness, and is characterized by gradual development, duration of each symptom no longer than 1 hour, a mix of positive and negative features and complete reversibility.

Diagnostic criteria:

- A. At least two attacks fulfilling criteria B and C
- B. Aura consisting of visual, sensory and/or speech/language symptoms, each fully reversible, but no motor, brainstem or retinal symptoms
- C. At least two of the following four characteristics:
 1. at least one aura symptom spreads gradually over ≥ 5 minutes, and/or two or more symptoms occur in succession

2. each individual aura symptom lasts 5-60 minutes¹
 3. at least one aura symptom is unilateral²
 4. the aura is accompanied, or followed within 60 minutes, by headache
- D. Not better accounted for by another ICHD-3 diagnosis, and transient ischaemic attack has been excluded.

Notes:

1. When for example three symptoms occur during an aura, the acceptable maximal duration is 3×60 minutes.
2. Aphasia is always regarded as a unilateral symptom; dysarthria may or may not be.

1.2.1.1 Typical aura with headache

Description:

Migraine with typical aura in which aura is accompanied or followed within 60 minutes by headache with or without migraine characteristics.

Diagnostic criteria:

- A. Fulfils criteria for 1.2.1 *Migraine with typical aura*
- B. Headache, with or without migraine characteristics, accompanies or follows the aura within 60 minutes.

1.2.1.2 Typical aura without headache

Description:

Migraine with typical aura in which aura is neither accompanied nor followed by headache of any sort.

Diagnostic criteria:

- A. Fulfils criteria for 1.2.1 *Migraine with typical aura*
- B. No headache accompanies or follows the aura within 60 minutes.

Comments:

In some patients, a typical aura is always followed by migraine headache, but many patients have, in addition, attacks with aura followed by a less distinct headache or even without headache. A number of patients have, exclusively, 1.2.1.2 *Typical aura without headache*.

In the absence of headache fulfilling criteria for 1.1 *Migraine without aura*, the precise diagnosis of aura and its distinction from mimics that may signal serious

disease (e.g. transient ischaemic attack) becomes more difficult and often requires investigation. When aura occurs for the first time after age 40, when symptoms are exclusively negative (e.g. hemianopia) or when aura is prolonged or very short, other causes, particularly transient ischaemic attacks, should be ruled out.

1.2.2 Migraine with brainstem aura

Previously used terms:

Basilar artery migraine; basilar migraine; basilar-type migraine.

Description:

Migraine with aura symptoms clearly originating from the brainstem, but no motor weakness.

Diagnostic criteria:

- A. At least two attacks fulfilling criteria B-D
- B. Aura consisting of visual, sensory and/or speech/language symptoms, each fully reversible, but no motor¹ or retinal symptoms
- C. At least two of the following brainstem symptoms:
 1. dysarthria
 2. vertigo
 3. tinnitus
 4. hypacusis
 5. diplopia
 6. ataxia
 7. decreased level of consciousness
- D. At least two of the following four characteristics:
 1. at least one aura symptom spreads gradually over ≥ 5 minutes, and/or two or more symptoms occur in succession
 2. each individual aura symptom lasts 5-60 minutes²
 3. at least one aura symptom is unilateral³
 4. the aura is accompanied, or followed within 60 minutes, by headache
- E. Not better accounted for by another ICHD-3 diagnosis, and transient ischaemic attack has been excluded.

Notes:

1. When motor symptoms are present, code as 1.2.3 *Hemiplegic migraine*.
2. When for example three symptoms occur during an aura, the acceptable maximal duration is 3×60 minutes.
3. Aphasia is always regarded as a unilateral symptom; dysarthria may or may not be.

Comments:

Originally the terms *basilar artery migraine* or *basilar migraine* were used but, as involvement of the basilar artery is unlikely, the term *migraine with brainstem aura* is preferred.

There are typical aura symptoms in addition to the brainstem symptoms during most attacks. Many patients who have attacks with brainstem aura also report other attacks with typical aura and should be coded for both 1.2.1 *Migraine with typical aura* and 1.2.2 *Migraine with brainstem aura*.

Many of the symptoms listed under criterion C may occur with anxiety and hyperventilation, and therefore are subject to misinterpretation.

1.2.3 Hemiplegic¹ migraine

Description:

Migraine with aura including motor weakness.

Diagnostic criteria:

- A. At least two attacks fulfilling criteria B and C
- B. Aura consisting of both of the following:
 1. fully reversible motor weakness
 2. fully reversible visual, sensory and/or speech/language symptoms
- C. At least two of the following four characteristics:
 1. at least one aura symptom spreads gradually over ≥ 5 minutes, and/or two or more symptoms occur in succession
 2. each individual non-motor aura symptom lasts 5-60 minutes, and motor symptoms last < 72 hours²
 3. at least one aura symptom is unilateral³
 4. the aura is accompanied, or followed within 60 minutes, by headache
- D. Not better accounted for by another ICHD-3 diagnosis, and transient ischaemic attack and stroke have been excluded.

Notes:

1. The term *plegic* means paralysis in most languages, but most attacks are characterized by motor weakness.
2. In some patients, motor weakness may last weeks.
3. Aphasia is always regarded as a unilateral symptom; dysarthria may or may not be.

Comment:

It may be difficult to distinguish weakness from sensory loss.

1.2.3.1 *Familial hemiplegic migraine (FHM)*

Description:

Migraine with aura including motor weakness, and at least one first- or second-degree relative has migraine aura including motor weakness.

Diagnostic criteria:

- A. Fulfils criteria for 1.2.3 *Hemiplegic migraine*
- B. At least one first- or second-degree relative has had attacks fulfilling criteria for 1.2.3 *Hemiplegic migraine*.

Comments:

New genetic data have allowed a more precise definition of 1.2.3.1 *Familial hemiplegic migraine (FHM)* than was possible previously. Specific genetic subtypes have been identified: in FHM1 there are mutations in the CACNA1A gene (coding for a calcium channel) on chromosome 19; in FHM2 there are mutations in the ATP1A2 gene (coding for a K/Na-ATPase) on chromosome 1; and in FHM3 there are mutations in the SCN1A gene (coding for a sodium channel) on chromosome 2. There may be other loci not yet identified. When genetic testing is done, the genetic subtype (if discovered) should be specified at the fifth digit.

It has been shown that 1.2.3.1 *Familial hemiplegic migraine (FHM)* very often presents with brainstem symptoms in addition to the typical aura symptoms, and that headache almost always occurs. Rarely, during FHM attacks, disturbances of consciousness (sometimes including coma), confusion, fever and CSF pleocytosis can occur.

1.2.3.1 *Familial hemiplegic migraine (FHM)* may be mistaken for epilepsy and (unsuccessfully) treated as such. FHM attacks can be triggered by (mild) head trauma. In approximately 50% of FHM families, chronic progressive cerebellar ataxia occurs independently of the migraine attacks.

1.2.3.1.1 *Familial hemiplegic migraine type 1 (FHM1)*

Diagnostic criteria:

- A. Fulfils criteria for 1.2.3.1 *Familial hemiplegic migraine*
- B. A causative mutation on the CACNA1A gene has been demonstrated.

1.2.3.1.2 *Familial hemiplegic migraine type 2 (FHM2)*

Diagnostic criteria:

- A. Fulfils criteria for 1.2.3.1 *Familial hemiplegic migraine*
- B. A causative mutation on the ATP1A2 gene has been demonstrated.

1.2.3.1.3 *Familial hemiplegic migraine type 3 (FHM3)*

Diagnostic criteria:

- A. Fulfils criteria for 1.2.3.1 *Familial hemiplegic migraine*
- B. A causative mutation on the SCN1A gene has been demonstrated.

1.2.3.1.4 *Familial hemiplegic migraine, other loci*

Diagnostic criteria:

- A. Fulfils criteria for 1.2.3.1 *Familial hemiplegic migraine*
- B. Genetic testing has demonstrated no mutation on the CACNA1A, ATP1A2 or SCN1A genes.

1.2.3.2 *Sporadic hemiplegic migraine*

Description:

Migraine with aura including motor weakness, and no first- or second-degree relative has migraine aura including motor weakness.

Diagnostic criteria:

- A. Fulfils criteria for 1.2.3 *Hemiplegic migraine*
- B. No first- or second-degree relative fulfils criteria for 1.2.3 *Hemiplegic migraine*.

Comments:

Epidemiological studies have shown that sporadic cases occur with approximately the same prevalence as familial cases.

The attacks in 1.2.3.2 *Sporadic hemiplegic migraine* have the same clinical characteristics as those in 1.2.3.1 *Familial hemiplegic migraine*. Some apparently sporadic cases have known FHM mutations, and in some a first- or second-degree relative later develops hemiplegic migraine, thus completing fulfilment of the criteria for 1.2.3.1 *Familial hemiplegic migraine* and requiring a change of diagnosis.

Sporadic cases usually require neuroimaging and other tests to rule out other causes. A lumbar puncture may be necessary to rule out 7.3.5 *Syndrome of transient Headache and Neurological Deficits with cerebrospinal fluid Lymphocytosis (HaNDL)*.

1.2.4 Retinal migraine

Description:

Repeated attacks of monocular visual disturbance, including scintillations, scotomata or blindness, associated with migraine headache.

Diagnostic criteria:

- A. At least two attacks fulfilling criteria B and C
- B. Aura consisting of fully reversible monocular positive and/or negative visual phenomena (e.g. scintillations, scotomata or blindness) confirmed during an attack by either or both of the following:
 1. clinical visual field examination
 2. the patient's drawing (made after clear instruction) of a monocular field defect
- C. At least two of the following three characteristics
 1. the aura spreads gradually over ≥ 5 minutes
 2. aura symptoms last 5-60 minutes
 3. the aura is accompanied, or followed within 60 minutes, by headache
- D. Not better accounted for by another ICHD-3 diagnosis, and other causes of amaurosis fugax have been excluded.

Comments:

Some patients who complain of monocular visual disturbance in fact have hemianopia. Some cases without headache have been reported, but migraine cannot be ascertained as the underlying aetiology.

1.2.4 *Retinal migraine* is an extremely rare cause of transient monocular visual loss. Cases of permanent monocular visual loss associated with migraine have been described. Appropriate investigations are required to exclude other causes of transient monocular blindness.

1.3 Chronic migraine^{1,2}

Description:

Headache occurring on 15 or more days per month for more than 3 months, which has the features of migraine headache on at least 8 days per month.

Diagnostic criteria:

- A. Headache (tension-type-like and/or migraine-like) on ≥ 15 days per month for >3 months² and fulfilling criteria B and C
- B. Occurring in a patient who has had at least five attacks fulfilling criteria B-D for 1.1 *Migraine without aura* and/or criteria B and C for 1.2 *Migraine with aura*
- C. On ≥ 8 days per month for >3 months, fulfilling any of the following³:
 1. criteria C and D for 1.1 *Migraine without aura*
 2. criteria B and C for 1.2 *Migraine with aura*
 3. believed by the patient to be migraine at onset and relieved by a triptan or ergot derivative
- D. Not better accounted for by another ICHD-3 diagnosis.

Notes:

1. The diagnosis of 1.3 *Chronic migraine* excludes the diagnosis of 2. *Tension-type headache* or its subtypes because tension-type-like headache is within the diagnostic criteria for 1.3 *Chronic migraine*.
2. The reason for singling out chronic from episodic migraine is that it is impossible to distinguish the individual episodes of headache in patients with such frequent or continuous headaches. In fact, the characteristics of the headache may change not only from day to day but even within the same day. It is extremely difficult to keep such patients medication-free in order to observe the natural history of the headache. In this situation, attacks with or without aura are both counted, as well as tension-type-like headaches. The most common cause of symptoms suggestive of chronic migraine is medication overuse, as defined under 8.2 *Medication-overuse headache*. Around 50% of patients apparently with 1.3 *Chronic migraine* revert to an episodic migraine subtype after drug withdrawal; such patients are in a sense wrongly diagnosed as 1.3 *Chronic migraine*. Equally, many patients apparently overusing medication do not improve after drug withdrawal, and the diagnosis of 8.2 *Medication-overuse headache* may in a sense be inappropriate (assuming that chronicity induced by drug overuse is always reversible). For these reasons, and because of the general rule, patients meeting criteria for 1.3 *Chronic migraine* and for 8.2 *Medication-overuse headache* should be given both diagnoses. After drug withdrawal, migraine will either revert to the episodic subtype or remain chronic, and be re-diagnosed accordingly; in the latter case, the diagnosis of 8.2 *Medication-overuse*

headache may be rescinded. In some countries, it is usual practice to diagnose 8.2 *Medication-overuse headache* only on discharge.

3. Characterization of frequently recurring headache generally requires a headache diary to record information on pain and associated symptoms day-by-day for at least 1 month. Sample diaries are available at <http://www.i-h-s.org>.

1.4 Complications of migraine

Comment:

Code separately for both the migraine subtype and for the complication.

1.4.1 *Status migrainosus*

Description:

A debilitating migraine attack lasting for more than 72 hours.

Diagnostic criteria:

- A. A headache attack fulfilling criteria B and C
- B. Occurring in a patient with 1.1 *Migraine without aura* and/or 1.2 *Migraine with aura*, and typical of previous attacks except for its duration and severity
- C. Both of the following characteristics:
 1. unremitting for >72 hours¹
 2. pain and/or associated symptoms are debilitating²
- D. Not better accounted for by another ICHD-3 diagnosis.

Notes:

1. Remissions of up to 12 hours because of medication or sleep are accepted.
2. Milder cases, not meeting criterion C2, are coded 1.5.1 *Probable migraine without aura*.

Comments:

Headache with the features of 1.4.1 *Status migrainosus* may often be caused by medication overuse. When headache in these circumstances meets the criteria for 8.2 *Medication-overuse headache*, code for 1.3 *Chronic migraine* and 8.2 *Medication-overuse headache* but not for 1.4.1 *Status migrainosus*. When overuse of medication is of shorter duration than 3 months, code for the appropriate migraine subtype(s) only.

1.4.2 *Persistent aura without infarction*

Description:

Aura symptoms persisting for 1 week or more without evidence of infarction on neuroimaging.

Diagnostic criteria:

- A. Aura fulfilling criterion B
- B. Occurring in a patient with 1.2 *Migraine with aura* and typical of previous auras except that one or more aura symptoms persists for ≥ 1 week
- C. Neuroimaging shows no evidence of infarction
- D. Not better accounted for by another ICHD-3 diagnosis.

Comments:

Persistent aura symptoms are rare but well documented. They are often bilateral and may last for months or years. The 1-week minimum in criterion B is based on the opinion of experts and should be formally studied.

Diagnostic work-up must distinguish 1.4.2 *Persistent aura without infarction* from 1.4.3 *Migrainous infarction*, and exclude symptomatic aura as a result of cerebral infarction of other causes. Attacks lasting more than 1 hour and less than 1 week and not fulfilling criteria for 1.2.1 *Migraine with typical aura* are coded 1.5.2 *Probable migraine with aura*.

1.4.3 *Migrainous infarction*

Description:

One or more migraine aura symptoms associated with an ischaemic brain lesion in the appropriate territory demonstrated by neuroimaging.

Diagnostic criteria:

- A. A migraine attack fulfilling criteria B and C
- B. Occurring in a patient with 1.2 *Migraine with aura* and typical of previous attacks except that one or more aura symptoms persists for >60 minutes
- C. Neuroimaging demonstrates ischaemic infarction in a relevant area
- D. Not better accounted for by another diagnosis.

Comments:

Ischaemic stroke in a migraine sufferer may be categorized as cerebral infarction of other cause coexisting with migraine, cerebral infarction of other cause presenting

with symptoms resembling migraine with aura, or cerebral infarction occurring during the course of a typical migraine with aura attack. Only the last fulfils criteria for 1.4.3 *Migrainous infarction*.

1.4.3 *Migrainous infarction* mostly occurs in the posterior circulation and in younger women

A two-fold increased risk of ischaemic stroke in patients with migraine with aura patients has been demonstrated in several population-based studies. However, it should be noted that these infarctions are not migrainous infarctions. The mechanisms of the increased risk of ischaemic stroke in migraine sufferers remain unclear; likewise, the relationship between frequency of aura and the nature of aura symptoms denoting the increase in risk is unknown. Most studies have shown a lack of association between migraine without aura and ischaemic stroke.

1.4.4 *Migraine aura-triggered seizure*

Description:

A seizure triggered by an attack of migraine with aura.

Diagnostic criteria:

- A. A seizure fulfilling diagnostic criteria for one type of epileptic attack, and criterion B below
- B. Occurring in a patient with 1.2 *Migraine with aura*, and during, or within 1 hour after, an attack of migraine with aura
- C. Not better accounted for by another diagnosis.

Comment:

Migraine and epilepsy are prototypical examples of paroxysmal brain disorders. Although migraine-like headaches are quite frequently seen in the epileptic postictal period, sometimes a seizure occurs during or following a migraine attack. This phenomenon, sometimes referred to as *migrainepoly*, is a rare event, originally described in patients with 1.2 *Migraine with aura*. Evidence for association with 1.1 *Migraine without aura* is still lacking.

1.5 Probable migraine

Previously used term:

Migrainous disorder.

Coded elsewhere:

Migraine-like headache secondary to another disorder (symptomatic migraine) is coded according to that disorder.

Description:

Migraine-like attacks missing one of the features required to fulfil all criteria for a subtype of migraine coded above, and not fulfilling criteria for another headache disorder.

Diagnostic criteria:

- A. Attacks fulfilling all but one of criteria A-D for 1.1 *Migraine without aura*, or all but one of criteria A-C for 1.2 *Migraine with aura*
- B. Not fulfilling ICHD-3 criteria for any other headache disorder
- C. Not better accounted for by another ICHD-3 diagnosis.

Comment:

In making a headache diagnosis, attacks that fulfil criteria for both 2. *Tension-type headache* and 1.5 *Probable migraine* are coded as the former in accordance with the general rule that a definite diagnosis always trumps a probable diagnosis. However, in patients who already have a migraine diagnosis, and where the issue is to count the number of attacks they are having (e.g. as an outcome measure in a drug trial), attacks fulfilling criteria for 1.5 *Probable migraine* should be counted as migraine. The reason for this is that mild migraine attacks, or attacks treated early, often do not achieve all characteristics necessary for a migraine attack diagnosis but nevertheless respond to specific migraine treatments.

1.5.1 *Probable migraine without aura*

Diagnostic criteria:

- A. Attacks fulfilling all but one of criteria A–D for 1.1 *Migraine without aura*
- B. Not fulfilling ICHD-3 criteria for any other headache disorder
- C. Not better accounted for by another ICHD-3 diagnosis.

1.5.2 *Probable migraine with aura*

Diagnostic criteria:

- A. Attacks fulfilling all but one of criteria A–C for 1.2 *Migraine with aura* or any of its subforms
- B. Not fulfilling ICHD-3 criteria for any other headache disorder
- C. Not better accounted for by another ICHD-3 diagnosis.

1.6 Episodic syndromes that may be associated with migraine

Previously used terms:

Childhood periodic syndromes; periodic syndromes of childhood.

Comments:

This group of disorders occurs in patients who also have 1.1 *Migraine without aura* or 1.2 *Migraine with aura*, or who have an increased likelihood to develop either of these disorders. Although historically noted to occur in childhood, they may also occur in adults.

Additional conditions that may also occur in these patients include episodes of motion sickness and periodic sleep disorders including sleep walking, sleep talking, night terrors and bruxism.

1.6.1 Recurrent gastrointestinal disturbance

Previously used terms:

Chronic abdominal pain; functional abdominal pain; functional dyspepsia; irritable bowel syndrome; functional abdominal pain syndrome.

Description:

Recurrent episodic attacks of abdominal pain and/or discomfort, nausea and/or vomiting, occurring infrequently, chronically or at predictable intervals, that may be associated with migraine.

Diagnostic criteria:

- A. At least five attacks with distinct episodes of abdominal pain and/or discomfort and/or nausea and/or vomiting
- B. Normal gastrointestinal examination and evaluation
- C. Not attributed to another disorder.

1.6.1.1 *Cyclic vomiting syndrome*

Description:

Recurrent episodic attacks of intense nausea and vomiting, usually stereotypical in the individual and with predictable timing of episodes. Attacks may be associated with pallor and lethargy. There is complete resolution of symptoms between attacks.

Diagnostic criteria:

- A. At least five attacks of intense nausea and vomiting, fulfilling criteria B and C

- B. Stereotypical in the individual patient and recurring with predictable periodicity
- C. All of the following:
 - 1. nausea and vomiting occur at least four times per hour
 - 2. attacks last ≥ 1 hour and up to 10 days
 - 3. attacks occur ≥ 1 week apart
- D. Complete freedom from symptoms between attacks
- E. Not attributed to another disorder.¹

Note:

- 1. In particular, history and physical examination do not show signs of gastrointestinal disease.

Comments:

1.6.1.1 *Cyclic vomiting syndrome* is typically a self-limiting episodic condition occurring in childhood, with periods of complete normality between episodes. The cyclic nature is the hallmark, and is predictable.

This disorder was not included as a childhood periodic syndrome in ICHD-I, but it was in ICHD-II. The clinical features of this syndrome resemble those found in association with migraine headaches, and multiple threads of research over the last years have suggested that cyclic vomiting syndrome is a condition related to migraine.

1.6.1.2 *Abdominal migraine*

Description:

An idiopathic disorder seen mainly in children as recurrent attacks of moderate to severe midline abdominal pain, associated with vasomotor symptoms, nausea and vomiting, lasting 2–72 hours and with normality between episodes. Headache does not occur during these episodes.

Diagnostic criteria:

- A. At least five attacks of abdominal pain, fulfilling criteria B–D
- B. Pain has at least two of the following three characteristics:
 - 1. midline location, periumbilical or poorly localized
 - 2. dull or ‘just sore’ quality
 - 3. moderate or severe intensity
- C. During attacks, at least two of the following:
 - 1. anorexia
 - 2. nausea
 - 3. vomiting
 - 4. pallor

- D. Attacks last 2-72 hours when untreated or unsuccessfully treated
- E. Complete freedom from symptoms between attacks
- F. Not attributed to another disorder.¹

Note:

1. In particular, history and physical examination do not show signs of gastrointestinal or renal disease, or such disease has been ruled out by appropriate investigations.

Comments:

Pain of 1.6.1.2 *Abdominal migraine* is severe enough to interfere with normal daily activities.

In young children the presence of headache is often overlooked. A careful history of presence or absence of headache must be taken and, if headache or head pain during attacks is identified, a diagnosis of 1.1 *Migraine without aura* should be considered.

Children may find it difficult to distinguish anorexia from nausea. Pallor is often accompanied by dark shadows under the eyes. In a few patients, flushing is the predominant vasomotor phenomenon.

Most children with abdominal migraine will develop migraine headache later in life.

1.6.2 *Benign paroxysmal vertigo*

Description:

A disorder characterized by recurrent brief attacks of vertigo, occurring without warning and resolving spontaneously, in otherwise healthy children.

Diagnostic criteria:

- A. At least five attacks fulfilling criteria B and C
- B. Vertigo¹ occurring without warning, maximal at onset and resolving spontaneously after minutes to hours without loss of consciousness
- C. At least one of the following associated symptoms or signs:
 1. nystagmus
 2. ataxia
 3. vomiting
 4. pallor
 5. fearfulness
- D. Normal neurological examination and audiometric and vestibular functions between attacks
- E. Not attributed to another disorder.

Note:

1. Young children with vertigo may not be able to describe vertiginous symptoms. Parental observation of episodic periods of unsteadiness may be interpreted as vertigo in young children.

Comments:

Posterior fossa tumours, seizures and vestibular disorders must be excluded.

The relationship between 1.6.2 *Benign paroxysmal vertigo* and A1.6.6 *Vestibular migraine* (see Appendix) needs to be further examined.

1.6.3 *Benign paroxysmal torticollis*

Description:

Recurrent episodes of head tilt to one side, perhaps with slight rotation, which remit spontaneously. The condition occurs in infants and small children, with onset in the first year.

Diagnostic criteria:

- A. Recurrent attacks¹ in a young child, fulfilling criteria B and C
- B. Tilt of the head to either side, with or without slight rotation, remitting spontaneously after minutes to days
- C. At least one of the following associated symptoms or signs:
 1. pallor
 2. irritability
 3. malaise
 4. vomiting
 5. ataxia²
- D. Normal neurological examination between attacks
- E. Not attributed to another disorder.

Notes:

1. Attacks tend to recur monthly.
2. Ataxia is more likely in older children within the affected age group.

Comments:

The child's head can be returned to the neutral position during attacks: some resistance may be encountered, but can be overcome.

The differential diagnosis includes gastro-oesophageal reflux, idiopathic torsional dystonia and complex partial seizure, but particular attention must be paid to

the posterior fossa and craniocervical junction where congenital or acquired lesions may produce torticollis. These observations need further validation by patient diaries, structured interviews and longitudinal data collection.

1.6.3 *Benign paroxysmal torticollis* may evolve into 1.6.2 *Benign paroxysmal vertigo* or 1.2 *Migraine with aura* (particularly 1.2.2 *Migraine with brainstem aura*), or cease without further symptoms.

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2. Tension-type headache (TTH)

- 2.1 Infrequent episodic tension-type headache
 - 2.1.1 Infrequent episodic tension-type headache associated with pericranial tenderness
 - 2.1.2 Infrequent episodic tension-type headache not associated with pericranial tenderness
- 2.2 Frequent episodic tension-type headache
 - 2.2.1 Frequent episodic tension-type headache associated with pericranial tenderness
 - 2.2.2 Frequent episodic tension-type headache not associated with pericranial tenderness
- 2.3 Chronic tension-type headache
 - 2.3.1 Chronic tension-type headache associated with pericranial tenderness
 - 2.3.2 Chronic tension-type headache not associated with pericranial tenderness
- 2.4 Probable tension-type headache
 - 2.4.1 Probable infrequent episodic tension-type headache
 - 2.4.2 Probable frequent episodic tension-type headache
 - 2.4.3 Probable chronic tension-type headache

Previously used terms:

Tension headache; muscle contraction headache; psychomyogenic headache; stress headache; ordinary headache; essential headache; idiopathic headache; psychogenic headache.

Coded elsewhere:

Tension-type-like headache attributed to another disorder is coded to that disorder.

General comment

Primary or secondary headache or both?

When a headache with the characteristics of tension-type characteristics occurs for the first time in close temporal relation to another disorder that is known to cause headache, or fulfils other criteria for causation by that disorder, the new headache is coded as a secondary headache attributed to the causative disorder. When *pre-existing* tension-type headache becomes *chronic* in close temporal relation to such a causative disorder, both the initial tension-type headache diagnosis and the secondary diagnosis should be given. When *pre-existing* tension-type headache is made significantly worse (usually meaning a two-fold or greater increase in frequency and/or severity) in close temporal relation to such a causative disorder, both the initial tension-type headache diagnosis and the secondary diagnosis should be given, provided that there is good evidence that the disorder can

cause headache. In the case of chronic tension-type headache with medication overuse, a close temporal relation is often difficult to establish. Therefore, both diagnoses, 2.3 *Chronic tension-type headache* and 8.2 *Medication-overuse headache*, should be given in all such cases.

Introduction

2. *Tension-type headache* is very common, with a lifetime prevalence in the general population ranging between 30% and 78% in different studies, and it has a very high socio-economic impact.

Although this type of headache was previously considered to be primarily psychogenic, a number of studies have appeared after publication of ICHD-I that strongly suggest a neurobiological basis, at least for the more severe subtypes of tension-type headache.

The division of 2. *Tension-type headache* into *episodic* and *chronic* subtypes, which was introduced in ICHD-I, has proved extremely useful. In ICHD-II, the episodic form was further subdivided into an *infrequent* subform with headache episodes less than once per month and a *frequent* subform. 2.3 *Chronic tension-type headache* is a serious disease, causing greatly decreased quality of life and high disability. 2.2 *Frequent episodic tension-type headache* can be associated with considerable disability, and sometimes warrants treatment with expensive drugs. In contrast, 2.1 *Infrequent episodic tension-type headache*, which occurs in almost the entire population, usually has very little impact on the individual and, in most instances, requires no attention from the medical profession. The distinction of 2.1 *Infrequent episodic tension-type headache* from 2.2 *Frequent episodic tension-type headache* thus separates individuals who typically do not require medical management, and avoids categorizing almost the entire population as having a significant headache disorder, yet allows their headaches to be classified.

The exact mechanisms of 2. *Tension-type headache* are not known. Peripheral pain mechanisms are most likely to play a role in 2.1 *Infrequent episodic tension-type headache* and 2.2 *Frequent episodic tension-type headache*, whereas central pain mechanisms play a more important role in 2.3 *Chronic tension-type headache*. Increased pericranial tenderness recorded by manual palpation is the most significant abnormal finding in patients with tension-type headache. The tenderness is typically present interictally, is further increased during actual headache and increases with the intensity and frequency of headaches. Pericranial tenderness is easily recorded by manual palpation by small rotating movements and a firm pressure (preferably aided by use

of a palpometer) with the second and third fingers on the frontal, temporal, masseter, pterygoid, sternocleidomastoid, splenius and trapezius muscles. Local tenderness scores of 0-3 for each muscle can be summed to yield a total tenderness score for each individual. Palpation is a useful guide for treatment strategy. It also adds value and credibility to the explanations given to the patient.

Increased tenderness is most likely of pathophysiological importance. The Classification Committee encourages further research into the pathophysiological mechanisms and treatment of 2. *Tension-type headache*. With this aim, ICHD-II distinguished patients with and without such disorder of the pericranial muscles, and this subdivision is maintained in ICHD-3 beta to stimulate further research in this area.

The diagnostic difficulty most often encountered among the primary headache disorders is to discriminate between tension-type headache and mild migraine without aura. This is more so because patients with frequent headaches often suffer from both disorders. It has been suggested to tighten the diagnostic criteria for 2. *Tension-type headache* in the hope of excluding migraine that phenotypically resembles tension-type headache. Such an increase in specificity would, at the same time, reduce the sensitivity of the criteria, resulting in a larger proportion of patients whose headaches could be classified only as 2.4 *Probable tension-type headache* or 1.5 *Probable migraine*. Stricter diagnostic criteria for 2. *Tension-type headache* were proposed in the Appendix of ICHD-II, as A2. *Tension-type headache*, but so far with no evidence that such a change would be beneficial. These stricter diagnostic criteria remain in the Appendix, for research purposes only. The Classification Committee recommends comparisons between patients diagnosed according to each set of criteria, not only for characterization of clinical features but also for enquiry into pathophysiological mechanisms and response to treatments.

2.1 Infrequent episodic tension-type headache

Description:

Infrequent episodes of headache, typically bilateral, pressing or tightening in quality and of mild to moderate intensity, lasting minutes to days. The pain does not worsen with routine physical activity and is not associated with nausea, but photophobia or phonophobia may be present.

Diagnostic criteria:

A. At least 10 episodes of headache occurring on <1 day per month on average (<12 days per year) and fulfilling criteria B-D

- B. Lasting from 30 minutes to 7 days
- C. At least two of the following four characteristics:
 1. bilateral location
 2. pressing or tightening (non-pulsating) quality
 3. mild or moderate intensity
 4. not aggravated by routine physical activity such as walking or climbing stairs
- D. Both of the following:
 1. no nausea or vomiting
 2. no more than one of photophobia or phonophobia
- E. Not better accounted for by another ICHD-3 diagnosis.

2.1.1 Infrequent episodic tension-type headache associated with pericranial tenderness

Diagnostic criteria:

- A. Episodes fulfilling criteria for 2.1 *Infrequent episodic tension-type headache*
- B. Increased pericranial tenderness on manual palpation.

2.1.2 Infrequent episodic tension-type headache not associated with pericranial tenderness

Diagnostic criteria:

- A. Episodes fulfilling criteria for 2.1 *Infrequent episodic tension-type headache*
- B. No increase in pericranial tenderness.

2.2 Frequent episodic tension-type headache

Description:

Frequent episodes of headache, typically bilateral, pressing or tightening in quality and of mild to moderate intensity, lasting minutes to days. The pain does not worsen with routine physical activity and is not associated with nausea, but photophobia or phonophobia may be present.

Diagnostic criteria:

- A. At least 10 episodes of headache occurring on 1-14 days per month on average for >3 months (≥ 12 and <180 days per year) and fulfilling criteria B-D
- B. Lasting from 30 minutes to 7 days
- C. At least two of the following four characteristics:
 1. bilateral location
 2. pressing or tightening (non-pulsating) quality

- 3. mild or moderate intensity
- 4. not aggravated by routine physical activity such as walking or climbing stairs
- D. Both of the following:
 - 1. no nausea or vomiting
 - 2. no more than one of photophobia or phonophobia
- E. Not better accounted for by another ICHD-3 diagnosis.

Comments:

2.2 *Frequent episodic tension-type headache* often coexists with 1.1 *Migraine without aura*. Coexisting tension-type headache in migraineurs should preferably be identified through use of a diagnostic headache diary. The treatment of migraine differs considerably from that of tension-type headache, and it is important to educate patients to distinguish between these headache types if they are to select the right treatment for each while avoiding medication overuse and the adverse consequence of 8.2 *Medication-overuse headache*.

When headache fulfils criteria for both 1.5 *Probable migraine* and 2. *Tension-type headache*, code as 2. *Tension-type headache* (or as any subtype of it for which the criteria are fulfilled) under the general rule that definite diagnoses always trump probable diagnoses. When headache fulfils criteria for both 1.5 *Probable migraine* and 2.4 *Probable tension-type headache*, code as the former under the general rule of hierarchy, which puts 1. *Migraine* and its subtypes before 2. *Tension-type headache* and its subtypes.

2.2.1 *Frequent episodic tension-type headache associated with pericranial tenderness*

Diagnostic criteria:

- A. Episodes fulfilling criteria for 2.2 *Frequent episodic tension-type headache*
- B. Increased pericranial tenderness on manual palpation.

2.2.2 *Frequent episodic tension-type headache not associated with pericranial tenderness*

Diagnostic criteria:

- A. Episodes fulfilling criteria for 2.2 *Frequent episodic tension-type headache*
- B. No increase in pericranial tenderness.

2.3 *Chronic tension-type headache*

Coded elsewhere:

4.10 *New daily persistent headache*.

Description:

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

Diagnostic criteria:

- A. Headache occurring on ≥ 15 days per month on average for > 3 months (≥ 180 days per year), fulfilling criteria B-D
- B. Lasting hours to days, or unremitting
- C. At least two of the following four characteristics:
 - 1. bilateral location
 - 2. pressing or tightening (non-pulsating) quality
 - 3. mild or moderate intensity
 - 4. not aggravated by routine physical activity such as walking or climbing stairs
- D. Both of the following:
 - 1. no more than one of photophobia, phonophobia or mild nausea
 - 2. neither moderate or severe nausea nor vomiting
- E. Not better accounted for by another ICHD-3 diagnosis.

Comments:

2.3 *Chronic tension-type headache* evolves over time from 2.2 *Frequent episodic tension-type headache*; when these criteria A-E are fulfilled by headache that, unambiguously, is daily and unremitting from less than 24 hours after its first onset, code as 4.10 *New daily persistent headache*. When the manner of onset is not remembered or is otherwise uncertain, code as 2.3 *Chronic tension-type headache*.

Both 2.3 *Chronic tension-type headache* and 1.3 *Chronic migraine* require headache on at least 15 days per month. For 2.3 *Chronic tension-type headache*, headache on at least 15 days must meet the criteria for 2. *Tension-type headache* and for 1.3 *Chronic migraine* headache on at least 8 days must meet the criteria for 1. *Migraine*. Therefore, a patient can fulfil criteria for both these diagnoses, for example by having headache on 25 days per month meeting migraine criteria on 8 days and tension-type headache criteria on 17 days. In these cases, only the diagnosis 1.3 *Chronic migraine* should be given.

In many uncertain cases there is overuse of medication. When this fulfils criterion B for any of the subforms of 8.2 *Medication-overuse headache* and the criteria for 2.3 *Chronic tension-type headache* are also fulfilled, the rule is to code for both 2.3 *Chronic tension-type headache* and 8.2 *Medication-overuse headache*. After drug withdrawal, the diagnosis should be re-evaluated: not uncommonly the criteria for 2.3 *Chronic tension-type headache* will no longer be fulfilled, with reversion to one or other episodic subtype.

2.3.1 *Chronic tension-type headache associated with pericranial tenderness*

Diagnostic criteria:

- A. Headache fulfilling criteria for 2.3 *Chronic tension-type headache*
- B. Increased pericranial tenderness on manual palpation.

2.3.2 *Chronic tension-type headache not associated with pericranial tenderness*

Diagnostic criteria:

- A. Headache fulfilling criteria for 2.3 *Chronic tension-type headache*
- B. No increase in pericranial tenderness.

2.4 Probable tension-type headache

Description:

Tension-type-like headache missing one of the features required to fulfil all criteria for a subtype of tension-type headache coded above, and not fulfilling criteria for another headache disorder.

Comment:

Patients meeting one of the sets of criteria below may also meet the criteria for one of the subforms of 1.6 *Probable migraine*. In such cases, all other available information should be used to decide which of the alternatives is the more likely.

2.4.1 *Probable infrequent episodic tension-type headache*

Diagnostic criteria:

- A. One or more episodes of headache fulfilling all but one of criteria A-D for 2.1 *Infrequent episodic tension-type headache*
- B. Not fulfilling ICHD-3 criteria for any other headache disorder

- C. Not better accounted for by another ICHD-3 diagnosis.

2.4.2 *Probable frequent episodic tension-type headache*

Diagnostic criteria:

- A. Episodes of headache fulfilling all but one of criteria A-D for 2.2 *Frequent episodic tension-type headache*
- B. Not fulfilling ICHD-3 criteria for any other headache disorder
- C. Not better accounted for by another ICHD-3 diagnosis.

2.4.3 *Probable chronic tension-type headache*

Diagnostic criteria:

- A. Headache fulfilling all but one of criteria A-D for 2.3 *Chronic episodic tension-type headache*
- B. Not fulfilling ICHD-3 criteria for any other headache disorder
- C. Not better accounted for by another ICHD-3 diagnosis.

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3. Trigeminal autonomic cephalalgias (TACs)

- 3.1 Cluster headache
 - 3.1.1 Episodic cluster headache
 - 3.1.2 Chronic cluster headache
- 3.2 Paroxysmal hemicrania
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 - 3.5.4 Probable hemicrania continua

General comment

Primary or secondary headache or both?

When a new headache with the characteristics of a trigeminal autonomic cephalalgia (TAC) occurs for the first time in close temporal relation to another disorder known to cause headache, or fulfils other criteria for causation by that disorder, the new headache is coded as a secondary headache attributed to the causative disorder. When a *pre-existing* TAC becomes *chronic* in close temporal relation to such a causative disorder, both the initial TAC diagnosis and the secondary diagnosis should be given. When a *pre-existing* TAC is made significantly worse (usually meaning a two-fold or greater increase in frequency and/or severity) in close temporal relation to such a causative disorder, both the initial TAC diagnosis and the secondary headache diagnosis should be given, provided that there is good evidence that the disorder can cause headache.

Introduction

The trigeminal autonomic cephalalgias (TACs) share the clinical features of headache, which is usually lateralized, and often prominent cranial parasympathetic

autonomic features, which are again lateralized and ipsilateral to the headache. Experimental and human functional imaging suggests that these syndromes activate a normal human trigeminal parasympathetic reflex, with clinical signs of cranial sympathetic dysfunction being secondary.

Typical migraine aura can be seen, rarely, in association with TACs.

3.1 Cluster headache

Previously used terms:

Ciliary neuralgia; erythro-melalgia of the head; erythroprosopalgia of Bing; hemicrania angioparalytica; hemicrania neuralgiformis chronica; histaminic cephalalgia; Horton's headache; Harris-Horton's disease; migrainous neuralgia (of Harris); petrosal neuralgia (of Gardner); Sluder's neuralgia; sphenopalatine neuralgia; vidian neuralgia.

Coded elsewhere:

Symptomatic cluster headache, secondary to another disorder, is coded as a secondary headache attributed to that disorder.

Description:

Attacks of severe, strictly unilateral pain which is orbital, supraorbital, temporal or in any combination of these sites, lasting 15–180 minutes and occurring from once every other day to eight times a day. The pain is associated with ipsilateral conjunctival injection, lacrimation, nasal congestion, rhinorrhoea, forehead and facial sweating, miosis, ptosis and/or eyelid oedema, and/or with restlessness or agitation.

Diagnostic criteria:

- A. At least five attacks fulfilling criteria B–D
- B. Severe or very severe unilateral orbital, supraorbital and/or temporal pain lasting 15–180 minutes (when untreated)¹
- C. Either or both of the following:
 - 1. at least one of the following symptoms or signs, ipsilateral to the headache:
 - a) conjunctival injection and/or lacrimation
 - b) nasal congestion and/or rhinorrhoea
 - c) eyelid oedema
 - d) forehead and facial sweating
 - e) forehead and facial flushing
 - f) sensation of fullness in the ear
 - g) miosis and/or ptosis
 - 2. a sense of restlessness or agitation
- D. Attacks have a frequency between one every other day and eight per day for more than half of the time when the disorder is active

E. Not better accounted for by another ICHD-3 diagnosis.

B. At least two cluster periods lasting from 7 days to 1 year (when untreated) and separated by pain-free remission periods of ≥ 1 month.

Note:

1. During part (but less than half) of the time-course of 3.1 *Cluster headache*, attacks may be less severe and/or of shorter or longer duration.

Comment:

Cluster periods usually last between 2 weeks and 3 months.

Comments:

Attacks occur in series lasting for weeks or months (so-called cluster periods) separated by remission periods usually lasting months or years. About 10–15% of patients have 3.1.2 *Chronic cluster headache*, without such remission periods. In a large series with good follow-up, one-quarter of patients had only a single cluster period. Such patients meet the criteria for and should be coded as 3.1 *Cluster headache*.

The pain of 3.1 *Cluster headache* is maximal orbitally, supraorbitally, temporally or in any combination of these sites, but may spread to other regions. During the worst attacks, the intensity of pain is excruciating. Patients are usually unable to lie down, and characteristically pace the floor. Pain usually recurs on the same side of the head during an individual cluster period. During a cluster period in 3.1.1 *Episodic cluster headache*, and at any time in 3.1.2 *Chronic cluster headache*, attacks occur regularly and may be provoked by alcohol, histamine or nitroglycerin.

Age at onset is usually 20–40 years. For unknown reasons, men are afflicted three times more often than women.

Acute attacks involve activation in the region of the posterior hypothalamic grey matter. 3.1 *Cluster headache* may be autosomal dominant in about 5% of cases.

Some patients have been described who have both 3.1 *Cluster headache* and 13.1 *Trigeminal neuralgia* (sometimes referred to as *cluster-tic syndrome*). They should receive both diagnoses. The importance of this observation is that both conditions must be treated for the patient to become headache-free.

3.1.1 Episodic cluster headache

Description:

Cluster headache attacks occurring in periods lasting from 7 days to 1 year, separated by pain-free periods lasting at least 1 month.

Diagnostic criteria:

- A. Attacks fulfilling criteria for 3.1 *Cluster headache* and occurring in bouts (cluster periods)

3.1.2 Chronic cluster headache

Description:

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

Diagnostic criteria:

- A. Attacks fulfilling criteria for 3.1 *Cluster headache*, and criterion B below
- B. Occurring without a remission period, or with remissions lasting < 1 month, for at least 1 year.

Comment:

3.1.2 *Chronic cluster headache* may arise de novo (previously referred to as *primary chronic cluster headache*), or evolve from 3.1.1 *Episodic cluster headache* (previously *secondary chronic cluster headache*). In some patients change occurs from 3.1.2 *Chronic cluster headache* to 3.1.1 *Episodic cluster headache*.

3.2 Paroxysmal hemicrania

Description:

Attacks of severe, strictly unilateral pain which is orbital, supraorbital, temporal or in any combination of these sites, lasting 2–30 minutes and occurring several or many times a day. The attacks are associated with ipsilateral conjunctival injection, lacrimation, nasal congestion, rhinorrhoea, forehead and facial sweating, miosis, ptosis and/or eyelid oedema. They respond absolutely to indomethacin.

Diagnostic criteria:

- A. At least 20 attacks fulfilling criteria B-E
- B. Severe unilateral orbital, supraorbital and/or temporal pain lasting 2–30 minutes
- C. At least one of the following symptoms or signs, ipsilateral to the pain:
 1. conjunctival injection and/or lacrimation
 2. nasal congestion and/or rhinorrhoea
 3. eyelid oedema
 4. forehead and facial sweating

- 5. forehead and facial flushing
- 6. sensation of fullness in the ear
- 7. miosis and/or ptosis
- D. Attacks have a frequency above five per day for more than half of the time
- E. Attacks are prevented absolutely by therapeutic doses of indomethacin¹
- F. Not better accounted for by another ICHD-3 diagnosis.

Note:

- 1. In an adult, oral indomethacin should be used initially in a dose of at least 150 mg daily and increased if necessary up to 225 mg daily. The dose by injection is 100–200 mg. Smaller maintenance doses are often employed.

Comment:

In contrast to cluster headache, there is no male predominance. Onset is usually in adulthood, although childhood cases are reported.

3.2.1 Episodic paroxysmal hemicrania

Description:

Attacks of paroxysmal hemicrania occurring in periods lasting from 7 days to 1 year, separated by pain-free periods lasting at least 1 month.

Diagnostic criteria:

- A. Attacks fulfilling criteria for 3.2 *Paroxysmal hemicrania* and occurring in bouts
- B. At least two bouts lasting from 7 days to 1 year (when untreated) and separated by pain-free remission periods of ≥ 1 month.

3.2.2 Chronic paroxysmal hemicrania

Description:

Attacks of paroxysmal hemicrania occurring for more than 1 year without remission, or with remission periods lasting less than 1 month.

Diagnostic criteria:

- A. Attacks fulfilling criteria for 3.2 *Paroxysmal hemicrania*, and criterion B below
- B. Occurring without a remission period, or with remissions lasting < 1 month, for at least 1 year.

Comment:

Patients who fulfil criteria for both 3.2.2 *Chronic paroxysmal hemicrania* and 13.1 *Trigeminal neuralgia* (sometimes referred to as *CPH-tic syndrome*) should receive both diagnoses. Their recognition is important, as both disorders require treatment. The pathophysiological significance of the association is not yet clear.

3.3 Short-lasting unilateral neuralgiform headache attacks

Description:

Attacks of moderate or severe, strictly unilateral head pain lasting seconds to minutes, occurring at least once a day and usually associated with prominent lacrimation and redness of the ipsilateral eye.

Diagnostic criteria:

- A. At least 20 attacks fulfilling criteria B–D
- B. Moderate or severe unilateral head pain, with orbital, supraorbital, temporal and/or other trigeminal distribution, lasting for 1–600 seconds and occurring as single stabs, series of stabs or in a saw-tooth pattern
- C. At least one of the following cranial autonomic symptoms or signs, ipsilateral to the pain:
 - 1. conjunctival injection and/or lacrimation
 - 2. nasal congestion and/or rhinorrhoea
 - 3. eyelid oedema
 - 4. forehead and facial sweating
 - 5. forehead and facial flushing
 - 6. sensation of fullness in the ear
 - 7. miosis and/or ptosis
- D. Attacks have a frequency of at least one a day for more than half of the time when the disorder is active
- E. Not better accounted for by another ICHD-3 diagnosis.

Comments:

Longer-duration attacks are characterized by multiple stabs or a saw-tooth pain pattern.

Two subtypes of 3.3 *Short-lasting unilateral neuralgiform headache attacks* are recognized in ICHD-3 beta: 3.3.1 *Short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing (SUNCT)* and 3.3.2 *Short lasting unilateral neuralgiform headache attacks with cranial autonomic symptoms (SUNA)*. 3.3.1 *SUNCT* may be a subform of 3.3.2 *SUNA*, although this requires further study. Meanwhile, each is classified as a separate subtype, described below.

3.3.1 *SUNCT* and 3.3.2 *SUNA* are usually triggered without a refractory period. This is in contrast to

13.1 *Trigeminal neuralgia*, which usually has a refractory period after each attack.

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

Diagnostic criteria:

- A. Attacks fulfilling criteria for 3.3 *Short-lasting unilateral neuralgiform headache attacks*
- B. Both of conjunctival injection and lacrimation (tearing).

Comments:

The literature suggests that the most common mimic of 3.3.1 *SUNCT* is a lesion in the posterior fossa.

Patients have been described in whom there is overlap between 3.3.1 *SUNCT* and 13.1 *Trigeminal neuralgia*. Differentiation is clinically complex. Such patients should receive both diagnoses.

Patients with both 3.3.1 *SUNCT* and 3.1 *Cluster headache* have been reported; the pathophysiological significance of this overlap is yet to be determined.

3.3.1.1 *Episodic SUNCT*

Description:

Attacks of *SUNCT* occurring in periods lasting from 7 days to 1 year, separated by pain-free periods lasting 1 month or more.

Diagnostic criteria:

- A. Attacks fulfilling criteria for 3.3.1 *Short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing* and occurring in bouts
- B. At least two bouts lasting from 7 days to 1 year and separated by pain-free remission periods of ≥ 1 month.

3.3.1.2 *Chronic SUNCT*

Description:

Attacks of *SUNCT* occurring for more than 1 year without remission, or with remission periods lasting less than 1 month.

Diagnostic criteria:

- A. Attacks fulfilling criteria for 3.3.1 *Short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing*, and criterion B below

- B. Occurring without a remission period, or with remissions lasting < 1 month, for at least 1 year.

3.3.2 *Short lasting unilateral neuralgiform headache attacks with cranial autonomic symptoms (SUNA)*

- A. Attacks fulfilling criteria for 3.3 *Short-lasting unilateral neuralgiform headache attacks*, and criterion B below
- B. Only one or neither of conjunctival injection and lacrimation (tearing).

3.3.2.1 *Episodic SUNA*

Description:

Attacks of *SUNA* occurring in periods lasting from 7 days to 1 year, separated by pain-free periods lasting at least 1 month.

Diagnostic criteria:

- A. Attacks fulfilling criteria for 3.3.2 *Short-lasting unilateral neuralgiform headache attacks with cranial autonomic symptoms* and occurring in bouts
- B. At least two bouts lasting from 7 days to 1 year and separated by pain-free remission periods of ≥ 1 month.

3.3.2.2 *Chronic SUNA*

Description:

Attacks of *SUNA* occurring for more than 1 year without remission, or with remission periods lasting less than 1 month.

Diagnostic criteria:

- A. Attacks fulfilling criteria for 3.3.2 *Short-lasting unilateral neuralgiform headache attacks with cranial autonomic symptoms*, and criterion B below
- B. Occurring without a remission period, or with remissions lasting < 1 month, for at least 1 year.

3.4 *Hemicrania continua*

Description:

Persistent, strictly unilateral headache, associated with ipsilateral conjunctival injection, lacrimation, nasal congestion, rhinorrhoea, forehead and facial sweating, miosis, ptosis and/or eyelid oedema, and/or with restlessness or agitation. The headache is absolutely sensitive to indomethacin.

Diagnostic criteria:

- A. Unilateral headache fulfilling criteria B-D
- B. Present for >3 months, with exacerbations of moderate or greater intensity
- C. Either or both of the following:
 - 1. at least one of the following symptoms or signs, ipsilateral to the headache:
 - a) conjunctival injection and/or lacrimation
 - b) nasal congestion and/or rhinorrhoea
 - c) eyelid oedema
 - d) forehead and facial sweating
 - e) forehead and facial flushing
 - f) sensation of fullness in the ear
 - g) miosis and/or ptosis
 - 2. a sense of restlessness or agitation, or aggravation of the pain by movement
- D. Responds absolutely to therapeutic doses of indomethacin¹
- E. Not better accounted for by another ICHD-3 diagnosis.

Note:

- 1. In an adult, oral indomethacin should be used initially in a dose of at least 150 mg daily and increased if necessary up to 225 mg daily. The dose by injection is 100–200 mg. Smaller maintenance doses are often employed.

Comments:

Migrainous symptoms such as photophobia and phonophobia are often seen in 3.4 *Hemicrania continua*.

3.4 *Hemicrania continua* has been included under 3. *Trigeminal autonomic cephalgias* in ICHD-3 beta (previously it was under 4. *Other primary headache disorders*) on the basis that the pain is typically unilateral, as are the cranial autonomic symptoms when present. Brain imaging studies show important overlaps between all disorders included here, notably activation in the region of the posterior hypothalamic grey. In addition, the absolute response to indomethacin of 3.4 *Hemicrania continua* is shared with 3.2 *Paroxysmal hemicrania*.

3.4.1 Hemicrania continua, remitting subtype

Description:

Hemicrania continua characterized by pain that is not continuous but is interrupted by remission periods of at least 1 day.

Diagnostic criteria:

- A. Headache fulfilling criteria for 3.4 *Hemicrania continua*, and criterion B below
- B. Headache is not daily or continuous, but interrupted by remission periods of ≥ 1 day without treatment.

Comment:

3.4.1 *Hemicrania continua, remitting subtype* can arise de novo or from 3.4.2 *Hemicrania continua, unremitting subtype*.

3.4.2 Hemicrania continua, unremitting subtype

Description:

Hemicrania continua characterized by continuous pain, without remission periods of at least 1 day, for at least 1 year.

Diagnostic criteria:

- A. Headache fulfilling criteria for 3.4 *Hemicrania continua*, and criterion B below
- B. Headache is daily and continuous for at least 1 year, without remission periods of ≥ 1 day.

Comment:

3.4.2 *Hemicrania continua, unremitting subtype* can arise de novo or evolve from 3.4.1 *Hemicrania continua, remitting subtype*. The majority of patients have the unremitting subtype from onset.

3.5 Probable trigeminal autonomic cephalgia

Description:

Headache attacks which are believed to be a type of 3. *Trigeminal autonomic cephalgia*, but which are missing one of the features required to fulfil all criteria for any of the subtypes coded above, and do not fulfil all criteria for another headache disorder.

Diagnostic criteria:

- A. Headache attacks fulfilling all but one of criteria A-D for 3.1 *Cluster headache*, criteria A-E for 3.2 *Paroxysmal hemicrania*, criteria A-D for 3.3 *Short-lasting unilateral neuralgiform headache attacks* or criteria A-D for 3.4 *Hemicrania continua*
- B. Not fulfilling ICHD-3 criteria for any other headache disorder
- C. Not better accounted for by another ICHD-3 diagnosis.

Comment:

Patients may be coded 3.5.1 *Probable cluster headache*, 3.5.2 *Probable paroxysmal hemicrania*, 3.5.3 *Probable short-lasting unilateral neuralgiform headache attacks* or 3.5.4 *Probable hemicrania continua*. Such patients either have not had a sufficient number of typical attacks (e.g. a first bout of cluster headache), or have had, but fail to fulfil one of the other criteria.

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4. Other primary headache disorders

- 4.1 Primary cough headache
 - 4.1.1 Probable primary cough headache
- 4.2 Primary exercise headache
 - 4.2.1 Probable primary exercise headache
- 4.3 Primary headache associated with sexual activity
 - 4.3.1 Probable primary headache associated with sexual activity
- 4.4 Primary thunderclap headache
- 4.5 Cold-stimulus headache
 - 4.5.1 Headache attributed to external application of a cold stimulus
 - 4.5.2 Headache attributed to ingestion or inhalation of a cold stimulus
 - 4.5.3 Probable cold-stimulus headache
 - 4.5.3.1 Headache probably attributed to external application of a cold stimulus
 - 4.5.3.2 Headache probably attributed to ingestion or inhalation of a cold stimulus
- 4.6 External-pressure headache
 - 4.6.1 External-compression headache
 - 4.6.2 External-traction headache
 - 4.6.3 Probable external-pressure headache
 - 4.6.3.1 Probable external-compression headache
 - 4.6.3.2 Probable external-traction headache
- 4.7 Primary stabbing headache
 - 4.7.1 Probable primary stabbing headache
- 4.8 Nummular headache
 - 4.8.1 Probable nummular headache
- 4.9 Hypnic headache
 - 4.9.1 Probable hypnic headache
- 4.10 New daily persistent headache (NDPH)
 - 4.10.1 Probable new daily persistent headache

General comment

Primary or secondary headache or both?

When a headache with the characteristics of any of the disorders classified here occurs for the first time in close temporal relation to another disorder that is known to cause headache, or fulfils other criteria for causation by that disorder, the new headache is coded as a secondary headache attributed to the causative disorder. When a *pre-existing* headache with the characteristics of any of the disorders classified here becomes *chronic*, or is made significantly worse (usually meaning a two-fold or greater increase in frequency and/or severity), in close temporal relation to such a causative disorder, both the initial headache diagnosis and the secondary headache

diagnosis should be given, provided that there is good evidence that the disorder can cause headache.

Introduction

This chapter includes a number of primary headache disorders that are clinically heterogeneous. Their pathogenesis is still poorly understood, and their treatments are suggested on the basis of anecdotal reports or uncontrolled trials. Headaches with similar characteristics to several of these disorders can be symptomatic of another disorder (i.e. secondary headaches); when they first present, they demand careful evaluation by imaging and/or other appropriate tests.

The onset of some of these headaches, for example, 4.2 *Primary exercise headache*, 4.3 *Primary headache associated with sexual activity* and 4.4 *Primary thunderclap headache*, can be acute, and affected patients are sometimes assessed in emergency departments. Appropriate and full investigation (neuroimaging, in particular) is mandatory in these cases.

This chapter also includes some clinical entities, such as 4.7 *Primary stabbing headache* and 4.9 *Hypnic headache*, that are primary in most cases. In addition, more and more evidence indicates that 4.8 *Nummular headache* is a primary headache disorder, and therefore it has been moved from the Appendix of ICHD-II to this chapter of ICHD-3 beta. Two headache disorders originally in chapter 13 of ICHD-II have also been moved to this chapter: 4.5 *Cold-stimulus headache* and 4.6 *External-pressure headache*. The latter includes 4.6.1 *External-compression headache* and the newly added entity, 4.6.2 *External-traction headache*, because these seem more likely to be primary headache disorders in that they are brought on by physiological (non-damaging) stimuli. In contrast, 3.4 *Hemicrania continua*, originally in this chapter in ICHD-II, is now moved to chapter 3 because evidence indicates that it rightly belongs to 3. *Trigeminal autonomic cephalalgias*.

The headache disorders in this chapter can be grouped into four categories: (1) headaches associated with physical exertion, including 4.1 *Primary cough headache*, 4.2 *Primary exercise headache*, 4.3 *Primary headache attributed to sexual activity* and 4.4 *Primary thunderclap headache*; (2) headaches attributed to direct physical stimuli, including 4.5 *Cold-stimulus headache* and 4.6 *External-pressure headache*; (3) epicranial headaches (i.e. head pain over the scalp), including 4.7 *Primary stabbing headache* and 4.8 *Nummular headache* (as well as A4.11 *Epicrania fugax* in the Appendix); and (4) other miscellaneous primary headache disorders including 4.9 *Hypnic headache* and 4.10 *New daily persistent headache*. Therefore, the coding sequence is rearranged in ICHD-3 beta according to these groupings.

4.1 Primary cough headache

Previously used terms:

Benign cough headache; Valsalva-manœuvre headache.

Description:

Headache precipitated by coughing or other Valsalva (straining) manœuvre, but not by prolonged physical exercise, in the absence of any intracranial disorder.

Diagnostic criteria:

- A. At least two headache episodes fulfilling criteria B-D
- B. Brought on by and occurring only in association with coughing, straining and/or other Valsalva manœuvre
- C. Sudden onset
- D. Lasting between 1 second and 2 hours
- E. Not better accounted for by another ICHD-3 diagnosis.

Comments:

4.1 *Primary cough headache* is a rare condition, accounting for 1% or fewer of all headache patients consulting neurological clinics. However, one report found one-fifth of patients with cough seen in a chest medicine clinic had cough headache.

4.1 *Primary cough headache* arises moments after the cough, reaches its peak almost immediately, and then subsides over several seconds to a few minutes (although some patients experience mild to moderate headache for 2 hours). It is usually bilateral and posterior, and predominantly affects patients older than 40 years of age. There is a significant correlation between the frequency of cough and the severity of the headache. Associated symptoms such as vertigo, nausea and sleep abnormality have been reported by up to two-thirds of patients with 4.1 *Primary cough headache*.

Although indomethacin (50–200 mg/day) is usually effective in treatment of 4.1 *Primary cough headache*, a few symptomatic cases have been reported to respond to this treatment. The syndrome of cough headache is symptomatic in about 40% of cases, and the majority of patients in whom this is so have Arnold-Chiari malformation type I. Other reported causes include CSF hypotension, carotid or vertebrobasilar diseases, middle cranial fossa or posterior fossa tumours, mid-brain cyst, basilar impression, platybasia, subdural haematoma, cerebral aneurysms and reversible cerebral vasoconstriction syndrome. Diagnostic neuroimaging plays an important role in the search for possible intracranial lesions or abnormalities. As subtentorial

tumours accounted for more than 50% of intracranial space-occupying lesions in children, cough headache in paediatric patients should be considered symptomatic until proved otherwise.

4.1.1 Probable primary cough headache

Diagnostic criteria:

- A. Either of the following:
 - 1. a single headache episode fulfilling criteria B-D
 - 2. at least two headache episodes fulfilling criterion B and either of criteria C and D
- B. Brought on by and occurring only in association with coughing, straining and/or other Valsalva manœuvre
- C. Sudden onset
- D. Lasting between 1 second and 2 hours
- E. Not fulfilling ICHD-3 criteria for any other headache disorder
- F. Not better accounted for by another ICHD-3 diagnosis.

4.2 Primary exercise headache

Previously used terms:

Primary exertional headache; benign exertional headache.

Coded elsewhere:

Exercise-induced migraine is coded under 1. *Migraine* according to its subtype.

Description:

Headache precipitated by any form of exercise in the absence of any intracranial disorder.

Diagnostic criteria:

- A. At least two headache episodes fulfilling criteria B and C
- B. Brought on by and occurring only during or after strenuous physical exercise
- C. Lasting <48 hours
- D. Not better accounted for by another ICHD-3 diagnosis.

Comments:

4.2 *Primary exercise headache* occurs particularly in hot weather or at high altitude. Subforms such as 'weight-lifters' headache are recognized but not individually classified. Unlike 4.1 *Primary cough headache*, which can be triggered by short-lasting trains of efforts (i.e. Valsalva-like manœuvres), 4.2 *Primary exercise*

headache is usually precipitated by sustained physically strenuous exercise.

Headache had a pulsating character in most respondents with exercise headache in the Vågå study (less so among adolescent sufferers, of whom almost half had headache durations of less than 5 minutes).

There are reports of prevention in some patients by ingestion of ergotamine tartrate. Indomethacin has been found effective in the majority of the cases.

The pathophysiological mechanisms underlying 4.2 *Primary exercise headache* are unknown. Most investigators believe it is vascular in origin, hypothesizing that venous or arterial distension, secondary to physical exercise, is the pain-inducing mechanism. The recent finding that patients with primary exercise headache have significantly higher prevalence of internal jugular venous valve incompetence (70% compared with 20% of controls), suggests that intracranial venous congestion caused by retrograde jugular venous flow may play a role in the pathophysiology of this disorder.

Symptomatic cases occur. On first occurrence of headache with these characteristics, it is mandatory to exclude subarachnoid haemorrhage, arterial dissection and reversible cerebral vasoconstriction syndrome (RCVS).

4.2.1 Probable primary exercise headache

Diagnostic criteria:

- A. Either of the following:
 1. a single headache episode fulfilling criteria B and C
 2. at least two headache episodes fulfilling criterion B but not criterion C
- B. Brought on by and occurring only during or after strenuous physical exercise
- C. Lasting <48 hours
- D. Not fulfilling ICHD-3 criteria for any other headache disorder
- E. Not better accounted for by another ICHD-3 diagnosis.

4.3 Primary headache associated with sexual activity

Previously used terms:

Benign sex headache; benign vascular sexual headache; coital cephalalgia; coital headache; intercourse headache; orgasmic cephalalgia; orgasmic headache; sexual headache.

Coded elsewhere:

Postural headache occurring after coitus should be coded as 7.2.3 *Headache attributed to spontaneous*

intracranial hypotension because it is most probably a result of CSF leakage.

Description:

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

Diagnostic criteria:

- A. At least two episodes of pain in the head and/or neck fulfilling criteria B-D
- B. Brought on by and occurring only during sexual activity
- C. Either or both of the following:
 1. increasing in intensity with increasing sexual excitement
 2. abrupt explosive intensity just before or with orgasm
- D. Lasting from 1 minute to 24 hours with severe intensity and/or up to 72 hours with mild intensity
- E. Not better accounted for by another ICHD-3 diagnosis.

Comments:

Two subforms (*preorgasmic headache* and *orgasmic headache*) were included in ICHD-I and ICHD-II, but clinical studies since have been unable to distinguish these; therefore, 4.3 *Headache associated with sexual activity* is now regarded as a single entity with variable presentation.

Recent studies have shown that up to 40% of all cases run a chronic course over more than a year.

Some patients experience only one attack of 4.3 *Primary headache attributed to sexual activity* during their lives; they should be diagnosed as 4.3.1 *Probable primary headache associated with sexual activity*. For further research on this headache type, it is recommended to include only patients with at least two attacks.

Epidemiological research has further shown that 4.3 *Primary headache associated with sexual activity* can occur at any sexually active age, is more prevalent in males than in females (ratios range from 1.2:1 to 3:1), occurs independently of the type of sexual activity, is not accompanied by autonomic or vegetative symptoms in most cases, is bilateral in two-thirds and unilateral in one-third of cases and is diffuse or occipitally localized in 80% of cases. Attack frequency of 4.3 *Primary headache attributed to sexual activity* should always be related to the frequency of sexual activity.

4.3 *Primary headache associated with sexual activity* is not associated with disturbance of consciousness,

vomiting or visual, sensory or motor symptoms (whereas symptomatic sexual headache may be). On the first onset of headache with sexual activity, it is mandatory to exclude subarachnoid haemorrhage, arterial dissection and reversible cerebral vasoconstriction syndrome (RCVS). Multiple explosive headaches during sexual activities should be considered as 6.7.3 *Headache attributed to reversible cerebral vasoconstriction syndrome (RCVS)* (qv) until proven otherwise by angiographic studies (including conventional, magnetic resonance or computed tomography angiography) or transcranial Doppler ultrasonography. Of note, vasoconstrictions may not be observed at the early stage of RCVS; therefore, follow-up studies may be needed.

4.3.1 *Probable primary headache associated with sexual activity*

Diagnostic criteria:

- A. Either of the following:
 1. a single headache episode fulfilling criteria B-D
 2. at least two headache episodes fulfilling criterion B and either but not both of criteria C and D
- B. Brought on by and occurring only during sexual activity
- C. Either or both of the following:
 1. increasing in intensity with increasing sexual excitement
 2. abrupt explosive intensity just before or with orgasm
- D. Lasting from 1 minute to 24 hours with severe intensity and/or up to 72 hours with mild intensity
- E. Not fulfilling ICHD-3 criteria for any other headache disorder
- F. Not better accounted for by another ICHD-3 diagnosis.

4.4 **Primary thunderclap headache**

Previously used term:

Benign thunderclap headache.

Coded elsewhere:

4.1 *Primary cough headache*, 4.2 *Primary exercise headache* and 4.3 *Primary headache associated with sexual activity* can all present as thunderclap headache. When such headache is attributed uniquely to one of these triggers, it should be coded accordingly as one of these headache types.

Description:

High-intensity headache of abrupt onset, mimicking that of ruptured cerebral aneurysm, in the absence of any intracranial pathology.

Diagnostic criteria:

- A. Severe head pain fulfilling criteria B and C
- B. Abrupt onset, reaching maximum intensity in <1 minute
- C. Lasting for ≥ 5 minutes
- D. Not better accounted for by another ICHD-3 diagnosis.

Comment:

Evidence that thunderclap headache exists as a primary disorder is poor: the search for an underlying cause should be expedited and exhaustive. Thunderclap headache is frequently associated with serious vascular intracranial disorders, particularly subarachnoid haemorrhage: it is mandatory to exclude this and a range of other such conditions including intracerebral haemorrhage, cerebral venous thrombosis, unruptured vascular malformation (mostly aneurysm), arterial dissection (intra- and extracranial), reversible cerebral vasoconstriction syndrome (RCVS) and pituitary apoplexy. Other organic causes of thunderclap headache are meningitis, colloid cyst of the third ventricle, CSF hypotension and acute sinusitis (particularly with barotrauma). 4.4 *Primary thunderclap headache* should be a diagnosis of last resort, reached only when all organic causes have been demonstrably excluded. This implies normal brain imaging, including the brain vessels, and/or normal CSF. Of note, vasoconstrictions may not be observed in the early stage of RCVS. For this reason, *probable primary thunderclap headache* is not a diagnosis that should be made, even temporarily.

4.5 **Cold-stimulus headache**

Description:

Headache brought on by a cold stimulus applied externally to the head or ingested or inhaled.

4.5.1 *Headache attributed to external application of a cold stimulus*

Description:

Generalized headache following exposure of the unprotected head to a very low environmental temperature.

Diagnostic criteria:

- A. At least two acute headache episodes fulfilling criteria B and C
- B. Brought on by and occurring only during application of an external cold stimulus to the head
- C. Resolving within 30 minutes after removal of the cold stimulus

D. Not better accounted for by another ICHD-3 diagnosis.

Comment:

This headache is a result of external cooling of the head, such as occurs during exposure in very cold weather, diving into cold water or receiving cryotherapy. Some patients develop intense, short-lasting, stabbing headache midfrontally, although the pain can be unilateral and temporal, frontal or retro-orbital.

4.5.2 Headache attributed to ingestion or inhalation of a cold stimulus

Previously used terms:

Ice-cream headache; brain-freeze headache.

Description:

Short-lasting frontal or temporal pain, which may be intense, induced in susceptible people by passage of cold material (solid, liquid or gaseous) over the palate and/or posterior pharyngeal wall.

Diagnostic criteria:

- A. At least two episodes of acute frontal or temporal headache fulfilling criteria B and C
- B. Brought on by and occurring immediately after a cold stimulus to the palate and/or posterior pharyngeal wall from ingestion of cold food or drink or inhalation of cold air
- C. Resolving within 10 minutes after removal of the cold stimulus
- D. Not better accounted for by another ICHD-3 diagnosis.

Comments:

4.5.2 *Headache attributed to ingestion or inhalation of a cold stimulus* is common in the general population, especially among those with 1. *Migraine*. Rapid ingestion of crushed ice slurry is particularly likely to provoke this headache, but eating ice-cream, even slowly, can do so.

Headache is frontal or temporal, and most commonly bilateral (but may be lateralized to the side of usual migraine headache in those who have unilateral headache as part of 1. *Migraine*).

4.5.3 Probable cold-stimulus headache

Diagnostic criteria:

- A. A single headache episode fulfilling criteria B and C

B. Brought on by and occurring only during or immediately after a cold stimulus applied externally to the head or ingested or inhaled

C. Resolving within 10 minutes after removal of the cold stimulus

D. Not fulfilling ICHD-3 criteria for any other headache disorder

E. Not better accounted for by another ICHD-3 diagnosis.

Comment:

Codable subforms are 4.5.3.1 *Headache probably attributed to external application of a cold stimulus* and 4.5.3.2 *Headache probably attributed to ingestion or inhalation of a cold stimulus*.

4.6 External-pressure headache

Description:

Headache resulting from sustained compression of or traction on pericranial soft tissues.

Comment:

4.6 *External-pressure headache* is a primary headache disorder because compression and traction are too subtle to cause damage to the scalp; in other words, they are physiological stimuli.

4.6.1 External-compression headache

Description:

Headache resulting from sustained compression of pericranial soft tissues, for example by a tight band around the head, hat or helmet, or goggles worn during swimming or diving, without damage to the scalp.

Diagnostic criteria:

- A. At least two episodes of headache fulfilling criteria B–D
- B. Brought on by and occurring within 1 hour during sustained external compression of the forehead or scalp
- C. Maximal at the site of external compression
- D. Resolving within 1 hour after external compression is relieved
- E. Not better accounted for by another ICHD-3 diagnosis.

4.6.2 External-traction headache

Previously used term:

Ponytail headache.

Description:

Headache resulting from sustained traction on pericranial soft tissues, without damage to the scalp.

Diagnostic criteria:

- A. At least two episodes of headache fulfilling criteria B–D
- B. Brought on by and occurring only during sustained external traction on the scalp
- C. Maximal at the traction site
- D. Resolving within 1 hour after traction is relieved
- E. Not better accounted for by another ICHD-3 diagnosis.

Comment:

The duration of headache varies with the severity and duration of the external traction. Although headache is maximal at the site of traction, it often extends to other areas of the head.

4.6.3 Probable external-pressure headache

Diagnostic criteria:

- A. Either of the following:
 - 1. a single episode of headache fulfilling criteria B–D
 - 2. at least two episodes of headache fulfilling criterion B and either but not both of criteria C and D
- B. Brought on by and occurring only during sustained external compression of or traction on the forehead and/or scalp
- C. Maximal at the compression or traction site
- D. Resolving within 1 hour after compression or traction is relieved
- E. Not fulfilling ICHD-3 criteria for any other headache disorder
- F. Not better accounted for by another ICHD-3 diagnosis.

Comment:

Codable subforms are 4.6.3.1 *Probable external-compression headache* and 4.6.3.2 *Probable external-traction headache*.

4.7 Primary stabbing headache

Previously used terms:

Ice-pick pains; jabs and jolts; needle-in-the-eye syndrome; ophthalmodynia periodica; sharp short-lived head pain.

Description:

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

Diagnostic criteria:

- A. Head pain occurring spontaneously as a single stab or series of stabs and fulfilling criteria B–D
- B. Each stab lasts for up to a few seconds
- C. Stabs recur with irregular frequency, from one to many per day
- D. No cranial autonomic symptoms
- E. Not better accounted for by another ICHD-3 diagnosis.

Comments:

Studies show 80% of stabs last 3 seconds or less; rarely, stabs last for 10–120 seconds. Attack frequency is generally low, with one or a few per day. In rare cases, stabs occur repetitively over days, and there has been one description of *status* lasting 1 week.

4.7 *Primary stabbing headache* involves extratrigeminal regions in 70% of cases. It may move from one area to another, in either the same or the opposite hemicranium: in only one-third of patients it has a fixed location. When stabs are strictly localized to one area, structural changes at this site and in the distribution of the affected cranial nerve must be excluded.

A few patients have accompanying symptoms, but not including cranial autonomic symptoms. The latter help to differentiate 4.7 *Primary stabbing headache* from 3.3 *Short-lasting unilateral neuralgiform headache attacks*.

4.7 *Primary stabbing headache* is more commonly experienced by people with 1. *Migraine*, in which cases stabs tend to be localized to the site habitually affected by migraine headaches.

4.7.1 Probable primary stabbing headache

Diagnostic criteria:

- A. Head pain occurring spontaneously as a single stab or series of stabs and fulfilling two only of criteria B–D
- B. Each stab lasts for up to a few seconds
- C. Stabs recur with irregular frequency, from one to many per day
- D. No cranial autonomic symptoms
- E. Not fulfilling ICHD-3 criteria for any other headache disorder
- F. Not better accounted for by another ICHD-3 diagnosis.

4.8 Nummular headache

Previously used term:

Coin-shaped headache.

Description:

Pain of highly variable duration, but often chronic, in a small circumscribed area of the scalp in the absence of any underlying structural lesion.

Diagnostic criteria:

- A. Continuous or intermittent head pain fulfilling criterion B
- B. Felt exclusively in an area of the scalp, with all of the following four characteristics:
 1. sharply contoured
 2. fixed in size and shape
 3. round or elliptical
 4. 1–6 cm in diameter
- C. Not better accounted for by another ICHD-3 diagnosis.

Comments:

The painful area may be localized in any part of the scalp, but is usually in the parietal region. Rarely, 4.8 *Nummular headache* is bi- or multifocal, each symptomatic area retaining all the characteristics of nummular headache. Pain intensity is generally mild to moderate, but occasionally severe. Superimposed on the background pain, spontaneous or triggered exacerbations may occur. Duration is highly variable: in up to 75% of published cases, the disorder has been chronic (present for longer than 3 months), but cases have also been described with durations of seconds, minutes, hours or days.

The affected area commonly shows variable combinations of hypaesthesia, dysaesthesia, paraesthesia, allodynia and/or tenderness.

Other causes, in particular structural and dermatological lesions, must be excluded by history, physical examination and appropriate investigations.

4.8.1 Probable nummular headache

Diagnostic criteria:

- A. Continuous or intermittent head pain fulfilling criterion B
- B. Felt exclusively in an area of the scalp, with three only of the following four characteristics:
 1. sharply contoured
 2. fixed in size and shape
 3. round or elliptical

4. 1–6 cm in diameter

- C. Not fulfilling ICHD-3 criteria for any other headache disorder
- D. Not better accounted for by another ICHD-3 diagnosis.

4.9 Hypnic headache

Previously used terms:

Hypnic headache syndrome; ‘alarm clock’ headache.

Description:

Frequently recurring headache attacks developing only during sleep, causing wakening and lasting for up to 4 hours, without characteristic associated symptoms and not attributed to other pathology.

Diagnostic criteria:

- A. Recurrent headache attacks fulfilling criteria B–E
- B. Developing only during sleep, and causing wakening
- C. Occurring on ≥ 10 days per month for > 3 months
- D. Lasting ≥ 15 minutes and for up to 4 hours after wakening
- E. No cranial autonomic symptoms or restlessness
- F. Not better accounted for by another ICHD-3 diagnosis.

Comments:

4.9 *Hypnic headache* usually begins after age 50 years, but may occur in younger people. The pain is usually mild to moderate, but severe pain is reported by one-fifth of patients. Pain is bilateral in about two-thirds of cases. Attacks usually last from 15 to 180 minutes, but longer durations have been described. Most cases are persistent, with daily or near daily headaches, but an episodic subform (on less than 15 days per month) may occur. Although it was thought that the features of 4.9 *Hypnic headache* were generally tension-type-like, recent studies found that patients could present with migraine-like features and some patients had nausea during attacks.

Onset of 4.9 *Hypnic headache* is probably not related to sleep stage. A recent MRI study showed grey matter volume reduction in the hypothalamus in patients with 4.9 *Hypnic headache*.

Lithium, caffeine, melatonin and indomethacin have been effective treatments in several reported cases. Distinction from one of the subtypes of 3. *Trigeminal autonomic cephalalgias*, especially 3.1 *Cluster headache*, is necessary for effective management.

Other possible causes of headache developing during and causing wakening from sleep should be ruled out,

with particular attention given to sleep apnoea, nocturnal hypertension, hypoglycaemia and medication overuse; intracranial disorders must also be excluded. However, the presence of sleep apnoea syndrome does not necessarily exclude the diagnosis of 4.9 *Hypnic headache*.

4.9.1 Probable hypnic headache

Diagnostic criteria:

- A. Recurrent headache attacks fulfilling criterion B and two only of criteria C-E
- B. Developing only during sleep, and causing wakening
- C. Occurring on ≥ 10 days per month for >3 months
- D. Lasting ≥ 15 minutes and for up to 4 hours after waking
- E. No cranial autonomic symptoms or restlessness
- F. Not fulfilling ICHD-3 criteria for any other headache disorder
- G. Not better accounted for by another ICHD-3 diagnosis.

4.10 New daily persistent headache (NDPH)

Previously used terms:

Chronic headache with acute onset; de novo chronic headache.

Description:

Persistent headache, daily from its onset, which is clearly remembered. The pain lacks characteristic features, and may be migraine-like or tension-type-like, or have elements of both.

Diagnostic criteria:

- A. Persistent headache fulfilling criteria B and C
- B. Distinct and clearly remembered onset, with pain becoming continuous and unremitting within 24 hours
- C. Present for >3 months
- D. Not better accounted for by another ICHD-3 diagnosis.

Comments:

4.10 *New daily persistent headache (NDPH)* is unique in that headache is daily from onset, and very soon unremitting, typically occurring in individuals without a prior headache history. Patients with this disorder invariably recall and can accurately describe such an

onset; if they cannot do so, another diagnosis should be made. Nevertheless, patients with prior headache (1. *Migraine* or 2. *Tension-type headache*) are not excluded from this diagnosis, but they should not describe increasing headache frequency prior to its onset. Similarly, patients with prior headache should not describe exacerbation followed by medication overuse.

4.10 *New daily persistent headache (NDPH)* may have features suggestive of either 1. *Migraine* or 2. *Tension-type headache*. Even though criteria for 1.3 *Chronic migraine* and/or 2.3 *Chronic tension-type headache* may also be fulfilled, the default diagnosis is 4.10 *New daily persistent headache (NDPH)* whenever the criteria for this disorder are met. In contrast, when the criteria for both 4.10 *New daily persistent headache (NDPH)* and 3.4 *Hemicrania continua* are met, then the latter is the default diagnosis.

Abortive drug use may exceed the limits defined as causative of 8.2 *Medication-overuse headache (qv)*. In such cases, the diagnosis of 4.10 *New daily persistent headache* cannot be made unless the onset of daily headache clearly predates the medication overuse. When this is so, both diagnoses, 4.10 *New daily persistent headache (NDPH)* and 8.2 *Medication-overuse headache*, should be given.

In all cases, other secondary headaches such as 7.1 *Headache attributed to increased cerebrospinal fluid pressure*, 7.2 *Headache attributed to low cerebrospinal fluid pressure* and 5.1 *Acute headache attributed to traumatic injury to the head* should be ruled out by appropriate investigations.

4.10 *New daily persistent headache (NDPH)* has two subforms: a self-limiting subform that typically resolves within several months without therapy, and a refractory form that is resistant to aggressive treatment regimens. These are not separately coded.

4.10.1 Probable new daily persistent headache

Diagnostic criteria:

- A. Persistent headache fulfilling criteria B and C
- B. Distinct and clearly remembered onset, with pain becoming continuous and unremitting within 24 hours
- C. Present for <3 months
- D. Not fulfilling ICHD-3 criteria for any other headache disorder
- E. Not better accounted for by another ICHD-3 diagnosis.

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Part two

The secondary headaches

5. Headache attributed to trauma or injury to the head and/or neck
6. Headache attributed to cranial or cervical vascular disorder
7. Headache attributed to non-vascular intracranial disorder
8. Headache attributed to a substance or its withdrawal
9. Headache attributed to infection
10. Headache attributed to disorder of homoeostasis
11. Headache or facial pain attributed to disorder of cranium, neck, eyes, ears, nose, sinuses, teeth, mouth or other facial or cranial structure
12. Headache attributed to psychiatric disorder

Introduction to the secondary headaches

When a patient has headache for the first time, or a new headache type, and at the same time develops a brain tumour, it is straightforward to conclude that headache is secondary to the tumour. Such patients shall be given only one headache diagnosis – 7.4 *Headache attributed to intracranial neoplasia* (or one of its subforms) – even when the headache phenomenologically is migraine, tension-type headache or cluster headache. In other words, a de novo headache occurring with another disorder recognized to be capable of causing it is always diagnosed as secondary.

The situation is different when the patient has previously had a type of primary headache that becomes worse in close temporal relation to the occurrence of another disorder. Three possible explanations for this worsening exist: that it is coincidental; that it is an aggravation of the primary headache, causally related to the other disorder; that it represents a new headache, again causally related to the other disorder. The rules for attribution developed in ICHD-II allowed one or two diagnoses in such circumstances, but relied on judgement. They have been modified in ICHD-3 beta in order to be less open to interpretation.

When a new headache occurs for the first time in close temporal relation to another disorder that is known to cause headache, or fulfils other criteria for causation by that disorder, the new headache is coded as a secondary headache attributed to the causative disorder. This remains true even when the headache has the characteristics of a primary headache (migraine, tension-type headache, cluster headache or one of the other trigeminal autonomic cephalalgias).

When a *pre-existing* primary headache becomes *chronic* in close temporal relation to such a causative disorder, both the primary and the secondary diagnoses should be given. When a *pre-existing* primary headache is made significantly worse (usually meaning a two-fold or greater increase in frequency and/or severity) in close temporal relation to such a causative disorder, both the primary and the secondary headache diagnoses should be given, provided that there is good evidence that the disorder can cause headache.

ICHD-II standardized the format of the diagnostic criteria for secondary headaches, but this was not without problems. A revision was necessary, and this revision is adopted in ICHD-3 beta:

General diagnostic criteria for secondary headaches:

A. Any headache fulfilling criterion C

- B. Another disorder scientifically documented to be able to cause headache has been diagnosed¹
- C. Evidence of causation demonstrated by at least two of the following²:
1. headache has developed in temporal relation to the onset of the presumed causative disorder
 2. one or both of the following:
 - a) headache has significantly worsened in parallel with worsening of the presumed causative disorder
 - b) headache has significantly improved in parallel with improvement of the presumed causative disorder
 3. headache has characteristics typical for the causative disorder³
 4. other evidence exists of causation⁴
- D. Not better accounted for by another ICHD-3 diagnosis.

Notes:

1. As headache is extremely prevalent, it can occur simultaneously with another disorder by chance and without a causal relation. Therefore, a secondary headache can be definitely diagnosed only when solid evidence exists from published scientific studies that the disorder specified in criterion B is capable of causing headache. Scientific evidence can come from large clinical studies observing close temporal relationships between the disorder and headache outcomes after treatment of the disorder, or from smaller studies using advanced scanning methods, blood tests or other paraclinical tests, even if these are not readily available to the diagnosing physician who will use these criteria. In other words, study methods that are not useful in routine use of the diagnostic criteria may nonetheless be useful for establishing general causal relationships as the basis of criterion B. Throughout ICHD-3 beta, however, diagnostic criteria restrict themselves to information reasonably available to the diagnosing physician in a typical clinical situation.
2. The general criteria require two separate evidential features to be present, and allow up to four types of evidence, as set out. Not all of these four types are appropriate for all disorders, and not all four need form part of the specific criteria for a particular secondary headache when this is so. There are a few secondary headaches for which evidence of causation depends very heavily on onset in temporal relation to the presumed cause. Examples are the subtypes of 7.2 *Headache attributed to low cerebrospinal fluid pressure*, which are usually orthostatic but not invariably, so that this characteristic

- cannot be relied on as a diagnostic criterion. In such cases, criterion D is of particular importance.
3. An example is very sudden (thunderclap) onset of headache in 6.2.2 *Headache attributed to non-traumatic subarachnoid haemorrhage (SAH)*. The characteristics (if any) must be specified for each secondary headache.
 4. This is to be specified (if appropriate) for each secondary headache. One example of this kind of

evidence is accordance between the site of the headache and the location of a presumed causative disorder. Another is variations in parallel between headache features (such as intensity) and markers of activity of the presumed causative disorder (e.g. changes on neuroimaging, or in other laboratory measures [such as erythrocyte sedimentation rate in 6.4.1 *Headache attributed to giant cell arteritis (GCA)*]).

5. Headache attributed to trauma or injury to the head and/or neck

- 5.1 Acute headache attributed to traumatic injury to the head
 - 5.1.1 Acute headache attributed to moderate or severe traumatic injury to the head
 - 5.1.2 Acute headache attributed to mild traumatic injury to the head
- 5.2 Persistent headache attributed to traumatic injury to the head
 - 5.2.1 Persistent headache attributed to moderate or severe traumatic injury to the head
 - 5.2.2 Persistent headache attributed to mild traumatic injury to the head
- 5.3 Acute headache attributed to whiplash
- 5.4 Persistent headache attributed to whiplash
- 5.5 Acute headache attributed to craniotomy
- 5.6 Persistent headache attributed to craniotomy

General comment

Primary or secondary headache or both?

When a headache occurs for the first time in close temporal relation to trauma or injury to the head and/or neck, it is coded as a secondary headache attributed to the trauma or injury. This remains true when the new headache has the characteristics of any of the primary headache disorders classified in Part one of ICHD-3 beta. When a *pre-existing* headache with the characteristics of a primary headache disorder becomes *chronic*, or is made significantly worse (usually meaning a two-fold or greater increase in frequency and/or severity), in close temporal relation to such trauma or injury, both the initial headache diagnosis and a diagnosis of 5. *Headache attributed to trauma or injury to the head and/or neck* (or one of its subtypes) should be given.

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The subtypes of 5. *Headache attributed to trauma or injury to the head and/or neck* are among the most common secondary headache disorders. During the first 3 months from onset they are considered *acute*; if they continue beyond that period they are designated *persistent*. This time period is consistent with ICHD-II diagnostic criteria, although the term *persistent* has been adopted in place of *chronic*.

There are no specific headache features known to distinguish the subtypes of 5. *Headache attributed to trauma or injury to the head and/or neck* from other headache types; most often these resemble tension-type headache or migraine. Consequently their diagnosis is largely dependent on the close temporal relation between the trauma

or injury and headache onset. Consistently with those of ICHD-II, the diagnostic criteria of ICHD-3 beta for all subtypes require that headache must be reported to have developed within 7 days of trauma or injury, or within 7 days after regaining consciousness and/or the ability to sense and report pain when these have been lost following trauma or injury. Although this 7-day interval is somewhat arbitrary, and although some experts argue that headache may develop after a longer interval in a minority of patients, there is not enough evidence at this time to change this requirement.

Headache may occur as an isolated symptom following trauma or injury or as one of a constellation of symptoms, commonly including dizziness, fatigue, reduced ability to concentrate, psychomotor slowing, mild memory problems, insomnia, anxiety, personality changes and irritability. When several of these symptoms follow head injury, the patient may be considered to have a post-concussion syndrome.

The pathogenesis of 5. *Headache attributed to trauma or injury to the head and/or neck* is often unclear. Numerous factors that may contribute to its development include, but are not limited to, axonal injury, alterations in cerebral metabolism, alterations in cerebral haemodynamics, underlying genetic predisposition, psychopathology and a patient's expectations of developing headache after head injury. Recent research, using advanced neuroimaging modalities, suggests a potential for detecting brain structural abnormalities following minor trauma that are not detectable through conventional diagnostic tests. Post-traumatic sleep disturbances, mood disturbances and psychosocial stressors can plausibly influence the development and perpetuation of headache. The overuse of abortive headache medications may contribute to the persistence of headache after head injury through the development of 8.2 *Medication-overuse headache*. Clinicians must consider this possibility whenever such headache persists beyond the initial post-trauma phase.

Risk factors for the development of 5. *Headache attributed to trauma or injury to the head and/or neck* may include a previous history of headache, less severe injury, female gender and the presence of comorbid psychiatric disorders. The association between repetitive head trauma and the development of headache should be investigated further. The degree to which a patient's expectation of headache following head injury and litigation regarding such headache promote its development and persistence is still widely debated. The majority of evidence suggests that malingering is a factor in only a small minority of patients. Those with pending litigation and those without are similar regarding headache characteristics, cognitive test results, treatment responses and improvement in symptoms over time. Furthermore, symptom resolution does not

typically occur following legal settlements. In Lithuania, for example, a country in which there is little expectation of developing headache after head injury, and a lack of insurance against personal injury, rates of 5.2 *Persistent headache attributed to traumatic injury to the head* are low.

5. *Headache attributed to trauma or injury to the head and/or neck* is also reported in children, although less often than in adults. The clinical presentations of the subtypes are similar in children and adults, and the diagnostic criteria in children are the same.

5.1 Acute headache attributed to traumatic injury to the head

Coded elsewhere:

Trauma as a result of acceleration/deceleration movements of the head, with flexion/extension of the neck, is classified as whiplash. Acute headache attributed to such trauma is coded as 5.3 *Acute headache attributed to whiplash*. Acute headache attributed to surgical craniotomy performed for reasons other than traumatic head injury is coded as 5.5 *Acute headache attributed to craniotomy*.

Description:

Headache of less than 3 months' duration caused by traumatic injury to the head.

Diagnostic criteria:

- A. Any headache fulfilling criteria C and D
- B. Traumatic injury to the head¹ has occurred
- C. Headache is reported to have developed within 7 days after one of the following:
 - 1. the injury to the head
 - 2. regaining of consciousness following the injury to the head
 - 3. discontinuation of medication(s) that impair ability to sense or report headache following the injury to the head
- D. Either of the following:
 - 1. headache has resolved within 3 months after the injury to the head
 - 2. headache has not yet resolved but 3 months have not yet passed since the injury to the head
- E. Not better accounted for by another ICHD-3 diagnosis.

Note:

- 1. Traumatic injury to the head is defined as a structural or functional injury resulting from the action of external forces on the head. These include

striking the head with or the head striking an object, penetration of the head by a foreign body, forces generated from blasts or explosions, and other forces yet to be defined.

Comment:

The stipulation that headache must be reported to have developed within 7 days is somewhat arbitrary (see Introduction). Compared with longer intervals, a 7-day interval yields diagnostic criteria with higher specificity for 5.1 *Acute headache attributed to traumatic injury to the head* (i.e. stronger evidence of causation) but a correlative loss of sensitivity. Further research is needed into whether or not a different interval might be more appropriate. In the meantime, Appendix criteria for A5.1.1.1 *Delayed-onset acute headache attributed to moderate or severe traumatic injury to the head* and A5.1.2.1 *Delayed-onset acute headache attributed to mild traumatic injury to the head* (qv) may be used when the interval between injury and headache onset is greater than 7 days.

5.1.1 Acute headache attributed to moderate or severe traumatic injury to the head

Diagnostic criteria:

- A. Headache fulfilling criteria for 5.1 *Acute headache attributed to traumatic injury to the head*
- B. Injury to the head associated with at least one of the following:
 - 1. loss of consciousness for >30 minutes
 - 2. Glasgow Coma Scale (GCS) score <13
 - 3. post-traumatic amnesia¹ lasting >24 hours
 - 4. alteration in level of awareness for >24 hours
 - 5. imaging evidence of a traumatic head injury such as intracranial haemorrhage and/or brain contusion.

Note:

- 1. The duration of post-traumatic amnesia is defined as the time between head injury and recovery of memory of current events and those occurring in the last 24 hours.

5.1.2 Acute post-traumatic headache attributed to mild traumatic injury to the head

Diagnostic criteria:

- A. Headache fulfilling criteria for 5.1 *Acute headache attributed to traumatic injury to the head*

- B. Injury to the head fulfilling both of the following:
1. associated with none of the following:
 - a) loss of consciousness for >30 minutes
 - b) Glasgow Coma Scale (GCS) score <13
 - c) post-traumatic amnesia lasting >24 hours
 - d) altered level of awareness for >24 hours
 - e) imaging evidence of a traumatic head injury such as intracranial haemorrhage and/or brain contusion
 2. associated, immediately following the head injury, with one or more of the following symptoms and/or signs:
 - a) transient confusion, disorientation or impaired consciousness
 - b) loss of memory for events immediately before or after the head injury
 - c) two or more other symptoms suggestive of mild traumatic brain injury: nausea, vomiting, visual disturbances, dizziness and/or vertigo, impaired memory and/or concentration.

Comment:

The diagnostic criteria for mild traumatic injury to the head and for moderate or severe traumatic injury to the head allow for substantial variability in the severity of head injury classified in each category. This has led some experts to suggest inclusion of additional categories: *headache attributed to very mild traumatic injury to the head* and *headache attributed to very severe traumatic injury to the head*. Although there is insufficient evidence for adding these categories at present, future studies should investigate the utility of doing so.

5.2 Persistent headache attributed to traumatic injury to the head

Coded elsewhere:

Trauma as a result of acceleration/deceleration movements of the head, with flexion/extension of the neck, is classified as whiplash. Persistent headache attributed to such trauma is coded as 5.4 *Persistent headache attributed to whiplash*. Persistent headache attributed to surgical craniotomy performed for reasons other than traumatic head injury is coded as 5.6 *Persistent headache attributed to craniotomy*.

Description:

Headache of greater than 3 months' duration caused by traumatic injury to the head.

Diagnostic criteria:

- A. Any headache fulfilling criteria C and D
- B. Traumatic injury to the head¹ has occurred
- C. Headache is reported to have developed within 7 days after one of the following:
 1. the injury to the head
 2. regaining of consciousness following the injury to the head
 3. discontinuation of medication(s) that impair ability to sense or report headache following the injury to the head
- D. Headache persists for >3 months after the injury to the head
- E. Not better accounted for by another ICHD-3 diagnosis.

Note:

1. Traumatic injury to the head is defined as a structural or functional injury resulting from the action of external forces on the head. These include striking the head with or the head striking an object, penetration of the head by a foreign body, forces generated from blasts or explosions, and other forces yet to be defined.

Comments:

The stipulation that headache must be reported to have developed within 7 days is somewhat arbitrary (see Introduction). Compared with longer intervals, a 7-day interval yields diagnostic criteria with higher specificity for 5.2 *Persistent headache attributed to traumatic injury to the head* (i.e. stronger evidence of causation) but a correlative loss of sensitivity. Further research is needed into whether or not a different interval might be more appropriate. In the meantime, Appendix criteria for A5.2.1.1 *Delayed-onset persistent headache attributed to moderate or severe traumatic injury to the head* and A5.2.2.1 *Delayed-onset persistent headache attributed to mild traumatic injury to the head* (qv) may be used when the interval between injury and headache onset is greater than 7 days.

To be consistent with ICHD-II diagnostic criteria for *chronic post-traumatic headache* and with the time interval used in the diagnoses of other secondary headache disorders, 3 months is the time interval after which headache attributed to head injury is considered persistent. Further research is needed to investigate whether shorter or longer intervals may be more appropriately adopted.

5.2.1 Persistent headache attributed to moderate or severe traumatic injury to the head

Diagnostic criteria:

- A. Headache fulfilling criteria for 5.2 Persistent headache attributed to traumatic injury to the head
- B. Injury to the head associated with at least one of the following:
 - 1. loss of consciousness for >30 minutes
 - 2. Glasgow Coma Scale (GCS) score <13
 - 3. post-traumatic amnesia¹ lasting >24 hours
 - 4. alteration in level of awareness for >24 hours
 - 5. imaging evidence of a traumatic head injury such as intracranial haemorrhage and/or brain contusion.

Note:

- 1. The duration of post-traumatic amnesia is defined as the time between head injury and recovery of memory of current events and of those occurring in the last 24 hours.

Comment:

When headache following head injury becomes persistent, the possibility of 8.2 Medication-overuse headache needs to be considered.

5.2.2 Persistent headache attributed to mild traumatic injury to the head

Diagnostic criteria:

- A. Headache fulfilling criteria for 5.2 Persistent headache attributed to traumatic injury to the head
- B. Head injury fulfilling both of the following:
 - 1. associated with none of the following:
 - a) loss of consciousness for >30 minutes
 - b) Glasgow Coma Scale (GCS) score <13
 - c) post-traumatic amnesia lasting >24 hours
 - d) altered level of awareness for >24 hours
 - e) imaging evidence of a traumatic head injury such as intracranial haemorrhage and/or brain contusion
 - 2. associated, immediately following the head injury, with one or more of the following symptoms and/or signs:
 - a) transient confusion, disorientation or impaired consciousness
 - b) loss of memory for events immediately before or after the head injury

- c) two or more other symptoms suggestive of mild traumatic brain injury: nausea, vomiting, visual disturbances, dizziness and/or vertigo, impaired memory and/or concentration.

Comment:

When headache following head injury becomes persistent, the possibility of 8.2 Medication-overuse headache needs to be considered.

5.3 Acute headache attributed to whiplash¹

Description:

Headache of less than 3 months' duration caused by whiplash.

Diagnostic criteria:

- A. Any headache fulfilling criteria C and D
- B. Whiplash¹, associated at the time with neck pain and/or headache, has occurred
- C. Headache has developed within 7 days after the whiplash
- D. Either of the following:
 - 1. headache has resolved within 3 months after the whiplash
 - 2. headache has not yet resolved but 3 months have not yet passed since the whiplash
- E. Not better accounted for by another ICHD-3 diagnosis.

Note:

- 1. Whiplash is defined as sudden and inadequately restrained acceleration/deceleration movements of the head with flexion/extension of the neck. Whiplash may occur after either high or low impact forces.

Comments:

Whiplash most commonly occurs in the context of a motor vehicle accident.

5.3 Acute headache attributed to whiplash may occur as an isolated symptom or with a constellation of other symptoms that relate to the neck, as well as somatic extracervical, neurosensory, behavioural, cognitive and/or mood symptoms. Whiplash itself may be classified according to the severity of the clinical presentation, using a scheme such as that presented by the Quebec Task Force on Whiplash-Associated Disorders.

5.4 Persistent headache attributed to whiplash

Description:

Headache of greater than 3 months' duration caused by whiplash.

Diagnostic criteria:

- A. Any headache fulfilling criteria C and D
- B. Whiplash¹, associated at the time with neck pain and/or headache, has occurred
- C. Headache has developed within 7 days after the whiplash
- D. Headache persists for >3 months after the whiplash
- E. Not better accounted for by another ICHD-3 diagnosis.

Note:

1. Whiplash is defined as sudden and inadequately restrained acceleration/deceleration movements of the head with flexion/extension of the neck. Whiplash may occur after either high or low impact forces.

Comment:

When post-whiplash headache becomes persistent, the possibility of 8.2 *Medication-overuse headache* needs to be considered.

5.5 Acute headache attributed to craniotomy

Description:

Headache of less than 3 months' duration caused by surgical craniotomy.

Diagnostic criteria:

- A. Any headache fulfilling criteria C and D
- B. Surgical craniotomy¹ has been performed
- C. Headache is reported to have developed within 7 days after one of the following:
 1. the craniotomy
 2. regaining of consciousness following the craniotomy
 3. discontinuation of medication(s) that impair ability to sense or report headache following the craniotomy
- D. Either of the following:
 1. headache has resolved within 3 months after the craniotomy

2. headache has not yet resolved but 3 months have not yet passed since the craniotomy

- E. Not better accounted for by another ICHD-3 diagnosis.

Note:

1. When the craniotomy was performed following head injury, code as 5.1.1 *Acute headache attributed to moderate or severe traumatic injury to the head*.

Comments:

5.5 *Acute headache attributed to craniotomy* may occur in more than two-thirds of patients following surgical craniotomy. In the majority of cases, it resolves within the acute post-operative period. It is more common after surgery of the skull base compared with other locations. Although the pain of 5.5 *Acute headache attributed to craniotomy* is often felt maximally at the site of craniotomy, it may be more diffuse and resemble tension-type headache or migraine.

Exclusion of other secondary headache disorders that may occur following craniotomy is necessary prior to assigning the diagnosis of 5.5 *Acute headache attributed to craniotomy*. Although there are numerous potential aetiologies of headache following craniotomy, considerations should include cervicogenic headache (as a result of positioning during surgery), headache from cerebrospinal fluid leak, infections, hydrocephalus and intracranial haemorrhage.

5.6 Persistent headache attributed to craniotomy

Description:

Headache of greater than 3 months' duration caused by surgical craniotomy.

Diagnostic criteria:

- A. Any headache fulfilling criteria C and D
- B. Surgical craniotomy¹ has been performed
- C. Headache is reported to have developed within 7 days after one of the following:
 1. the craniotomy
 2. regaining of consciousness following the craniotomy
 3. discontinuation of medication(s) that impairs ability to sense or report headache following the craniotomy

- D. Headache persists for >3 months after the craniotomy
- E. Not better accounted for by another ICHD-3 diagnosis.

Note:

1. When the craniotomy was performed following head injury, code as 5.2.1 *Persistent headache attributed to moderate or severe traumatic injury to the head.*

Comments:

About a quarter of patients who develop 5.5 *Acute headache attributed to craniotomy* go on to experience 5.6 *Persistent headache attributed to craniotomy.*

When headache following craniotomy becomes persistent, the possibility of 8.2 *Medication-overuse headache* needs to be considered.

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6. Headache attributed to cranial or cervical vascular disorder

- 6.1 Headache attributed to ischaemic stroke or transient ischaemic attack
 - 6.1.1 Headache attributed to ischaemic stroke (cerebral infarction)
 - 6.1.2 Headache attributed to transient ischaemic attack (TIA)
- 6.2 Headache attributed to non-traumatic intracranial haemorrhage
 - 6.2.1 Headache attributed to non-traumatic intracerebral haemorrhage
 - 6.2.2 Headache attributed to non-traumatic subarachnoid haemorrhage (SAH)
 - 6.2.3 Headache attributed to non-traumatic acute subdural haemorrhage (ASDH)
- 6.3 Headache attributed to unruptured vascular malformation
 - 6.3.1 Headache attributed to unruptured saccular aneurysm
 - 6.3.2 Headache attributed to arteriovenous malformation (AVM)
 - 6.3.3 Headache attributed to dural arteriovenous fistula (DAVF)
 - 6.3.4 Headache attributed to cavernous angioma
 - 6.3.5 Headache attributed to encephalotrigeminal or leptomeningeal angiomatosis (Sturge Weber syndrome)
- 6.4 Headache attributed to arteritis
 - 6.4.1 Headache attributed to giant cell arteritis (GCA)
 - 6.4.2 Headache attributed to primary angiitis of the central nervous system (PACNS)
 - 6.4.3 Headache attributed to secondary angiitis of the central nervous system (SACNS)
- 6.5 Headache attributed to cervical carotid or vertebral artery disorder
 - 6.5.1 Headache or facial or neck pain attributed to cervical carotid or vertebral artery dissection
 - 6.5.2 Post-endarterectomy headache
 - 6.5.3 Headache attributed to carotid or vertebral angioplasty
- 6.6 Headache attributed to cerebral venous thrombosis (CVT)
- 6.7 Headache attributed to other acute intracranial arterial disorder
 - 6.7.1 Headache attributed to an intracranial endovascular procedure
 - 6.7.2 Angiography headache
 - 6.7.3 Headache attributed to reversible cerebral vasoconstriction syndrome (RCVS)
 - 6.7.3.1 Headache probably attributed to reversible cerebral vasoconstriction syndrome (RCVS)

6.7.4 Headache attributed to intracranial arterial dissection

- 6.8 Headache attributed to genetic vasculopathy
 - 6.8.1 Cerebral Autosomal Dominant Arteriopathy with Subcortical Infarcts and Leukoencephalopathy (CADASIL)
 - 6.8.2 Mitochondrial Encephalopathy, Lactic Acidosis and Stroke-like episodes (MELAS)
 - 6.8.3 Headache attributed to another genetic vasculopathy
- 6.9 Headache attributed to pituitary apoplexy

General comment

Primary or secondary headache or both?

When a headache occurs for the first time in close temporal relation to a cranial or cervical vascular disorder, it is coded as a secondary headache attributed to that disorder. This remains true when the new headache has the characteristics of any of the primary headache disorders classified in Part one of ICHD-3 beta. When a *pre-existing* headache with the characteristics of a primary headache disorder becomes *chronic*, or is made significantly worse (usually meaning a two-fold or greater increase in frequency and/or severity), in close temporal relation to a cranial or cervical vascular disorder, both the initial headache diagnosis and a diagnosis of 6. *Headache attributed to cranial or cervical vascular disorder* (or one of its subtypes) should be given, provided that there is good evidence that the disorder can cause headache.

Introduction

The diagnosis of headache and its causal link is easy in most of the vascular conditions listed below because the headache presents both acutely and with neurological signs and because it often remits rapidly. The close temporal relationship between the headache and these neurological signs is therefore crucial to establishing causation.

In many of these conditions, such as ischaemic or haemorrhagic stroke, headache is overshadowed by focal signs and/or disorders of consciousness. In others, such as subarachnoid haemorrhage, headache is usually the prominent symptom. In a number of other conditions that can induce both headache and stroke, such as dissections, cerebral venous thrombosis, giant cell arteritis and central nervous system angiitis, headache is often an initial warning symptom. It is therefore crucial to recognize the association of headache with these disorders in order to diagnose correctly the underlying vascular disease and start appropriate treatment as early as possible, thus preventing potentially devastating neurological consequences.

All of these conditions can occur in patients who have previously suffered a primary headache of any type. A clue that points to an underlying vascular condition is the onset, usually sudden, of a *new* headache, so far unknown to the patient. Whenever this occurs, vascular conditions should urgently be looked for.

For headache attributed to any of the vascular disorders listed here, the diagnostic criteria include whenever possible:

- A. Headache fulfilling criterion C
- B. A cranial or cervical vascular disorder known to be able to cause headache has been demonstrated
- C. Evidence of causation demonstrated by at least two of the following:
 1. headache has developed in temporal relation to the onset of the cranial or cervical vascular disorder
 2. either or both of the following:
 - a) headache has significantly worsened in parallel with worsening of the cranial or cervical vascular disorder
 - b) headache has significantly improved in parallel with improvement of the cranial or cervical vascular disorder
 3. headache has characteristics typical for the cranial or cervical vascular disorder
 4. other evidence exists of causation
- D. Not better accounted for by another ICHD-3 diagnosis.

6.1 Headache attributed to ischaemic stroke or transient ischaemic attack

6.1.1 Headache attributed to ischaemic stroke (cerebral infarction)

Description:

Headache caused by ischaemic stroke, usually with acute onset and associated with focal neurological signs. It has a self-limited course, and is very rarely the presenting or a prominent feature of ischaemic stroke.

Diagnostic criteria:

- A. Any new headache fulfilling criterion C
- B. Acute ischaemic stroke has been diagnosed
- C. Evidence of causation demonstrated by at least one of the following:
 1. headache has developed in very close temporal relation to other symptoms and/or clinical signs of ischaemic stroke, or has led to the diagnosis of ischaemic stroke

2. headache has significantly improved in parallel with stabilization or improvement of other symptoms or clinical or radiological signs of ischaemic stroke
- D. Not better accounted for by another ICHD-3 diagnosis.

Comments:

6.1.1 *Headache attributed to ischaemic stroke (cerebral infarction)* is accompanied by focal neurological signs and/or alterations in consciousness, which in most cases allows easy differentiation from the primary headaches. It is usually of moderate intensity, and has no specific characteristics. It can be bilateral or unilateral ipsilateral to the stroke. Rarely, an acute ischaemic stroke, notably a cerebellar infarction, can present with an isolated sudden (even thunderclap) headache.

Headache accompanies ischaemic stroke in up to one-third of cases; it is more frequent in basilar- than in carotid-territory strokes. It is of little practical value in establishing stroke aetiology except that headache is very rarely associated with lacunar infarcts but extremely common in acute arterial wall disorders such as dissection or reversible cerebral vasoconstriction syndrome. In these latter conditions, headache may be directly caused by the arterial wall lesions and may precede ischaemic stroke.

6.1.2 Headache attributed to transient ischaemic attack (TIA)

Description:

Headache caused by a transient ischaemic attack (TIA) and accompanied by the sudden-onset transient focal signs of a TIA. It lasts less than 24 hours.

Diagnostic criteria:

- A. Any new headache fulfilling criterion C
- B. Transient ischaemic attack (TIA) has been diagnosed
- C. Evidence of causation demonstrated by both of the following:
 1. headache has developed simultaneously with other symptoms and/or clinical signs of TIA
 2. headache resolves within 24 hours
- D. Not better accounted for by another ICHD-3 diagnosis.

Comments:

A TIA is a transient episode of neurological dysfunction caused by focal brain or retinal ischaemia without clinical, imaging or other evidence of acute cerebral or retinal infarction. Symptoms of a TIA typically, but not

invariably, last less than 1 hour. Although more common with basilar- than carotid-territory TIA, headache is very rarely a prominent symptom of TIA.

The differential diagnosis between 6.1.2 *Headache attributed to transient ischaemic attack* and an attack of 1.2 *Migraine with aura* may be particularly difficult. The mode of onset is crucial: the focal deficit is typically sudden in a TIA and more frequently progressive in a migrainous aura. Furthermore, positive phenomena (e.g. scintillating scotoma) are far more common in migrainous aura than in TIA, whereas negative phenomena are more usual in TIA.

The coincidence of an otherwise typical TIA and a severe headache should prompt the search for some arterial disorders that can directly induce severe headache (arterial dissection, among others).

6.2 Headache attributed to non-traumatic intracranial haemorrhage

Coded elsewhere:

Headache attributed to traumatic intracerebral and/or subarachnoid haemorrhage or to traumatic intracerebral, subdural or epidural haematoma is coded as 5.1.1 *Acute headache attributed to moderate or severe traumatic injury to the head* or 5.2.1 *Persistent headache attributed to moderate or severe traumatic injury to the head*.

Description:

Headache caused by non-traumatic intracranial haemorrhage, with, generally, sudden (even thunderclap) onset. Depending on the type of haemorrhage, it may be isolated or associated with focal neurological deficits.

6.2.1 Headache attributed to non-traumatic intracerebral haemorrhage

Description:

Headache caused by non-traumatic intracerebral haemorrhage, usually with acute onset and associated with focal neurological signs. It can, rarely, be the presenting and prominent feature of non-traumatic intracerebral haemorrhage.

Diagnostic criteria:

- A. Any new headache fulfilling criterion C
- B. Intracerebral haemorrhage (ICH)¹ in the absence of head trauma has been diagnosed
- C. Evidence of causation demonstrated by at least two of the following:
 1. headache has developed in close temporal relation to other symptoms and/or clinical signs of ICH, or has led to the diagnosis of ICH

2. headache has significantly improved in parallel with stabilization or improvement of other symptoms or clinical or radiological signs of ICH
3. headache has at least one of the following three characteristics:
 - a) sudden or thunderclap onset
 - b) maximal on the day of its onset
 - c) localized in accordance with the site of the haemorrhage
- D. Not better accounted for by another ICHD-3 diagnosis.

Note:

1. Through usage, the term *intracerebral* is taken in this context to include *intracerebellar*.

Comments:

6.2.1 *Headache attributed to non-traumatic intracerebral haemorrhage* is more often a result of associated subarachnoid blood and local compression than intracranial hypertension. Headache is more usual and more severe in haemorrhagic than in ischaemic stroke, and 6.2.1 *Headache attributed to non-traumatic intracerebral haemorrhage* can occasionally present as thunderclap headache.

The headache is usually overshadowed by focal deficits or coma, but it can be the prominent early feature of some intracerebral haemorrhages, notably cerebellar haemorrhage, which may require emergency surgical decompression.

6.2.2 Headache attributed to non-traumatic subarachnoid haemorrhage (SAH)

Description:

Headache caused by non-traumatic subarachnoid haemorrhage (SAH), typically severe and sudden, peaking in seconds (thunderclap headache) or minutes. It can be the sole symptom of SAH.

Diagnostic criteria:

- A. Any new headache fulfilling criterion C
- B. Subarachnoid haemorrhage (SAH) in the absence of head trauma has been diagnosed
- C. Evidence of causation demonstrated by at least two of the following:
 1. headache has developed in close temporal relation to other symptoms and/or clinical signs of SAH, or has led to the diagnosis of SAH

- 2. headache has significantly improved in parallel with stabilization or improvement of other symptoms or clinical or radiological signs of SAH
- 3. headache has sudden or thunderclap onset
- D. Not better accounted for by another ICHD-3 diagnosis.

Comments:

SAH is the most common cause of persistent, intense and incapacitating headache of abrupt onset (thunderclap headache), and is a serious condition (mortality rate is 40–50% and 10–20% of patients die before arriving at hospital; 50% of survivors are left disabled).

6.2.2 *Headache attributed to non-traumatic subarachnoid haemorrhage (SAH)* may nonetheless be moderate and without any associated signs. The abrupt onset is the key feature. Any patient with headache of abrupt onset or thunderclap headache should be evaluated for SAH. Diagnosis is confirmed by non-contrast-enhanced CT scan, which has a sensitivity of 98% in the first 12 hours after onset (dropping to 93% at 24 hours and 50% at 7 days). If CT results are non-diagnostic, a lumbar puncture is essential. Xanthochromia is present in 100% of cases with aneurysmal SAH when cerebrospinal fluid (CSF) is collected between 12 hours and 2 weeks after the onset of symptoms and analysed spectrophotometrically. MRI is not indicated as an initial diagnostic test for SAH; however, FLAIR and gradient-echo T2-weighted images may be useful when the CT is normal and the CSF abnormal.

Initial misdiagnosis occurs in one-quarter to one-half of patients; the most common specific misdiagnosis is migraine, but often, in these cases, no cause is identified. The most common reasons for misdiagnosis are failure to obtain appropriate neuroimaging, or misinterpretation, or failure to perform a lumbar puncture in cases where this is required. Delayed diagnosis often has a catastrophic outcome.

SAH is a neurointerventional emergency. After diagnosis of SAH, the next urgent step is to identify a ruptured aneurysm (80% of cases of spontaneous SAH result from ruptured saccular aneurysms).

6.2.3 *Headache attributed to non-traumatic acute subdural haemorrhage (ASDH)*

Description:

Headache caused by non-traumatic acute subdural haemorrhage (ASDH), typically severe and sudden, peaking in seconds (thunderclap headache) or minutes. It is usually accompanied or rapidly followed by focal signs and decrease in consciousness.

Diagnostic criteria:

- A. Any new headache fulfilling criterion C
- B. Acute subdural haemorrhage (ASDH) in the absence of head trauma has been diagnosed
- C. Evidence of causation demonstrated by at least two of the following:
 - 1. headache has developed in very close temporal relation to other symptoms and/or clinical signs of ASDH, or has led to the diagnosis of ASDH
 - 2. either or both of the following:
 - a) headache has significantly worsened in parallel with worsening of ASDH
 - b) headache has significantly improved in parallel with improvement of other symptoms or clinical or radiological signs of ASDH
 - 3. headache has either or both of the following two characteristics:
 - a) sudden or thunderclap onset
 - b) localized in accordance with the site of the haemorrhage
- D. Not better accounted for by another ICHD-3 diagnosis.

Comments:

Most cases of ASDH occur after head trauma and should be coded accordingly. Non-traumatic ASDH without other intracranial haemorrhage ('pure ASDH') is rare and represents a life-threatening condition. It is a neurosurgical emergency.

The bleeding may be from arterial or venous origin. Reported causes include 'spontaneous' cortical artery rupture, aneurysm rupture, arteriovenous malformations and dural arteriovenous fistulae, tumours or metastasis, coagulopathies, moyo-moya disease, cerebral venous thrombosis and intracranial hypotension. Isolated cases or small series have mostly been reported by neurosurgeons. Headache is described in 25–100% of cases depending on the series and the underlying cause. Isolated headache can be the presenting sign; but usually it is associated or followed by a rapid neurological deterioration.

6.3 *Headache attributed to unruptured vascular malformation*

Coded elsewhere:

Headache attributed to ruptured vascular malformation is coded as 6.2.1 *Headache attributed to intracerebral haemorrhage* or 6.2.2 *Headache attributed to subarachnoid haemorrhage* or, rarely, 6.2.3 *Headache attributed to non traumatic acute subdural haemorrhage*.

Description:

Headache secondary to an unruptured intracranial vascular malformation (occurring without haemorrhage). Depending on the type of malformation, the headache may have a chronic course with recurrent attacks mimicking episodic primary headaches, or an acute and self-limited course.

6.3.1 Headache attributed to unruptured saccular aneurysm

Diagnostic criteria:

- A. Any new headache fulfilling criterion C
- B. An unruptured saccular aneurysm has been diagnosed
- C. Evidence of causation demonstrated by at least two of the following:
 1. headache has developed in close temporal relation to other symptoms and/or clinical signs of unruptured saccular aneurysm, or has led to its diagnosis
 2. either or both of the following:
 - a) headache has significantly worsened in parallel with other symptoms or clinical or radiological signs of growth of the saccular aneurysm
 - b) headache has resolved after treatment of the saccular aneurysm
 3. either or both of the following:
 - a) headache has sudden or thunderclap onset
 - b) headache is associated with a painful IIIrd nerve palsy
- D. Not better accounted for by another ICHD-3 diagnosis, and intracranial haemorrhage and reversible cerebral vasoconstriction syndrome have been excluded by appropriate investigations.

Comments:

Headache is reported by approximately one-fifth of patients with unruptured cerebral aneurysm, but whether this association is incidental or causal is an unresolved issue.

6.3.1 *Headache attributed to unruptured saccular aneurysm* usually has no specific features. On the other hand, a new-onset headache can reveal a symptomatic but unruptured saccular aneurysm. A classic variety is acute IIIrd nerve palsy with retro-orbital pain and a dilated pupil, indicating an aneurysm of the posterior communicating cerebral artery or termination of the carotid artery. Such painful IIIrd nerve palsy is an emergency, signalling impending rupture or progressive enlargement of the arterial malformation.

Several retrospective studies have shown that about half of patients with an aneurysmal subarachnoid

haemorrhage reported the occurrence of a sudden and severe headache within the 4 weeks prior to the diagnosis of aneurysmal rupture. Setting aside the possibility of memory biases, this suggests these headaches are a result of sudden enlargement of the arterial malformation ('sentinel headache') or to mild subarachnoid haemorrhage that is not diagnosed as such ('warning leak'). Evidence for the existence of sentinel headaches is poor. Moreover, the term *warning leak* should not be used, because a leak indicates a subarachnoid haemorrhage. Given that at least one in three patients with aneurysmal subarachnoid haemorrhage is initially misdiagnosed, and given the risks of re-bleeding, patients with sudden severe headaches should undergo complete investigation, including cerebral imaging, CSF study and cerebral angiography (MRA or CT angiography).

6.3.2 Headache attributed to arteriovenous malformation (AVM)

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. An arteriovenous malformation (AVM) has been diagnosed
- C. Evidence of causation demonstrated by at least two of the following:
 1. headache has developed in close temporal relation to other symptoms and/or clinical signs of AVM, or has led to the discovery of an AVM
 2. either or both of the following:
 - a) headache has significantly worsened in parallel with worsening of the AVM
 - b) headache has significantly improved in parallel with improvement of the AVM
 3. headache is localized to the site of the AVM
- D. Not better accounted for by another ICHD-3 diagnosis, and intracranial haemorrhage has been excluded by appropriate investigations.

Comments:

Cases have been reported highlighting the association of AVM with a variety of headaches such as 3.1 *Cluster headache*, 3.2.2 *Chronic paroxysmal hemicrania* and 3.3.1 *Short-lasting unilateral neuralgiform headache with conjunctival injection and tearing (SUNCT)*, but these cases had atypical features. There is no good evidence of a relationship between AVM and these primary headache disorders.

1.2 *Migraine with aura* has been reported in up to 58% of women with AVM. A strong argument in favour of a causal relationship is the overwhelming correlation between the side of the headache, or of the aura, and the side of the AVM. There is thus a

strong suggestion that AVM can cause attacks of migraine with aura (symptomatic migraine). Yet in a large AVM series, presenting features frequently included epilepsy or focal deficits with or without haemorrhage and, much more rarely, migraine-like symptoms.

6.3.3 Headache attributed to dural arteriovenous fistula (DAVF)

Diagnostic criteria:

- A. Any new headache fulfilling criterion C
- B. A dural arteriovenous fistula (DAVF) has been diagnosed
- C. Evidence of causation demonstrated by at least two of the following:
 1. headache has developed in close temporal relation to other symptoms and/or clinical signs of DAVF, or has led to the diagnosis of DAVF
 2. either or both of the following:
 - a) headache has significantly worsened in parallel with other symptoms or clinical or radiological signs of growth of the DAVF
 - b) headache has significantly improved after treatment of the DAVF
 3. at least one of the following:
 - a) headache is accompanied by pulsatile tinnitus
 - b) headache is accompanied by ophthalmoplegia
 - c) headache is both progressive and worse in the morning and/or during coughing and/or bending over
 4. headache is localized to the site of the DAVF
- D. Not better accounted for by another ICHD-3 diagnosis, and intracerebral haemorrhage and cerebral venous thrombosis have been excluded by appropriate investigations.

Comment:

Studies devoted to 6.3.3 *Headache attributed to dural arteriovenous fistula* are lacking. A painful pulsatile tinnitus can be a presenting symptom, as well as headache with features of intracranial hypertension as a result of decrease in venous outflow and sometimes to sinus thrombosis. Carotidocavernous fistulae may present as painful ophthalmoplegia.

6.3.4 Headache attributed to cavernous angioma

Coded elsewhere:

Headache attributed to cerebral haemorrhage or seizure secondary to cavernous angioma is coded as 6.2.1 *Headache attributed to intracerebral haemorrhage* or 7.6 *Headache attributed to epileptic seizure*.

Diagnostic criteria:

- A. Any new headache fulfilling criterion C
- B. A cavernous angioma has been diagnosed
- C. Evidence of causation demonstrated by at least two of the following:
 1. headache has developed in close temporal relation to other symptoms and/or clinical signs of cavernous angioma
 2. either or both of the following:
 - a) headache has significantly worsened in parallel with other symptoms or clinical or radiological signs of growth of the cavernous angioma
 - b) headache has significantly improved or resolved after removal of the cavernous angioma
 3. headache is localized to the site of the cavernous angioma
- D. Not better accounted for by another ICHD-3 diagnosis, and intracerebral haemorrhage has been excluded by appropriate investigations.

Comments:

Cavernous angiomas are increasingly recognized on MRI. Isolated case reports suggest that some cavernous angiomas may trigger SUNCT-like or migraine-like attacks. However, there is still no good study devoted to 6.3.4 *Headache attributed to cavernous angioma*.

In a series of 126 symptomatic patients with cavernous angiomas and *KRIT 1* mutations, only 4% reported headache as a presenting symptom. On the contrary, headache is commonly reported as a consequence of cerebral haemorrhage or of seizures, which are the two main manifestations of cavernous angiomas; such headache should be coded to either of these accordingly.

6.3.5 Headache attributed to encephalotrigeminal or leptomeningeal angiomatosis (Sturge Weber syndrome)

Coded elsewhere:

Headache attributed to seizure secondary to Sturge Weber syndrome is coded as 7.6 *Headache attributed to epileptic seizure*.

Diagnostic criteria:

- A. Any new headache fulfilling criterion C
- B. Facial angioma is present, together with neuroimaging evidence of meningeal angioma ipsilateral to it
- C. Evidence of causation demonstrated by at least two of the following:
 1. headache has developed in close temporal relation to other symptoms and/or clinical signs

and/or imaging evidence of the meningeal angioma

2. headache has significantly worsened in parallel with other symptoms or clinical or radiological signs of growth of the meningeal angioma
3. headache is migraine-like, either bilateral or localized to the site of the angioma, and associated with aura contralateral to the site of the angioma

D. Not better accounted for by another ICHD-3 diagnosis.

Comment:

6.3.5 *Headache attributed to encephalotrigeminal or leptomeningeal angiomatosis (Sturge Weber syndrome)* is poorly documented. More than 90% of cases of Sturge Weber syndrome have seizures, and half report post-seizure headaches, which should be coded accordingly. Isolated reports suggest that encephalotrigeminal or leptomeningeal angiomatosis may be a cause of symptomatic migraine, particularly of attacks with prolonged auras (possibly related to chronic oligoemia).

6.4 Headache attributed to arteritis

Description:

Headache caused by and symptomatic of an inflammation of cervical, cranial and/or brain arteries. Headache may be the sole symptom of arteritis.

Diagnostic criteria:

- A. Any new headache fulfilling criterion C
- B. Arteritis has been diagnosed
- C. Evidence of causation demonstrated by either or both of the following:
 1. headache has developed in close temporal relation to other symptoms and/or clinical signs of onset of arteritis, or has led to the diagnosis of arteritis
 2. either or both of the following:
 - a) headache has significantly worsened in parallel with worsening of arteritis
 - b) headache has significantly improved in parallel with improvement of arteritis
- D. Not better accounted for by another ICHD-3 diagnosis.

6.4.1 *Headache attributed to giant cell arteritis (GCA)*

Previously used term:

Headache attributed to temporal arteritis.

Description:

Headache caused by and symptomatic of giant cell arteritis. Headache may be the sole symptom of giant cell arteritis, a disease most conspicuously associated with headache, which is a result of inflammation of cranial arteries, especially branches of the external carotid artery. The features of the headache are variable.

Diagnostic criteria:

- A. Any new headache fulfilling criterion C
- B. Giant cell arteritis (GCA) has been diagnosed
- C. Evidence of causation demonstrated by at least two of the following:
 1. headache has developed in close temporal relation to other symptoms and/or clinical or biological signs of onset of GCA, or has led to the diagnosis of GCA
 2. either or both of the following:
 - a) headache has significantly worsened in parallel with worsening of GCA
 - b) headache has significantly improved or resolved within 3 days of high-dose steroid treatment
 3. headache is associated with scalp tenderness and/or jaw claudication
- D. Not better accounted for by another ICHD-3 diagnosis.

Comments:

Of all arteritides and collagen vascular diseases, giant cell arteritis is the disease most conspicuously associated with headache, which is a result of inflammation of cranial arteries, especially branches of the external carotid artery. The variability in the features of 6.4.1 *Headache attributed to giant cell arteritis* and in the other symptoms of GCA (polymyalgia rheumatica, jaw claudication) are such that any recent persisting headache in a patient over 60 years of age should suggest GCA and lead to appropriate investigations.

Recent repeated attacks of amaurosis fugax associated with headache are strongly suggestive of GCA and should prompt urgent investigations. The major risk is of blindness as a result of anterior ischaemic optic neuropathy, which can be prevented by immediate steroid treatment; the time interval between visual loss in one eye and in the other is usually less than 1 week. Patients with GCA are also at risk of cerebral ischaemic events and of dementia.

Histological diagnosis can be difficult, because the temporal artery may appear uninvolved in some areas (skip lesions), pointing to the necessity of serial sectioning.

6.4.2 Headache attributed to primary angiitis of the central nervous system (PACNS)

Previously used term:

Headache attributed to isolated CNS angiitis or granulomatous CNS angiitis.

Description:

Headache caused by and symptomatic of primary angiitis of the central nervous system. Headache is the dominant symptom of this disorder, but lacks specific features.

Diagnostic criteria:

- A. Any new headache fulfilling criterion C
- B. Primary angiitis of the central nervous system (PACNS) has been diagnosed
- C. Evidence of causation demonstrated by either or both of the following:
 - 1. headache has developed in close temporal relation to other symptoms and/or clinical signs of onset of PACNS, or has led to the diagnosis of PACNS
 - 2. either or both of the following:
 - a) headache has significantly worsened in parallel with worsening of PACNS
 - b) headache has significantly improved in parallel with improvement in PACNS resulting from steroid and/or immunosuppressive treatment
- D. Not better accounted for by another ICHD-3 diagnosis, and CNS infection, CNS neoplasia and reversible cerebral vasoconstriction syndrome have been excluded by appropriate investigations.

Comments:

Headache is the dominant symptom in CNS angiitis (either primary or secondary). It is present in 50–80% of cases according to the diagnostic methods used, angiography and histology, respectively. Nevertheless it has no specific features and is therefore of little diagnostic value until other signs are present, such as focal deficits, seizures, altered cognition or disorders of consciousness. However, the absence of both headache and CSF pleocytosis makes CNS angiitis unlikely.

The pathogenesis of 6.4.2 *Headache attributed to primary angiitis of the central nervous system* is multifactorial: inflammation, stroke (ischaemic or haemorrhagic), raised intracranial pressure and/or subarachnoid haemorrhage.

The effect of treatment is far less dramatic than in 6.4.1 *Headache attributed to giant cell arteritis*.

Histologically proven primary CNS angiitis remains a serious and not infrequently lethal condition.

6.4.3 Headache attributed to secondary angiitis of the central nervous system (SACNS)

Description:

Headache caused by and symptomatic of secondary angiitis of the central nervous system. Headache is the dominant symptom of this disorder, but lacks specific features.

Diagnostic criteria:

- A. Any new headache fulfilling criterion C
- B. Secondary angiitis of the central nervous system (SACNS) (angiitis of the CNS in the presence of systemic angiitis) has been diagnosed
- C. Evidence of causation demonstrated by either or both of the following:
 - 1. headache has developed in close temporal relation to other symptoms and/or clinical signs of onset of SACNS
 - 2. either or both of the following:
 - a) headache has significantly worsened in parallel with worsening of the systemic angiitis
 - b) headache has significantly improved in parallel with improvement in the systemic angiitis resulting from steroid and/or immunosuppressive treatment
- D. Not better accounted for by another ICHD-3 diagnosis.

Comments:

Headache is the dominant symptom in CNS angiitis (either primary or secondary). It is present in 50–80% of cases according to the diagnostic methods used, angiography and histology, respectively. Nevertheless it has no specific features and is therefore of little diagnostic value until other signs are present such as focal deficits, seizures, altered cognition or disorders of consciousness. However, the absence of both headache and CSF pleocytosis makes CNS angiitis unlikely.

The difficulty here is two-fold: 1) diagnosing CNS angiitis in a patient known to have one of the many conditions that can cause angiitis; 2) finding the underlying condition (inflammatory, infectious, malignant, toxic) in a patient presenting with CNS angiitis.

The pathogenesis of 6.4.3 *Headache attributed to secondary angiitis of the central nervous system* is multifactorial: inflammation, stroke (ischaemic or haemorrhagic), raised intracranial pressure and/or subarachnoid haemorrhage.

6.5 Headache attributed to cervical carotid or vertebral artery disorder

Description:

Headache and/or pain in the face and/or neck caused by non-inflammatory lesions affecting the cervical carotid and/or vertebral arteries. The pain generally has a sudden (even thunderclap) onset. It can remain isolated or be a warning symptom preceding the focal deficits of ischaemic stroke.

Diagnostic criteria:

- A. Any new headache and/or facial or neck pain fulfilling criterion C
- B. A cervical artery lesion has been demonstrated, or a surgical or radiological intervention has been performed on a cervical artery
- C. Evidence of causation demonstrated by at least two of the following:
 1. pain has developed in close temporal relation to other local signs of cervical artery disorder, or has led to the diagnosis of cervical artery disorder
 2. either or both of the following:
 - a) pain has significantly worsened in parallel with other signs of the cervical artery lesion
 - b) pain has significantly improved or resolved within 1 month of its onset
 3. pain is unilateral and ipsilateral to the affected cervical artery
- D. Not better accounted for by another ICHD-3 diagnosis.

6.5.1 Headache or facial or neck pain attributed to cervical arterial dissection

Description:

Headache and/or pain in the face and/or neck caused by dissection of a cervical carotid or vertebral artery. The pain is usually ipsilateral to the dissected vessel and generally has a sudden (even thunderclap) onset. It can remain isolated or be a warning symptom preceding ischaemic stroke.

Diagnostic criteria:

- A. Any new headache and/or facial or neck pain fulfilling criterion C
- B. Cervical carotid or vertebral dissection has been diagnosed
- C. Evidence of causation demonstrated by at least two of the following:
 1. pain has developed in close temporal relation to other local signs of cervical artery dissection, or

has led to the diagnosis of cervical artery dissection

2. either or both of the following:
 - a) pain has significantly worsened in parallel with other signs of the cervical artery lesion
 - b) pain has significantly improved or resolved within 1 month of its onset
3. either or both of the following:
 - a) pain is severe and continuous for days or longer
 - b) pain precedes signs of acute retinal and/or cerebral ischaemia
4. pain is unilateral and ipsilateral to the affected cervical artery
- D. Not better accounted for by another ICHD-3 diagnosis.

Comments:

Headache with or without neck pain can be the only manifestation of cervical artery dissection. It is by far the most frequent symptom (55–100% of cases), and the most frequent inaugural symptom (33–86% of cases), of this disorder.

6.5.1 *Headache or facial or neck pain attributed to cervical arterial dissection* is usually unilateral (ipsilateral to the dissected artery), severe and persistent (for a mean of 4 days). However, it has no constant specific pattern and it can sometimes be very misleading, mimicking other headaches such as 1. *Migraine*, 3.1 *Cluster headache* or 4.4 *Primary thunderclap headache*. Associated signs (of cerebral or retinal ischaemia and local signs) are common: a painful Horner's syndrome, painful tinnitus of sudden onset or painful XIIth nerve palsy are highly suggestive of carotid artery dissection.

Cervical artery dissection may be associated with intracranial artery dissection, which is a potential cause of subarachnoid haemorrhage. 6.7.4 *Headache attributed to intracranial arterial dissection* may be present in addition to 6.5.1 *Headache or facial or neck pain attributed to cervical arterial dissection*.

6.5.1 *Headache or facial or neck pain attributed to cervical arterial dissection* usually precedes the onset of ischaemic signs, and therefore requires early diagnosis and treatment. Diagnosis is based on cervical MRI with fat suppression, Duplex scanning, MRA and/or CTA and, in doubtful cases, conventional angiography. Several of these investigations are commonly needed as any of them can be normal. There have been no randomized trials of treatment, but there is a consensus in favour of heparin followed by warfarin for 3–6 months according to the quality of the arterial recovery.

6.5.2 *Post-endarterectomy headache*

Description:

Headache caused by the surgical procedure of carotid endarterectomy. Pain can also involve the neck and face. It can remain isolated or be a warning symptom preceding the focal deficits of (mostly haemorrhagic) stroke.

Diagnostic criteria:

- A. Any new headache fulfilling criterion C
- B. Carotid endarterectomy has been performed
- C. Evidence of causation demonstrated by at least two of the following:
 1. headache develops within 1 week of carotid endarterectomy
 2. headache resolves within 1 month after carotid endarterectomy
 3. headache is unilateral, on the side of the carotid endarterectomy, and has one of the following three characteristics:
 - a) diffuse mild pain
 - b) cluster headache-like pain occurring once or twice a day in attacks lasting 2–3 hours
 - c) pulsating severe pain
- D. Not better accounted for by another ICHD-3 diagnosis, and arterial dissection has been excluded by appropriate investigations.

Comment:

Three subforms of 6.5.2 *Post-endarterectomy headache* have been described (but are not separately coded). The most frequent (up to 60% of cases) is a diffuse, mild isolated headache occurring in the first few days after surgery. It is a benign self-limiting condition. The second subform (reported in up to 38% of cases) is a unilateral cluster headache-like pain with attacks, lasting 2–3 hours, occurring once or twice a day. It resolves in about 2 weeks. The third subform is part of the rare hyperperfusion syndrome, with a unilateral pulsating and severe pain occurring 3 days after surgery. It often precedes a rise in blood pressure and the onset of seizures or neurological deficits on or about the seventh day. Urgent treatment is required, as these symptoms can herald cerebral haemorrhage.

6.5.3 *Headache attributed to carotid or vertebral angioplasty*

Description:

Headache caused by the surgical procedure of cervical angioplasty. Pain can also involve the neck and face. It can remain isolated or be a warning symptom preceding the focal deficits of (mostly haemorrhagic) stroke.

Diagnostic criteria:

- A. Any new headache, fulfilling criterion C
- B. Carotid or vertebral angioplasty has been performed
- C. Evidence of causation demonstrated by all of the following:
 1. headache has developed within 1 week of the angioplasty
 2. headache has resolved within 1 month after the angioplasty
 3. headache is on the same side as the angioplasty
- D. Not better accounted for by another ICHD-3 diagnosis, and arterial dissection has been excluded by appropriate investigations.

Comments:

Percutaneous transluminal angioplasty (PTA) and stenting versus surgery are presently undergoing randomized trials. Data on headache are still scarce, and headache is not mentioned in large series of carotid PTA. In a small series of 53 patients, cervical pain occurred during balloon inflation in one-half of patients and head pain in one-third, mostly disappearing within seconds of balloon deflation.

6.5.3 *Headache attributed to carotid or vertebral angioplasty* has been reported as part of the rare hyperperfusion syndrome.

6.6 *Headache attributed to cerebral venous thrombosis (CVT)*

Description:

Headache caused by cerebral venous thrombosis. It has no specific characteristics: it is most often diffuse, progressive and severe, but can be unilateral and sudden (even thunderclap), or mild, and sometimes is migraine-like.

Diagnostic criteria:

- A. Any new headache, fulfilling criterion C
- B. Cerebral venous thrombosis (CVT) has been diagnosed
- C. Evidence of causation demonstrated by both of the following:
 1. headache has developed in close temporal relation to other symptoms and/or clinical signs of CVT, or has led to the discovery of CVT
 2. either or both of the following:
 - a) headache has significantly worsened in parallel with clinical or radiological signs of extension of the CVT

- b) headache has significantly improved or resolved after improvement of the CVT
- D. Not better accounted for by another ICHD-3 diagnosis.

Comments:

Headache is by far the most frequent symptom of cerebral venous thrombosis (CVT), present in 80–90% of cases, and also the most frequent inaugural symptom. 6.6 *Headache attributed to cerebral venous thrombosis* has no specific characteristics, but most often is diffuse, progressive and severe, and associated with other signs of intracranial hypertension. It can also be unilateral and sudden, and sometimes very misleading, mimicking 1. *Migraine*, 4.4 *Primary thunderclap headache*, 7.2 *Headache attributed to low cerebrospinal fluid pressure* or 6.2.2 *Headache attributed to non-traumatic subarachnoid haemorrhage* (CVT can be a cause of SAH).

Headache can be the only manifestation of CVT but, in over 90% of cases, it is associated with focal signs (neurological deficits or seizures) and/or signs of intracranial hypertension, subacute encephalopathy or cavernous sinus syndrome.

Given the absence of specific characteristics of 6.6 *Headache attributed to cerebral venous thrombosis*, any recent persisting headache should raise suspicion, particularly in the presence of an underlying prothrombotic condition. Diagnosis is based on neuroimaging (MRI with T2*-weighted images plus MRA, or CT scan plus CT angiography, and intra-arterial angiography in doubtful cases). Treatment should be started as early as possible and includes symptomatic treatment, heparin followed by at least 6 months of oral anticoagulation and, whenever indicated, treatment of the underlying cause.

6.7 Headache attributed to other acute intracranial arterial disorder

6.7.1 Headache attributed to an intracranial endovascular procedure

Description:

Unilateral headache caused directly by an intracranial endovascular procedure, ipsilateral to the procedure and lasting less than 24 hours.

Diagnostic criteria:

- A. Any new headache fulfilling criterion C
- B. Intracranial angioplasty or embolization has been performed

- C. Evidence of causation demonstrated by all of the following:
1. headache has developed within seconds of the procedure
 2. headache has resolved within 24 hours after the end of the procedure
 3. headache is severe, unilateral and ipsilateral to the procedure
- D. Not better accounted for by another ICHD-3 diagnosis, and arterial dissection has been excluded by appropriate investigations.

Comment:

A very specific subform of 6.7.1 *Headache attributed to an intracranial endovascular procedure* has been reported after balloon inflation or embolization of an AVM or aneurysm. It is a severe pain of abrupt onset, localized in a specific area according to the artery involved, occurring within a few seconds of the procedure and disappearing rapidly.

6.7.2 Angiography headache

Description:

Headache caused directly by cerebral angiography, either diffuse, burning and severe or, in people with migraine, with the clinical features of a migraine attack.

Diagnostic criteria:

- A. Any new headache fulfilling criterion C
- B. Intra-arterial carotid or vertebral angiography has been performed
- C. Evidence of causation demonstrated by at least two of the following:
1. headache has developed during the angiography
 2. headache has resolved within 72 hours after the end of the angiography
 3. headache is either of the following:
 - a) diffuse, burning and severe
 - b) in a patient with migraine, having the features of 1.1 *Migraine without aura* or 1.2 *Migraine with aura*
- D. Not better accounted for by another ICHD-3 diagnosis.

Comments:

Intracarotid or intravertebral injection of contrast induces a diffuse severe headache with a burning sensation, which resolves spontaneously. It can also trigger a migraine attack in a person who has 1. *Migraine*. In the latter case, the patient should have both diagnoses: the

appropriate subtype of 1. *Migraine* and 6.7.2 *Angiography headache*.

Contrast angiography is contraindicated in patients affected by any subform of 1.2.3 *Hemiplegic migraine* because it may trigger a life-threatening attack, with prolonged hemiplegia and coma.

6.7.3 *Headache attributed to reversible cerebral vasoconstriction syndrome (RCVS)*

Description:

Headache caused by reversible cerebral vasoconstriction syndrome, typically thunderclap headache recurring over 1–2 weeks, often triggered by sexual activity, exertion, Valsalva manœuvres and/or emotion. Headache can remain the sole symptom of RCVS.

Diagnostic criteria:

- A. Any new headache fulfilling criterion C
- B. Reversible cerebral vasoconstriction syndrome (RCVS) has been diagnosed
- C. Evidence of causation demonstrated by at least one of the following:
 - 1. headache, with or without focal deficits and/or seizures, has led to angiography (with ‘strings and beads’ appearance) and diagnosis of RCVS
 - 2. headache has either or both of the following characteristics:
 - a) recurrent during ≤ 1 month, and with thunderclap onset
 - b) triggered by sexual activity, exertion, Valsalva manœuvres, emotion, bathing and/or showering
 - 3. no new significant headache occurs > 1 month after onset
- D. Not better accounted for by another ICHD-3 diagnosis, and aneurysmal subarachnoid haemorrhage has been excluded by appropriate investigations.

Comments:

Reversible cerebral vasoconstriction syndrome (RCVS) is a poorly understood condition, characterized clinically by severe diffuse headaches that typically are of the thunderclap type, mimicking aneurysmal SAH. RCVS is the most frequent cause of thunderclap headache recurring over a few days or weeks. 6.7.3 *Headache attributed to reversible cerebral vasoconstriction syndrome* may rarely have other modes of onset: progressing rapidly over hours or more slowly over days. Headache is often the only symptom of RCVS, but the condition can be associated with fluctuating focal neurological deficits and sometimes seizures.

Angiography is, by definition, abnormal, with alternating segments of arterial constriction and dilatation (‘strings and beads’ appearance). However, MR-, CT- and even catheter-angiography can be normal during the first week after clinical onset. Patients with recurring thunderclap headache and a normal angiogram, but fulfilling all other criteria for RCVS, should be considered as having 6.7.3.1 *Headache probably attributed to reversible cerebral vasoconstriction syndrome*. Brain MRI is abnormal in 30-80% of cases, showing various patterns of lesions including intracranial haemorrhages (convexity subarachnoid, intracerebral and/or subdural), cerebral infarctions and/or cerebral oedema corresponding to ‘posterior reversible encephalopathy syndrome’.

At least half of cases of RCVS are secondary, mainly postpartum and/or following exposure to vasoactive substances including illicit drugs, alpha-sympathomimetics and serotonergic drugs. The disease is self-limiting in 1–3 months, with resolution of the headache and disappearance of the arterial abnormalities (hence ‘reversible’). However, strokes as a result of RCVS can produce permanent impairment.

6.7.3.1 *Headache probably attributed to reversible cerebral vasoconstriction syndrome (RCVS)*

Description:

Headache typical for reversible cerebral vasoconstriction syndrome (RCVS), namely thunderclap headache recurring over 1–2 weeks and triggered by sexual activity, exertion, Valsalva manœuvres and/or emotion, but the intracranial arterial beading typical of RCVS has not been demonstrated by cerebral angiography.

Diagnostic criteria:

- A. Any new headache fulfilling criterion C
- B. Reversible cerebral vasoconstriction syndrome (RCVS) is suspected, but cerebral angiography is normal
- C. Probability of causation demonstrated by all of the following:
 - 1. at least two headaches within 1 month, with all three of the following characteristics:
 - a) thunderclap onset, and peaking in < 1 minute
 - b) severe intensity
 - c) lasting ≥ 5 minutes
 - 2. at least one thunderclap headache has been triggered by one of the following:
 - a) sexual activity (just before or at orgasm)
 - b) exertion
 - c) Valsalva-like manœuvre
 - d) emotion
 - e) bathing and/or showering

- f) bending
- 3. no new thunderclap or other significant headache occurs >1 month after onset
- D. Not fulfilling ICHD-3 criteria for any other headache disorder
- E. Not better accounted for by another ICHD-3 diagnosis, and aneurysmal subarachnoid haemorrhage has been excluded by appropriate investigations.

Comment:

Large series of patients with confirmed RCVS have shown that up to 75% of patients presented with isolated headaches. The arterial abnormalities of RCVS may be difficult to demonstrate. Some RCVS cases need repeated CT- or MR-angiography 2-3 weeks after headache onset and others need invasive conventional angiography to be detected. In patients who have recurrent, triggered thunderclap headaches typical for RCVS over a period of less than 1 month and normal initial cerebral angiography, and in whom another cause of the headaches has been excluded by appropriate investigations, a diagnosis of 6.7.3.1 *Headache probably attributed to reversible cerebral vasoconstriction syndrome (RCVS)* can be made.

6.7.4 Headache attributed to intracranial arterial dissection

Description:

Headache caused by dissection of an intracranial artery. The pain is mostly unilateral, ipsilateral to the dissected vessel, and generally has a sudden (even thunderclap) onset. It can remain isolated or be a warning symptom preceding (mostly haemorrhagic) stroke.

Diagnostic criteria:

- A. Any new headache fulfilling criterion C
- B. An intracranial arterial dissection has been diagnosed
- C. Evidence of causation demonstrated by at least two of the following:
 - 1. headache has developed in close temporal relation to other symptoms and/or clinical signs of intracranial dissection, or has led to the diagnosis of intracranial dissection
 - 2. headache resolves within 1 month of its onset
 - 3. headache has either or both of the following characteristics:
 - a) sudden or thunderclap onset
 - b) severe intensity
 - 4. headache is unilateral and ipsilateral to the dissection

- D. Not better accounted for by another ICHD-3 diagnosis.

Comment:

Dissection can affect any intracranial artery and may induce ischaemic infarcts, compression of adjacent structures or intracranial haemorrhages (subarachnoid or intracerebral). Acute headache is often the presenting symptom and can be the sole symptom of this disorder.

6.8 Headache attributed to genetic vasculopathy

Description:

Headache occurring as part of the phenotypic spectrum of the genetic cerebral vasculopathies, mostly manifesting as recurrent attacks of headache, which may have the features of migraine with or without aura. Attacks can recur for years, and are usually associated from onset, or after a variable delay, with other manifestations of the causative mutation.

Diagnostic criteria:

- A. Recurrent attacks of headache fulfilling criterion C
- B. A genetic vasculopathy has been diagnosed by appropriate genetic testing
- C. Headache is either:
 - 1. migraine-like
 - 2. the presenting symptom of stroke-like episodes
- D. Not better accounted for by another ICHD-3 diagnosis.

6.8.1 Cerebral Autosomal Dominant Arteriopathy with Subcortical Infarcts and Leukoencephalopathy (CADASIL)

Description:

An autosomal dominant (with some sporadic cases) small-artery disease of the brain characterized clinically by recurrent small deep infarcts, subcortical dementia, mood disturbances and, in one-third of cases, by migraine with aura (which is usually the first symptom of the disease).

Diagnostic criteria:

- A. Recurrent attacks of migraine with typical, hemiplegic or prolonged aura, fulfilling criterion C
- B. Cerebral Autosomal Dominant Arteriopathy with Subcortical Infarcts and Leukoencephalopathy (CADASIL) has been demonstrated by genetic testing for NOTCH-3 mutations and/or skin biopsy evidence

- C. Either or both of the following:
 1. migraine with aura was the earliest clinical manifestation of CADASIL
 2. attacks of migraine with aura improve or cease when other manifestations of CADASIL (e.g. ischaemic stroke, mood disturbances and/or cognitive dysfunction) appear and worsen
- D. Not better accounted for by another ICHD-3 diagnosis.

Comments:

CADASIL is an autosomal dominant disease, with some sporadic cases, involving the smooth muscle cells in the media of small arteries of the brain. It is a result of mutations of the NOTCH-3 gene; the diagnosis is made by screening for NOTCH-3 mutations or by a simple skin biopsy with immunostaining of NOTCH-3 antibodies.

CADASIL is characterized clinically by recurrent small deep infarcts, subcortical dementia, mood disturbances and, in one-third of cases, by migraine with aura. In such cases, this is usually the first symptom of the disease, appearing at a mean age of 30 years, some 15 years before ischaemic strokes and 20–30 years before death. Migraine attacks are typical of 1.2 *Migraine with aura* except for an unusual frequency of prolonged aura.

MRI is always abnormal, with striking white matter changes on T2-weighted images.

6.8.2 Mitochondrial Encephalopathy, Lactic Acidosis and Stroke-like episodes (MELAS)

Description:

A genetically heterogeneous mitochondrial disorder with a variable clinical phenotype, including features of central nervous system involvement (seizures, hemiparesis, hemianopia, cortical blindness, sensorineural deafness and/or episodic vomiting) and, frequently, headache, which is either recurrent in migraine-like attacks or a presenting symptom of stroke-like episodes.

Diagnostic criteria:

- A. Recurrent attacks of headache fulfilling criterion C
- B. A mitochondrial genetic abnormality associated with MELAS has been demonstrated
- C. Either or both of the following:
 1. recurrent migraine attacks with or without aura
 2. acute headache preceding focal neurological deficits and/or seizures
- D. Not better accounted for by another ICHD-3 diagnosis.

Comment:

MELAS syndrome, comprising mitochondrial myopathy, encephalopathy, lactic acidosis and stroke-like episodes, is a genetically heterogeneous mitochondrial disorder with a variable clinical phenotype. The disorder is accompanied by features of central nervous system involvement, including seizures, hemiparesis, hemianopia, cortical blindness, sensorineural deafness and episodic vomiting. Headache is frequent in MELAS, either as recurrent migraine-like attacks or as the presenting symptom of stroke-like episodes. The high frequency of migraine-like attacks as part of MELAS has led to the hypothesis that mitochondrial mutations may play a role in migraine with aura, but the 3243 mutation was not detected in two groups of subjects with 1.2 *Migraine with aura*. Other yet-undetected mutations may play a role in both migraine and ischaemic stroke, as migraine attacks, mostly with aura, also occur in other mitochondrial disorders.

6.8.3 Headache attributed to other genetic vasculopathy

Description:

Migraine with or without aura occurring as part of the phenotypic spectrum of a genetic vasculopathy other than those described above.

Diagnostic criteria:

- A. Recurrent attacks of migraine with or without aura, fulfilling criterion C
- B. A genetic vasculopathy has been demonstrated by appropriate genetic testing
- C. Migraine attacks are understood to be part of the syndrome associated with the genetic vasculopathy
- D. Not better accounted for by another ICHD-3 diagnosis.

Comment:

Recurrent migraine attacks have been reported as part of the clinical spectrum of the autosomal dominant disorder, retinal vasculopathy with cerebral leukodystrophy (RVCL), which is caused by TREX1 mutations, and of hereditary infantile hemiparesis, retinal arterial tortuosity and leukoencephalopathy (HIHRATL), a condition occurring as a result of COL4A1 mutations. Only a few families with either disorder have been reported. Because of the presence of other severe manifestations, migraine has not been systematically investigated in these pedigrees. It appears that RVCL was mainly associated with attacks of 1.1 *Migraine without aura* and HIHRATL with attacks of 1.2 *Migraine with aura*.

6.9 Headache attributed to pituitary apoplexy

Description:

Headache caused by pituitary apoplexy, usually with sudden (even thunderclap) onset and severe intensity, and accompanied from onset or later by visual symptoms and/or hypopituitarism.

Diagnostic criteria:

- A. Any new headache fulfilling criterion C
- B. Acute haemorrhagic pituitary infarction has been diagnosed
- C. Evidence of causation demonstrated by at least two of the following:
 1. headache has developed in close temporal relation to other symptoms and/or clinical signs of pituitary apoplexy, or has led to the diagnosis of pituitary apoplexy
 2. either or both of the following:
 - a) headache has significantly worsened in parallel with other symptoms and/or clinical signs of pituitary apoplexy
 - b) headache has significantly improved in parallel with other symptoms and/or clinical signs of improvement of pituitary apoplexy
 3. headache is severe and of sudden or thunderclap onset
- D. Not better accounted for by another ICHD-3 diagnosis.

Comments:

The rare clinical syndrome of pituitary apoplexy is an acute, life-threatening condition. It is one of the causes of thunderclap headache. Most cases occur as the first presentation of rapid enlargement of non-functioning pituitary macroadenomas as a result of haemorrhage and/or infarction. It is one of the causes of non-aneurysmal subarachnoid haemorrhage.

MRI is more sensitive than CT scan for detecting intrasellar pathology.

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6.9 Headache attributed to pituitary apoplexy

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7. Headache attributed to non-vascular intracranial disorder

- 7.1 Headache attributed to increased cerebrospinal fluid pressure
 - 7.1.1 Headache attributed to idiopathic intracranial hypertension (IIH)
 - 7.1.2 Headache attributed to intracranial hypertension secondary to metabolic, toxic or hormonal causes
 - 7.1.3 Headache attributed to intracranial hypertension secondary to hydrocephalus
- 7.2 Headache attributed to low cerebrospinal fluid pressure
 - 7.2.1 Post-dural puncture headache
 - 7.2.2 CSF fistula headache
 - 7.2.3 Headache attributed to spontaneous intracranial hypotension
- 7.3 Headache attributed to non-infectious inflammatory intracranial disease
 - 7.3.1 Headache attributed to neurosarcoidosis
 - 7.3.2 Headache attributed to aseptic (non-infectious) meningitis
 - 7.3.3 Headache attributed to other non-infectious inflammatory intracranial disease
 - 7.3.4 Headache attributed to lymphocytic hypophysitis
 - 7.3.5 Syndrome of transient Headache and Neurological Deficits with cerebrospinal fluid Lymphocytosis (HaNDL)
- 7.4 Headache attributed to intracranial neoplasia
 - 7.4.1 Headache attributed to intracranial neoplasm
 - 7.4.1.1 Headache attributed to colloid cyst of the third ventricle
 - 7.4.2 Headache attributed to carcinomatous meningitis
 - 7.4.3 Headache attributed to hypothalamic or pituitary hyper- or hyposecretion
- 7.5 Headache attributed to intrathecal injection
- 7.6 Headache attributed to epileptic seizure
 - 7.6.1 Hemicrania epileptica
 - 7.6.2 Post-ictal headache
- 7.7 Headache attributed to Chiari malformation type I (CM1)
- 7.8 Headache attributed to other non-vascular intracranial disorder

General comment

Primary or secondary headache or both?

When a headache occurs for the first time in close temporal relation to a non-vascular intracranial disorder, it is coded as a secondary headache attributed to that disorder. This remains true when the new headache

has the characteristics of any of the primary headache disorders classified in Part one of ICHD-3 beta. When a *pre-existing* headache with the characteristics of a primary headache disorder becomes *chronic*, or is made significantly worse (usually meaning a two-fold or greater increase in frequency and/or severity), in close temporal relation to a non-vascular intracranial disorder, both the initial headache diagnosis and a diagnosis of 7. *Headache attributed to non-vascular intracranial disorder* (or one of its subtypes) should be given, provided that there is good evidence that the disorder can cause headache.

Introduction

In this chapter, the headaches are attributed to changes in intracranial pressure. Both increased and decreased cerebrospinal fluid (CSF) pressure can lead to headache. Other causes of headache here are non-infectious inflammatory diseases, intracranial neoplasia, seizures, rare conditions such as intrathecal injections and Chiari malformation type I, and other non-vascular intracranial disorders.

Compared with those on primary headaches, there are few epidemiological studies of these headache types. Controlled trials of therapy are almost non-existent.

For headache attributed to any of the non-vascular intracranial disorders listed here, the diagnostic criteria include whenever possible:

- A. Headache fulfilling criterion C
- B. A non-vascular intracranial disorder known to be able to cause headache has been diagnosed
- C. Evidence of causation demonstrated by at least two of the following:
 1. headache has developed in temporal relation to the onset of the non-vascular intracranial disorder
 2. either or both of the following:
 - a) headache has significantly worsened in parallel with worsening of the non-vascular intracranial disorder
 - b) headache has significantly improved in parallel with improvement in the non-vascular intracranial disorder
 3. headache has characteristics typical for the non-vascular intracranial disorder
 4. other evidence exists of causation
- D. Not better accounted for by another ICHD-3 diagnosis.

Headache persisting for more than 1 month after successful treatment or spontaneous resolution of the intracranial disorder usually has other mechanisms. Headache persisting for more than 3 months after treatment or remission of intracranial disorders is defined in

the Appendix for research purposes. Such headache exists but has been poorly studied; Appendix entries are intended to stimulate further research into such headaches and their mechanisms.

7.1 Headache attributed to increased cerebrospinal fluid pressure

Coded elsewhere:

Headache attributed to intracranial pressure or hydrocephalus secondary to an intracranial neoplasm is coded as 7.4.1 *Headache attributed to intracranial neoplasm*.

Description:

Headache caused by increased cerebrospinal fluid (CSF) pressure, usually accompanied by other symptoms and/or clinical signs of intracranial hypertension. It remits after normalization of CSF pressure.

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. Increased CSF pressure (>250 mm CSF) measured by lumbar puncture (performed in the lateral decubitus position, without sedative medications), epidural or intraventricular monitoring, with normal CSF chemistry and cellularity
- C. Evidence of causation demonstrated by either or both of the following:
 1. headache has developed in temporal relation to intracranial hypertension
 2. headache is relieved by reducing intracranial pressure
- D. Not better accounted for by another ICHD-3 diagnosis.

7.1.1 *Headache attributed to idiopathic intracranial hypertension (IIH)*

Previously used terms:

Headache attributed to benign intracranial hypertension (BIH); pseudotumour cerebri; meningeal hydrops; serous meningitis.

Description:

Headache caused by idiopathic intracranial hypertension (IIH), usually accompanied by other symptoms and/or clinical signs of IIH. It remits after normalization of cerebrospinal fluid pressure.

Diagnostic criteria:

- A. Any headache fulfilling criterion C

- B. Idiopathic intracranial hypertension (IIH) has been diagnosed, with CSF pressure >250 mm CSF (measured by lumbar puncture performed in the lateral decubitus position, without sedative medications, or by epidural or intraventricular monitoring)
- C. Evidence of causation demonstrated by at least two of the following:
 1. headache has developed in temporal relation to IIH, or led to its discovery
 2. headache is relieved by reducing intracranial hypertension
 3. headache is aggravated in temporal relation to increase in intracranial pressure
- D. Not better accounted for by another ICHD-3 diagnosis.

Comments:

Idiopathic intracranial hypertension (IIH) most commonly occurs in young obese women.

IIH should be diagnosed with caution in those with altered mental status and in patients with CSF pressure below 250 mm CSF. In some patients, especially children, an opening pressure of up to 280 mm CSF is normal, but, for most, an opening pressure above 280 mm CSF should be considered elevated.

Body mass index is only weakly related to CSF pressure, and a mildly elevated CSF pressure should not be dismissed in obese patients.

CSF pressure varies when lumbar epidural pressure monitoring is done for 1 hour or more, so a single measurement performed within minutes may not be indicative of the average CSF pressure over 24 hours. Diagnostic CSF pressure measurement should be made when the patient is not receiving treatment to lower the intracranial pressure. Neuroimaging findings consistent with the diagnosis of IIH include empty sella turcica, distension of the perioptic subarachnoid space, flattening of the posterior sclerae, protrusion of the optic nerve papillae into the vitreous and transverse cerebral venous sinus stenosis.

Although the majority of patients with IIH have papilloedema, IIH without papilloedema has been observed. Other symptoms or signs of IIH include pulse-synchronous tinnitus, transient visual obscurations, neck or back pain and diplopia. 7.1.1 *Headache attributed to idiopathic intracranial hypertension (IIH)* lacks specific features. It is frequently described as frontal, retro-orbital, 'pressure like' or explosive; migraine-like headache may also occur.

7.1.2 Headache attributed to intracranial hypertension secondary to metabolic, toxic or hormonal causes

Coded elsewhere:

Headache attributed to increased intracranial pressure as a result of head trauma, vascular disorder or intracranial infection is coded to whichever of these is the cause. Headache attributed to raised intracranial pressure occurring as a side effect of medication is coded as 8.1.11 Headache attributed to long-term use of non-headache medication.

Description:

Headache caused by intracranial hypertension secondary to a variety of systemic disorders and accompanied by other symptoms and/or clinical signs of intracranial hypertension. It remits with resolution of the systemic disorder.

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. A metabolic, toxic or hormonal disorder has been diagnosed, with CSF pressure >250 mm CSF (measured by lumbar puncture performed in the lateral decubitus position, without sedative medications, or by epidural or intraventricular monitoring) and with normal CSF chemistry and cellularity
- C. Evidence of causation demonstrated by either or both of the following:
 - 1. headache has developed in temporal relation to the metabolic, toxic or hormonal disorder
 - 2. either or both of the following:
 - a) headache has significantly worsened in parallel with worsening of the metabolic, toxic or hormonal disorder
 - b) headache has significantly improved in parallel with improvement in the metabolic, toxic or hormonal disorder
- D. Not better accounted for by another ICHD-3 diagnosis.

Comments:

Potential causes of intracranial hypertension include acute hepatic failure, hypercarbia, acute hypertensive crisis, Reye's hepatocerebral syndrome and heart failure.

Removal of the inciting agent or treatment of the secondary cause may not be sufficient to normalize the high intracranial pressure; additional treatment is often required to prevent visual loss, and to relieve headache and other symptoms.

7.1.3 Headache attributed to intracranial hypertension secondary to hydrocephalus

Description:

Headache caused by hydrocephalus, accompanied by other symptoms and/or clinical signs of increased cerebrospinal fluid pressure or hydrocephalus. It remits after resolution of the hydrocephalus.

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. Hydrocephalus has been diagnosed, with CSF pressure >250 mm CSF (measured by lumbar puncture performed in the lateral decubitus position, without sedative medications, or by epidural or intraventricular monitoring)
- C. Evidence of causation demonstrated by either or both of the following:
 - 1. headache has developed in temporal relation to the hydrocephalus
 - 2. either or both of the following:
 - a) headache has significantly worsened in parallel with worsening of the hydrocephalus
 - b) headache has significantly improved in parallel with improvement in the hydrocephalus
- D. Not better accounted for by another ICHD-3 diagnosis.

Comment:

Normal-pressure hydrocephalus usually does not cause headache; occasionally, mild dull headache is reported.

7.2 Headache attributed to low cerebrospinal fluid pressure

Description:

Orthostatic headache in the presence of low cerebrospinal fluid (CSF) pressure (either spontaneous or secondary), or CSF leakage, usually accompanied by neck pain, tinnitus, changes in hearing, photophobia and/or nausea. It remits after normalization of CSF pressure or successful sealing of the CSF leak.

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. Low CSF pressure (<60 mm CSF) and/or evidence of CSF leakage on imaging
- C. Headache has developed in temporal relation to the low CSF pressure or CSF leakage, or led to its discovery
- D. Not better accounted for by another ICHD-3 diagnosis.

Comment:

7.2 Headache attributed to low cerebrospinal fluid pressure is usually but not invariably orthostatic. Headache that significantly worsens soon after sitting upright or standing and/or improves after lying horizontally is likely to be caused by low CSF pressure, but this cannot be relied on as a diagnostic criterion. Evidence of causation may depend on onset in temporal relation to the presumed cause together with exclusion of other diagnoses.

7.2.1 *Post-dural puncture headache**Previously used term:*

Post-lumbar puncture headache.

Description:

Headache occurring within 5 days of a lumbar puncture, caused by cerebrospinal fluid (CSF) leakage through the dural puncture. It is usually accompanied by neck stiffness and/or subjective hearing symptoms. It remits spontaneously within 2 weeks, or after sealing of the leak with autologous epidural lumbar patch.

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. Dural puncture has been performed
- C. Headache has developed within 5 days of the dural puncture
- D. Not better accounted for by another ICHD-3 diagnosis.

Comment:

Independent risk factors for 7.2.1 *Post-dural puncture headache* have recently been demonstrated: female gender, age between 31 and 50 years, a previous history of 7.2.1 *Post-dural puncture headache* and orientation of the needle bevel perpendicular to the long axis of the spinal column at the time of the dural puncture.

7.2.2 *CSF fistula headache**Description:*

Orthostatic headache occurring after a procedure or trauma causing a persistent cerebrospinal fluid (CSF) leakage resulting in low intracranial pressure. It remits after successful sealing of the CSF leak.

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. Both of the following:

1. a procedure has been performed, or trauma has occurred, known sometimes to cause persistent CSF leakage (CSF fistula)
 2. low CSF pressure (<60 mm CSF) and/or evidence of low CSF pressure and/or of CSF leakage on MRI, myelography, CT myelography or radionuclide cisternography
- C. Headache has developed in temporal relation to the procedure or trauma
 - D. Not better accounted for by another ICHD-3 diagnosis.

7.2.3 *Headache attributed to spontaneous intracranial hypotension**Previously used terms:*

Headache attributed to spontaneous low CSF pressure or primary intracranial hypotension; low CSF-volume headache; hypoliquorrhoeic headache.

Description:

Orthostatic headache caused by low cerebrospinal fluid (CSF) pressure of spontaneous origin. It is usually accompanied by neck stiffness and subjective hearing symptoms. It remits after normalization of CSF pressure.

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. Low CSF pressure (<60 mm CSF) and/or evidence of CSF leakage on imaging
- C. Headache has developed in temporal relation to the low CSF pressure or CSF leakage, or has led to its discovery
- D. Not better accounted for by another ICHD-3 diagnosis.

Comments:

7.2.3 *Headache attributed to spontaneous intracranial hypotension* cannot be diagnosed in a patient who has had a dural puncture within the prior month.

The headache in patients with spontaneous CSF leaks or spontaneously low CSF pressure may resemble 7.2.1 *Post-dural puncture headache*, occurring immediately or within seconds of assuming an upright position and resolving quickly (within 1 minute) after lying horizontally. Alternatively it may show delayed response to postural change, worsening after minutes or hours of being upright and improving, but not necessarily resolving, after minutes or hours of being horizontal. Although there is a clear postural component in most cases of 7.2.3 *Headache attributed to spontaneous*

intracranial hypotension, it may not be as dramatic or immediate as in 7.2.1 *Post-dural puncture headache*. The orthostatic nature of the headache at its onset should be sought when eliciting a history, as this feature may become much less obvious over time.

Although autologous epidural blood patches (EBPs) are frequently effective in sealing CSF leaks, the response to a single EBP may not be permanent, and complete relief of symptoms may not be achieved until two or more EBPs have been performed. However, some degree of sustained improvement, beyond a few days, is generally expected. In some cases, sustained improvement cannot be achieved with EBPs and surgical intervention may be required.

In patients with typical orthostatic headache and no apparent cause, after exclusion of postural orthostatic tachycardia syndrome (POTS) it is reasonable in clinical practice to provide autologous lumbar EBP.

It is not clear that all patients have an active CSF leak, despite a compelling history or brain imaging signs compatible with CSF leakage. Cisternography is an outdated test, now infrequently used; it is significantly less sensitive than other imaging modalities (MRI, CT or digital subtraction myelography). Dural puncture to measure CSF pressure directly is not necessary in patients with positive MRI signs such as dural enhancement with contrast.

The underlying disorder in 7.2.3 *Headache attributed to spontaneous intracranial hypotension* may be low CSF volume. A history of a trivial increase in intracranial pressure (e.g. on vigorous coughing) is sometimes elicited. Postural headache has been reported after coitus: such headache should be coded as 7.2.3 *Headache attributed to spontaneous intracranial hypotension* because it is most probably a result of CSF leakage.

7.3 Headache attributed to non-infectious inflammatory intracranial disease

Description:

Headache in the presence of a non-infectious inflammatory intracranial disease, usually with lymphocytic pleocytosis in the cerebrospinal fluid. It remits after resolution of the inflammatory disorder.

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. A non-infectious inflammatory disease known to be able to cause headache has been diagnosed
- C. Evidence of causation demonstrated by either or both of the following:
 - 1. headache has developed in temporal relation to the onset of the non-infectious inflammatory disease

- 2. either or both of the following:
 - a) headache has significantly worsened in parallel with worsening of the non-infectious inflammatory disease
 - b) headache has significantly improved in parallel with improvement of the non-infectious inflammatory disease
- D. Not better accounted for by another ICHD-3 diagnosis.

7.3.1 Headache attributed to neurosarcoidosis

Description:

Headache caused by neurosarcoidosis, associated with aseptic meningitis, cranial nerve lesions, intracranial space-occupying lesion(s) on brain MRI, periventricular inflammatory focal lesions and/or homogeneously enhancing mass lesions on brain or spinal MRI that are confirmed on biopsy as non-caseating granulomas.

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. Neurosarcoidosis has been diagnosed
- C. Evidence of causation demonstrated by at least two of the following:
 - 1. headache has developed in temporal relation to the onset of neurosarcoidosis
 - 2. either or both of the following:
 - a) headache has significantly worsened in parallel with worsening of neurosarcoidosis
 - b) headache has significantly improved in parallel with improvement in neurosarcoidosis
 - 3. headache is accompanied by one or more cranial nerve palsies
- D. Not better accounted for by another ICHD-3 diagnosis.

Comment:

Other manifestations of neurosarcoidosis include aseptic meningitis, cranial nerve lesions, intracranial space-occupying lesion(s) on brain MRI, periventricular inflammatory focal lesions and/or homogeneously enhancing mass lesions on brain or spinal MRI that are confirmed on biopsy as non-caseating granulomas.

7.3.2 Headache attributed to aseptic (non-infectious) meningitis

Description:

Headache caused by aseptic meningitis, associated with other symptoms and/or clinical signs of meningeal irritation. It resolves after resolution of the meningitis.

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. Aseptic meningitis has been diagnosed by CSF examination
- C. Evidence of causation demonstrated by at least two of the following:
 1. headache has developed in temporal relation to the onset of aseptic meningitis, or led to its discovery
 2. either or both of the following:
 - a) headache has significantly worsened in parallel with worsening of aseptic meningitis
 - b) headache has significantly improved in parallel with improvement in aseptic meningitis
 3. headache is accompanied by other symptoms and/or clinical signs of meningeal inflammation including neck stiffness (meningismus) and/or photophobia
- D. Not better accounted for by another ICHD-3 diagnosis.

Comments:

The CSF in patients with aseptic meningitis shows lymphocytic pleocytosis, mildly elevated protein and normal glucose in the absence of infectious organisms.

Aseptic meningitis may occur after exposure to certain drugs, including ibuprofen or other NSAIDs, immunoglobulins, penicillin or trimethoprim, intrathecal injections and/or insufflations.

7.3.3 Headache attributed to other non-infectious inflammatory intracranial disease

Description:

Headache caused by any of a variety of autoimmune disorders, associated with other symptoms and/or clinical signs of the causative disorder. It remits after successful treatment of the autoimmune disorder.

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. A non-infectious inflammatory disease known to be able to cause headache, other than those described above, has been diagnosed
- C. Evidence of causation demonstrated by either or both of the following:
 1. headache has developed in temporal relation to the onset of the non-infectious inflammatory disease
 2. either or both of the following:
 - a) headache has significantly worsened in parallel with worsening of the non-infectious inflammatory disease

- b) headache has significantly improved in parallel with improvement in the non-infectious inflammatory disease
- D. Not better accounted for by another ICHD-3 diagnosis.

Comment:

Headache can be causally associated with, but is not usually a presenting or prominent symptom of, acute demyelinating encephalomyelitis (ADEM), systemic lupus erythematosus (SLE), Behçet's syndrome and other systemic or focal (e.g. limbic encephalitis) autoimmune syndromes.

7.3.4 Headache attributed to lymphocytic hypophysitis

Description:

Headache caused by lymphocytic hypophysitis, associated with pituitary enlargement and, in half of cases, with hyperprolactinaemia. It remits after successful treatment of the lymphocytic hypophysitis.

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. Lymphocytic hypophysitis has been diagnosed
- C. Evidence of causation demonstrated by either or both of the following:
 1. headache has developed in temporal relation to the onset of the lymphocytic hypophysitis
 2. either or both of the following:
 - a) headache has significantly worsened in parallel with worsening of the lymphocytic hypophysitis
 - b) headache has significantly improved in parallel with improvement in the lymphocytic hypophysitis
- D. Not better accounted for by another ICHD-3 diagnosis.

Comments:

Lymphocytic hypophysitis is associated with pituitary enlargement and homogeneous contrast enhancement on brain MRI. It is accompanied by hyperprolactinaemia in 50% of cases or autoantibodies against hypophysial cytosol protein in 20% of cases.

The disorder typically develops at the end of pregnancy or during the post-partum period, but it can also occur in men.

7.3.5 Syndrome of transient Headache and Neurological Deficits with cerebrospinal fluid Lymphocytosis (HaNDL)

Previously used terms:

Migraine with cerebrospinal pleocytosis; pseudomigraine with lymphocytic pleocytosis.

Description:

Migraine-like headache episodes (typically one to twelve) accompanied by neurological deficits including hemiparaesthesia, hemiparesis and/or dysphasia, but positive visual symptoms only uncommonly, lasting several hours. There is lymphocytic pleocytosis. The disorder resolves spontaneously within 3 months.

Diagnostic criteria:

- A. Episodes of migraine-like headache fulfilling criteria B and C
- B. Both of the following:
 - 1. accompanied or shortly preceded by the onset of at least one of the following transient neurological deficits lasting >4 hours
 - a) hemiparaesthesia
 - b) dysphasia
 - c) hemiparesis
 - 2. associated with CSF lymphocytic pleocytosis (>15 white cells per µl), with negative aetiological studies
- C. Evidence of causation demonstrated by either or both of the following:
 - 1. headache and transient neurological deficits have developed or significantly worsened in temporal relation to the CSF lymphocytic pleocytosis, or led to its discovery
 - 2. headache and transient neurological deficits have significantly improved in parallel with improvement in the CSF lymphocytic pleocytosis
- D. Not better accounted for by another ICHD-3 diagnosis.

Comments:

The clinical picture of 7.3.5 *Syndrome of transient Headache and Neurological Deficits with cerebrospinal fluid Lymphocytosis (HaNDL)* is of 1–12 discrete episodes of transient neurological deficits accompanied or followed by moderate to severe headache. Most of the episodes last hours, but some may last for more than 24 hours. The neurological manifestations include sensory symptoms in about three-quarters of cases, aphasia in two-thirds and motor deficits in a little over half. Migraine-aura-like visual symptoms are relatively

uncommon (fewer than 20% of cases). The syndrome resolves within 3 months.

In addition to CSF lymphocytosis (up to 760 cells/µl), there are elevations of CSF total protein (up to 250 mg/dl) in >90% of cases and of CSF pressure (up to 400 mm CSF) in more than 50% of cases. The presence of a viral prodrome in at least one-quarter of cases has raised the possibility of an autoimmune pathogenesis of 7.3.5 *Syndrome of transient Headache and Neurological Deficits with cerebrospinal fluid Lymphocytosis (HaNDL)*. A recent description of antibodies to a subunit of the T-type voltage-gated calcium channel CACNA1H in the sera of two patients with this disorder supports this view.

Papilloedema is occasionally present. Routine CT and MRI scans (with or without intravenous contrast) and angiography are invariably normal when performed outside of an episode. Ictal brain imaging may show delayed brain perfusion without increased diffusion-weighted imaging changes, and narrowing of cerebral arteries. Also, grey matter oedema and sulcal enhancement have been described in a single patient. Microbiological studies have been uniformly normal. EEG and SPECT scans may show focally abnormal areas consistent with the focal neurological deficits.

Most patients with this syndrome have no prior history of migraine. The clinician must consider other diagnoses that may share some of its clinical features, including 1.2.3 *Hemiplegic migraine* (although mutations of the CACNA1A gene, the cause of 1.2.3.1.1 *Familial hemiplegic migraine type 1 (FHM1)*, have been excluded in several patients with 7.3.5 *Syndrome of transient Headache and Neurological Deficits with cerebrospinal fluid Lymphocytosis (HaNDL)*), neuroborreliosis, neurosyphilis, neurobrucellosis, mycoplasma, granulomatous and neoplastic arachnoiditis, encephalitis and CNS vasculitis.

7.4 Headache attributed to intracranial neoplasia

Description:

Headache caused by intracranial neoplasia.

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. Intracranial neoplasia has been diagnosed
- C. Evidence of causation demonstrated by at least one of the following:
 - 1. headache has developed in temporal relation to the intracranial neoplasia, or led to its discovery
 - 2. headache has significantly worsened in parallel with worsening of the intracranial neoplasia

3. headache has significantly improved in temporal relation to successful treatment of the intracranial neoplasia
- D. Not better accounted for by another ICHD-3 diagnosis.

7.4.1 Headache attributed to intracranial neoplasm

Description:

Headache, usually progressive, worse in the morning and aggravated by Valsalva-like manœuvres, caused by one or more space-occupying intracranial tumours.

Diagnostic criteria:

- A. Headache fulfilling criterion C
- B. A space-occupying intracranial neoplasm has been demonstrated
- C. Evidence of causation demonstrated by at least two of the following:
 1. headache has developed in temporal relation to development of the neoplasm, or led to its discovery
 2. either or both of the following:
 - a) headache has significantly worsened in parallel with worsening of the neoplasm
 - b) headache has significantly improved in temporal relation to successful treatment of the neoplasm
 3. headache has at least one of the following three characteristics:
 - a) progressive
 - b) worse in the morning or after daytime napping
 - c) aggravated by Valsalva-like manœuvres
- D. Not better accounted for by another ICHD-3 diagnosis.

7.4.1.1 Headache attributed to colloid cyst of the third ventricle

Description:

Headache caused by colloid cyst of the third ventricle, presenting very characteristically as recurrent attacks with thunderclap onset and reduced level or loss of consciousness.

Diagnostic criteria:

- A. Headache fulfilling criterion C
- B. A colloid cyst of the third ventricle has been demonstrated

- C. Evidence of causation demonstrated by both of the following:
 1. headache has developed in temporal relation to development of the colloid cyst, or led to its discovery
 2. either or both of the following:
 - a) headache is recurrent, with thunderclap onset and accompanied by reduced level or loss of consciousness
 - b) headache has significantly improved or resolved in temporal relation to successful treatment of the colloid cyst

- D. Not better accounted for by another ICHD-3 diagnosis.

Comment:

The vast majority of colloid cysts of the third ventricle are discovered incidentally, having been asymptomatic. Nevertheless, their position immediately adjacent to the foramen of Monro can, on occasion, result in sudden obstructive hydrocephalus, causing headache with thunderclap onset and reduced level or loss of consciousness. This highly characteristic presentation should lead to rapid diagnosis. 7.4.1.1 *Headache attributed to colloid cyst of the third ventricle* signals a life-threatening emergency.

7.4.2 Headache attributed to carcinomatous meningitis

Description:

Headache caused by carcinomatous meningitis, usually accompanied by signs of encephalopathy and/or cranial nerve palsies.

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. Carcinomatous meningitis (in the presence of systemic neoplasia known to be associated with carcinomatous meningitis) has been demonstrated
- C. Evidence of causation demonstrated by at least two of the following:
 1. headache has developed in temporal relation to development of the carcinomatous meningitis
 2. either or both of the following:
 - a) headache has significantly worsened in parallel with worsening of the carcinomatous meningitis
 - b) headache has significantly improved in parallel with improvement in the carcinomatous meningitis
 3. headache is associated with cranial nerve palsies and/or encephalopathy

D. Not better accounted for by another ICHD-3 diagnosis.

7.4.3 Headache attributed to hypothalamic or pituitary hyper- or hyposecretion

Description:

Headache caused by a pituitary adenoma and hypothalamic or pituitary hyper- or hyposecretion, usually accompanied by disorder of temperature regulation, abnormal emotional state and/or altered thirst or appetite. It remits after successful treatment of the underlying disorder.

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. Hypothalamic or pituitary hyper- or hyposecretion (including prolactin, growth hormone (GH) and/or adrenocorticotrophic hormone (ACTH) hypersecretion), associated with pituitary adenoma, has been demonstrated
- C. Evidence of causation demonstrated by at least two of the following:
 - 1. headache has developed in temporal relation to onset of hypothalamic or pituitary hyper- or hyposecretion
 - 2. either or both of the following:
 - a) headache has significantly worsened in parallel with worsening of the hypothalamic or pituitary hyper- or hyposecretion
 - b) headache has significantly improved in parallel with improvement in the hypothalamic or pituitary hyper- or hyposecretion
 - 3. headache is associated with at least one of the following:
 - a) disorder of temperature regulation
 - b) abnormal emotional state
 - c) altered thirst and/or appetite
- D. Not better accounted for by another ICHD-3 diagnosis.

7.5 Headache attributed to intrathecal injection

Description:

Headache experienced in both upright and recumbent postures, caused by and occurring within 4 days of an intrathecal injection and remitting within 14 days.

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. An intrathecal injection has been given

C. Evidence of causation demonstrated by at least two of the following:

- 1. headache has developed within 4 days of the intrathecal injection
- 2. headache has significantly improved within 14 days after the intrathecal injection
- 3. signs of meningeal irritation

D. Not better accounted for by another ICHD-3 diagnosis.

Comments:

Headache usually develops within 4 days after intrathecal injection, and is present in both upright and recumbent postures.

When headache persists beyond 14 days, alternative diagnoses should be considered, such as 7.2.2 *CSF fistula headache*, meningitis or leptomenigeal disease.

7.6 Headache attributed to epileptic seizure

Coded elsewhere:

There is a complex and bidirectional association between migraine and epilepsy. Where the two coexist, without either being a risk factor for the other, migraine is coded under 1. *Migraine* according to its subtype. Where migraine is comorbid with certain forms of epilepsy, such as benign occipital epilepsy, benign rolandic epilepsy and corticoreticular epilepsy with absence seizures, again it is coded under 1. *Migraine* according to its subtype. Where migraine-like or other headache and epilepsy are both part of a specific brain disorder (e.g. MELAS), the headache is coded to that disorder. Where a seizure occurs during or immediately following a migraine aura (*migrainepsy*), it is coded as 1.4.4 *Migraine aura-triggered seizure*.

Description:

Headache caused by an epileptic seizure, occurring during and/or after the seizure and remitting spontaneously within hours or up to 3 days.

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. The patient is having or has recently had an epileptic seizure
- C. Evidence of causation demonstrated by both of the following:
 - 1. headache has developed simultaneously with onset of the seizure
 - 2. headache has resolved spontaneously after the seizure has terminated

D. Not better accounted for by another ICHD-3 diagnosis.

Comment:

Pre-ictal headache has also been evaluated in a small study of 11 patients with intractable focal epilepsy. Headache was frontotemporal, ipsilateral to the focus in nine patients with temporal lobe epilepsy (TLE) and contralateral in one with TLE and one with frontal lobe epilepsy. More studies are needed to establish the existence of pre-ictal headache, and determine its prevalence and clinical features, in patients with partial and generalized epilepsy.

7.6.1 Hemicrania epileptica

Description:

Headache occurring during a partial epileptic seizure, ipsilateral to the epileptic discharge, and remitting immediately or soon after the seizure has terminated.

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. The patient is having a partial epileptic seizure
- C. Evidence of causation demonstrated by both of the following:
 1. headache has developed simultaneously with onset of the partial seizure
 2. either or both of the following:
 - a) headache has significantly improved immediately after the partial seizure has terminated
 - b) headache is ipsilateral to the ictal discharge
- D. Not better accounted for by another ICHD-3 diagnosis.

7.6.2 Post-ictal headache

Description:

Headache caused by and occurring within 3 hours after an epileptic seizure, and remitting spontaneously within 72 hours after seizure termination.

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. The patient has recently had a partial or generalized epileptic seizure
- C. Evidence of causation demonstrated by both of the following:
 1. headache has developed within 3 hours after the epileptic seizure has terminated

2. headache has resolved within 72 hours after the epileptic seizure has terminated

D. Not better accounted for by another ICHD-3 diagnosis.

Comment:

7.6.2 *Post-ictal headache* occurs in over 40% of patients with either temporal lobe epilepsy or frontal lobe epilepsy and in up to 60% of patients with occipital lobe epilepsy. It occurs more frequently after generalized tonic-clonic seizures than other seizure types.

7.7 Headache attributed to Chiari malformation type I (CMI)

Description:

Headache caused by Chiari type I malformation, usually occipital or suboccipital, of short duration (less than 5 minutes) and provoked by cough or other Valsalva-like manœuvres. It remits after the successful treatment of the Chiari malformation.

Diagnostic criteria:

- A. Headache fulfilling criterion C
- B. Chiari malformation type I (CM1) has been demonstrated¹
- C. Evidence of causation demonstrated by at least two of the following:
 1. either or both of the following:
 - a) headache has developed in temporal relation to the CM1
 - b) headache has resolved within 3 months after successful treatment of the CM1
 2. headache has at least one of the following three characteristics:
 - a) precipitated by cough or other Valsalva-like manœuvre
 - b) occipital or suboccipital location
 - c) lasting <5 minutes
 3. headache is associated with other symptoms and/or clinical signs of brainstem, cerebellar, lower cranial nerve and/or cervical spinal cord dysfunction
- D. Not better accounted for by another ICHD-3 diagnosis.²

Notes:

1. Diagnosis of Chiari malformation by MRI requires a 5-mm caudal descent of the cerebellar tonsils or 3-mm caudal descent of the cerebellar tonsils plus crowding of the subarachnoid space at the

craniocervical junction as evidenced by compression of the CSF spaces posterior and lateral to the cerebellum, or reduced height of the supraocciput, or increased slope of the tentorium, or kinking of the medulla oblongata.

2. Patients with spontaneous intracranial hypotension secondary to CSF leak may demonstrate MRI evidence of secondary tonsillar descent and CM1. These patients may also present with headache related to cough or other Valsalva-like manoeuvre (and are correctly coded as 7.2.3 *Headache attributed to spontaneous intracranial hypotension*). Therefore, in all patients presenting with headache and CM1, CSF leak must be excluded.

Comments:

7.7 *Headache attributed to Chiari malformation type I (CM1)* is often descriptively similar to 4.1 *Primary cough headache* with the exception, sometimes, of longer duration (minutes rather than seconds).

Almost all (95%) patients with CM1 report a constellation of five or more distinct symptoms.

An MRI database showed tonsillar herniation of at least 5 mm in 0.7% of the population. *The clinical context of CM1 is important as many of these subjects can be asymptomatic.* Some patients exhibit ‘Chiari-like’ symptoms with minimal cerebellar tonsillar herniation, whereas others may be asymptomatic with large herniations. No correlation exists between the amount of herniation and the severity of headache or level of disability in presenting patients. Rigid adherence to the clinical and radiological criteria described above is recommended prior to surgical intervention, to avoid an unnecessary surgical procedure that has significant potential for surgical morbidity.

These criteria for 7.7 *Headache attributed to Chiari malformation type I (CM1)* require validation. Prospective studies with long-term surgical outcome are needed.

7.8 Headache attributed to other non-vascular intracranial disorder

Description:

Headache caused by a non-vascular intracranial disorder other than those described above.

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. A non-vascular intracranial disorder known to be able to cause headache, other than those described above, has been demonstrated

- C. Evidence of causation demonstrated by at least two of the following:
 1. headache has developed in temporal relation to the onset of the non-vascular intracranial disorder
 2. either or both of the following:
 - a) headache has developed or significantly worsened in parallel with worsening of the non-vascular intracranial disorder
 - b) headache has significantly improved in parallel with improvement of the non-vascular intracranial disorder
 3. headache has characteristics typical for the non-vascular intracranial disorder
 4. other evidence exists of causation

- D. Not better accounted for by another ICHD-3 diagnosis.

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8. Headache attributed to a substance or its withdrawal

- 8.1 Headache attributed to use of or exposure to a substance
 - 8.1.1 Nitric oxide (NO) donor-induced headache
 - 8.1.1.1 Immediate NO donor-induced headache
 - 8.1.1.2 Delayed NO donor-induced headache
 - 8.1.2 Phosphodiesterase (PDE) inhibitor-induced headache
 - 8.1.3 Carbon monoxide (CO)-induced headache
 - 8.1.4 Alcohol-induced headache
 - 8.1.4.1 Immediate alcohol-induced headache
 - 8.1.4.2 Delayed alcohol-induced headache
 - 8.1.5 Headache induced by food and/or additive
 - 8.1.5.1 Monosodium glutamate (MSG)-induced headache
 - 8.1.6 Cocaine-induced headache
 - 8.1.7 Histamine-induced headache
 - 8.1.7.1 Immediate histamine-induced headache
 - 8.1.7.2 Delayed histamine-induced headache
 - 8.1.8 Calcitonin gene-related peptide (CGRP)-induced headache
 - 8.1.8.1 Immediate CGRP-induced headache
 - 8.1.8.2 Delayed CGRP-induced headache
 - 8.1.9 Headache attributed to exogenous acute pressor agent
 - 8.1.10 Headache attributed to occasional use of non-headache medication
 - 8.1.11 Headache attributed to long-term use of non-headache medication
 - 8.1.12 Headache attributed to exogenous hormone
 - 8.1.13 Headache attributed to use of or exposure to other substance
- 8.2 Medication-overuse headache (MOH)
 - 8.2.1 Ergotamine-overuse headache
 - 8.2.2 Triptan-overuse headache
 - 8.2.3 Simple analgesic-overuse headache
 - 8.2.3.1 Paracetamol (acetaminophen)-overuse headache
 - 8.2.3.2 Acetylsalicylic acid-overuse headache
 - 8.2.3.3 Other non-steroidal anti-inflammatory drug (NSAID)-overuse headache
 - 8.2.4 Opioid-overuse headache
 - 8.2.5 Combination-analgesic-overuse headache
 - 8.2.6 Medication-overuse headache attributed to multiple drug classes not individually overused
 - 8.2.7 Medication-overuse headache attributed to unverified overuse of multiple drug classes
 - 8.2.8 Medication-overuse headache attributed to other medication
- 8.3 Headache attributed to substance withdrawal
 - 8.3.1 Caffeine-withdrawal headache
 - 8.3.2 Opioid-withdrawal headache
 - 8.3.3 Oestrogen-withdrawal headache
 - 8.3.4 Headache attributed to withdrawal from chronic use of other substance

Coded elsewhere:

7.1.2 Headache attributed to intracranial hypertension secondary to metabolic, toxic or hormonal causes; 7.3.2 Headache attributed to aseptic (non-infectious) meningitis.

General comment

Primary or secondary headache or both?

When a headache occurs for the first time in close temporal relation to exposure to or withdrawal from a substance, it is coded as a secondary headache attributed to exposure to or withdrawal from that substance. This remains true when the new headache has the characteristics of any of the primary headache disorders classified in Part one of ICHD-3 beta. When a *pre-existing* headache with the characteristics of a primary headache disorder becomes *chronic*, or is made significantly worse (usually meaning a two-fold or greater increase in frequency and/or severity), in close temporal relation to exposure to or withdrawal from a substance, both the initial headache diagnosis and a diagnosis of 8. *Headache attributed to a substance or its withdrawal* (or one of its subtypes) should be given, provided that there is good evidence that exposure to or withdrawal from that substance can cause headache.

Introduction

People with 1. *Migraine* are physiologically and perhaps psychologically hyperresponsive to a variety of internal and external stimuli. Alcohol, food and food additives, and chemical and drug ingestion and withdrawal, have all been reported to provoke or activate migraine in susceptible individuals. The association is often based on anecdotal data and reports of adverse drug reactions. The fact that these stimuli are associated with headache does not prove causation or eliminate the need to consider other aetiologies. Because common events happen commonly, the association between a headache and an exposure to a substance may be mere coincidence. Headache can occur on the basis of chance. Headache can be a symptom of a systemic disease, and drugs given to treat such a condition will be associated with headache. In acute migraine drug trials, headache, as well as associated symptoms, is listed as an adverse drug reaction despite that it is a symptom of the treated disorder and not the result of treatment. Some

disorders may predispose to drug-related headache. Alone, neither the drug nor the condition would produce headache.

The general criteria for the headache disorders listed here are:

- A. Headache fulfilling criterion C
- B. Use of or exposure to a substance known to be able to cause headache has occurred
- C. Evidence of causation demonstrated by two of the following:
 1. headache has developed in temporal relation to use of or exposure to the substance
 2. headache has significantly improved or resolved after removal of the substance
 3. headache has characteristics typical for use of or exposure to the substance
 4. other evidence exists of causation
- D. Not better accounted for by another ICHD-3 diagnosis.

8.1 Headache attributed to use of or exposure to a substance

Description:

Headache caused by use of or exposure to a substance, with onset immediately or within hours.

Comments:

8.1 *Headache attributed to use of or exposure to a substance* can be caused by a toxic substance, as an unwanted effect of a substance in normal therapeutic use or in experimental studies.

Headache as a side effect has been recorded with many drugs, often merely reflecting the high prevalence of headache. Only when it occurs more often after an active drug than after placebo in double-blind controlled trials can headache be regarded as a true side effect. The double-blind design can also be used experimentally to study the relationship between drug effects and headache. In some cases, for example nitric oxide (NO) donors, such studies have led to a deeper understanding of the involvement of neurotransmitter mechanisms in primary headaches.

In general, people with 1. *Migraine* are much more susceptible to such headaches than other individuals, and the same may be true for people with 2. *Tension-type headache* or 3.1 *Cluster headache*. A number of substances, such as NO donors and histamine, induce an immediate headache in both normal volunteers and in migraineurs. However, it is now clear that people who have primary headache disorders also develop a delayed headache, one to several hours after the substance has been cleared from the blood. Knowledge of

the potential headache-inducing effects of substances in clinical use is important in order to label these substances appropriately. Combinations such as alcohol and disulfiram may cause headache when individual agents might not.

Paradoxically, the headache encountered by most people after heavy alcohol use may be a positive feature because it encourages avoidance of excessive drinking.

Substances that cause headache through their toxic effects, such as carbon monoxide, cannot be studied experimentally and the causal relationship between exposure and headache has therefore to be demonstrated in clinical cases where the substance has been used accidentally or for suicide attempt.

8.1.1 Nitric oxide (NO) donor-induced headache

Description:

Headache caused immediately, or after a delay, by acute exposure to a nitric oxide donor.

Comments:

8.1.1 *Nitric oxide (NO) donor-induced headache* is typically frontotemporal and pulsating. All NO donors (e.g. amyl nitrate, erythryl tetranitrate, pentaerythryl tetranitrate, glyceryl trinitrate [GTN], isosorbide mono- or dinitrate, sodium nitroprusside, mannitol hexanitrate) can cause headache of this subform.

GTN induces immediate headache in most normal people, but can also cause a delayed headache in migraineurs which fulfils the diagnostic criteria for 1.1 *Migraine without aura*. In people with 2.3 *Chronic tension-type headache*, GTN has been shown to induce a delayed headache which has the characteristics of 2. *Tension-type headache* (the effect is unknown in those with 2.1 *Infrequent episodic tension-type headache* or 2.2 *Frequent episodic tension-type headache*). These delayed headaches occur, on average, 5–6 hours after exposure. People with 3. *Cluster headache* develop delayed headache only during cluster periods: GTN usually induces a cluster headache attack 1–2 hours after intake.

Headache is a side effect of therapeutic use of nitroglycerine. With chronic use, tolerance develops within a week, and GTN-induced headache disappears in most patients within that time. Other NO donors used therapeutically may also produce headache. Isosorbide mononitrate has been the subject of one formal double-blind placebo-controlled study, and causes a much longer-lasting headache than GTN owing to its slow release of NO.

8.1.1.1 Immediate NO donor-induced headache

Previously used terms:

Nitroglycerine headache; dynamite headache; hot dog headache.

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. Absorption of a nitric oxide (NO) donor has occurred
- C. Evidence of causation demonstrated by all of the following:
 1. headache has developed within 1 hour after absorption of the NO donor
 2. headache has resolved within 1 hour after release of NO has ended
 3. headache has at least one of the following four characteristics:
 - a) bilateral
 - b) mild to moderate intensity
 - c) pulsating quality
 - d) aggravated by physical activity
- D. Not better accounted for by another ICHD-3 diagnosis.

8.1.1.2 Delayed NO donor-induced headache

Diagnostic criteria:

- A. Headache, in a person affected by a primary headache disorder and with the characteristics of that headache type, fulfilling criterion C
- B. Absorption of a nitric oxide (NO) donor has occurred
- C. Evidence of causation demonstrated by both of the following:
 1. headache has developed within 2–12 hours after exposure to the NO donor, and after NO is cleared from the blood
 2. headache has resolved within 72 hours after exposure
- D. Not better accounted for by another ICHD-3 diagnosis.

8.1.2 Phosphodiesterase (PDE) inhibitor-induced headache

Description:

Headache caused by intake of a phosphodiesterase inhibitor, resolving spontaneously within 72 hours.

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. A single dose of a phosphodiesterase (PDE) inhibitor has been taken
- C. Evidence of causation demonstrated by all of the following:
 1. headache has developed within 5 hours of intake of the PDE inhibitor
 2. headache has resolved within 72 hours of onset
 3. headache has at least one of the following four characteristics:
 - a) bilateral
 - b) mild to moderate intensity
 - c) pulsating quality
 - d) aggravated by physical activity
- D. Not better accounted for by another ICHD-3 diagnosis.

Comment:

Phosphodiesterases (PDEs) are enzymes that break down cGMP and cAMP. The PDE-5 inhibitors, sildenafil and dipyridamole, increase levels of cGMP and/or cAMP. The resultant headache usually has the characteristics of tension-type headache, but in people with 1.1 *Migraine* (who should be warned of this side effect) it has the characteristics of 1.1 *Migraine without aura*.

8.1.3 Carbon monoxide (CO)-induced headache

Previously used term:

Warehouse workers' headache.

Description:

Headache caused by exposure to carbon monoxide, resolving spontaneously within 72 hours after its elimination.

Diagnostic criteria:

- A. Bilateral headache fulfilling criterion C
- B. Exposure to carbon monoxide (CO) has occurred
- C. Evidence of causation demonstrated by all of the following:
 1. headache has developed within 12 hours of exposure to CO
 2. headache intensity varies with the severity of CO intoxication
 3. headache has resolved within 72 hours of elimination of CO
- D. Not better accounted for by another ICHD-3 diagnosis.

Comments:

Typically, carboxyhaemoglobin levels of 10–20% cause a mild headache without gastrointestinal or neurological symptoms, levels of 20–30% cause a moderate pulsating headache and irritability, and levels of 30–40% cause a severe headache with nausea, vomiting and blurred vision. At levels above 40%, headache is usually not a complaint because of the change in consciousness.

There are no good studies of the long-term effects of CO intoxication on headache, but there is some evidence of chronic post-CO intoxication headache.

*8.1.4 Alcohol-induced headache**Description:*

Headache caused immediately, or after a delay, by ingestion of alcohol (usually in the form of alcoholic beverages).

*8.1.4.1 Immediate alcohol-induced headache**Previously used term:*

Cocktail headache.

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. Alcohol has been ingested
- C. Evidence of causation demonstrated by all of the following:
 1. headache has developed within 3 hours of alcohol ingestion
 2. headache has resolved within 72 hours after alcohol ingestion has ceased
 3. headache has at least one of the following three characteristics:
 - a) bilateral
 - b) pulsating quality
 - c) aggravated by physical activity
- D. Not better accounted for by another ICHD-3 diagnosis.

Comment:

The effective dose of alcohol to cause 8.1.4.1 *Immediate alcohol-induced headache* is variable, and can be very small in people with 1. *Migraine* (who, at other times, may tolerate alcohol at the same level as non-migraineurs). 8.1.4.1 *Immediate alcohol-induced headache* is much rarer than 8.1.4.2 *Delayed alcohol-induced headache*.

*8.1.4.2 Delayed alcohol-induced headache**Previously used term:*

Hangover headache.

Description:

Headache caused, after a delay of hours, by ingestion of alcohol (usually in the form of alcoholic beverages). It resolves spontaneously within 72 hours.

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. Alcohol has been ingested
- C. Evidence of causation demonstrated by all of the following:
 1. headache has developed within 5–12 hours after ingestion of alcohol
 2. headache has resolved within 72 hours of onset
 3. headache has at least one of the following three characteristics:
 - a) bilateral
 - b) pulsating quality
 - c) aggravated by physical activity
- D. Not better accounted for by another ICHD-3 diagnosis.

Comment:

8.1.4.2 *Delayed alcohol-induced headache* is one of the commonest types of secondary headache. It remains unclear whether additional substances play a role, such as nicotine in cigarette smoke, which is often inhaled at the same time as alcohol ingestion. Whether the delayed headache is a toxic effect or a manifestation of mechanisms similar to those in 8.1.1.2 *Delayed NO donor-induced headache* is an unresolved question.

*8.1.5 Headache induced by food and/or additive**Previously used term:*

Dietary headache.

Coded elsewhere:

An episode of migraine triggered by a specific food or additive is coded as the appropriate subtype of 1. *Migraine*.

Description:

Headache caused by ingestion of a food or an additive containing one or more specific substances, which may not be identified, to which the patient is sensitive.

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. A food or an additive containing one or more specific substances, not necessarily identified but

capable of causing headache in sensitive subjects, has been ingested

- C. Evidence of causation demonstrated by all of the following:
 1. headache has developed within 12 hours of ingestion of the food or additive
 2. headache has resolved within 72 hours after ingestion of the food or additive
 3. headache has at least one of the following four characteristics:
 - a) bilateral
 - b) mild to moderate intensity
 - c) pulsating quality
 - d) aggravated by physical activity
- D. Not better accounted for by another ICHD-3 diagnosis.

Comment:

Monosodium glutamate, which is a well-established cause of headache, has a separate subcoding below. Phenylethylamine, tyramine and aspartame have been incriminated, but without conclusive evidence.

8.1.5.1 Monosodium glutamate (MSG)-induced headache

Previously used term:

Chinese restaurant syndrome.

Coded elsewhere:

An episode of migraine triggered by monosodium glutamate ingestion is coded as the appropriate subtype of 1. *Migraine*.

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. Monosodium glutamate (MSG) has been ingested
- C. Evidence of causation demonstrated by all of the following:
 1. headache has developed within 1 hour of MSG ingestion
 2. headache has resolved within 72 hours after MSG ingestion
 3. headache has at least one of the following five characteristics:
 - a) bilateral
 - b) mild to moderate intensity
 - c) pulsating quality
 - d) associated with flushing of the face, pressure in the face and chest, burning sensations in the neck, shoulders and/or chest, dizziness and abdominal discomfort
 - e) aggravated by physical activity

- D. Not better accounted for by another ICHD-3 diagnosis.

Comment:

8.1.5.1 *Monosodium glutamate (MSG)-induced headache* is typically pressing/tightening or burning in quality, but may be pulsating in people with 1. *Migraine*. It is commonly associated with flushing of the face, pressure in the face and chest, burning sensations in the neck, shoulders and/or chest, dizziness and abdominal discomfort.

8.1.6 Cocaine-induced headache

Description:

Headache caused by administration of cocaine by any route.

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. Cocaine has been administered by any route
- C. Evidence of causation demonstrated by all of the following:
 1. headache has developed within 1 hour of cocaine administration
 2. headache has resolved within 72 hours after cocaine administration
 3. headache has at least one of the following four characteristics:
 - a) bilateral
 - b) mild to moderate intensity
 - c) pulsating quality
 - d) aggravated by physical activity
- D. Not better accounted for by another ICHD-3 diagnosis.

Comment:

The principal routes of cocaine administration are oral ('chewing'), intranasal ('snorting'), intravenous ('mainlining') and inhalation (smoking).

8.1.7 Histamine-induced headache

Description:

Headache caused immediately, or after a delay, by acute exposure to histamine.

Comments:

Histamine has similar effect whether administered subcutaneously, by inhalation or intravenously. The mechanism is primarily mediated via the H1 receptor, and is almost completely blocked by mepyramine.

Histamine causes an immediate headache in most people, but can also cause a delayed headache in migraineurs, which fulfils the diagnostic criteria for 1.1 *Migraine without aura*. In people with 2. *Tension-type headache*, histamine may induce a delayed headache which has the characteristics of that disorder. These delayed headaches occur, on average, 5–6 hours after exposure. People with 3. *Cluster headache* develop delayed headache with the characteristics of that disorder only during cluster periods, usually 1–2 hours after exposure.

8.1.7.1 Immediate histamine-induced headache

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. Histamine has been administered
- C. Evidence of causation demonstrated by all of the following:
 1. headache has developed within 1 hour of histamine absorption
 2. headache has resolved within 1 hour after absorption of histamine has ceased
 3. headache has at least one of the following four characteristics:
 - a) bilateral
 - b) mild to moderate intensity
 - c) pulsating quality
 - d) aggravated by physical activity
- D. Not better accounted for by another ICHD-3 diagnosis.

8.1.7.2 Delayed histamine-induced headache

Diagnostic criteria:

- A. Headache, in a person affected by a primary headache disorder and with the characteristics of that headache type, fulfilling criterion C
- B. Histamine has been administered
- C. Evidence of causation demonstrated by both of the following:
 1. headache has developed within 2–12 hours after administration of histamine
 2. headache has resolved within 72 hours after administration of histamine
- D. Not better accounted for by another ICHD-3 diagnosis.

8.1.8 Calcitonin gene-related peptide (CGRP)-induced headache

Description:

Headache caused immediately, or after a delay, by acute exposure to calcitonin gene-related peptide (CGRP).

Comments:

Calcitonin gene-related peptide (CGRP), administered by infusion, causes an immediate headache. It can also cause a delayed headache in migraineurs, on average 5–6 hours after exposure, which fulfils the diagnostic criteria for 1.1 *Migraine without aura*.

The CGRP antagonist, telcagepant, is effective in the acute treatment of migraine.

8.1.8.1 Immediate CGRP-induced headache

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. Calcitonin gene-related peptide (CGRP) has been administered
- C. Evidence of causation demonstrated by all of the following:
 1. headache has developed within 1 hour of CGRP absorption
 2. headache has resolved within 1 hour after absorption of CGRP has ceased
 3. headache has at least one of the following four characteristics:
 - a) bilateral
 - b) mild to moderate intensity
 - c) pulsating quality
 - d) aggravated by physical activity
- D. Not better accounted for by another ICHD-3 diagnosis.

8.1.8.2 Delayed CGRP-induced headache

Diagnostic criteria:

- A. Headache, in a person affected by a primary headache disorder and with the characteristics of that headache type, fulfilling criterion C
- B. Calcitonin gene-related peptide (CGRP) has been administered
- C. Evidence of causation demonstrated by both of the following:
 1. headache has developed within 2–12 hours after administration of CGRP
 2. headache has resolved within 72 hours after administration of CGRP has ceased

D. Not better accounted for by another ICHD-3 diagnosis.

8.1.9 Headache attributed to exogenous acute pressor agent

Description:

Headache occurring during, and caused by, an acute rise in blood pressure induced by an exogenous pressor agent.

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. An acute rise in blood pressure has followed administration of an exogenous pressor agent
- C. Evidence of causation demonstrated by both of the following:
 - 1. headache has occurred within 1 hour of administration of the pressor agent
 - 2. headache has resolved within 72 hours after administration of the pressor agent has ceased
- D. Not better accounted for by another ICHD-3 diagnosis.

8.1.10 Headache attributed to occasional use of non-headache medication

Description:

Headache occurring as an acute adverse event after occasional use of a medication taken for purposes other than the treatment of headache.

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. One or more doses of medication have been taken for purposes other than the treatment of headache
- C. Evidence of causation demonstrated by both of the following:
 - 1. headache has developed within minutes to hours of intake
 - 2. headache has resolved within 72 hours after intake has ceased
- D. Not better accounted for by another ICHD-3 diagnosis.

Comments:

8.1.10 *Headache attributed to occasional use of non-headache medication* has been reported as an adverse event after use of many drugs. The following are the most commonly incriminated: atropine, digitalis,

disulfiram, hydralazine, imipramine, nicotine, nifedipine, nimodipine.

The headache characteristics are not very well defined in the literature, and probably depend on the drug, but in most cases headache is dull, continuous, diffuse and of moderate to severe intensity.

8.1.11 Headache attributed to long-term use of non-headache medication

Coded elsewhere:

Headache developing as an adverse event during hormone therapy is coded as 8.1.12 *Headache attributed to exogenous hormone*. Headache developing as a complication of long-term overuse of acute headache medication by a person with a headache disorder is coded as 8.2 *Medication-overuse headache* or one of its subtypes.

Description:

Headache developing as an adverse event during long-term use of a medication taken for purposes other than the treatment of headache, and not necessarily reversible.

Diagnostic criteria:

- A. Headache present on ≥ 15 days per month and fulfilling criterion C
- B. Long-term use of a medication taken for purposes other than the treatment of headache
- C. Evidence of causation demonstrated by at least two of the following:
 - 1. headache has developed in temporal relation to the commencement of medication intake
 - 2. one or more of the following:
 - a) headache has significantly worsened after an increase in dosage of the medication
 - b) headache has significantly improved or resolved after a reduction in dosage of the medication
 - c) headache has resolved after cessation of the medication
 - 3. the medication is recognized to cause headache, in at least some people, during long-term use
- D. Not better accounted for by another ICHD-3 diagnosis.

Comments:

The dosage and duration of exposure that may result in headache during long-term use varies from medication to medication. Similarly, the time required for resolution varies – if the effect is reversible.

8.1.11 *Headache attributed to long-term use of non-headache medication* can be a result of direct

pharmacological effect of the medication, such as vasoconstriction producing malignant hypertension, or to a secondary effect such as drug-induced intracranial hypertension. The latter is a recognized complication of long-term use of anabolic steroids, amiodarone, lithium carbonate, nalidixic acid, thyroid hormone replacement therapy, tetracycline and minocycline.

8.1.12 Headache attributed to exogenous hormone

Description:

Headache developing as an adverse event during regular intake of exogenous hormones, usually for contraception or as hormone replacement therapy.

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. Regular intake of one or more exogenous hormones
- C. Evidence of causation demonstrated by both of the following:
 1. headache has developed in temporal relation to the commencement of hormone intake
 2. one or more of the following:
 - a) headache has significantly worsened after an increase in dosage of the hormone
 - b) headache has significantly improved or resolved after a reduction in dosage of the hormone
 - c) headache has resolved after cessation of hormone intake
- D. Not better accounted for by another ICHD-3 diagnosis.

Comments:

Regular use of exogenous hormones, typically for contraception or hormone replacement therapy, can be associated with an increase in frequency or new development of migraine or other headache. The general rule is applied that when a headache occurs for the first time in close temporal relation to regular use of exogenous hormones, it is coded as 8.1.12 *Headache attributed to exogenous hormone*. When a pre-existing headache with the characteristics of a primary headache disorder becomes chronic, or is made significantly worse (usually meaning a two-fold or greater increase in frequency and/or severity), in close temporal relation to regular use of exogenous hormones, both the initial headache diagnosis and a diagnosis of 8.1.12 *Headache attributed to exogenous hormone* should be given.

When a woman with 8.1.12 *Headache attributed to exogenous hormone* also experiences 8.3.3 *Oestrogen withdrawal headache*, both diagnoses should be given.

8.1.13 Headache attributed to use of or exposure to other substance

Description:

Headache occurring during or soon after, and caused by, use of or exposure to a substance other than those described above, including herbal, animal or other organic or inorganic substances given by physicians or non-physicians with medicinal intent although not licensed as medicinal products.

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. Exposure to a substance other than those described above
- C. Evidence of causation demonstrated by both of the following:
 1. headache has developed within 12 hours of exposure
 2. headache has resolved within 72 hours after exposure
- D. Not better accounted for by another ICHD-3 diagnosis.

Comments:

8.1.13 *Headache attributed to use of or exposure to other substance* includes headache caused by herbal, animal or other organic or inorganic substances given by physicians or non-physicians with medicinal intent although not licensed as medicinal products.

8.1.13 *Headache attributed to use of or exposure to other substance* has been reported after exposure to a number of other organic and inorganic substances. The following are most commonly incriminated:

Inorganic compounds:

arsenic, borate, bromate, chlorate, copper, iodine, lead, lithium, mercury, tolazoline hydrochloride.

Organic compounds:

aniline, balsam, camphor, carbon disulfide, carbon tetrachloride, clordecane, EDTA, heptachlor, hydrogen sulfide, kerosene, long-chain alcohols, methyl alcohol, methyl bromide, methyl chloride, methyl iodine, naphthalene, organophosphorous compounds (parathion, pyrethrum).

The characteristics of 8.1.13 *Headache attributed to use of or exposure to other substance* are not well defined in the literature, and almost certainly vary with the agent. In most cases it is dull, diffuse, continuous and of moderate to severe intensity.

8.2 Medication-overuse headache (MOH)

Previously used terms:

Rebound headache; drug-induced headache; medication-misuse headache.

Coded elsewhere:

Patients with a pre-existing primary headache who, in association with medication overuse, develop a new type of headache or a marked worsening of their pre-existing headache that, in either case, meets the criteria for 8.2 *Medication-overuse headache* (or one of its subtypes), should be given both this diagnosis and the diagnosis of the pre-existing headache. Patients who meet criteria for both 1.3 *Chronic migraine* and 8.2 *Medication-overuse headache* should be given both diagnoses.

Description:

Headache occurring on 15 or more days per month developing as a consequence of regular overuse of acute or symptomatic headache medication (on 10 or more, or 15 or more days per month, depending on the medication) for more than 3 months. It usually, but not invariably, resolves after the overuse is stopped.

General comment:

In the criteria set out below for the various subtypes, the specified numbers of days of medication use considered to constitute overuse are based on expert opinion rather than on formal evidence.

Diagnostic criteria:

- A. Headache occurring on ≥ 15 days per month in a patient with a pre-existing headache disorder
- B. Regular overuse for >3 months of one or more drugs that can be taken for acute and/or symptomatic treatment of headache¹
- C. Not better accounted for by another ICHD-3 diagnosis.

Note:

- 1. Patients should be coded for one or more subtypes of 8.2 *Medication-overuse headache* according to the specific medication(s) overused and the criteria for each below. For example, a patient who fulfils the criteria for 8.2.2 *Triptan-overuse headache* and the criteria for one of the subforms of 8.2.3 *Simple analgesic-overuse headache* should receive both these codes. The exception occurs when patients overuse combination-analgesic medications, who are coded 8.2.5 *Combination-analgesic-overuse*

headache and not according to each constituent of the combination-analgesic medication.

Patients who use multiple drugs for acute or symptomatic treatment of headache may do so in a manner that constitutes overuse even though no individual drug or class of drug is overused; such patients should be coded 8.2.6 *Medication-overuse headache attributed to multiple drug classes not individually overused*.

Patients who are clearly overusing multiple drugs for acute or symptomatic treatment of headache but cannot give an adequate account of their names and/or quantities are coded 8.2.7 *Medication-overuse headache attributed to unverified overuse of multiple drug classes* until better information is available. In almost all cases, this necessitates diary follow-up.

Comments:

8.2 *Medication-overuse headache* is an interaction between a therapeutic agent used excessively and a susceptible patient. Among those with a previous primary headache diagnosis, most have 1. *Migraine* or 2. *Tension-type headache* (or both); only a small minority have other primary headache diagnoses such as 3.3 *Chronic cluster headache* or 4.10 *New daily persistent headache*.

The diagnosis of 8.2 *Medication-overuse headache* is extremely important clinically. Approximately half of people with headache on 15 or more days per month for more than 3 months have 8.2 *Medication-overuse headache*. Evidence shows that the majority of patients with this disorder improve after discontinuation of the overused medication, as does their responsiveness to preventative treatment. Simple advice on the causes and consequences of 8.2 *Medication-overuse headache* is an essential part of its management. An explanatory brochure is often all that is necessary to prevent or discontinue medication overuse. Prevention is especially important in patients prone to frequent headache.

However, the behaviour of some patients with 8.2 *Medication-overuse headache* is similar to that seen with other drug addictions, and the Severity of Dependence Scale (SDS) score is a significant predictor of medication overuse among headache patients.

8.2.1 Ergotamine-overuse headache

Diagnostic criteria:

- A. Headache fulfilling criteria for 8.2 *Medication-overuse headache*
- B. Regular intake of ergotamine on ≥ 10 days per month for >3 months.

Comments:

Bioavailability of ergots is so variable that a minimum dose cannot be defined.

A patient who fulfils the criteria for 8.2.1 *Ergotamine-overuse headache* and has regularly used or overused other drug(s) for the acute or symptomatic treatment of headache for more than 3 months should be given all other applicable codes.

8.2.2 *Triptan-overuse headache**Diagnostic criteria:*

- A. Headache fulfilling criteria for 8.2 *Medication-over-use headache*
- B. Regular intake of one or more triptans,¹ in any formulation, on ≥ 10 days per month for >3 months.

Note:

1. The triptan(s) will usually be specified in parenthesis.

Comments:

Triptan overuse may increase migraine frequency to that of 1.3 *Chronic migraine*. Evidence suggests that this occurs sooner with triptan overuse than with ergotamine overuse.

A patient who fulfils the criteria for 8.2.2 *Triptan-overuse headache* and has regularly used or overused other drug(s) for the acute or symptomatic treatment of headache for more than 3 months should be given all other applicable codes.

8.2.3 *Simple analgesic-overuse headache**Comment:*

A patient who fulfils the criteria for 8.2.3 *Simple analgesic-overuse headache* (or one of its subtypes) and has regularly used or overused other drug(s) for the acute or symptomatic treatment of headache for more than 3 months should be given all other applicable codes.

8.2.3.1 *Paracetamol (acetaminophen)-overuse headache**Diagnostic criteria:*

- A. Headache fulfilling criteria for 8.2 *Medication-over-use headache*
- B. Regular intake of paracetamol on ≥ 15 days per month for >3 months.

8.2.3.2 *Acetylsalicylic acid-overuse headache**Diagnostic criteria:*

- A. Headache fulfilling criteria for 8.2 *Medication-over-use headache*
- B. Regular intake of acetylsalicylic acid on ≥ 15 days per month for >3 months.

8.2.3.3 *Other non-steroidal anti-inflammatory drug (NSAID)-overuse headache**Diagnostic criteria:*

- A. Headache fulfilling criteria for 8.2 *Medication-over-use headache*
- B. Regular intake of one or more NSAIDs¹ other than acetylsalicylic acid on ≥ 15 days per month for >3 months.

Note:

1. The NSAID(s) will usually be specified in parenthesis.

8.2.4 *Opioid-overuse headache**Diagnostic criteria:*

1. Headache fulfilling criteria for 8.2 *Medication-over-use headache*
2. Regular intake of one or more opioids¹ on ≥ 10 days per month for >3 months.

Note:

1. The opioid(s) will usually be specified in parenthesis.

Comments:

Prospective studies indicate that patients overusing opioids have the highest relapse rate after withdrawal treatment.

A patient who fulfils the criteria for 8.2.4 *Opioid-overuse headache* (or one of its subtypes) and has regularly used or overused other drug(s) for the acute or symptomatic treatment of headache for more than 3 months should be given all other applicable codes.

8.2.5 *Combination-analgesic-overuse¹ headache*

Diagnostic criteria:

- A. Headache fulfilling criteria for 8.2 *Medication-overuse headache*
- B. Regular intake of one or more combination-analgesic medications^{1,2} on ≥ 10 days/month for >3 months.

Notes:

- 1. The term *combination-analgesic* is used specifically for formulations combining drugs of two or more classes, each with analgesic effect or acting as adjuvants.
- 2. The combination-analgesic(s) will usually be specified in parenthesis.

Comments:

Many combination-analgesics are marketed. They tend to be widely used by people with headache, and are very commonly implicated in medication-overuse headache. For this reason, 8.2.5 *Combination-analgesic-overuse headache* has a separate coding.

The most commonly overused combination-analgesics are tablets combining simple analgesics with opioids, butalbital and/or caffeine.

8.2.6 *Medication-overuse headache attributed to multiple drug classes not individually overused*

Diagnostic criteria:

- A. Headache fulfilling criteria for 8.2 *Medication-overuse headache*
- B. Regular intake of any combination of ergotamine, triptans, simple analgesics, NSAIDs and/or opioids¹ on a total of ≥ 10 days per month for >3 months without overuse of any single drug or drug class alone.²

Notes:

- 1. The drugs or drug classes will usually be specified in parenthesis.
- 2. 'Without overuse of any single drug or drug class alone' means criterion B has not been fulfilled for any of the specific subforms 8.2.1–8.2.5.

8.2.7 *Medication-overuse headache attributed to unverified overuse of multiple drug classes*

Diagnostic criteria:

- A. Headache fulfilling criteria for 8.2 *Medication-overuse headache*
- B. Both of the following:
 - 1. regular intake of any combination of ergotamine, triptans, simple analgesics, NSAIDs and/or opioids on ≥ 10 days per month for >3 months
 - 2. the identity, quantity and/or pattern of use or overuse of these classes of drug cannot be reliably established.

Comment:

Patients who are clearly overusing multiple medications for acute or symptomatic treatment of headache, but cannot give an accurate account of what, when or how much, are encountered not uncommonly. Although a prospective diary record over several weeks might provide the information, it would also delay withdrawal, which is clearly required.

8.2.8 *Medication-overuse headache attributed to other medication*

Diagnostic criteria:

- A. Headache fulfilling criteria for 8.2 *Medication-overuse headache*
- B. Regular overuse, on ≥ 10 days per month for >3 months, of one or more medications other than those described above,¹ taken for acute or symptomatic treatment of headache.

Note:

- 1. The medication(s) will usually be specified in parenthesis.

8.3 *Headache attributed to substance withdrawal*

Description:

Headache following, and caused by, withdrawal from exposure to a medication or other substance.

8.3.1 *Caffeine-withdrawal headache*

Description:

Headache developing within 24 hours after regular consumption of caffeine in excess of 200 mg/day for more

than 2 weeks, which has been interrupted. It resolves spontaneously within 7 days in the absence of further consumption.

Diagnostic criteria:

- A. Headache fulfilling criterion C
- B. Caffeine consumption of >200 mg/day for >2 weeks, which has been interrupted or delayed
- C. Evidence of causation demonstrated by both of the following:
 1. headache has developed within 24 hours after last caffeine intake
 2. either or both of the following:
 - a) headache is relieved within 1 hour by intake of caffeine 100 mg
 - b) headache has resolved within 7 days after total caffeine withdrawal
- D. Not better accounted for by another ICHD-3 diagnosis.

8.3.2 Opioid-withdrawal headache

Description:

Headache developing within 24 hours after daily consumption of opioid(s) for more than 3 months, which has been interrupted. It resolves spontaneously within 7 days in the absence of further consumption.

Diagnostic criteria:

- A. Headache fulfilling criterion C
- B. Opioid intake daily for >3 months, which has been interrupted
- C. Evidence of causation demonstrated by both of the following:
 1. headache has developed within 24 hours after last opioid intake
 2. headache has resolved within 7 days after total opioid withdrawal
- D. Not better accounted for by another ICHD-3 diagnosis.

8.3.3 Oestrogen-withdrawal headache

Description:

Headache or migraine developing within 5 days after daily consumption of exogenous oestrogen for 3 weeks or longer, which has been interrupted (usually during the pill-free interval of combined oral contraception or following a course of replacement or supplementary oestrogen). It resolves

spontaneously within 3 days in the absence of further consumption.

Diagnostic criteria:

- A. Headache or migraine fulfilling criterion C
- B. Daily use of exogenous oestrogen for ≥ 3 weeks, which has been interrupted
- C. Evidence of causation demonstrated by both of the following:
 1. headache or migraine has developed within 5 days after the last use of oestrogen
 2. headache or migraine has resolved within 3 days of its onset
- D. Not better accounted for by another ICHD-3 diagnosis.

Comment:

Oestrogen-withdrawal following cessation of a course of exogenous estrogens (such as during the pill-free interval of combined oral contraceptives or following a course of replacement or supplementary oestrogen) can induce headache and/or migraine.

8.3.4 Headache attributed to withdrawal from chronic use of other substance

Description:

Headache following, and caused by, withdrawal from chronic use of or exposure to a medication or substance other than those described above.

Diagnostic criteria:

- A. Headache fulfilling criterion C
- B. Daily intake of a substance other than those described above for >3 months, which has been interrupted
- C. Evidence of causation demonstrated by both of the following:
 1. headache has developed in close temporal relation to withdrawal from use of the substance
 2. headache has resolved within 3 months after total withdrawal from use of the substance
- D. Not better accounted for by another ICHD-3 diagnosis.

Comment:

It has been suggested, but without sufficient evidence, that withdrawal from chronic use of the following substances may cause headache: corticosteroids, tricyclic antidepressants, selective serotonin reuptake inhibitors

(SSRIs), non-steroidal anti-inflammatory drugs (NSAIDs).

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8.3 Headache attributed to substance withdrawal

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9. Headache attributed to infection

- 9.1 Headache attributed to intracranial infection
 - 9.1.1 Headache attributed to bacterial meningitis or meningoencephalitis
 - 9.1.1.1 Acute headache attributed to bacterial meningitis or meningoencephalitis
 - 9.1.1.2 Chronic headache attributed to bacterial meningitis or meningoencephalitis
 - 9.1.1.3 Persistent headache attributed to past bacterial meningitis or meningoencephalitis
 - 9.1.2 Headache attributed to viral meningitis or encephalitis
 - 9.1.2.1 Headache attributed to viral meningitis
 - 9.1.2.2 Headache attributed to viral encephalitis
 - 9.1.3 Headache attributed to intracranial fungal or other parasitic infection
 - 9.1.3.1 Acute headache attributed to intracranial fungal or other parasitic infection
 - 9.1.3.2 Chronic headache attributed to intracranial fungal or other parasitic infection
 - 9.1.4 Headache attributed to brain abscess
 - 9.1.5 Headache attributed to subdural empyema
- 9.2 Headache attributed to systemic infection
 - 9.2.1 Headache attributed to systemic bacterial infection
 - 9.2.1.1 Acute headache attributed to systemic bacterial infection
 - 9.2.1.2 Chronic headache attributed to systemic bacterial infection
 - 9.2.2 Headache attributed to systemic viral infection
 - 9.2.2.1 Acute headache attributed to systemic viral infection
 - 9.2.2.2 Chronic headache attributed to systemic viral infection
 - 9.2.3 Headache attributed to other systemic infection
 - 9.2.3.1 Acute headache attributed to other systemic infection
 - 9.2.3.2 Chronic headache attributed to other systemic infection

Coded elsewhere:

Headache disorders attributed to extracranial infections of the head (such as ear, eye and sinus infections) are coded as subtypes of 11. *Headache or facial pain attributed to disorder of the cranium, neck, eyes, ears, nose, sinuses, teeth, mouth or other facial or cervical structure.*

General comment

Primary or secondary headache or both?

When a headache occurs for the first time in close temporal relation to an infection, it is coded as a secondary headache attributed to that infection. This remains true when the new headache has the characteristics of any of the primary headache disorders classified in Part one of ICHD-3 beta. When a *pre-existing* headache with the characteristics of a primary headache disorder becomes *chronic*, or is made significantly worse (usually meaning a two-fold or greater increase in frequency and/or severity), in close temporal relation to an infection, both the initial headache diagnosis and a diagnosis of 9. *Headache attributed to infection* (or one of its subtypes) should be given, provided that there is good evidence that that infection can cause headache.

Acute, chronic or persistent?

9. *Headache attributed to infection* is usually the consequence of active infection, resolving within 3 months of eradication of the infection. In some cases, depending on the pathogenic agent, the infection cannot be treated effectively and remains active. The headache in these cases may not abate, because the cause remains present, and after 3 months is referred to as *chronic*. In other, rarer cases, the infection resolves or is eradicated but the headache does not remit; after 3 months, such headache is termed *persistent* (in keeping with other secondary headaches). Accordingly, *acute* and *chronic* subforms of headache attributed to *active or recent* infection have been defined, in some cases in contrast to *persistent* subforms of *post-infectious* headache (see for example 9.1.1.1 *Acute headache attributed to bacterial meningitis or meningoencephalitis*, 9.1.1.2 *Chronic headache attributed to bacterial meningitis or meningoencephalitis* and 9.1.1.3 *Persistent headache attributed to past bacterial meningitis or meningoencephalitis*). The purpose is to distinguish and keep separate two probably different causative mechanisms and two different management approaches.

Introduction

Headache is a common accompaniment of systemic viral infections such as influenza. It is also common with sepsis; more rarely it may accompany other systemic infections.

In intracranial infections, headache is usually the first and the most frequently encountered symptom. Occurrence of a new type of headache which is diffuse and associated with focal neurological signs and/or altered mental state, a general feeling of illness and/or fever should direct attention towards an intracranial

infection even in the absence of neck stiffness. Unfortunately, there are no good prospective studies of the headaches associated with intracranial infection and the diagnostic criteria for some of the subtypes of 9.1 *Headache attributed to intracranial infection* are at least partly reliant on expert consensus (including the views of experts in neuroinfection) when evidence is lacking.

The general criteria for this chapter, adhered to as far as possible, are as follows:

- A. Headache fulfilling criterion C
- B. An infection, or sequela of an infection, known to be able to cause headache has been diagnosed
- C. Evidence of causation demonstrated by at least two of the following:
 - 1. headache has developed in temporal relation to the onset of the infection
 - 2. either or both of the following:
 - a) headache has significantly worsened in parallel with worsening of the infection
 - b) headache has significantly improved or resolved in parallel with improvement in or resolution of the infection
 - 3. headache has characteristics typical for the infection
- D. Not better accounted for by another ICHD-3 diagnosis.

9.1 Headache attributed to intracranial infection

Description:

Headache of variable duration, and in rare cases persistent, caused by intracranial bacterial, viral, fungal or other parasitic infection or by a sequela of any of these.

9.1.1 Headache attributed to bacterial meningitis or meningoencephalitis

Description:

Headache of variable duration caused by bacterial meningitis or meningoencephalitis. It may develop in a context of mild flu-like symptoms. It is typically acute and associated with neck stiffness, nausea, fever and changes in mental state and/or other neurological symptoms and/or signs. In most cases it resolves once the infection has been eradicated, but rarely it becomes persistent.

Diagnostic criteria:

- A. Headache of any duration fulfilling criterion C
- B. Bacterial meningitis or meningoencephalitis has been diagnosed
- C. Evidence of causation demonstrated by at least two of the following:

- 1. headache has developed in temporal relation to the onset of the bacterial meningitis or meningoencephalitis
- 2. headache has significantly worsened in parallel with worsening of the bacterial meningitis or meningoencephalitis
- 3. headache has significantly improved in parallel with improvement in the bacterial meningitis or meningoencephalitis
- 4. headache is either or both of the following:
 - a) holocranial
 - b) located in the nuchal area and associated with neck stiffness
- D. Not better accounted for by another ICHD-3 diagnosis.

Comments:

Headache is the commonest and may be the first symptom of these infections. 9.1.1 *Headache attributed to bacterial meningitis or meningoencephalitis* should be suspected whenever headache is associated with fever, altered mental state (including reduced vigilance), focal neurological deficits or generalized seizures. In the case of encephalitis, associated deficits include disturbances of speech or hearing, double vision, loss of sensation in some parts of the body, muscle weakness, partial paralysis in the arms and legs, hallucinations, personality changes, impaired judgement, loss of consciousness, sudden severe dementia and/or memory loss.

Nevertheless, in most cases of intracranial bacterial infection it is extremely difficult to distinguish involvement purely of the meninges from involvement purely of the encephalon. Furthermore, this distinction does not lead to different approaches to evaluation or choice of treatment. Therefore, headache attributed to bacterial meningitis and headache attributed to bacterial encephalitis have been included in the same subgroup of 9.1.1 *Headache attributed to bacterial meningitis or meningoencephalitis*.

A variety of microorganisms may cause meningitis and/or encephalitis, including *Streptococcus pneumoniae*, *Neisseria meningitidis* and *Listeria monocytogenes*.

Direct stimulation of the sensory terminals located in the meninges by the bacterial infection causes the onset of headache. Bacterial products (toxins), mediators of inflammation such as bradykinin, prostaglandins and cytokines and other agents released by inflammation not only directly cause pain but also induce pain sensitization and neuropeptide release. In the case of encephalitis, increased intracranial pressure may also play a role in causing headache.

In most cases, headache remits with resolution of the infection. However, the infection may remain active for

months, leading to chronic headache. In a minority of cases, headache persists for more than 3 months after resolution of the causative infection. Three separate subforms of 9.1.1 *Headache attributed to bacterial meningitis or meningoencephalitis* are therefore described because pathophysiology and treatment are different depending on whether the infection has been completely eradicated or remains active.

9.1.1.1 Acute headache attributed to bacterial meningitis or meningoencephalitis

Diagnostic criteria:

- A. Headache fulfilling criteria for 9.1.1 *Headache attributed to bacterial meningitis or meningoencephalitis*, and criterion C below
- B. Bacterial meningitis or meningoencephalitis remains active or has recently resolved
- C. Headache has been present for <3 months.

9.1.1.2 Chronic headache attributed to bacterial meningitis or meningoencephalitis

Diagnostic criteria:

- A. Headache fulfilling criteria for 9.1.1 *Headache attributed to bacterial meningitis or meningoencephalitis*, and criterion C below
- B. Bacterial meningitis or meningoencephalitis remains active or has resolved within the last 3 months
- C. Headache has been present for >3 months.

9.1.1.3 Persistent headache attributed to past bacterial meningitis or meningoencephalitis

Diagnostic criteria:

- A. Headache previously fulfilling criteria for 9.1.1 *Headache attributed to bacterial meningitis or meningoencephalitis*, and fulfilling criterion C below
- B. Bacterial meningitis or meningoencephalitis has resolved
- C. Headache has persisted for >3 months after resolution of the bacterial meningitis or meningoencephalitis
- D. Not better accounted for by another ICHD-3 diagnosis.

9.1.2 Headache attributed to viral meningitis or encephalitis

Description:

Headache caused by viral meningitis or encephalitis, typically with neck stiffness and fever and variably associated, according to the extent of the infection, with neurological symptoms and/or signs including changes in mental state.

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. Viral meningitis or encephalitis has been diagnosed
- C. Evidence of causation demonstrated by at least two of the following:
 1. headache has developed in temporal relation to the onset of the viral meningitis or encephalitis
 2. headache has significantly worsened in parallel with worsening of the viral meningitis or encephalitis
 3. headache has significantly improved in parallel with improvement in the viral meningitis or encephalitis
 4. headache is either or both of the following:
 - a) holocranial
 - b) located in the nuchal area and associated with neck stiffness
- D. Not better accounted for by another ICHD-3 diagnosis.

Comments:

9.1.2 *Headache attributed to viral meningitis or encephalitis* should be suspected whenever headache is associated with fever, stiff neck, light sensitivity, nausea and/or vomiting.

Enteroviruses account for most cases of 9.1.2 *Headache attributed to viral meningitis or encephalitis*; Herpes simplex, adenovirus, mumps and others may also be responsible. CSF polymerase chain reaction (PCR) gives the specific diagnosis in the majority of cases. Positive CSF PCR for Herpes simplex virus (HSV) types 1 or 2 and serology for HSV-1&2 DNA presume the diagnosis of Herpes simplex encephalitis. In some cases, CSF PCR is positive for Human Herpes virus (HHV) types 6 or 7. It has been documented that PCR sensitivity is reduced by more than half when the test is performed 1 week after the onset of the symptoms, causing false negatives. When PCR performed after 1 week is negative, the diagnosis can be made on the basis of an altered CSF/blood antibody ratio.

As with 9.1.1 *Headache attributed to bacterial meningitis or meningoencephalitis*, it may be difficult to distinguish involvement purely of the meninges from involvement purely of the encephalon. The distinction is nonetheless important to make and

maintain, because the two conditions differ prognostically, the expectation being worse with encephalitic involvement. For this reason, separate criteria are given for 9.1.2.1 *Headache attributed to viral meningitis* and 9.1.2.2 *Headache attributed to viral encephalitis*.

Also at variance from 9.1.1 *Headache attributed to bacterial meningitis or meningoencephalitis*, a persistent post-infectious subform of 9.1.2 *Headache attributed to viral meningitis or encephalitis* is not supported by evidence and has not, therefore, been contemplated.

9.1.2.1 *Headache attributed to viral meningitis*

Diagnostic criteria:

- A. Headache fulfilling criteria for 9.1.2 *Headache attributed to viral meningitis or encephalitis*
- B. Neuroimaging shows enhancement of the leptomeninges.

9.1.2.2 *Headache attributed to viral encephalitis*

Diagnostic criteria:

- A. Headache fulfilling criteria for 9.1.2 *Headache attributed to viral meningitis or encephalitis*
- B. Either or both of the following:
 - 1. neuroimaging shows diffuse brain oedema
 - 2. at least one of the following:
 - a) altered mental state
 - b) focal neurological deficits
 - c) seizures.

Comment:

9.1.2.2 *Headache attributed to viral encephalitis* should be suspected whenever headache is associated with altered mental state (including impaired vigilance), focal neurological deficits and/or seizures. Pain is usually diffuse, with the focus in frontal and/or retro-orbital areas, severe or extremely severe, throbbing or pressing type. Other commonly associated neurological deficits are disturbances of speech or hearing, double vision, loss of sensation in some parts of the body, muscle weakness, partial paralysis in the arms and legs, ataxia, hallucinations, personality changes, loss of consciousness and/or memory loss.

9.1.3 *Headache attributed to intracranial fungal or other parasitic infection*

Description:

Headache of variable duration caused by intracranial fungal or other parasitic infection. It is usually observed in a context of congenital or acquired immunosuppression. In most cases it resolves once the infection has been eradicated, but rarely it becomes persistent.

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. Intracranial fungal or other parasitic infection has been diagnosed
- C. Evidence of causation demonstrated by at least two of the following:
 - 1. headache has developed in temporal relation to the onset of the intracranial fungal or other parasitic infection
 - 2. headache has significantly worsened in parallel with worsening of the intracranial fungal or other parasitic infection
 - 3. headache has significantly improved in parallel with improvement in the intracranial fungal or other parasitic infection
 - 4. headache develops progressively,¹ and is either or both of the following:
 - a) holocranial
 - b) located in the nuchal area and associated with neck stiffness
- D. Not better accounted for by another ICHD-3 diagnosis.

Note:

- 1. The clinical symptoms tend to evolve over weeks, in parallel with the level of immunosuppression.

Comments:

9.1.3 *Headache attributed to intracranial fungal or other parasitic infection* should be suspected whenever headache is associated with fever, progressively altered mental state (including impaired vigilance) and/or multiple focal neurological deficits of increasing severity, and neuroimaging shows enhancement of the leptomeninges and/or diffuse brain oedema.

Early diagnosis is best made by CT or MRI. Besides CSF culture and CSF PCR investigations, other tests on CSF and blood are available. These include direct detection of the pathogen (cytological detection, microscopic visualization, culture and identification of fungal elements in the biological materials under observation)

and tests for indirect detection of the pathogen (identification of an antigen or another element of the capsule). In the case of aspergillosis, the galattomannan antigen can be detected in biological fluids (serum, bronchoalveolar washing liquid or CSF). In other systemic fungal infections, serum 1,3- β -D-glucan may be diagnostically helpful. The India ink test enables staining of the capsule of cryptococcus.

It is noteworthy that fungal and parasitic infections of the meninges or encephalon are almost exclusively observed in immunodepressed patients or old people. More specifically, the following groups are to be considered at risk:

1. people with significant neutropaenia (<500 neutrophils/mm³) detected in close temporal relation to the infection
2. people who have undergone allogenic graft of stem cells
3. people undergoing chronic steroid therapy (prednisone 0.3 mg/kg/day or equivalent for more than 3 weeks)
4. people with ongoing or recent (within the previous 90 days) treatment with immunosuppressor drugs (cyclosporine, TNF blockers, monoclonal antibodies, analogues of nucleosides)
5. people with severe hereditary immunodeficiency.

A persistent post-infectious subform of 9.1.3 *Headache attributed to intracranial fungal or other parasitic infection* is not well supported by evidence; it appears only in the Appendix as A9.1.3.3 *Persistent headache attributed to past intracranial fungal or other parasitic infection*.

9.1.3.1 Acute headache attributed to intracranial fungal or other parasitic infection

Diagnostic criteria:

- A. Headache fulfilling criteria for 9.1.3 *Headache attributed to intracranial fungal or other parasitic infection*, and criterion C below
- B. Intracranial fungal or other parasitic infection remains active or has recently resolved
- C. Headache has been present for <3 months.

9.1.3.2 Chronic headache attributed to intracranial fungal or other parasitic infection

Diagnostic criteria:

- A. Headache fulfilling criteria for 9.1.3 *Headache attributed to intracranial fungal or other parasitic infection*, and criterion C below
- B. Intracranial fungal or other parasitic infection remains active or has resolved within the last 3 months

- C. Headache has been present for >3 months.

9.1.4 Headache attributed to brain abscess

Description:

Headache caused by brain abscess, usually associated with fever, focal neurological deficit(s) and/or altered mental state (including impaired vigilance).

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. A brain abscess has been demonstrated
- C. Evidence of causation demonstrated by at least two of the following:
 1. headache has developed in temporal relation to development of the abscess, or led to its discovery
 2. headache has significantly worsened in parallel with deterioration of the abscess shown by any of the following:
 - a) worsening of other symptoms and/or clinical signs arising from the abscess
 - b) evidence of enlargement of the abscess
 - c) evidence of rupture of the abscess
 3. headache has significantly improved in parallel with improvement in the abscess
 4. headache has at least one of the following three characteristics:
 - a) intensity increasing gradually, over several hours or days, to moderate or severe
 - b) aggravated by straining or other Valsalva manoeuvre
 - c) accompanied by nausea
- D. Not better accounted for by another ICHD-3 diagnosis.

Comments:

The most common organisms causing brain abscess include streptococcus, staphylococcus aureus, bacteroides species and enterobacter. Recently, brain abscesses have also been reported with aspergillosis and blastomycosis.

Predisposing factors include infections of the paranasal sinuses, ears, jaws, teeth or lungs.

Direct compression and irritation of the meningeal and/or arterial structures and increased intracranial pressure are the mechanisms causing 9.1.4 *Headache attributed to brain abscess*.

9.1.5 Headache attributed to subdural empyema

Description:

Headache caused by a subdural empyema, usually associated with fever and symptoms and/or clinical

signs of meningeal irritation and increased intracranial pressure.

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. Subdural empyema has been demonstrated
- C. Evidence of causation demonstrated by at least two of the following:
 - 1. headache has developed in temporal relation to development of the empyema, or led to its discovery
 - 2. headache has significantly worsened in parallel with deterioration of the empyema shown by any of the following:
 - a) worsening of other symptoms and/or clinical signs arising from the empyema
 - b) evidence of enlargement of the empyema
 - c) evidence of rupture of the empyema
 - 3. headache has significantly improved in parallel with improvement in the empyema
 - 4. headache has either or both of the following characteristics:
 - a) unilateral, or more intense on one side
 - b) associated with tenderness of the skull
- D. Not better accounted for by another ICHD-3 diagnosis.

Comments:

Subdural empyema is often secondary to sinusitis or otitis media. It may also be a complication of meningitis.

9.1.5 *Headache attributed to subdural empyema* is caused by meningeal irritation, increased intracranial pressure and/or fever.

9.2 Headache attributed to systemic infection

Coded elsewhere:

Headache attributed to meningitis or encephalitis accompanying systemic infection should be coded accordingly under 9.1 *Headache attributed to intracranial infection*.

Description:

Headache of variable duration caused by systemic infection, usually accompanied by other symptoms and/or clinical signs of the infection.

Comments:

Headache in systemic infections is usually a relatively inconspicuous symptom, and diagnostically unhelpful. These conditions are mostly dominated by fever, general malaise and other systemic symptoms.

Nevertheless, some systemic infections, particularly influenza, have headache as a prominent symptom along with fever and others. When systemic infection is accompanied by meningitis or encephalitis, any headache attributed to the infection should be coded to these disorders as a subtype of 9.1 *Headache attributed to intracranial infection*.

In infectious disease, headache commonly coexists with fever and may be dependent on it, but headache can also occur in the absence of fever. The exact nature of these mechanisms remains to be investigated. Meanwhile, the great variability in their propensity for causing headache indicates that systemic infections do not have this effect simply through fever and exogenous or endogenous pyrogens. The mechanisms causing headache include direct effects of the microorganisms themselves. Several cells are likely to be involved (activated microglia and monocytic macrophages, activated astrocytes and blood-brain barrier and endothelial cells), along with several immunoinflammatory mediators (cytokines, glutamate, COX-2/PGE2 system, NO-iNOS system and reactive oxygen species system).

9.2.1 Headache attributed to systemic bacterial infection

Description:

Headache caused by and occurring in association with other symptoms and/or clinical signs of a systemic bacterial infection, in the absence of meningitis or meningoencephalitis.

Diagnostic criteria:

- A. Headache of any duration fulfilling criterion C
- B. Both of the following:
 - 1. systemic bacterial infection has been diagnosed
 - 2. there is no evidence of meningitic or meningoencephalitic involvement
- C. Evidence of causation demonstrated by at least two of the following:
 - 1. headache has developed in temporal relation to onset of the systemic bacterial infection
 - 2. headache has significantly worsened in parallel with worsening of the systemic bacterial infection
 - 3. headache has significantly improved or resolved in parallel with improvement in or resolution of the systemic bacterial infection
 - 4. headache has either or both of the following characteristics:
 - a) diffuse pain
 - b) moderate or severe intensity
- D. Not better accounted for by another ICHD-3 diagnosis.

9.2.1.1 *Acute headache attributed to systemic bacterial infection*

Diagnostic criteria:

- A. Headache fulfilling criteria for 9.2.1 *Headache attributed to systemic bacterial infection*, and criterion C below
- B. The systemic bacterial infection remains active or has recently resolved
- C. Headache has been present for <3 months.

9.2.1.2 *Chronic headache attributed to systemic bacterial infection*

Diagnostic criteria:

- A. Headache fulfilling criteria for 9.2.1 *Headache attributed to systemic bacterial infection*, and criterion C below
- B. The systemic bacterial infection remains active or has resolved within the last 3 months
- C. Headache has been present for >3 months.

9.2.2 *Headache attributed to systemic viral infection*

Description:

Headache caused by and occurring in association with other symptoms and/or clinical signs of a systemic viral infection, in the absence of meningitis or encephalitis.

Diagnostic criteria:

- A. Headache of any duration fulfilling criterion C
- B. Both of the following:
 1. systemic viral infection has been diagnosed
 2. there is no evidence of meningitic or encephalitic involvement
- C. Evidence of causation demonstrated by at least two of the following:
 1. headache has developed in temporal relation to onset of the systemic viral infection
 2. headache has significantly worsened in parallel with worsening of the systemic viral infection
 3. headache has significantly improved or resolved in parallel with improvement in or resolution of the systemic viral infection
 4. headache has either or both of the following characteristics:
 - a) diffuse pain

- b) moderate or severe intensity
- D. Not better accounted for by another ICHD-3 diagnosis.

9.2.2.1 *Acute headache attributed to systemic viral infection*

Diagnostic criteria:

- A. Headache fulfilling criteria for 9.2.2 *Headache attributed to systemic viral infection*, and criterion C below
- B. The systemic viral infection remains active or has recently resolved
- C. Headache has been present for <3 months.

9.2.2.2 *Chronic headache attributed to systemic viral infection*

Diagnostic criteria:

- A. Headache fulfilling criteria for 9.2.2 *Headache attributed to systemic viral infection*, and criterion C below
- B. The systemic viral infection remains active or has resolved within the last 3 months
- C. Headache has been present for >3 months.

9.2.3 *Headache attributed to other systemic infection*

Description:

Headache caused by and occurring in association with other symptoms and/or clinical signs of a systemic fungal infection or infestation by protozoal or other parasites, in the absence of meningitis or meningoencephalitis.

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. Both of the following:
 1. systemic fungal infection, or infestation by protozoal or other parasites, has been diagnosed
 2. there is no evidence of meningitic or meningoencephalitic involvement
- C. Evidence of causation demonstrated by at least two of the following:
 1. headache has developed in temporal relation to onset of the systemic infection or infestation

2. headache has significantly worsened in parallel with worsening of the systemic infection or infestation
 3. headache has significantly improved in parallel with improvement in the systemic infection or infestation
 4. headache has either or both of the following characteristics:
 - a) diffuse pain
 - b) moderate or severe intensity
- D. Not better accounted for by another ICHD-3 diagnosis.

Comment:

This is a heterogenous and ill-defined group of systemic infections, most frequently seen in immunosuppressed patients or in specific geographical areas. The fungi most commonly involved are the pathogenic fungi (*Cryptococcus neoformans*, *Histoplasma capsulatum* and *Coccidioides immitis*) and the opportunistic fungi (*Candida* species, *Aspergillus* species and others). Among protozoa, *Pneumocystis carinii* and *Toxoplasma gondii* infestations may be associated with headache. Headache has also been reported with the nematode *Strongyloides stercoralis*.

9.2.3.1 Acute headache attributed to other systemic infection

Diagnostic criteria:

- A. Headache fulfilling criteria for 9.2.3 *Headache attributed to other systemic infection*, and criterion C below
- B. The systemic infection remains active or has recently resolved
- C. Headache has been present for <3 months.

9.2.3.2 Chronic headache attributed to other systemic infection

Diagnostic criteria:

- A. Headache fulfilling criteria for 9.2.3 *Headache attributed to other systemic infection*, and criterion C below
- B. The systemic infection remains active or has resolved within the last 3 months
- C. Headache has been present for >3 months.

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9.1.2 Headache attributed to viral meningitis or encephalitis

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9.1.5 Headache attributed to subdural empyema

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10. Headache attributed to disorder of homoeostasis

- 10.1 Headache attributed to hypoxia and/or hypercapnia
 - 10.1.1 High-altitude headache
 - 10.1.2 Headache attributed to aeroplane travel
 - 10.1.3 Diving headache
 - 10.1.4 Sleep apnoea headache
- 10.2 Dialysis headache
- 10.3 Headache attributed to arterial hypertension
 - 10.3.1 Headache attributed to pheochromocytoma
 - 10.3.2 Headache attributed to hypertensive crisis without hypertensive encephalopathy
 - 10.3.3 Headache attributed to hypertensive encephalopathy
 - 10.3.4 Headache attributed to pre-eclampsia or eclampsia
 - 10.3.5 Headache attributed to autonomic dysreflexia
- 10.4 Headache attributed to hypothyroidism
- 10.5 Headache attributed to fasting
- 10.6 Cardiac cephalgia
- 10.7 Headache attributed to other disorder of homoeostasis

Coded elsewhere:

7.1.2 Headache attributed to intracranial hypertension secondary to metabolic, toxic or hormonal causes.

General comment

Primary or secondary headache or both?

When a headache occurs for the first time in close temporal relation to a disorder of homoeostasis, it is coded as a secondary headache attributed to that disorder. This remains true when the new headache has the characteristics of any of the primary headache disorders classified in Part one of ICHD-3 beta. When a *pre-existing* headache with the characteristics of a primary headache disorder becomes *chronic*, or is made significantly worse (usually meaning a two-fold or greater increase in frequency and/or severity), in close temporal relation to a disorder of homoeostasis, both the initial headache diagnosis and a diagnosis of 10. *Headache attributed to a disorder of homoeostasis* (or one of its subtypes) should be given, provided that there is good evidence that that disorder can cause headache.

Introduction

The mechanisms behind causation of the various subtypes of 10. *Headache attributed to disorder of*

homoeostasis are various. Nevertheless, it is possible to set out general diagnostic criteria, applicable in most cases, as follows:

- A. Headache fulfilling criterion C
- B. A disorder of homoeostasis known to be able to cause headache has been diagnosed
- C. Evidence of causation demonstrated by at least two of the following:
 - 1. headache has developed in temporal relation to the onset of the disorder of homoeostasis
 - 2. either or both of the following:
 - a) headache has significantly worsened in parallel with worsening of the disorder of homoeostasis
 - b) headache has significantly improved after resolution of the disorder of homoeostasis
 - 3. headache has characteristics typical for the disorder of homoeostasis
- D. Not better accounted for by another ICHD-3 diagnosis.

10.1 Headache attributed to hypoxia and/or hypercapnia

Description:

Headache caused by hypoxia and/or hypercapnia and occurring in conditions of exposure to one or both.

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. Exposure to conditions of hypoxia and/or hypercapnia
- C. Evidence of causation demonstrated by at least one of the following:
 - 1. headache has developed in temporal relation to the exposure
 - 2. either or both of the following:
 - a) headache has significantly worsened in parallel with increasing exposure to hypoxia and/or hypercapnia
 - b) headache has significantly improved in parallel with improvement in hypoxia and/or hypercapnia
- D. Not better accounted for by another ICHD-3 diagnosis.

10.1.1 High-altitude headache

Description:

Headache, usually bilateral and aggravated by exertion, caused by ascent above 2500 metres. It resolves spontaneously within 24 hours after descent.

Diagnostic criteria:

- A. Headache fulfilling criterion C
- B. Ascent to altitude above 2500 m has taken place
- C. Evidence of causation demonstrated by at least two of the following:
 1. headache has developed in temporal relation to the ascent
 2. either or both of the following:
 - a) headache has significantly worsened in parallel with continuing ascent
 - b) headache has resolved within 24 hours after descent to below 2500 m
 3. headache has at least two of the following three characteristics:
 - a) bilateral location
 - b) mild or moderate intensity
 - c) aggravated by exertion, movement, straining, coughing and/or bending
- D. Not better accounted for by another ICHD-3 diagnosis.

Comments:

10.1.1 *High-altitude headache* is a frequent complication of ascent to altitude, occurring in more than 30% of mountaineers. Risk factors include a history of 1. *Migraine*, low arterial oxygen saturation, high perceived degree of exertion and fluid intake below 2 litres in 24 hours.

Most cases of 10.1.1 *High-altitude headache* respond to simple analgesics such as paracetamol (acetaminophen) or ibuprofen. However, acute mountain sickness (AMS) consists of at least moderate headache combined with one or more of nausea, anorexia, fatigue, photophobia, dizziness and sleep disturbances. Acetazolamide (125 mg, two to three times daily) and steroids may reduce susceptibility to AMS. Other preventative strategies include 2 days of acclimatization prior to engaging in strenuous exercise at high altitudes, liberal fluid intake and avoidance of alcohol.

*10.1.2 Headache attributed to aeroplane travel**Description:*

Headache, often severe, usually unilateral and periorbital and without autonomic symptoms, occurring during and caused by aeroplane travel. It remits after landing.

Diagnostic criteria:

- A. At least two episodes of headache fulfilling criterion C
- B. The patient is travelling by aeroplane

- C. Evidence of causation demonstrated by at least two of the following:
 1. headache has developed exclusively during aeroplane travel
 2. either or both of the following:
 - a) headache has worsened in temporal relation to ascent after take-off and/or descent prior to landing of the aeroplane
 - b) headache has spontaneously improved within 30 minutes after the ascent or descent of the aeroplane is completed
 3. headache is severe, with at least two of the following three characteristics:
 - a) unilateral location
 - b) orbitofrontal location (parietal spread may occur)
 - c) jabbing or stabbing quality (pulsation may also occur)
- D. Not better accounted for by another ICHD-3 diagnosis.

Comments:

10.1.2 *Headache attributed to aeroplane travel* occurs during landing in more than 85% of patients. Side-shift between different flights occurs in around 10% of cases. Nasal congestion, a stuffy feeling of the face or tearing may occur ipsilaterally, but these have been described in fewer than 5% of cases.

The presence of a sinus disorder should be excluded.

*10.1.3 Diving headache**Coded elsewhere:*

1. *Migraine*, 2. *Tension-type headache*, 4.2 *Primary exercise headache*, 4.5 *Cold-stimulus headache*, 4.6.1 *External compression headache* and 11.2.1 *Cervicogenic headache* can occur during a dive. In these instances, diving should be considered a precipitating factor rather than the cause, and the headache should be coded as these disorders accordingly.

Description:

Headache caused by diving below 10 metres, occurring during the dive and often intensified on resurfacing, in the absence of decompression illness. It is usually accompanied by symptoms of carbon dioxide intoxication. It remits quickly with oxygen or, if this is not given, spontaneously within 3 days after the dive has ended.

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. Both of the following:

1. the patient is diving at a depth greater than 10 m
2. there is no evidence of decompression illness
- C. Evidence of causation demonstrated by at least one of the following:
 1. headache has developed during the dive
 2. either or both of the following:
 - a) headache has worsened as the dive is continued
 - b) either of the following:
 - (i) headache has spontaneously resolved within 3 days of completion of the dive
 - (ii) headache has remitted within 1 hour after treatment with 100% oxygen
 3. at least one of the following symptoms of CO₂ intoxication:
 - a) mental confusion
 - b) light-headedness
 - c) motor incoordination
 - d) dyspnoea
 - e) facial flushing
- D. Not better accounted for by another ICHD-3 diagnosis.

Comment:

There is some evidence that hypercapnia in the absence of hypoxia is associated with headache. The best clinical example of headache attributed to hypercapnia is 10.1.3 *Diving headache*. Hypercapnia (arterial pCO₂ >50 mmHg) is known to cause relaxation of cerebrovascular smooth muscle, leading to intracranial vasodilatation and increased intracranial pressure. Carbon dioxide may accumulate in a diver who intentionally holds his or her breath intermittently (skip breathing) in a mistaken attempt to conserve air, or takes shallow breaths to minimize buoyancy variations in the narrow passages of a wreck or cave. Divers may also hypoventilate unintentionally when a tight wetsuit or buoyancy compensator jacket restricts chest wall expansion, or when ventilation is inadequate in response to physical exertion. Strenuous exercise increases the rate of CO₂ production more than 10-fold, resulting in a transient elevation of pCO₂ to more than 60 mmHg. 10.1.3 *Diving headache* usually intensifies during the decompression phase of the dive or on resurfacing.

10.1.4 *Sleep apnoea headache*

Description:

Morning headache, usually bilateral and with a duration of less than 4 hours, caused by sleep apnoea. The disorder resolves with successful treatment of the sleep apnoea.

Diagnostic criteria:

- A. Headache present on awakening after sleep and fulfilling criterion C
- B. Sleep apnoea (apnoea-hypopnoea index ≥ 5) has been diagnosed¹
- C. Evidence of causation demonstrated by at least two of the following:
 1. headache has developed in temporal relation to the onset of sleep apnoea
 2. either or both of the following:
 - a) headache has worsened in parallel with worsening of sleep apnoea
 - b) headache has significantly improved or remitted in parallel with improvement in or resolution of sleep apnoea
 3. headache has at least one of the following three characteristics:
 - a) recurs on >15 days per month
 - b) all of the following:
 - (i) bilateral location
 - (ii) pressing quality
 - (iii) not accompanied by nausea, photophobia or phonophobia
 - c) resolves within 4 hours
- D. Not better accounted for by another ICHD-3 diagnosis.

Note:

1. The apnoea-hypopnoea index is calculated by dividing the number of apnoeic events by the number of hours of sleep (5–15/hours = mild; 15–30/hours = moderate; >30/hours = severe).

Comments:

10.1.4 *Sleep apnoea headache* seems to be less frequent and of longer duration than previously assumed. A definitive diagnosis requires overnight polysomnography. Although morning headache is significantly more common in patients with sleep apnoea than in the general population, headache present on awakening is a non-specific symptom which occurs in a variety of primary and secondary headache disorders, in sleep-related respiratory disorders other than sleep apnoea (e.g. Pickwickian syndrome, chronic obstructive pulmonary disorder), and in other primary sleep disorders such as periodic leg movements of sleep.

It is unclear whether the mechanism of 10.1.4 *Sleep apnoea headache* is related to hypoxia, hypercapnia or disturbance in sleep.

10.2 Dialysis headache

Description:

Headache with no specific characteristics occurring during and caused by haemodialysis. It resolves spontaneously within 72 hours after the haemodialysis session has ended.

Diagnostic criteria:

- A. At least three episodes of acute headache fulfilling criterion C
- B. The patient is on haemodialysis
- C. Evidence of causation demonstrated by at least two of the following:
 1. each headache has developed during a session of haemodialysis
 2. either or both of the following:
 - a) each headache has worsened during the dialysis session
 - b) each headache has resolved within 72 hours after the end of the dialysis session
 3. headache episodes cease altogether after successful kidney transplantation and termination of haemodialysis
- D. Not better accounted for by another ICHD-3 diagnosis.

Comments:

10.2 *Dialysis headache* commonly occurs in association with hypotension and dialysis disequilibrium syndrome. This syndrome may begin as headache and then progress to obtundation and finally coma, with or without seizures. It is relatively rare, and may be prevented by changing dialysis parameters.

Low magnesium and high sodium levels may be risk factors for developing 10.2 *Dialysis headache*.

As caffeine is rapidly removed by dialysis, 8.3.1 *Caffeine-withdrawal headache* should be considered in patients who consume large quantities of caffeine.

10.3 Headache attributed to arterial hypertension

Description:

Headache, often bilateral and pulsating, caused by arterial hypertension, usually during an acute rise in systolic (to ≥ 180 mmHg) and/or diastolic (to ≥ 120 mmHg) blood pressure. It remits after normalization of blood pressure.

Diagnostic criteria:

- A. Any headache fulfilling criterion C

- B. Hypertension defined as systolic pressure ≥ 180 mmHg and/or diastolic pressure ≥ 120 mmHg has been demonstrated
- C. Evidence of causation demonstrated by either or both of the following:
 1. headache has developed in temporal relation to the onset of hypertension
 2. either or both of the following:
 - a) headache has significantly worsened in parallel with worsening hypertension
 - b) headache has significantly improved in parallel with improvement in hypertension
- D. Not better accounted for by another ICHD-3 diagnosis.

Comments:

Mild (140–159/90–99 mmHg) or moderate (160–179/100–109 mmHg) chronic arterial hypertension does not appear to *cause* headache. Whether moderate hypertension *predisposes* to headache at all remains controversial, but there is some evidence that it does.

Ambulatory blood pressure monitoring in patients with mild and moderate hypertension has shown no convincing relationship between blood pressure fluctuations over a 24-hour period and presence or absence of headache.

10.3.1 Headache attributed to pheochromocytoma

Coded elsewhere:

When hypertensive encephalopathy is present, headache is coded as 10.3.3 *Headache attributed to hypertensive encephalopathy*. When the diagnosis of pheochromocytoma has not yet been made, and hypertensive encephalopathy is not present, patients may meet the diagnostic criteria for 10.3.2 *Headache attributed to hypertensive crisis without hypertensive encephalopathy*.

Description:

Headache attacks, usually severe and of short duration (less than 1 hour) and accompanied by sweating, palpitations, pallor and/or anxiety, caused by pheochromocytoma.

Diagnostic criteria:

- A. Recurrent discrete short-lasting headache episodes fulfilling criterion C
- B. Pheochromocytoma has been demonstrated
- C. Evidence of causation demonstrated by at least two of the following:
 1. headache episodes have commenced in temporal relation to development of the pheochromocytoma, or led to its discovery

2. either or both of the following:
 - a) individual headache episodes develop in temporal relation to abrupt rises in blood pressure
 - b) individual headache episodes remit in temporal relation to normalization of blood pressure
 3. headache is accompanied by at least one of the following:
 - a) sweating
 - b) palpitations
 - c) anxiety
 - d) pallor
 4. headache episodes remit entirely after removal of the phaeochromocytoma
- D. Not better accounted for by another ICHD-3 diagnosis.

Comments:

10.3.1 *Headache attributed to phaeochromocytoma* occurs as a paroxysmal headache in 51–80% of patients with phaeochromocytoma. It is often severe, frontal or occipital and usually described as either pulsating or constant in quality. An important feature of the headache is its short duration: less than 15 minutes in 50% and less than 1 hour in 70% of patients. Associated features include apprehension and/or anxiety, often with a sense of impending death, tremor, visual disturbances, abdominal or chest pain, nausea, vomiting and occasionally paraesthesia. The face can blanch or flush during the attack.

The diagnosis of phaeochromocytoma is established by the demonstration of increased excretion of catecholamines or catecholamine metabolites, and can usually be secured by analysis of a single 24-hour urine sample collected when the patient is hypertensive or symptomatic.

10.3.2 *Headache attributed to hypertensive crisis without hypertensive encephalopathy*

Coded elsewhere:

10.3.1 *Headache attributed to phaeochromocytoma.*

Description:

Headache, usually bilateral and pulsating, caused by a paroxysmal rise of arterial hypertension (systolic ≥ 180 mmHg and/or diastolic ≥ 120 mmHg). It remits after normalization of blood pressure.

Diagnostic criteria:

- A. Headache fulfilling criterion C
- B. Both of the following:

1. a hypertensive crisis¹ is occurring
 2. there are no clinical features or other evidence of hypertensive encephalopathy
- C. Evidence of causation demonstrated by at least two of the following:
1. headache has developed during the hypertensive crisis
 2. either or both of the following:
 - a) headache has significantly worsened in parallel with increasing hypertension
 - b) headache has significantly improved or resolved in parallel with improvement in or resolution of the hypertensive crisis
 3. headache has at least one of the following three characteristics:
 - a) bilateral location
 - b) pulsating quality
 - c) precipitated by physical activity
- D. Not better accounted for by another ICHD-3 diagnosis.

Note:

1. A hypertensive crisis is defined as a paroxysmal rise in systolic (to ≥ 180 mmHg) and/or diastolic (to ≥ 120 mmHg) blood pressure.

Comment:

Paroxysmal hypertension may occur in association with failure of baroreceptor reflexes (after carotid endarterectomy or subsequent to irradiation of the neck) or in patients with enterochromaffin cell tumours.

10.3.3 *Headache attributed to hypertensive encephalopathy*

Description:

Headache, usually bilateral and pulsating, caused by persistent blood pressure elevation to 180/120 mmHg or above and accompanied by symptoms of encephalopathy such as confusion, lethargy, visual disturbances or seizures. It improves after normalization of blood pressure.

Diagnostic criteria:

- A. Headache fulfilling criterion C
- B. Hypertensive encephalopathy has been diagnosed
- C. Evidence of causation demonstrated by at least two of the following:
 1. headache has developed in temporal relation to the onset of the hypertensive encephalopathy
 2. either or both of the following:

- a) headache has significantly worsened in parallel with worsening of the hypertensive encephalopathy
- b) headache has significantly improved or resolved in parallel with improvement in or resolution of the hypertensive encephalopathy
- 3. headache has at least two of the following three characteristics:
 - a) diffuse pain
 - b) pulsating quality
 - c) aggravated by physical activity
- D. Not better accounted for by another ICHD-3 diagnosis.

Comments:

Hypertensive encephalopathy presents with persistent elevation of blood pressure to $\geq 180/120$ mmHg and at least two of confusion, reduced level of consciousness, visual disturbances including blindness, and seizures. It is thought to occur when compensatory cerebrovascular vasoconstriction can no longer prevent cerebral hyperperfusion as blood pressure rises. As normal cerebral autoregulation of blood flow is overwhelmed, endothelial permeability increases and cerebral oedema occurs. On MRI, this is often most prominent in the parieto-occipital white matter.

Although hypertensive encephalopathy in patients with chronic arterial hypertension is usually accompanied by a diastolic blood pressure of >120 mmHg, and by grade III or IV hypertensive retinopathy (Keith-Wagener-Barker classification), previously normotensive individuals may develop signs of encephalopathy with blood pressures as low as 160/100 mmHg. Hypertensive retinopathy may not be present at the time of clinical presentation.

Any cause of hypertension can lead to hypertensive encephalopathy. Headache attributed to hypertensive encephalopathy should be coded as 10.3.3 *Headache attributed to hypertensive encephalopathy*, regardless of the underlying cause.

10.3.4 Headache attributed to pre-eclampsia or eclampsia

Description:

Headache, usually bilateral and pulsating, occurring in women during pregnancy or the immediate puerperium with pre-eclampsia or eclampsia. It remits after resolution of the pre-eclampsia or eclampsia.

Diagnostic criteria:

- A. Headache, in a woman who is pregnant or in the puerperium (up to 4 weeks postpartum), fulfilling criterion C

- B. Pre-eclampsia or eclampsia has been diagnosed
- C. Evidence of causation demonstrated by at least two of the following:
 - 1. headache has developed in temporal relation to the onset of the pre-eclampsia or eclampsia
 - 2. either or both of the following:
 - a) headache has significantly worsened in parallel with worsening of the pre-eclampsia or eclampsia
 - b) headache has significantly improved or resolved in parallel with improvement in or resolution of the pre-eclampsia or eclampsia
 - 3. headache has at least two of the following three characteristics:
 - a) bilateral location
 - b) pulsating quality
 - c) aggravated by physical activity
- D. Not better accounted for by another ICHD-3 diagnosis.

Comments:

Pre-eclampsia and eclampsia appear to involve a strong maternal inflammatory response, with broad immunological systemic activity. A placenta appears essential for their development, although case reports indicate that eclampsia can occur in the puerperium as well as during pregnancy.

Pre-eclampsia and eclampsia are multi-system disorders with various forms. Their diagnosis requires hypertension ($>140/90$ mmHg) documented on two blood pressure readings at least 4 hours apart, or a rise in diastolic pressure of ≥ 15 mmHg or in systolic pressure of ≥ 30 mmHg, coupled with urinary protein excretion >0.3 g/24 hours. In addition, tissue oedema, thrombocytopenia and abnormalities in liver function can occur.

10.3.5 Headache attributed to autonomic dysreflexia

Description:

Throbbing severe headache, with sudden onset, in patients with spinal cord injury and autonomic dysreflexia. The latter, which can be life-threatening, manifests as a paroxysmal rise in blood pressure among other symptoms and clinical signs, and is often triggered by bladder or bowel irritation (by infection, distension or impaction).

Diagnostic criteria:

- A. Headache of sudden onset, fulfilling criterion C
- B. Presence of spinal cord injury and autonomic dysreflexia documented by a paroxysmal rise above baseline in systolic pressure of ≥ 30 mmHg and/or diastolic pressure ≥ 20 mmHg

- C. Evidence of causation demonstrated by at least two of the following:
1. headache has developed in temporal relation to the rise in blood pressure
 2. either or both of the following:
 - a) headache has significantly worsened in parallel with increase in blood pressure
 - b) headache has significantly improved in parallel with decrease in blood pressure
 3. headache has at least two of the following four characteristics:
 - a) severe intensity
 - b) pounding or throbbing (pulsating) quality
 - c) accompanied by diaphoresis cranial to the level of the spinal cord injury
 - d) triggered by bladder or bowel reflexes
- D. Not better accounted for by another ICHD-3 diagnosis.

Comments:

The time to onset of autonomic dysreflexia after spinal cord injury is variable and has been reported from 4 days to 15 years.

Given that autonomic dysreflexia can be a life-threatening condition, its prompt recognition and adequate management are critical. Typically, 10.3.5 *Headache attributed to autonomic dysreflexia* is a sudden-onset, severe headache accompanied by several other symptoms and clinical signs including increased blood pressure, altered heart rate and diaphoresis cranial to the level of spinal cord injury. These are triggered by noxious or non-noxious stimuli, usually of visceral origin (bladder distension, urinary tract infection, bowel distension or impaction, urological procedures, gastric ulcer and others) but also of somatic origin (pressure ulcers, ingrown toenail, burns, trauma or surgical or invasive diagnostic procedures).

10.4 Headache attributed to hypothyroidism

Description:

Headache, usually bilateral and non-pulsatile, in patients with hypothyroidism and remitting after normalization of thyroid hormone levels.

Diagnostic criteria:

- A. Headache fulfilling criterion C
- B. Hypothyroidism has been demonstrated
- C. Evidence of causation demonstrated by at least two of the following:
 1. headache has developed in temporal relation to the onset of hypothyroidism, or led to its discovery

2. either or both of the following:
 - a) headache has significantly worsened in parallel with worsening of the hypothyroidism
 - b) headache has significantly improved or resolved in parallel with improvement in or resolution of the hypothyroidism
 3. headache has at least one of the following three characteristics:
 - a) bilateral location
 - b) non-pulsatile quality
 - c) constant over time
- D. Not better accounted for by another ICHD-3 diagnosis.

Comments:

It has been estimated that approximately 30% of patients with hypothyroidism suffer from 10.4 *Headache attributed to hypothyroidism*. Its mechanism is unclear. There is a female preponderance and often a history of migraine in childhood. The headache is not associated with nausea or vomiting.

In the presence of hypothyroidism, headache can also be a manifestation of pituitary adenoma (coded 7.4.3 *Headache attributed to hypothalamic or pituitary hyper- or hyposecretion*).

10.5 Headache attributed to fasting

Coded elsewhere:

An episode of migraine triggered by fasting is coded as 1. *Migraine* or one of its subtypes.

Description:

Diffuse non-pulsating headache, usually mild to moderate, occurring during and caused by fasting for at least 8 hours. It is relieved after eating.

Diagnostic criteria:

- A. Diffuse headache not fulfilling the criteria for 1. *Migraine* or any of its subtypes but fulfilling criterion C below
- B. The patient has fasted for ≥ 8 hours
- C. Evidence of causation demonstrated by both of the following:
 1. headache has developed during fasting
 2. headache has significantly improved after eating
- D. Not better accounted for by another ICHD-3 diagnosis.

Comments:

10.5 *Headache attributed to fasting* is significantly more common in people who have a prior history of

headache. Even though the typical headache attributed to fasting is diffuse, non-pulsating and mild to moderate in intensity, in those with a prior history of migraine the headache may resemble 1.1 *Migraine without aura*. If the criteria for this disorder are met, the headache should be coded accordingly (fasting being a precipitating factor).

The likelihood of headache developing as a result of a fast increases with the duration of the fast. Nevertheless, 10.5 *Headache attributed to fasting* does not appear to be related to duration of sleep, to caffeine withdrawal or to hypoglycaemia. Although headache may occur under conditions of hypoglycaemia-induced brain dysfunction, there is no conclusive evidence to support a causal association. 10.5 *Headache attributed to fasting* can occur in the absence of hypoglycaemia, insulin-induced hypoglycaemia does not precipitate headache in migraine sufferers, and headache is not a complaint of patients presenting to the emergency department with symptomatic hypoglycaemia.

10.6 Cardiac cephalalgia

Description:

Migraine-like headache, usually but not always aggravated by exercise, occurring during an episode of myocardial ischaemia. It is relieved by nitroglycerine.

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. Acute myocardial ischaemia has been demonstrated
- C. Evidence of causation demonstrated by at least two of the following:
 1. headache has developed in temporal relation to onset of acute myocardial ischaemia
 2. either or both of the following:
 - a) headache has significantly worsened in parallel with worsening of the myocardial ischaemia
 - b) headache has significantly improved or resolved in parallel with improvement in or resolution of the myocardial ischaemia
 3. headache has at least two of the following four characteristics:
 - a) moderate to severe intensity
 - b) accompanied by nausea
 - c) not accompanied by photophobia or phonophobia
 - d) aggravated by exertion
 4. headache is relieved by nitroglycerine or derivatives of it

- D. Not better accounted for by another ICHD-3 diagnosis.

Comments:

Diagnosis must include careful documentation of headache and simultaneous cardiac ischaemia during treadmill or nuclear cardiac stress testing. However, 10.6 *Cardiac cephalalgia* occurring at rest has been described.

Failure to recognize and correctly diagnose 10.6 *Cardiac cephalalgia* can have serious consequences. Therefore, distinguishing this disorder from 1.1 *Migraine without aura* is of crucial importance, particularly as vasoconstrictor medications (e.g. triptans, ergots) are indicated in the treatment of migraine but contraindicated in patients with ischaemic heart disease. Both disorders can produce severe head pain accompanied by nausea, and both can be triggered by exertion. Migraine-like headache may be triggered by angina treatment such as nitroglycerine.

10.7 Headache attributed to other disorder of homoeostasis

Description:

Headache caused by any disorder of homoeostasis not described above.

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. A disorder of homoeostasis other than those described above, and known to be able to cause headache, has been diagnosed
- C. Evidence of causation demonstrated by either or both of the following:
 1. headache has developed in temporal relation to the onset of the disorder of homoeostasis
 2. either or both of the following:
 - a) headache has significantly worsened in parallel with worsening of the disorder of homoeostasis
 - b) headache has significantly improved or resolved in parallel with improvement in or resolution of the disorder of homoeostasis
- D. Not better accounted for by another ICHD-3 diagnosis.

Comment:

Although relationships between headache and a variety of systemic and metabolic diseases have been proposed,

systematic evaluation of these relationships has not been performed and there is insufficient evidence on which to build operational diagnostic criteria.

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11. Headache or facial pain attributed to disorder of the cranium, neck, eyes, ears, nose, sinuses, teeth, mouth or other facial or cervical structure

- 11.1 Headache attributed to disorder of cranial bone
- 11.2 Headache attributed to disorder of the neck
 - 11.2.1 Cervicogenic headache
 - 11.2.2 Headache attributed to retropharyngeal tendonitis
 - 11.2.3 Headache attributed to craniocervical dystonia
- 11.3 Headache attributed to disorder of the eyes
 - 11.3.1 Headache attributed to acute glaucoma
 - 11.3.2 Headache attributed to refractive error
 - 11.3.3 Headache attributed to heterophoria or heterotropia (latent or persistent squint)
 - 11.3.4 Headache attributed to ocular inflammatory disorder
 - 11.3.5 Headache attributed to trochleitis
- 11.4 Headache attributed to disorder of the ears
- 11.5 Headache attributed to disorder of the nose or paranasal sinuses
 - 11.5.1 Headache attributed to acute rhinosinusitis
 - 11.5.2 Headache attributed to chronic or recurring rhinosinusitis
- 11.6 Headache attributed to disorder of the teeth or jaw
- 11.7 Headache attributed to temporomandibular disorder (TMD)
- 11.8 Head or facial pain attributed to inflammation of the stylohyoid ligament
- 11.9 Headache or facial pain attributed to other disorder of cranium, neck, eyes, ears, nose, sinuses, teeth, mouth or other facial or cervical structure

Coded elsewhere:

Headaches that are caused by head or neck trauma are classified under 5. *Headache attributed to trauma or injury to the head and/or neck.* This is true in particular for post-whiplash headache, despite the likely possibility that these headaches are attributable to pathology in the neck. Neuralgiform headaches manifesting with facial, neck and/or head pain are classified under 13. *Painful cranial neuropathies and other facial pains.*

General comment

Primary or secondary headache or both?

When a headache occurs for the first time in close temporal relation to a cranial, cervical, facial, neck, eye, ear, nose, sinus, dental or mouth disorder known to cause headache, it is coded as a secondary headache attributed to that disorder. This remains true when the new

headache has the characteristics of any of the primary headache disorders classified in Part one of ICHD-3 beta. When a *pre-existing* headache with the characteristics of a primary headache disorder becomes *chronic*, or is made significantly worse (usually meaning a two-fold or greater increase in frequency and/or severity), in close temporal relation to a cranial, cervical, facial, neck, eye, ear, nose, sinus, dental or mouth disorder, both the initial headache diagnosis and a diagnosis of 11. *Headache or facial pain attributed to disorder of the cranium, neck, eyes, ears, nose, sinuses, teeth, mouth or other facial or cervical structure* (or one of its subtypes) should be given, provided that there is good evidence that that disorder can cause headache.

Introduction

Disorders of the cervical spine and of other structures of the neck and head have not infrequently been regarded as common causes of headache, as many headaches seem to originate from the cervical, nuchal or occipital regions or are localized there. Degenerative changes in the cervical spine can be found in virtually all people over 40 years of age. However, large-scale controlled studies have shown that such changes are equally widespread among people with and people without headache. Spondylosis or osteochondrosis are therefore not conclusive as the explanation of headache. A similar situation applies to other widespread disorders: chronic sinusitis, temporomandibular disorders and refractive errors of the eyes.

Without specific criteria it would be possible for virtually any type of headache to be classified as 11. *Headache or facial pain attributed to disorder of the cranium, neck, eyes, ears, nose, sinuses, teeth, mouth or other facial or cervical structure.* It is not sufficient merely to list manifestations of headaches in order to define them, as these manifestations are not unique. The purpose of the criteria in this chapter is not to describe headaches in all their possible subforms, but rather to establish specific causal relationships between headaches and facial pain and the disorders of the cranium, neck, eyes, ears, nose, sinuses, teeth, mouth and other facial or cranial structures where these exist. For this reason it has been necessary to identify strict specific operational criteria for cervicogenic headache and other causes of headache described in this chapter. It is not possible here to take account of diagnostic tests that are unconfirmed or for which quality criteria have not been investigated. Instead, the aim of the revised criteria is to motivate the development of reliable and valid operational tests to establish specific causal relationships between headaches and craniocervical disorders.

For these reasons, and because of the variety of causative disorders dealt with in this chapter, it is difficult to describe a general set of criteria for headache and/or facial pain attributed to them. However, in most cases there is conformity with the following:

- A. Headache or facial pain fulfilling criterion C
- B. Clinical, laboratory and/or imaging evidence of a disorder or lesion of the cranium, neck, eyes, ears, nose, sinuses, teeth, mouth or other facial or cervical structure known to be able to cause headache
- C. Evidence that the pain can be attributed to the disorder or lesion
- D. Not better accounted for by another ICHD-3 diagnosis.

11.1 Headache attributed to disorder of cranial bone

Description:

Headache caused by a disorder or lesion of the cranial bones.

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. Clinical, laboratory and/or imaging evidence of a disorder or lesion of the cranial bones known to be able to cause headache
- C. Evidence of causation demonstrated by at least two of the following:
 1. headache has developed in temporal relation to the onset of the cranial bone disorder or appearance of the lesion
 2. either or both of the following:
 - a) headache has significantly worsened in parallel with worsening of the cranial bone disorder or lesion
 - b) headache has significantly improved in parallel with improvement in the cranial bone disorder or lesion
 3. headache is exacerbated by pressure applied to the cranial bone lesion
 4. headache is localized to the site of the cranial bone lesion
- D. Not better accounted for by another ICHD-3 diagnosis.

Comment:

Most disorders of the skull (e.g. congenital abnormalities, fractures, tumours, metastases) are usually not accompanied by headache. Exceptions of importance are osteomyelitis, multiple myeloma and Paget's disease. Headache may also be caused by lesions of the mastoid, and by petrositis.

11.2 Headache attributed to a disorder of the neck

Coded elsewhere:

Headache caused by neck trauma is classified under 5. *Headache attributed to trauma or injury to the head and/or neck* or one of its subtypes.

Description:

Headache caused by a disorder involving any structure in the neck, including bony, muscular and other soft tissue elements.

11.2.1 Cervicogenic headache

Coded elsewhere:

Headache causally associated with cervical myofascial pain sources (myofascial trigger points) may, if it meets other criteria, be coded as 2.1.1 *Infrequent episodic tension-type headache associated with pericranial tenderness*, 2.2.1 *Frequent episodic tension-type headache associated with pericranial tenderness* or 2.3.1 *Chronic tension-type headache associated with pericranial tenderness*. It seems appropriate to add an Appendix diagnosis A11.2.5 *Headache attributed to cervical myofascial pain*, and await evidence that this type of headache is more closely related to other cervicogenic headaches than to 2. *Tension-type headache*. Clearly, there are many cases that overlap these two categories, for which diagnosis can be challenging.

Description:

Headache caused by a disorder of the cervical spine and its component bony, disc and/or soft tissue elements, usually but not invariably accompanied by neck pain.

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. Clinical, laboratory and/or imaging evidence of a disorder or lesion within the cervical spine or soft tissues of the neck, known to be able to cause headache
- C. Evidence of causation demonstrated by at least two of the following:
 1. headache has developed in temporal relation to the onset of the cervical disorder or appearance of the lesion
 2. headache has significantly improved or resolved in parallel with improvement in or resolution of the cervical disorder or lesion
 3. cervical range of motion is reduced and headache is made significantly worse by provocative manoeuvres

4. headache is abolished following diagnostic blockade of a cervical structure or its nerve supply
- D. Not better accounted for by another ICHD-3 diagnosis.

Comments:

Features that tend to distinguish 11.2.1 *Cervicogenic headache* from 1. *Migraine* and 2. *Tension-type headache* include side-locked pain, provocation of typical headache by digital pressure on neck muscles and by head movement, and posterior-to-anterior radiation of pain. However, although these may be features of 11.2.1 *Cervicogenic headache*, they are not unique to it, and they do not necessarily define causal relationships. Migrainous features such as nausea, vomiting and photo/phonophobia may be present with 11.2.1 *Cervicogenic headache*, although to a generally lesser degree than in 1. *Migraine*, and may differentiate some cases from 2. *Tension-type headache*.

Tumours, fractures, infections and rheumatoid arthritis of the upper cervical spine have not been validated formally as causes of headache, but are nevertheless accepted as such when demonstrated to be so in individual cases. Cervical spondylosis and osteochondritis may or may not be valid causes fulfilling criterion B, depending on the individual case. When cervical myofascial pain is the cause, the headache should probably be coded under 2. *Tension-type headache*. However, awaiting further evidence, an alternative diagnosis of A11.2.5 *Headache attributed to cervical myofascial pain* is included in the Appendix.

Headache caused by upper cervical radiculopathy has been postulated and, considering the now well-understood convergence between upper cervical and trigeminal nociception, this is a logical cause of headache. Pending further evidence, this diagnosis is found in the Appendix as A11.2.4 *Headache attributed to upper cervical radiculopathy*.

11.2.2 Headache attributed to retropharyngeal tendonitis

Description:

Headache caused by inflammation or calcification in the retropharyngeal soft tissues, and usually brought on by stretching or compression of upper cervical prevertebral muscles.

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. Retropharyngeal tendonitis has been demonstrated by imaging evidence of abnormal swelling of prevertebral soft tissues at upper cervical spine levels

- C. Evidence of causation demonstrated by at least two of the following:
1. headache has developed in temporal relation to the onset of the retropharyngeal tendonitis
 2. either or both of the following:
 - a) headache has significantly worsened in parallel with progression of the retropharyngeal tendonitis
 - b) headache has significantly improved or resolved in parallel with improvement in or resolution of the retropharyngeal tendonitis
 3. headache is made significantly worse by extension of the neck, rotation of the head and/or swallowing
 4. there is tenderness over the spinous processes of the upper three cervical vertebrae
- D. Not better accounted for by another ICHD-3 diagnosis.

Comments:

Body temperature and erythrocyte sedimentation rate (ESR) are usually elevated in retropharyngeal tendonitis. Although retroflexion of the neck most consistently aggravates pain, the same usually occurs also with rotation of the head and swallowing. Tissues over the transverse processes of the upper three vertebrae are usually tender to palpation.

Calcification in prevertebral tissues is best seen on CT or MRI, but plain films of the neck can also reveal this. In several cases, amorphous calcific material has been aspirated from the swollen prevertebral tissues.

Upper carotid dissection (or another lesion in or around the carotid artery) should be ruled out before the diagnosis of 11.2.2 *Headache attributed to retropharyngeal tendonitis* is confirmed.

11.2.3 Headache attributed to craniocervical dystonia

Description:

Headache caused by dystonia involving neck muscles, with abnormal movements or defective posturing of the neck or head as a result of muscular hyperactivity.

Diagnostic criteria:

- A. Neck and posterior head pain fulfilling criterion C
- B. Craniocervical dystonia is demonstrated by abnormal movements or defective posturing of the neck or head as a result of muscular hyperactivity
- C. Evidence of causation demonstrated by at least two of the following:
1. headache has developed in temporal relation to the onset of craniocervical dystonia

2. headache has significantly worsened in parallel with progression of the craniocervical dystonia
 3. headache has significantly improved or resolved in parallel with improvement in or resolution of the craniocervical dystonia
 4. headache location corresponds to the location of the dystonic muscle(s)
- D. Not better accounted for by another ICHD-3 diagnosis.

Comments:

Focal dystonias of the head and neck accompanied by 11.2.3 *Headache attributed to craniocervical dystonia* are pharyngeal dystonia, spasmodic torticollis, mandibular dystonia, lingual dystonia and a combination of the cranial and cervical dystonias (segmental craniocervical dystonia).

Pain is presumably caused by local muscle contraction and secondary changes in sensitization.

11.3 Headache attributed to disorder of the eyes

Description:

Headache caused by a disorder involving one or both eyes.

11.3.1 Headache attributed to acute glaucoma

Description:

Headache, usually unilateral, caused by acute narrow-angle glaucoma and associated with other symptoms and clinical signs of this disorder.

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. Acute narrow-angle glaucoma has been diagnosed
- C. Evidence of causation demonstrated by at least two of the following:
 1. headache has developed in temporal relation to the onset of glaucoma
 2. headache has significantly worsened in parallel with progression of glaucoma
 3. headache has significantly improved or resolved in parallel with improvement in or resolution of glaucoma
 4. pain location includes the affected eye
- D. Not better accounted for by another ICHD-3 diagnosis.

Comment:

Acute glaucoma generally causes eye and/or periorbital pain, visual acuity loss (blurring), nausea and vomiting.

When intraocular pressure rises above 30 mmHg, the risk of permanent visual loss rises dramatically, which makes early diagnosis essential.

11.3.2 Headache attributed to refractive error

Description:

Headache caused by ocular refractive error(s), generally symptomatic after prolonged visual tasks.

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. Uncorrected or miscorrected refractive error(s) in one or both eyes
- C. Evidence of causation demonstrated by at least two of the following:
 1. headache has developed and/or significantly worsened in temporal relation to the onset or worsening of the refractive error(s)
 2. headache has significantly improved after correction of the refractive error(s)
 3. headache is aggravated by prolonged visual tasks at an angle or distance at which vision is impaired
 4. headache significantly improves when the visual task is discontinued
- D. Not better accounted for by another ICHD-3 diagnosis.

Comment:

Most patients with 11.3.2 *Headache attributed to refractive error* will seek advice from an ophthalmologist. Although refractive error is much less commonly a cause of headache than is generally believed, there is some evidence for it in children, as well as a number of supportive cases in adults.

11.3.3 Headache attributed to heterophoria or heterotropia (latent or persistent squint)

Description:

Headache caused by latent or persistent strabismus, usually occurring after prolonged visual tasks.

Diagnostic criteria:

- A. Frontal headache fulfilling criterion C
- B. Strabismus has been identified, with at least one of the following symptoms:
 1. blurred vision
 2. diplopia
 3. difficulty switching from near to far focus and/or vice versa

- C. Evidence of causation demonstrated by at least two of the following:
1. headache has developed in temporal relation to the onset of the strabismus, or led to its discovery
 2. headache has significantly improved after correction of the strabismus
 3. headache is aggravated by sustained visual tasks
 4. headache is alleviated by closing one eye and/or discontinuation of the visual task
- D. Not better accounted for by another ICHD-3 diagnosis.

Comment:

Most patients with 11.3.3 *Headache attributed to heterophoria or heterotropia* will seek advice from an ophthalmologist. There is little evidence for this cause of headache other than a number of supportive cases.

11.3.4 Headache attributed to ocular inflammatory disorder

Description:

Headache caused by ocular inflammatory conditions such as iritis, uveitis, scleritis or conjunctivitis and associated with other symptoms and clinical signs of the disorder.

Diagnostic criteria:

- A. Periorbital headache and eye pain fulfilling criterion C
- B. Clinical, laboratory and/or imaging evidence of ocular inflammatory disease such as iritis, uveitis, cyclitis, scleritis, choroiditis, conjunctivitis or corneal inflammation
- C. Evidence of causation demonstrated by at least two of the following:
 1. headache has developed in temporal relation to the onset of the ocular disorder
 2. either or both of the following:
 - a) headache has significantly worsened in parallel with worsening of the ocular disorder
 - b) headache has significantly improved or resolved in parallel with improvement in or resolution of the ocular disorder
 3. either or both of the following:
 - a) headache significantly improves with topical application of local anaesthetic agent to the eye
 - b) headache is aggravated by pressure applied to the eye
 4. in the case of a unilateral eye disorder, headache is localized ipsilateral to it

- D. Not better accounted for by another ICHD-3 diagnosis.

Comments:

Ocular inflammation takes many forms, and may be categorized variously by anatomical site (e.g. iritis, cyclitis, choroiditis), by course (i.e. acute, subacute, chronic), by presumed cause (e.g. endogenous or exogenous infectious agents, lens-related, traumatic) or by type of inflammation (granulomatous, non-granulomatous).

Because of nociceptive field overlap and convergence (leading to complex pain referral), any ocular source of pain may lead to headache in any region. Nevertheless, if the eye disorder is unilateral, headache is likely to be localized ipsilateral to it.

11.3.5 Headache attributed to trochleitis

Coded elsewhere:

An episode of migraine triggered by trochleitis is coded as 1. *Migraine* or one of its subtypes.

Description:

Headache, usually frontal and/or periorbital in location, with or without eye pain, caused by peritrochlear inflammation. It is often exacerbated by downward movements of the eye.

Diagnostic criteria:

- A. Periorbital and/or frontal headache fulfilling criterion C
- B. Clinical and/or imaging evidence of trochlear inflammation
- C. Evidence of causation demonstrated by at least two of the following:
 1. unilateral ocular pain
 2. headache is exacerbated by movement of the eye, particularly downward in adduction
 3. headache is significantly improved by injection of local anaesthetic or steroid agent into the peritrochlear region
 4. in the case of a unilateral trochleitis, headache is localized ipsilateral to it
- D. Not better accounted for by another ICHD-3 diagnosis.

Comments:

Trochleitis, defined as inflammation of the trochlea and/or sheath of the superior oblique muscle, can lead to eye pain and frontal headache that are aggravated by movements of the eye involving the superior

oblique muscle. Although not common, it is not rare, and must be considered when evaluating unilateral peri-orbital head pain.

Trochleitis can also trigger an episode of 1. *Migraine*, which is coded accordingly.

11.4 Headache attributed to disorder of the ears

Description:

Headache caused by an inflammatory, neoplastic or other disorder of one or both ears and associated with other symptoms and/or clinical signs of the disorder.

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. Clinical, laboratory and/or imaging evidence of an infectious, neoplastic or other irritative disorder or lesion of one or both ears, known to be able to cause headache
- C. Evidence of causation demonstrated by at least two of the following:
 1. headache has developed in temporal relation to the onset of the ear disorder or appearance of the ear lesion
 2. either or both of the following:
 - a) headache has significantly worsened in parallel with worsening or progression of the ear disorder or lesion
 - b) headache has significantly improved or resolved in parallel with improvement in or resolution of the ear disorder or lesion
 3. headache is exacerbated by pressure applied to the affected ear(s) or periauricular structures
 4. in the case of a unilateral ear disorder or lesion, headache is localized ipsilateral to it
- D. Not better accounted for by another ICHD-3 diagnosis.

Comment:

Because of nociceptive field overlap and convergence in the nociceptive pathways of the head and neck, it seems clear that a painful disorder or lesion of the ear may lead to headache. It is highly unlikely that headache in such conditions can occur in the absence of ear pain, the typical manifestation of otological pathology.

11.5 Headache attributed to disorder of the nose or paranasal sinuses

Previously used term:

The term 'sinus headache' is outmoded because it has been applied both to primary headaches and headache

supposedly attributed to various conditions involving nasal or sinus structures.

Description:

Headache caused by a disorder of the nose and/or paranasal sinuses and associated with other symptoms and/or clinical signs of the disorder.

11.5.1 Headache attributed to acute rhinosinusitis

Description:

Headache caused by acute rhinosinusitis and associated with other symptoms and/or clinical signs of this disorder.

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. Clinical, nasal endoscopic and/or imaging evidence of acute rhinosinusitis
- C. Evidence of causation demonstrated by at least two of the following:
 1. headache has developed in temporal relation to the onset of the rhinosinusitis
 2. either or both of the following:
 - a) headache has significantly worsened in parallel with worsening of the rhinosinusitis
 - b) headache has significantly improved or resolved in parallel with improvement in or resolution of the rhinosinusitis
 3. headache is exacerbated by pressure applied over the paranasal sinuses
 4. in the case of a unilateral rhinosinusitis, headache is localized ipsilateral to it
- D. Not better accounted for by another ICHD-3 diagnosis.

Comments:

1. *Migraine* and 2. *Tension-type headache* can be mistaken for 11.5.1 *Headache attributed to acute rhinosinusitis* because of similarity in location of the headache and, in the case of migraine, because of the commonly accompanying nasal autonomic symptoms. The presence or absence of purulent nasal discharge and/or other features diagnostic of acute rhinosinusitis help to differentiate these conditions. However, an episode of 1. *Migraine* may be triggered or exacerbated by nasal or sinus pathology.

Pain as a result of pathology in the nasal mucosa or related structures is usually perceived as frontal or facial, but may be referred more posteriorly. Simply finding pathological changes on imaging of acute rhinosinusitis, correlating with the patient's pain description, is not enough to secure the diagnosis of 11.5.1

Headache attributed to acute rhinosinusitis. Treatment response to local anaesthesia is compelling evidence, but may also not be pathognomonic.

11.5.2 Headache attributed to chronic or recurring rhinosinusitis

Description:

Headache caused by a chronic infectious or inflammatory disorder of the paranasal sinuses and associated with other symptoms and/or clinical signs of the disorder.

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. Clinical, nasal endoscopic and/or imaging evidence of current or past infection or other inflammatory process within the paranasal sinuses
- C. Evidence of causation demonstrated by at least two of the following:
 - 1. headache has developed in temporal relation to the onset of chronic rhinosinusitis
 - 2. headache waxes and wanes in parallel with the degree of sinus congestion, drainage and other symptoms of chronic rhinosinusitis
 - 3. headache is exacerbated by pressure applied over the paranasal sinuses
 - 4. in the case of a unilateral rhinosinusitis, headache is localized ipsilateral to it
- D. Not better accounted for by another ICHD-3 diagnosis.

Comment:

It has been controversial whether or not chronic sinus pathology can produce persistent headache. Recent studies seem to support such causation.

11.6 Headache attributed to disorder of the teeth or jaw

Description:

Headache caused by a disorder involving the teeth and/or jaw.

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. Clinical and/or imaging evidence of a disorder or lesion of one or more teeth and/or the jaw, known to be able to cause headache
- C. Evidence of causation demonstrated by at least two of the following:
 - 1. headache has developed in temporal relation to the onset of the disorder or appearance of the lesion

- 2. either or both of the following:
 - a) headache has significantly worsened in parallel with worsening or progression of the disorder or lesion
 - b) headache has significantly improved or resolved in parallel with improvement in or resolution of the disorder or lesion
- 3. headache is exacerbated by pressure applied to the lesion
- 4. in the case of a unilateral disorder or lesion, headache is localized ipsilateral to it
- D. Not better accounted for by another ICHD-3 diagnosis.

Comment:

Disorders of the teeth usually cause toothache and/or facial pain, and those causing headache are rare. Pain from the teeth may be referred, however, and cause diffuse headache. The most common cause of 11.6 *Headache attributed to disorder of the teeth or jaw* is periodontitis or pericoronitis as the result of infection or traumatic irritation around a partially erupted lower wisdom tooth.

11.7 Headache attributed to temporomandibular disorder (TMD)

Description:

Headache caused by a disorder involving structures in the temporomandibular region.

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. Clinical and/or imaging evidence of a pathological process affecting the temporomandibular joint (TMJ), muscles of mastication and/or associated structures
- C. Evidence of causation demonstrated by at least two of the following:
 - 1. headache has developed in temporal relation to the onset of the temporomandibular disorder
 - 2. either or both of the following:
 - a) headache has significantly worsened in parallel with progression of the temporomandibular disorder
 - b) headache has significantly improved or resolved in parallel with improvement in or resolution of the temporomandibular disorder
 - 3. the headache is produced or exacerbated by active jaw movements, passive movements through the range of motion of the jaw and/or provocative manœuvres applied to

temporomandibular structures such as pressure on the TMJ and surrounding muscles of mastication

4. headache, when unilateral, is ipsilateral to the side of the temporomandibular disorder

D. Not better accounted for by another ICHD-3 diagnosis.

Comments:

11.7 *Headache attributed to temporomandibular disorder (TMD)* is usually most prominent in the preauricular areas of the face, masseter muscles and/or temporal regions. Pain generators include disk displacements, joint osteoarthritis, joint hypermobility and regional myofascial pain. 11.7 *Headache attributed to temporomandibular disorder (TMD)* tends to be unilateral when the temporomandibular complex is the generator of pain, but may be bilateral when muscular involvement is present. Pain referral to the face is common.

Diagnosis of TMD can be difficult, with some controversy regarding the relative importance of clinical and radiographic evidence. The use of diagnostic criteria evolved by the International RDC/TMD Consortium Network and Orofacial Pain Special Interest Group is recommended.

There is some overlap between 11.7 *Headache attributed to temporomandibular disorder (TMD)* as a result of muscular tension and 2. *Tension-type headache*. When the diagnosis of TMD is uncertain, the headache should be coded as 2. *Tension-type headache* or one of its subtypes (presumably with pericranial muscle tenderness).

11.8 Headache or facial pain attributed to inflammation of the stylohyoid ligament

Previously used term:
Eagle's syndrome.

Description:

Unilateral headache, with neck, pharyngeal and/or facial pain, caused by inflammation of the stylohyoid ligament and usually provoked or exacerbated by head turning.

Diagnostic criteria:

- A. Any head, neck, pharyngeal and/or facial pain fulfilling criterion C
- B. Radiological evidence of calcified or elongated stylohyoid ligament
- C. Evidence of causation demonstrated by at least two of the following:
 1. pain is provoked or exacerbated by digital palpation of the stylohyoid ligament

2. pain is provoked or exacerbated by head turning

3. pain is significantly improved by local injection of local anaesthetic agent to the stylohyoid ligament, or by styloidectomy

4. pain is ipsilateral to the inflamed stylohyoid ligament

D. Not better accounted for by another ICHD-3 diagnosis.

Comment:

11.8 *Headache or facial pain attributed to inflammation of the stylohyoid ligament* is generally perceived in the oropharynx, neck and/or face, but some patients experience more diffuse headache.

11.9 Headache or facial pain attributed to other disorder of cranium, neck, eyes, ears, nose, sinuses, teeth, mouth or other facial or cervical structure

Description:

Headache and/or facial pain caused by a disorder of the cranium, neck, eyes, ears, nose, sinuses, teeth, mouth or other facial or cervical structure not described above.

Diagnostic criteria:

- A. Any headache and/or facial pain fulfilling criterion C
- B. A disorder or lesion of cranium, neck, eyes, ears, nose, sinuses, teeth, mouth or other facial or cervical structure not described above but known to be able to cause headache has been diagnosed
- C. Evidence of causation demonstrated by at least two of the following:
 1. headache and/or facial pain has developed in temporal relation to the onset of the disorder or appearance of the lesion
 2. either or both of the following:
 - a) headache and/or facial pain has significantly worsened in parallel with progression of the disorder or lesion
 - b) headache and/or facial pain has significantly improved or resolved in parallel with improvement in or resolution of the disorder or lesion
 3. headache and/or facial pain is exacerbated by pressure applied to the lesion
 4. headache and/or facial pain is localized in accordance with the site of the lesion
- D. Not better accounted for by another ICHD-3 diagnosis.

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11.8 Head or facial pain attributed to inflammation of the stylohyoid ligament

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12. Headache attributed to psychiatric disorder

12.1 Headache attributed to somatization disorder

12.2 Headache attributed to psychotic disorder

Coded elsewhere:

Headache attributed to a substance use disorder (e.g. dependence), headache attributed to substance withdrawal, headache attributed to acute intoxication and headache attributed to medication overuse are all coded under 8. *Headache attributed to a substance or its withdrawal.*

General comment

Primary or secondary headache or both?

Headaches are common, and so are psychiatric disorders. Therefore, frequent coexistence by chance alone is expected.

When a headache occurs for the first time in close temporal relation to a psychiatric disorder, however, a causal relationship may be present. If causation is confirmed, the headache must be coded as a secondary headache attributed to that disorder. This remains true when the new headache has the characteristics of any of the primary headache disorders classified in Part one of ICHD-3 beta. When a *pre-existing* headache with the characteristics of a primary headache disorder becomes chronic, or is made significantly worse (usually meaning a two-fold or greater increase in frequency and/or severity), in close temporal relation to a psychiatric disorder, both the initial headache diagnosis and a diagnosis of 12. *Headache attributed to psychiatric disorder* (or one of its subtypes) should be given, provided that there is good evidence that that disorder can cause headache. When a causal relationship cannot be confirmed, the pre-existing primary headache and the psychiatric disorder are diagnosed separately.

Chronic headache attributed to and persisting after resolution of a psychiatric disorder has not yet been described.

Introduction

Evidence supporting psychiatric causes of headache remains scarce. Thus, the diagnostic categories in this section of the classification are limited to those few cases in which a headache occurs in the context and as a direct consequence of a psychiatric condition known to be symptomatically manifested by headache.

Diagnostic criteria must be restrictive enough not to include false positive cases, but must set the threshold low enough to admit the majority of affected patients.

In the vast majority of cases of 12. *Headache attributed to psychiatric disorder*, the diagnosis is based on personal evaluation of case histories and physical examinations rather than objective diagnostic biomarkers.

Headache disorders may, of course, occur in association with psychiatric disorders without any causal connection. Headache disorders occur coincidentally with a number of psychiatric disorders, including depressive disorders (major depressive disorders, single episode or recurrent; persistent depressive disorder), anxiety disorders (separation anxiety disorder, panic disorder, social anxiety disorder and generalized anxiety disorder) and trauma- and stress-related disorders (reactive attachment disorder, acute stress disorder, post-traumatic stress disorder, adjustment disorders). In such cases, when there is no evidence of a causal relationship, both a primary headache diagnosis and a separate psychiatric diagnosis should be made.

Epidemiological data nonetheless show that headache and psychiatric disorders occur together at frequencies higher than would be expected by chance. Confounding factors may in part explain these apparent comorbidities. For example, patients who have one diagnosis are more likely to be diagnosed with other conditions simply because they receive more medical scrutiny. Genuine comorbidities also are possible, such as between migraine and depression, indicating the likelihood of an underlying association. Putative causal associations include the headache causing the psychiatric condition, the psychiatric condition causing the headache, reciprocal influence between the headache and the psychiatric condition and a common underlying factor causing both.

Although it is suggested that headache occurring exclusively in association with some common psychiatric disorders such as depressive disorders, anxiety disorders and trauma/stress-related disorders might be considered as attributed to these disorders, because of uncertainties concerning the causal relationships and relative lack of evidence in this context, criteria for headaches attributed to these psychiatric disorders have been included only in the Appendix. Further clarification of the mechanisms underlying these causal associations is necessary for sturdy conclusions.

Evidence suggests that the presence of a comorbid psychiatric disorder tends to worsen the course of 1. *Migraine* and/or 2. *Tension-type headache* by increasing the frequency and severity of the headache and/or making it less responsive to treatment. Thus, identification and treatment of any comorbid psychiatric condition is important for the proper management of these headaches. In children and adolescents, primary headache disorders (migraine, episodic tension-type headache and especially chronic tension-type headache) are often comorbid with psychiatric disorder.

Sleep disorders, post-traumatic stress disorder, social anxiety disorder (school phobia) attention-deficit/hyperactivity disorder (ADHD), conduct disorder, learning disorder, enuresis, encopresis and tic disorder should be carefully looked for and treated when found, considering their negative burden in disability and prognosis of paediatric headache.

To ascertain whether a headache should be attributed to a psychiatric disorder, it is necessary to determine whether or not there is a concomitant psychiatric disorder. It is recommended to inquire about commonly comorbid psychiatric symptoms such as depressive and anxiety disorders in all headache patients. When a psychiatric disorder is suspected to be a possible cause of the headache condition, then an evaluation by an experienced psychiatrist or psychologist is recommended.

12.1 Headache attributed to somatization disorder

Description:

Headache occurring as part of the symptomatic presentation of a somatization disorder.

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. A diagnosis has been made of somatization disorder characterized by both of the following:
 - 1. a history of multiple physical symptoms beginning before age 30 years, which either have not been fully explained by a known medical condition or, when there is a related medical condition, are in excess of what would be expected from the history, physical examination, or laboratory findings
 - 2. during the course of the disorder, all of the following:
 - a) at least four pain symptoms from or during four different sites or functions (e.g. from head, chest, back, abdomen, joints, extremities and/or rectum, and/or during menstruation, sexual intercourse and/or urination)
 - b) at least two gastrointestinal symptoms other than pain (e.g. nausea, bloating, vomiting other than during pregnancy, diarrhoea and/or intolerance of several different foods)
 - c) at least one sexual symptom other than pain (e.g. sexual indifference, erectile or ejaculatory dysfunction, irregular menses, excessive menstrual bleeding and/or vomiting throughout pregnancy)
 - d) at least one pseudoneurological symptom not limited to pain (e.g. conversion symptoms such as impaired coordination or balance,

paralysis or localized weakness, difficulty swallowing or lump in the throat, aphonia, urinary retention, hallucinations, loss of touch or pain sensation, double vision, blindness, deafness, seizures, dissociation symptoms such as amnesia and/or loss of consciousness other than fainting)

- C. Evidence of causation demonstrated by at least one of the following:
 - 1. headache has evolved or significantly worsened in intensity in parallel with the development of other somatic symptoms attributed to somatization disorder
 - 2. constant or remitting headache parallels in time the fluctuation of other somatic symptoms attributed to somatization disorder
 - 3. headache has remitted in parallel with remission of the other somatic symptoms attributed to somatization disorder
- D. Not better accounted for by another ICHD-3 diagnosis.

Comments:

Somatization disorder is characterized by a combination of multiple distressing symptoms and an excessive or maladaptive response to these symptoms or associated health concerns. Symptoms include gastric and/or other intestinal problems or dysfunctions, back pain, pain in the arms, legs or joints, headaches, chest pain and/or dyspnoea, dizziness, feeling tired and/or having low energy, and sleep troubles. The patient's suffering is authentic, whether or not it is medically explained. Patients typically experience distress and a high level of functional impairment. The symptoms may or may not accompany diagnosed general medical disorders or psychiatric disorders. There may be a high level of medical care utilization, which rarely alleviates the patient's concerns. From the clinician's point of view, many of these patients seem unresponsive to therapies, and new interventions or therapies may only exacerbate the presenting symptoms or lead to new side effects and complications. Some patients feel that their medical assessment and treatment have been inadequate.

It should be noted that somatization disorder per se is not included in the fifth edition of the *Diagnostic and Statistical Manual of Mental Disorders (DSM-5)*, the latest revision of the American Psychiatric Association's diagnostic manual, scheduled for release in May 2013; it has been replaced by the category *Somatic Symptom Disorder*, characterized by one or more somatic symptoms associated with disproportionate and persistent thoughts about the seriousness of one's symptoms, persistently high level of anxiety about health or symptoms or excessive time and

energy devoted to these symptoms or health concerns. Given the enormous heterogeneity of this category (i.e. it includes both individuals with headaches who have disproportionate concerns about the seriousness of the headache as well as classic cases of somatization disorder with a lifelong pattern of multiple somatic symptoms including headache), it was decided that it would be possible to assert attribution only when headache was part of a larger pattern of multiple somatic complaints. Thus, ICHD-3 beta continues to refer to the DSM-IV definition of somatization disorder.

12.2 Headache attributed to psychotic disorder

Description:

Headache as a manifestation of a delusion whose content involves a mechanism that the patient believes explains the headache (e.g. headache is the result of a device implanted in the head by aliens).

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. Presence of a delusion whose content involves a mechanism that would explain the headache (e.g. the patient believes that a device has been implanted into his or her head, which is causing a headache, or that he or she has a brain tumour causing headache despite irrefutable proof to the contrary)
- C. Evidence of causation demonstrated by either or both of the following:
 1. headache has developed with or after the onset of the delusion
 2. headache has remitted after remission of the delusion
- D. Not better accounted for by another ICHD-3 diagnosis.

Comments:

Delusions are false fixed beliefs, based on incorrect inferences about reality, that are firmly held despite obvious proof to the contrary. They may involve a false belief that a serious medical condition (e.g. brain tumour or aneurysm) is present and causes the headache, despite repeated proofs and appropriate authoritative reassurances that no such medical condition is present. The content of the delusion may be more bizarre, such as the idea of a transmitter being surgically implanted into one's head and causing the headache.

When the patient first develops a headache (e.g. one of the primary headache disorders classified in Part one of ICHD-3 beta) and then develops a delusional explanation for the headache, such as its being a

result of a brain tumour despite no medical evidence in support of that belief, the headache may not be *attributed* to the psychiatric disorder; instead, the headache should be coded as a primary headache disorder and the patient given the additional psychiatric diagnosis of *delusional disorder, somatic type*.

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Part three

Painful cranial neuropathies, other facial pains and other headaches

13. Painful cranial neuropathies and other facial pains
14. Other headache disorders

13. Painful cranial neuropathies and other facial pains

- 13.1 Trigeminal neuralgia
 - 13.1.1 Classical trigeminal neuralgia
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Introduction

Pain in the head and neck is mediated by afferent fibres in the trigeminal nerve, nervus intermedius, glossopharyngeal and vagus nerves and the upper cervical roots via the occipital nerves. Stimulation of these nerves by compression, distortion, exposure to cold or other forms of irritation or by a lesion in central pathways may give rise to stabbing or constant pain felt in the area innervated.

The cause may be clear, such as infection by Herpes zoster or a structural abnormality demonstrated by imaging, but in some cases there may be no cause apparent for neuralgic pain.

Trigeminal and glossopharyngeal neuralgias present a problem of terminology. When pain is found to result from compression of the nerve by a vascular loop at operation, the neuralgia should strictly be regarded as secondary. As many patients do not come to operation, it remains uncertain as to whether they have primary or secondary neuralgias. For this reason the term *classical* rather than *primary* has been applied to those patients with a typical history even though a vascular source of compression may be discovered during its course. The term *secondary* can then be reserved for those patients in whom a neuroma or similar lesion is demonstrated.

Definitions of terms used in this chapter¹:

Pain: An unpleasant sensory and emotional experience associated with actual or potential tissue damage, or described in terms of such damage.

Neuropathic pain: Pain (qv) caused by a lesion or disease of the somatosensory nervous system.

Central neuropathic pain: Pain (qv) caused by a lesion or disease of the central somatosensory nervous system.

Peripheral neuropathic pain: Pain (qv) caused by a lesion or disease of the peripheral somatosensory nervous system.

Neuropathy: A disturbance of function or pathological change in a nerve or nerves (in one nerve: mononeuropathy; in several nerves: mononeuropathy multiplex; when diffuse and bilateral: polyneuropathy). The term *neuropathy* is not intended to cover neurapraxia, neurotmesis, section of a nerve, disturbances of a nerve as a result of transient impact such as a blow, stretching or epileptic discharge (the term *neurogenic* applies to pain attributed to such temporary perturbations).

Neuralgia: Pain in the distribution of a nerve or nerves. (Common usage, especially in Europe, often implies a paroxysmal quality, but the term *neuralgia* should not be reserved for paroxysmal pains.)

Note:

1. International Association for the Study of Pain: Taxonomy. URL: <http://www.iasp-pain.org>.

13.1 Trigeminal neuralgia

Description:

A disorder characterized by recurrent unilateral brief electric shock-like pains, abrupt in onset and termination, limited to the distribution of one or more divisions

of the trigeminal nerve and triggered by innocuous stimuli. It may develop without apparent cause or be a result of another diagnosed disorder. There may or may not be, additionally, persistent background facial pain of moderate intensity.

13.1.1 Classical trigeminal neuralgia

Previously used term:

Tic douloureux.

Description:

Trigeminal neuralgia developing without apparent cause other than neurovascular compression.

Diagnostic criteria:

- A. At least three attacks of unilateral facial pain fulfilling criteria B and C
- B. Occurring in one or more divisions of the trigeminal nerve, with no radiation beyond the trigeminal distribution
- C. Pain has at least three of the following four characteristics:
 - 1. recurring in paroxysmal attacks lasting from a fraction of a second to 2 minutes
 - 2. severe intensity
 - 3. electric shock-like, shooting, stabbing or sharp in quality
 - 4. precipitated by innocuous stimuli to the affected side of the face¹
- D. No clinically evident neurological deficit²
- E. Not better accounted for by another ICHD-3 diagnosis.

Notes:

- 1. Some attacks may be, or appear to be, spontaneous, but there must be at least three that are precipitated in this way to meet this criterion.
- 2. Hypoaesthesia or hypoalgesia in the affected trigeminal region always indicates axonal damage. When either is present, there is trigeminal neuropathy and extensive diagnostic work-up is necessary to exclude symptomatic cases. There are some patients with hyperalgesia in the painful region, which should not necessarily lead to a diagnosis of trigeminal neuropathy because it may reflect the patient's increased attention to the painful side.

Comments:

The term *classical* (rather than *primary*) trigeminal neuralgia is used because, according to current

evidence, 13.1.1 *Classical trigeminal neuralgia* is caused by neurovascular compression, most frequently by the superior cerebellar artery. Imaging (preferably MRI) should be done to exclude secondary causes and, in the majority of patients, to demonstrate neurovascular compression of the trigeminal nerve.

Many patients with 13.1.1 *Classical trigeminal neuralgia* have a memorable onset of pain.

13.1.1 *Classical trigeminal neuralgia* usually appears in the second or third divisions. The pain never crosses to the opposite side but it may rarely occur bilaterally. Following a painful paroxysm there is usually a refractory period during which pain cannot be triggered. When very severe, the pain often evokes contraction of the muscle of the face on the affected side (*tic douloureux*). Mild autonomic symptoms such as lacrimation and/or redness of the eye may be present.

The duration of pain attacks can change over time and become more prolonged as well as severe. They can result in psychosocial dysfunction, significantly impairing quality of life and often leading to weight loss.

Between paroxysms, most patients are asymptomatic. In the subform 13.1.1.2 *Classical trigeminal neuralgia with concomitant persistent facial pain*, there is prolonged background pain in the affected area.

13.1.1 *Classical trigeminal neuralgia* may be preceded by a period of atypical continuous pain termed *pre-trigeminal neuralgia* in the literature.

13.1.1.1 Classical trigeminal neuralgia, purely paroxysmal

Description:

Trigeminal neuralgia without persistent background facial pain.

Diagnostic criteria:

- A. Recurrent attacks of unilateral facial pain fulfilling criteria for 13.1.1 *Classical trigeminal neuralgia*
- B. No persistent facial pain between attacks
- C. Not better accounted for by another ICHD-3 diagnosis.

Comment:

13.1.1.1 *Classical trigeminal neuralgia, purely paroxysmal* is usually responsive, at least initially, to pharmacotherapy (especially carbamazepine or oxcarbazepine).

13.1.1.2 Classical trigeminal neuralgia with concomitant persistent facial pain

Previously used terms:

Atypical trigeminal neuralgia; trigeminal neuralgia type 2.

Description:

Trigeminal neuralgia with persistent background facial pain.

Diagnostic criteria:

- A. Recurrent attacks of unilateral facial pain fulfilling criteria for 13.1.1 *Classical trigeminal neuralgia*
- B. Persistent facial pain of moderate intensity in the affected area
- C. Not better accounted for by another ICHD-3 diagnosis.

Comments:

13.1.1.2 *Classical trigeminal neuralgia with concomitant persistent facial pain* has been referred to as atypical trigeminal neuralgia or, recently, as trigeminal neuralgia type 2.

Central sensitization may account for the persistent facial pain. Neurovascular compression on MRI is less likely to be demonstrated. 13.1.1.2 *Classical trigeminal neuralgia with concomitant persistent facial pain* responds poorly to conservative treatment and to neurosurgical interventions. It is less likely to be triggered by innocuous stimuli.

13.1.2 *Painful trigeminal neuropathy**Description:*

Head and/or facial pain in the distribution of one or more branches of the trigeminal nerve caused by another disorder and indicative of neural damage. The pain is highly variable in quality and intensity according to the cause.

13.1.2.1 *Painful trigeminal neuropathy attributed to acute Herpes zoster**Description:*

Unilateral head and/or facial pain of less than 3 months' duration in the distribution of one or more branches of the trigeminal nerve, caused by and associated with other symptoms and/or clinical signs of acute Herpes zoster.

Diagnostic criteria:

- A. Unilateral head and/or facial pain lasting <3 months and fulfilling criterion C
- B. Either or both of the following:
 - 1. herpetic eruption has occurred in the territory of a trigeminal nerve branch or branches
 - 2. varicella zoster virus DNA has been detected in the CSF by polymerase chain reaction

- C. Evidence of causation demonstrated by both of the following:
 - 1. pain preceded the herpetic eruption by <7 days
 - 2. pain is located in the distribution of the same trigeminal nerve branch or branches
- D. Not better accounted for by another ICHD-3 diagnosis.

Comments:

Herpes zoster affects the trigeminal ganglion in 10–15% of cases, with the ophthalmic division being singled out in some 80% of patients. Rarely, pain is not followed by an eruption or rash (*zoster sine herpette*). The diagnosis in such cases is confirmed by polymerase chain reaction detection of varicella zoster virus DNA in the cerebrospinal fluid.

13.1.2.1 *Painful trigeminal neuropathy attributed to acute Herpes zoster* is usually burning, stabbing/shooting, tingling or aching, and accompanied by cutaneous allodynia.

Ophthalmic herpes may be associated with IIIrd, IVth and VIth cranial nerve palsies. Herpes zoster is common in immunocompromised patients, occurring in about 10% of those with lymphoma and 25% of patients with Hodgkin's disease.

13.1.2.2 *Post-herpetic trigeminal neuropathy**Previously used term:*

Post-herpetic trigeminal neuralgia.

Description:

Unilateral head and/or facial pain persisting or recurring for at least 3 months in the distribution of one or more branches of the trigeminal nerve, with variable sensory changes, caused by Herpes zoster.

Diagnostic criteria:

- A. Unilateral head and/or facial pain persisting or recurring for ≥ 3 months and fulfilling criterion C
- B. History of acute Herpes zoster affecting a trigeminal nerve branch or branches
- C. Evidence of causation demonstrated by both of the following:
 - 1. pain developed in temporal relation to the acute Herpes zoster
 - 2. pain is located in the distribution of the same trigeminal nerve branch or branches
- D. Not better accounted for by another ICHD-3 diagnosis.

Comments:

Following acute Herpes zoster, post-herpetic neuralgia is more prevalent in the elderly.

The first division of the trigeminal nerve is most commonly affected in 13.1.2.2 *Post-herpetic trigeminal neuropathy*, but the second and third divisions can be involved also. Typically the pain is burning and itching. Itching of affected areas may be very prominent and extremely bothersome. Sensory abnormalities and allodynia are usually present in the territory involved. Pale or light purple scars may be present as sequelae of the herpetic eruption.

13.1.2.3 *Painful post-traumatic trigeminal neuropathy*

Previously used term:

Anaesthesia dolorosa.

Coded elsewhere:

Here are described *painful* post-traumatic neuropathies; most trigeminal nerve injuries do not result in pain and therefore have no place in ICHD-3 beta.

Description:

Unilateral facial or oral pain following trauma to the trigeminal nerve, with other symptoms and/or clinical signs of trigeminal nerve dysfunction.

Diagnostic criteria:

- A. Unilateral facial and/or oral pain fulfilling criterion C
- B. History of an identifiable traumatic event¹ to the trigeminal nerve, with clinically evident positive (hyperalgesia, allodynia) and/or negative (hypoesthesia, hypoalgesia) signs of trigeminal nerve dysfunction
- C. Evidence of causation demonstrated by both of the following:
 - 1. pain is located in the distribution of the same trigeminal nerve
 - 2. pain has developed within 3–6 months of the traumatic event
- D. Not better accounted for by another ICHD-3 diagnosis.

Note:

- 1. The traumatic event may be mechanical, chemical, thermal or caused by radiation.

Comment:

Pain duration ranges widely from paroxysmal to constant, and may be mixed. Specifically following

radiation-induced postganglionic injury, neuropathy may appear after more than 3 months.

13.1.2.4 *Painful trigeminal neuropathy attributed to multiple sclerosis (MS) plaque*

Description:

Unilateral head and/or facial pain in the distribution of a trigeminal nerve and with the characteristics of classical trigeminal neuralgia, induced by a multiple sclerosis plaque affecting the trigeminal nerve root and associated with other symptoms and/or clinical signs of multiple sclerosis.

Diagnostic criteria:

- A. Head and/or facial pain with the characteristics of 13.1.1 *Classical trigeminal neuralgia* with or without concomitant persistent facial pain, but not necessarily unilateral
- B. Multiple sclerosis (MS) has been diagnosed
- C. An MS plaque affecting the trigeminal nerve root has been demonstrated by MRI or by routine electrophysiological studies (blink reflex or trigeminal evoked potentials) indicating impairment of the affected trigeminal nerve(s)
- D. Not better accounted for by another ICHD-3 diagnosis.

Comments:

Current studies indicate that about 7% of MS patients have a syndrome that is similar to 13.1.1 *Classical trigeminal neuralgia*. However, symptoms of trigeminal neuralgia are very rarely a presenting feature of MS.

Symptoms of 13.1.2.4 *Painful trigeminal neuropathy attributed to multiple sclerosis (MS) plaque* are more likely to be bilateral than those of 13.1.1 *Classical trigeminal neuralgia*.

Patients with 13.1.2.4 *Painful trigeminal neuropathy attributed to multiple sclerosis (MS) plaque* benefit less from pharmacological interventions than those with 13.1.1 *Classical trigeminal neuralgia*.

13.1.2.5 *Painful trigeminal neuropathy attributed to space-occupying lesion*

Description:

Unilateral head and/or facial pain in the distribution of a trigeminal nerve and with the characteristics of classical trigeminal neuralgia, induced by contact between the affected trigeminal nerve and a space-occupying lesion.

Diagnostic criteria:

- A. Unilateral head and/or facial pain with the characteristics of 13.1.1 *Classical trigeminal neuralgia* with or without concomitant persistent facial pain and fulfilling criterion C
- B. A space-occupying lesion, and contact between the lesion and the affected trigeminal nerve, have been demonstrated by imaging
- C. Pain has developed after contact occurred between the lesion and the trigeminal nerve, or led to its discovery
- D. Not better accounted for by another ICHD-3 diagnosis.

Comment:

Patients with 13.1.2.5 *Painful trigeminal neuropathy attributed to space-occupying lesion* have clinically detectable sensory signs or electrophysiological abnormalities.

13.1.2.6 *Painful trigeminal neuropathy attributed to other disorder*

Diagnostic criteria:

- A. Head and/or facial pain with the characteristics of 13.1.1 *Classical trigeminal neuralgia* with or without concomitant persistent facial pain, but not necessarily unilateral
- B. A disorder, other than those described above but known to be capable of causing painful trigeminal neuropathy, has been diagnosed
- C. Pain has developed after onset of the disorder, or led to its discovery
- D. Not better accounted for by another ICHD-3 diagnosis.

13.2 Glossopharyngeal neuralgia

Previously used term:

Vagoglossopharyngeal neuralgia.

Description:

A severe, transient, stabbing, unilateral pain experienced in the ear, base of the tongue, tonsillar fossa and/or beneath the angle of the jaw. It is commonly provoked by swallowing, talking and/or coughing, and may remit and relapse in the fashion of classical trigeminal neuralgia.

Diagnostic criteria:

- A. At least three attacks of unilateral pain fulfilling criteria B and C

- B. Pain is located in the posterior part of the tongue, tonsillar fossa, pharynx, beneath the angle of the lower jaw and/or in the ear
- C. Pain has at least three of the following four characteristics:
 1. recurring in paroxysmal attacks lasting from a few seconds to 2 minutes
 2. severe intensity
 3. shooting, stabbing or sharp in quality
 4. precipitated by swallowing, coughing, talking or yawning
- D. No clinically evident neurological deficit
- E. Not better accounted for by another ICHD-3 diagnosis.

Comments:

13.2 *Glossopharyngeal neuralgia* is felt in the distributions of the auricular and pharyngeal branches of the vagus nerve as well as branches of the glossopharyngeal nerve. Prior to its development, unpleasant sensations can be experienced in affected areas for weeks to several months.

13.2 *Glossopharyngeal neuralgia* is less severe than 13.1.1 *Classical trigeminal neuralgia* but can be bad enough for patients to lose weight. These two disorders can occur together.

In rare cases, attacks of pain are associated with vagal symptoms such as cough, hoarseness, syncope and/or bradycardia. Some authors have proposed distinguishing between pharyngeal, otalgic and vagal subtypes of neuralgia, and suggested using the term *vagoglossopharyngeal neuralgia* when pain is accompanied by asystole, convulsions and syncope.

Imaging may show neurovascular compression of the glossopharyngeal nerve. There are single reports of secondary glossopharyngeal neuropathy caused by neck trauma, multiple sclerosis, tonsillar or regional tumours, cerebello-pontine angle tumours and Arnold-Chiari malformation.

13.2 *Glossopharyngeal neuralgia* is usually responsive, at least initially, to pharmacotherapy, especially antiepileptics. It has been suggested that application of local anaesthetic to the tonsil and pharyngeal wall can prevent attacks for a few hours.

13.3 Nervus intermedius (facial nerve) neuralgia

Description:

A rare disorder characterized by brief paroxysms of pain felt deeply in the auditory canal, sometimes radiating to the parieto-occipital region. It may develop without apparent cause or as a complication of Herpes zoster.

13.3.1 *Classical nervus intermedius neuralgia*

Description:

Nervus intermedius neuralgia developing without apparent cause.

Diagnostic criteria:

- A. At least three attacks of unilateral pain fulfilling criteria B and C
- B. Pain is located in the auditory canal, sometimes radiating to the parieto-occipital region
- C. Pain has at least three of the following four characteristics:
 1. recurring in paroxysmal attacks lasting from a few seconds to minutes
 2. severe intensity
 3. shooting, stabbing or sharp in quality
 4. precipitated by stimulation of a trigger area in the posterior wall of the auditory canal and/or periauricular region
- D. No clinically evident neurological deficit
- E. Not better accounted for by another ICHD-3 diagnosis.

Comments:

Disorders of lacrimation, salivation and/or taste sometimes accompany the pain of 13.3.1 *Classical nervus intermedius neuralgia*. In view of the complex and overlapping innervation of the external ear, deriving from the trigeminal (auriculotemporal nerve), facial (nervus intermedius), glossopharyngeal, vagus and second cranial nerves, attribution of neuralgias to a single nerve may not be easy in this body region if a specific neurovascular contact cannot be visualized.

The pain of 13.3.1 *Classical nervus intermedius neuralgia* can result in psychological effects and significantly impair quality of life.

13.3.2 *Secondary nervus intermedius neuropathy attributed to acute Herpes zoster*

Previously used term:

Ramsay Hunt syndrome.

Description:

Unilateral pain felt deeply in the auditory canal, sometimes radiating to the parieto-occipital region, associated with facial paresis and caused by Herpes zoster of the nervus intermedius.

Diagnostic criteria:

- A. Unilateral facial pain fulfilling criterion C

- B. Herpetic eruption has occurred in the ear and/or oral mucosa, in the territory of the nervus intermedius
- C. Evidence of causation demonstrated by both of the following:
 1. pain has preceded the herpetic eruption by <7 days
 2. pain is localized to the distribution of the nervus intermedius
- D. Clinical features of peripheral facial paresis
- E. Not better accounted for by another ICHD-3 diagnosis.

Comments:

The most frequent cause of secondary nervus intermedius neuropathy is Herpes zoster. A very few cases are described resulting from other disorders such as neurovascular compression, and there are rare familial cases associated with occipital neuralgia.

In Ramsay Hunt syndrome, zoster lesions in the ear or oral mucosa accompanied by facial paresis are pathognomonic, but the original description pointed to additional symptoms such as vertigo, tinnitus, acoustic disturbances and nausea.

13.3.2 *Secondary nervus intermedius neuropathy attributed to Herpes zoster* should be treated with cortisone and acyclovir as early as possible.

13.4 *Occipital neuralgia*

Description:

Unilateral or bilateral paroxysmal, shooting or stabbing pain in the posterior part of the scalp, in the distribution of the greater, lesser or third occipital nerves, sometimes accompanied by diminished sensation or dysaesthesia in the affected area and commonly associated with tenderness over the involved nerve(s).

Diagnostic criteria:

- A. Unilateral or bilateral pain fulfilling criteria B-E
- B. Pain is located in the distribution of the greater, lesser and/or third occipital nerves
- C. Pain has two of the following three characteristics:
 1. recurring in paroxysmal attacks lasting from a few seconds to minutes
 2. severe intensity
 3. shooting, stabbing or sharp in quality
- D. Pain is associated with both of the following:
 1. dysaesthesia and/or allodynia apparent during innocuous stimulation of the scalp and/or hair
 2. either or both of the following:
 - a) tenderness over the affected nerve branches

b) trigger points at the emergence of the greater occipital nerve or in the area of distribution of C2

- E. Pain is eased temporarily by local anaesthetic block of the affected nerve
- F. Not better accounted for by another ICHD-3 diagnosis.

Comments:

The pain of 13.4 *Occipital neuralgia* may reach the fronto-orbital area through trigeminocervical inter-neuronal connections in the trigeminal spinal nuclei.

13.4 *Occipital neuralgia* must be distinguished from occipital referral of pain arising from the atlantoaxial or upper zygapophyseal joints or from tender trigger points in neck muscles or their insertions.

13.5 Optic neuritis

Previously used term:
Retrobulbar neuritis.

Description:

Pain behind one or both eyes caused by demyelination of the optic nerve(s) and accompanied by impairment of central vision.

Diagnostic criteria:

- A. Unilateral or bilateral headache fulfilling criterion C
- B. Clinical, electrophysiological, imaging and/or laboratory evidence confirming the presence of optic neuritis
- C. Evidence of causation demonstrated by both of the following:
 1. headache has developed in temporal relationship to optic neuritis
 2. headache has either or both of the following features:
 - a) localized in retro-orbital, orbital, frontal and/or temporal regions
 - b) aggravated by eye movement
- D. Not better accounted for by another ICHD-3 diagnosis.

Comments:

13.5 *Optic neuritis* is often a manifestation of multiple sclerosis. Pain may precede impairment of vision. Clinical series report the prevalence of head pain in optic neuritis to be about 90%.

There is a high incidence (90%) of pain with eye movement when there is an orbital segment enhancement in cranial MRI, and a high

probability (70%) of no such pain when there is no enhancement.

13.6 Headache attributed to ischaemic ocular motor nerve palsy

Description:

Unilateral frontal and/or periorbital pain caused by and associated with other symptoms and/or clinical signs of ischaemic paresis of the ipsilateral IIIrd, IVth or VIth cranial nerve.

Diagnostic criteria:

- A. Unilateral headache fulfilling criterion C
- B. Clinical and imaging findings confirming an ischaemic ocular motor nerve palsy
- C. Evidence of causation demonstrated by both of the following:
 1. headache has developed in temporal relation to the motor nerve palsy
 2. headache is localized around the ipsilateral brow and eye
- D. Not better accounted for by another ICHD-3 diagnosis.

Comments:

The majority of ocular motor nerve palsies are painful, regardless of the presence or absence of diabetes. 13.6 *Headache attributed to ischaemic ocular motor nerve palsy* can occur prior to or concurrently with the onset of diplopia.

Pain is most frequent in patients with IIIrd nerve palsies, less so in VIth nerve paresis and least frequent in cases of IVth nerve paresis.

13.7 Tolosa-Hunt syndrome

Description:

Unilateral orbital pain associated with paresis of one or more of the IIIrd, IVth and/or VIth cranial nerves caused by a granulomatous inflammation in the cavernous sinus, superior orbital fissure or orbit.

Diagnostic criteria:

- A. Unilateral headache fulfilling criterion C
- B. Both of the following:
 1. granulomatous inflammation of the cavernous sinus, superior orbital fissure or orbit, demonstrated by MRI or biopsy
 2. paresis of one or more of the ipsilateral IIIrd, IVth and/or VIth cranial nerves

- C. Evidence of causation demonstrated by both of the following:
 1. headache has preceded paresis of the IIIrd, IVth and/or VIth nerves by ≤ 2 weeks, or developed with it
 2. headache is localized around the ipsilateral brow and eye
- D. Not better accounted for by another ICHD-3 diagnosis.

Comments:

Some reported cases of 13.7 *Tolosa-Hunt syndrome* had additional involvement of the Vth nerve (commonly the first division) or optic, VIIth or VIIIth nerves. Sympathetic innervation of the pupil is occasionally affected. The syndrome has been caused by granulomatous material in the cavernous sinus, superior orbital fissure or orbit in some biopsied cases.

Careful follow-up is required to exclude other causes of painful ophthalmoplegia such as tumours, vasculitis, basal meningitis, sarcoid or diabetes mellitus.

Pain and paresis of 13.7 *Tolosa-Hunt syndrome* resolve when it is treated adequately with corticosteroids.

13.8 Paratrigeminal oculosympathetic (Raeder's) syndrome

Description:

Constant, unilateral pain in the distribution of the ophthalmic division of the trigeminal nerve, sometimes extending to the maxillary division, accompanied by Horner's syndrome and caused by a disorder in the middle cranial fossa or of the carotid artery.

Diagnostic criteria:

- A. Constant, unilateral headache fulfilling criterion C
- B. Imaging evidence of underlying disease of either the middle cranial fossa or of the ipsilateral carotid artery
- C. Evidence of causation demonstrated by both of the following:
 1. headache has developed in temporal relation to the onset of the underlying disorder
 2. headache has either or both of the following features:
 - a) localized to the distribution of the ophthalmic division of the trigeminal nerve, with or without spread to the maxillary division
 - b) aggravated by eye movement
- D. Ipsilateral Horner's syndrome
- E. Not better accounted for by another ICHD-3 diagnosis.

Comment:

The original description of 13.8 *Paratrigeminal oculosympathetic (Raeder's) syndrome* was useful because the involvement of oculosympathetic fibres indicated a lesion of the middle cranial fossa. It is regarded as a classical example of clinico-anatomical methodology in the early 20th century. Whether the term *Raeder's syndrome* should be used today is heavily debated, but painful Horner's syndrome is still considered by some authors to be a diagnostically useful indication of a middle cranial fossa lesion or of carotid artery dissection.

13.9 Recurrent painful ophthalmoplegic neuropathy

Previously used term:

Ophthalmoplegic migraine.

Description:

Repeated attacks of paresis of one or more ocular cranial nerves (commonly the IIIrd), with ipsilateral headache.

Diagnostic criteria:

- A. At least two attacks fulfilling criterion B
- B. Unilateral headache accompanied by ipsilateral paresis of one, two or all three ocular motor nerves
- C. Orbital, parasellar or posterior fossa lesion has been excluded by appropriate investigation
- D. Not better accounted for by another ICHD-3 diagnosis.

Comments:

The old and inappropriate term *ophthalmoplegic migraine* was rejected because this syndrome is not migrainous but rather a recurrent painful neuropathy.

Recent data suggest that headache can develop up to 14 days prior to ocular motor paresis. Gadolinium enhancement or nerve thickening can be demonstrated using MRI. Treatment with corticosteroids is beneficial in some patients.

13.10 Burning mouth syndrome (BMS)

Previously used terms:

Stomatodynia, or glossodynia when confined to the tongue.

Description:

An intraoral burning or dysaesthetic sensation, recurring daily for more than 2 hours per day over more than 3 months, without clinically evident causative lesions.

Diagnostic criteria:

- A. Oral pain fulfilling criteria B and C
- B. Recurring daily for >2 hours per day for >3 months
- C. Pain has both of the following characteristics:
 1. burning quality
 2. felt superficially in the oral mucosa
- D. Oral mucosa is of normal appearance and clinical examination including sensory testing is normal
- E. Not better accounted for by another ICHD-3 diagnosis.

Comments:

The pain of 13.10 *Burning mouth syndrome (BMS)* is usually bilateral and its intensity fluctuates. The most common site is the tip of the tongue. Subjective dryness of the mouth, dysaesthesia and altered taste may be present.

There is a high menopausal female prevalence, and some studies show comorbid psychosocial and psychiatric disorders. Recent laboratory and brain imaging investigations have indicated changes in central and peripheral nervous systems.

Whether *secondary burning mouth syndrome attributed to a local* (candidiasis, lichen planus, hyposalivation) *or systemic disorder* (medication induced, anaemia, deficiencies of vitamin B12 or folic acid, Sjögren's syndrome, diabetes) should be considered as an entity is a matter for debate. Current evidence does not justify inclusion even in the Appendix.

13.11 Persistent idiopathic facial pain (PIFP)

Previously used term:
Atypical facial pain.

Description:

Persistent facial and/or oral pain, with varying presentations but recurring daily for more than 2 hours per day over more than 3 months, in the absence of clinical neurological deficit.

Diagnostic criteria:

- A. Facial and/or oral pain fulfilling criteria B and C
- B. Recurring daily for >2 hours per day for >3 months
- C. Pain has both of the following characteristics:
 1. poorly localized, and not following the distribution of a peripheral nerve
 2. dull, aching or nagging quality
- D. Clinical neurological examination is normal
- E. A dental cause has been excluded by appropriate investigations

- F. Not better accounted for by another ICHD-3 diagnosis.

Comments:

A wide variety of words are used to describe the character of 13.11 *Persistent idiopathic facial pain (PIFP)* but it is most often depicted as dull, nagging or aching. It can have sharp exacerbations, and is aggravated by stress. Pain may be described as either deep or superficial. With time, it may spread to a wider area of the craniocervical region.

13.11 *Persistent idiopathic facial pain (PIFP)* may be comorbid with other pain conditions such as chronic widespread pain and irritable bowel syndrome. In addition, it presents with high levels of psychiatric comorbidity and psychosocial disability.

A continuum seems to exist from 13.11 *Persistent idiopathic facial pain (PIFP)* induced by insignificant trauma to 13.1.2.3 *Painful post-traumatic trigeminal neuropathy* caused obviously by significant insult to the peripheral nerves. 13.11 *Persistent idiopathic facial pain (PIFP)* may originate from a minor operation or injury to the face, maxillae, teeth or gums but persist after healing of the initial noxious event and without any demonstrable local cause. However, psychophysical or neurophysiological tests may demonstrate sensory abnormalities.

The term *atypical odontalgia* has been applied to a continuous pain in one or more teeth or in a tooth socket after extraction, in the absence of any usual dental cause. This is thought to be a subform of 13.11 *Persistent idiopathic facial pain (PIFP)*, although it is more localized, the mean age at onset is younger and genders are more balanced. Based on the history of trauma, atypical odontalgia may also be a subform of 13.1.2.3 *Painful post-traumatic trigeminal neuropathy*. These subforms, if they exist, have not been sufficiently studied to propose diagnostic criteria.

13.12 Central neuropathic pain*Description:*

Unilateral or bilateral craniocervical pain with variable presentation, with or without sensory changes, of central origin. Depending on the cause, it may be constant or remitting and relapsing.

*13.12.1 Central neuropathic pain attributed to multiple sclerosis (MS)**Description:*

Unilateral or bilateral craniocervical pain with variable presentation, with or without sensory changes, attributed to a demyelinating lesion of the central ascending

connections of the trigeminal nerve in a person with multiple sclerosis. It commonly remits and relapses.

Diagnostic criteria:

- A. Facial and/or head pain fulfilling criterion C
- B. Multiple sclerosis (MS) has been diagnosed, with MRI demonstration of a demyelinating lesion in the brain stem or ascending projections of the trigeminal nuclei
- C. Pain has developed in temporal relation to the demyelinating lesion, or led to its discovery
- D. Not better accounted for by another ICHD-3 diagnosis.

Comment:

Non-painful sensory abnormalities (usually dysaesthesia but also hypoaesthesia, anaesthesia, hypoalgesia, paraesthesia, etc.) may coexist with pain in 13.12.1 *Central neuropathic pain attributed to multiple sclerosis (MS)*. Pain may be paroxysmal, as in 13.1.2.4 *Painful trigeminal neuropathy attributed to multiple sclerosis (MS) plaque*, or continuous.

13.12.2 *Central post-stroke pain (CPSP)*

Description:

Usually unilateral facial and/or head pain, with varying presentations involving parts or all of the cranio-cervical region and associated with impaired sensation, occurring within 6 months of and caused by stroke. It is not explicable by a lesion of the peripheral trigeminal or other cranial or cervical nerves.

Diagnostic criteria:

- A. Facial and/or head pain fulfilling criterion C
- B. Ischaemic or haemorrhagic stroke has occurred
- C. Evidence of causation demonstrated by both of the following:
 - 1. pain has developed within 6 months after the stroke
 - 2. imaging (usually MRI) has demonstrated a vascular lesion in an appropriate site
- D. Not better accounted for by another ICHD-3 diagnosis.

Comments:

13.12.2 *Central post-stroke pain (CPSP)* is attributed to a lesion of the ascending projections of the trigeminal nuclei. Cervical spinothalamic pathways and cortical processing may also play a significant role.

Therefore, symptoms may also involve the trunk and limbs of the affected side.

Cranio-cervical pain following a thalamic lesion is part of a hemisindrome. With lateral medullary lesions, hemifacial pain may occur in isolation but is more often accompanied by crossed hemidysaesthesia.

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14. Other headache disorders

14.1 Headache not elsewhere classified

14.2 Headache unspecified

Introduction

In order to make this classification exhaustive there are, in appropriate cases, subcategories for conditions that fulfil all but one criterion for specific disorders. Still there may be headaches that cannot fit into any of the existing chapters because they are being described for the first time, or because there simply is not enough information available. This chapter is intended for these types or subtypes of headaches.

14.1 Headache not elsewhere classified

Previously used term:

Headache not classifiable.

Diagnostic criteria:

- A. Headache with characteristic features suggesting that it is a unique diagnostic entity
- B. Does not fulfil criteria for any of the headache disorders described above.

Comment:

Several new headache entities have been described in the time between the first edition of *The International Classification of Headache Disorders* and this third edition. It is anticipated that there are more entities still to be described. Such headaches, until classified, can be coded as 14.1 *Headache not elsewhere classified*.

14.2 Headache unspecified

Previously used term:

Headache not classifiable.

Diagnostic criteria:

- A. Headache is or has been present
- B. Not enough information is available to classify the headache at any level of this classification.

Comment:

It is also apparent that a diagnosis must be made in a large number of patients where very little information is available, allowing only to state that they have headache but not which type of headache. Such patients are coded as 14.2 *Headache unspecified*. This code, however, must never be used as an excuse for not gathering detailed information about a headache when such information is available. It should be used only in situations where information cannot be obtained because the patient is dead, unable to communicate or unavailable.

Appendix

- A1. Migraine
- A2. Tension-type headache (alternative criteria)
- A3. Trigeminal-autonomic cephalalgias (TACs)
- A4. Other primary headache disorders
- A5. Headache attributed to trauma or injury to the head and/or neck
- A6. Headache attributed to cranial or cervical vascular disorder
- A7. Headache attributed to non-vascular intracranial disorder
- A8. Headache attributed to a substance or its withdrawal
- A9. Headache attributed to infection
- A10. Headache attributed to disorder of homeostasis
- A11. Headache or facial pain attributed to disorder of the cranium, neck, eyes, ears, nose, sinuses, teeth, mouth or other facial or cervical structure
- A12. Headache attributed to psychiatric disorder

- A1. Migraine**
- A1.1 Migraine without aura
 - A1.1.1 Pure menstrual migraine without aura
 - A1.1.2 Menstrually related migraine without aura
 - A1.1.3 Non-menstrual migraine without aura
- A1.2 Migraine with aura (alternative criteria)
 - A1.2.1 Migraine with typical aura (alternative criteria)
- A1.3 Chronic migraine (alternative criteria)
 - A1.3.1 Chronic migraine with pain-free periods
 - A1.3.2 Chronic migraine with continuous pain
- A1.4 Complications of migraine
 - A1.4.5 Migraine aura status
- A1.6 Episodic syndromes that may be associated with migraine
 - A1.6.4 Infantile colic
 - A1.6.5 Alternating hemiplegia of childhood
 - A1.6.6 Vestibular migraine
- A2. Tension-type headache (alternative criteria)**
- A3. Trigeminal-autonomic cephalalgias (TACs)**
- A3.6 Undifferentiated trigeminal autonomic cephalalgia
- A4. Other primary headache disorders**
- A4.11 Epicrania fugax
- A5. Headache attributed to trauma or injury to the head and/or neck**
- A5.1 Acute headache attributed to traumatic injury to the head
 - A5.1.1.1 Delayed-onset acute headache attributed to moderate or severe traumatic injury to the head
 - A5.1.2.1 Delayed-onset acute headache attributed to mild traumatic injury to the head
- A5.2 Persistent headache attributed to traumatic injury to the head
 - A5.2.1.1 Delayed-onset persistent headache attributed to moderate or severe traumatic injury to the head
 - A5.2.2.1 Delayed-onset persistent headache attributed to mild traumatic injury to the head
- A5.7 Headache attributed to radiosurgery of the brain
- A5.8 Acute headache attributed to other trauma or injury to the head and/or neck
- A5.9 Persistent headache attributed to other trauma or injury to the head and/or neck
- A6. Headache attributed to cranial or cervical vascular disorder**
- A6.10 Persistent headache attributed to past cranial or cervical vascular disorder
- A7. Headache attributed to non-vascular intracranial disorder**
- A7.6 Headache attributed to epileptic seizure
 - A7.6.3 Post-electroconvulsive therapy (ECT) headache
- A7.9 Persistent headache attributed to past non-vascular intracranial disorder
- A8. Headache attributed to a substance or its withdrawal**
- A8.4 Persistent headache attributed to past use of or exposure to a substance
- A9. Headache attributed to infection**
- A9.1 Headache attributed to intracranial infection
 - A9.1.3.3 Persistent headache attributed to past intracranial fungal or other parasitic infection
 - A9.1.6 Headache attributed to other infective space-occupying lesion
- A9.3 Headache attributed to human immunodeficiency virus (HIV) infection
- A10. Headache attributed to disorder of homeostasis**
- A10.7 Head and/or neck pain attributed to orthostatic (postural) hypotension
- A10.8 Headache attributed to other disorder of homeostasis
 - A10.8.1 Headache attributed to travel in space
 - A10.8.2 Headache attributed to other metabolic or systemic disorder
- A10.9 Persistent headache attributed to past disorder of homeostasis

- A11. Headache or facial pain attributed to disorder of the cranium, neck, eyes, ears, nose, sinuses, teeth, mouth or other facial or cervical structure**
- A11.2 Headache attributed to disorder of the neck
 - A11.2.4 Headache attributed to upper cervical radiculopathy
 - A11.2.5 Headache attributed to cervical myofascial pain
 - A11.5 Headache attributed to disorder of the nose or paranasal sinuses
 - A11.5.3 Headache attributed to disorder of the nasal mucosa, turbinates or septum
- A12. Headache attributed to psychiatric disorder**
- A12.3 Headache attributed to depressive disorder
 - A12.4 Headache attributed to separation anxiety disorder
 - A12.5 Headache attributed to panic disorder
 - A12.6 Headache attributed to specific phobia
 - A12.7 Headache attributed to social anxiety disorder (social phobia)
 - A12.8 Headache attributed to generalized anxiety disorder
 - A12.9 Headache attributed to post-traumatic stress disorder
 - A12.10 Headache attributed to acute stress disorder

Introduction

An Appendix was first added to the second edition of *The International Classification of Headache Disorders* (ICHD-II). It had several purposes, which are retained in ICHD-3 beta.

The primary purpose of the Appendix is to present research criteria for a number of novel entities that have not been sufficiently validated by research conducted so far. The experience of the experts in the Classification Committee, and publications of variable quality, suggest that there are still a number of diagnostic entities that are believed to be real but for which better scientific evidence must be presented before they can be formally accepted. Therefore, as happened between ICHD-II and ICHD-3 beta, it is anticipated

that some disorders now in the Appendix will move into the main body of the classification at the next revision.

In a few places the Appendix presents alternative sets of diagnostic criteria to those in the main body of the classification. This is again because clinical experience and a certain amount of published evidence suggest that the alternative criteria may be preferable, but the committee does not yet feel that the evidence is sufficient to change the main classification.

Finally, the Appendix is used as a first step in eliminating disorders historically included as diagnostic entities in previous editions of ICHD, but for which sufficient evidence has still not been published.

A1. Migraine

A1.1 Migraine without aura

A1.1.1 Pure menstrual migraine without aura

Diagnostic criteria:

- A. Attacks, in a menstruating woman,¹ fulfilling criteria for 1.1 *Migraine without aura* and criterion B below
- B. Documented and prospectively recorded evidence over at least three consecutive cycles has confirmed that attacks occur exclusively on day 1 ± 2 (i.e. days -2 to $+3$)² of menstruation¹ in at least two out of three menstrual cycles and at no other times of the cycle.

Notes:

1. For the purposes of ICHD-3 beta, menstruation is considered to be endometrial bleeding resulting from either the normal menstrual cycle or from the withdrawal of exogenous progestogens, as in the use of combined oral contraceptives or cyclical hormone replacement therapy.
2. The first day of menstruation is day 1 and the preceding day is day -1 ; there is no day 0.

A1.1.2 Menstrually related migraine without aura

Diagnostic criteria:

- A. Attacks, in a menstruating woman,¹ fulfilling criteria for 1.1 *Migraine without aura* and criterion B below
- B. Documented and prospectively recorded evidence over at least three consecutive cycles has confirmed that attacks occur on day 1 ± 2 (i.e. days -2 to $+3$)² of menstruation¹ in at least two out of three menstrual cycles, and additionally at other times of the cycle.

Notes:

1. For the purposes of ICHD-3 beta, menstruation is considered to be endometrial bleeding resulting from either the normal menstrual cycle or from the withdrawal of exogenous progestogens, as in the use of combined oral contraceptives or cyclical hormone replacement therapy.
2. The first day of menstruation is day 1 and the preceding day is day -1 ; there is no day 0.

A1.1.3 Non-menstrual migraine without aura

Diagnostic criteria:

- A. Attacks, in a menstruating woman,¹ fulfilling criteria for 1.1 *Migraine without aura* and criterion B below
- B. Attacks do not fulfil criterion B for A1.1.1 *Pure menstrual migraine without aura* or A1.1.2 *Menstrually related migraine without aura*.

Note:

1. For the purposes of ICHD-3 beta, menstruation is considered to be endometrial bleeding resulting from either the normal menstrual cycle or from the withdrawal of exogenous progestogens, as in the use of combined oral contraceptives or cyclical hormone replacement therapy.

Comments:

This subclassification of 1.1 *Migraine without aura* is clearly applicable only to menstruating women as defined above.

The importance of distinguishing between A1.1.1 *Pure menstrual migraine without aura* and A1.1.2 *Menstrually related migraine without aura* is that hormone prophylaxis is more likely to be effective for the former. Documented prospectively recorded evidence, kept for a minimum of three cycles, is necessary to confirm the diagnosis because many women over-report an association between attacks and menstruation.

Menstrual attacks are mostly migraine without aura. In women who have both 1.1 *Migraine without aura* and 1.2 *Migraine with aura*, the latter does not appear to be associated with menstruation.

The mechanism(s) of migraine may be different with endometrial bleeding resulting from the normal menstrual cycle and bleeding as a result of the withdrawal of exogenous progestogens (as occurs with combined oral contraception and cyclical hormone replacement therapy). For example, the endogenous menstrual cycle results from complex hormonal changes in the hypothalamic-pituitary-ovarian axis resulting in ovulation, which is suppressed by use of combined oral contraceptives. Therefore research should separate these subpopulations. Management strategies may also differ for these distinct subpopulations.

There is some evidence that menstrual migraine attacks, at least in some women, result from oestrogen withdrawal, although other hormonal and biochemical

changes at this time of the cycle may also be relevant. When pure menstrual migraine or menstrually related migraine is considered to be associated with exogenous oestrogen withdrawal, both codes A1.1.1 *Pure menstrual migraine without aura* or A1.1.2 *Menstrually related migraine without aura* and 8.3.3 *Oestrogen-withdrawal headache* should be used.

The menstrual relation may change over a woman's reproductive lifetime.

A1.2 Migraine with aura (alternative criteria)

Alternative diagnostic criteria:

- A. At least two attacks fulfilling criteria B and C
- B. One or more of the following fully reversible aura symptoms:
 1. visual
 2. sensory
 3. speech and/or language
 4. motor
 5. brainstem
 6. retinal
- C. At least three of the following six characteristics:
 1. at least one aura symptom spreads gradually over ≥ 5 minutes
 2. two or more aura symptoms occur in succession
 3. each individual aura symptom lasts 5–60 min¹
 4. at least one aura symptom is unilateral²
 5. at least one aura symptom is positive³
 6. the aura is accompanied, or followed within 60 minutes, by headache
- D. Not better accounted for by another ICHD-3 diagnosis.

Notes:

1. When, for example, three symptoms occur during an aura, the acceptable maximal duration is 3×60 minutes. Motor symptoms may last up to 72 hours.
2. Aphasia is always regarded as a unilateral symptom; dysarthria may or may not be.
3. Scintillations and pins and needles are positive symptoms of aura.

A1.2.1 Migraine with typical aura (alternative criteria)

Alternative diagnostic criteria:

- A. At least two attacks fulfilling criteria B and C
- B. Aura consisting of visual, sensory and or speech/language symptoms, each fully reversible, but no motor, brainstem or retinal symptoms

- C. At least three of the following six characteristics:
 1. at least one aura symptom spreads gradually over ≥ 5 minutes
 2. two or more aura symptoms occur in succession
 3. each individual aura symptom lasts 5–60 minutes¹
 4. at least one aura symptom is unilateral²
 5. at least one aura symptom is positive³
 6. the aura is accompanied, or followed within 60 minutes, by headache
- D. Not better accounted for by another ICHD-3 diagnosis.

Notes:

1. When for example three symptoms occur during an aura, the acceptable maximal duration is 3×60 minutes.
2. Aphasia is always regarded as a unilateral symptom; dysarthria may or may not be.
3. Scintillations and pins and needles are positive symptoms of aura.

A1.3 Chronic migraine (alternative criteria)

Alternative diagnostic criteria:

- A. Headache (tension-type-like and/or migraine-like) on ≥ 15 days per month for >3 months and fulfilling criteria B and C
- B. Occurring in a patient who has had at least five attacks fulfilling criteria B–D for 1.1 *Migraine without aura* and/or criteria B and C for 1.2 *Migraine with aura*
- C. On ≥ 8 days per month for >3 months fulfilling any of the following:
 1. criteria C and D for 1.1 *Migraine without aura*
 2. criteria B and C for 1.2 *Migraine with aura*
 3. criteria A and B for 1.5 *Probable migraine*
- D. Not better accounted for by another ICHD-3 diagnosis.

A1.3.1 Chronic migraine with pain-free periods

Diagnostic criteria:

- A. Headache fulfilling criteria for 1.3 *Chronic migraine* and criterion B below
- B. Interrupted by pain-free periods of >3 hours on ≥ 5 days per month which are not attributed to drug treatment.

A1.3.2 Chronic migraine with continuous pain

Diagnostic criteria:

- A. Headache fulfilling criteria for 1.3 *Chronic migraine* and criterion B below
- B. Not interrupted by pain-free periods of >3 hours on ≥ 5 days per month unless these are attributed to drug treatment.

A1.4 Complications of migraine

A1.4.5 Migraine aura status

Diagnostic criteria:

- A. Migraine fulfilling criteria for 1.2 *Migraine with aura* or one of its subtypes
- B. At least two auras occur per day for ≥ 3 days.

Comment:

Other neurological disorders including reversible cerebral vasoconstriction syndrome, posterior reversible encephalopathy syndrome and arterial dissection should be excluded by appropriate investigation.

A1.6 Episodic syndromes that may be associated with migraine

A1.6.4 Infantile colic

Description:

Excessive, frequent crying in a baby who appears to be otherwise healthy and well fed.

Diagnostic criteria:

- A. Recurrent episodes of irritability, fussing or crying from birth to 4 months of age, fulfilling criterion B
- B. Both of the following:
 1. episodes last for ≥ 3 hours per day
 2. episodes occur on ≥ 3 days per week for ≥ 3 weeks
- C. Not attributed to another disorder.

Comments:

Infantile colic affects one baby in five, but failure to thrive needs to be excluded.

Infants with colic have a higher likelihood of developing 1.1 *Migraine without aura* or 1.2 *Migraine with aura* later in life. Mothers with 1. *Migraine* have been found to be 2.5 times more likely to have infants with colic than mothers without. For fathers with 1.

Migraine, the likelihood of an infant with colic was increased two-fold.

A1.6.5 Alternating hemiplegia of childhood

Description:

Infantile attacks of hemiplegia involving each side alternately, associated with a progressive encephalopathy, other paroxysmal phenomena and mental impairment.

Diagnostic criteria:

- A. Recurrent attacks of hemiplegia alternating between the two sides of the body and fulfilling criteria B and C
- B. Onset before the age of 18 months
- C. At least one other paroxysmal phenomenon is associated with the bouts of hemiplegia or occurs independently, such as tonic spells, dystonic posturing, choreoathetoid movements, nystagmus or other ocular motor abnormalities and/or autonomic disturbances
- D. Evidence of mental and/or neurological deficit(s)
- E. Not attributed to another disorder.

Comment:

This is a heterogeneous neurodegenerative disorder. A relationship with migraine is suggested on clinical grounds. The possibility that it is an unusual form of epilepsy cannot be ruled out. Mutations in the ATP1A3 gene (encoding the sodium-potassium $[Na^+/K^+]ATPase$ $\alpha 3$ subunit) are likely to be responsible for at least 70% of cases.

A1.6.5 Vestibular migraine

Previously used terms:

Migraine-associated vertigo/dizziness; migraine-related vestibulopathy; migrainous vertigo.

Diagnostic criteria:

- A. At least five episodes fulfilling criteria C and D
- B. A current or past history of 1.1 *Migraine without aura* or 1.2 *Migraine with aura*¹
- C. Vestibular symptoms² of moderate or severe intensity,³ lasting between 5 minutes and 72 hours⁴
- D. At least 50% of episodes are associated with at least one of the following three migrainous features⁵:
 1. headache with at least two of the following four characteristics:
 - a) unilateral location
 - b) pulsating quality
 - c) moderate or severe intensity

- d) aggravation by routine physical activity
- 2. photophobia and phonophobia⁶
- 3. visual aura⁷
- E. Not better accounted for by another ICHD-3 diagnosis or by another vestibular disorder⁸.

Notes:

1. Code also for the underlying migraine diagnosis.
2. Vestibular symptoms, as defined by the Bárány Society's Classification of Vestibular Symptoms and qualifying for a diagnosis of A1.6.5 *Vestibular migraine*, include:
 - a) spontaneous vertigo:
 - (i) internal vertigo (a false sensation of self-motion);
 - (ii) external vertigo (a false sensation that the visual surround is spinning or flowing);
 - b) positional vertigo, occurring after a change of head position;
 - c) visually induced vertigo, triggered by a complex or large moving visual stimulus;
 - d) head motion-induced vertigo, occurring during head motion;
 - e) head motion-induced dizziness with nausea (dizziness is characterized by a sensation of disturbed spatial orientation; other forms of dizziness are currently not included in the classification of vestibular migraine).
3. Vestibular symptoms are rated *moderate* when they interfere with but do not prevent daily activities and *severe* when daily activities cannot be continued.
4. Duration of episodes is highly variable. About 30% of patients have episodes lasting minutes, 30% have attacks for hours and another 30% have attacks over several days. The remaining 10% have attacks lasting seconds only, which tend to occur repeatedly during head motion, visual stimulation or after changes of head position. In these patients, episode duration is defined as the total period during which short attacks recur. At the other end of the spectrum, there are patients who may take 4 weeks to recover fully from an episode. However, the core episode rarely exceeds 72 hours.
5. One symptom is sufficient during a single episode. Different symptoms may occur during different episodes. Associated symptoms may occur before, during or after the vestibular symptoms.
6. Phonophobia is defined as sound-induced discomfort. It is a transient and bilateral phenomenon that must be differentiated from recruitment, which is often unilateral and persistent. Recruitment leads

to an enhanced perception and often distortion of loud sounds in an ear with decreased hearing.

7. Visual auras are characterized by bright scintillating lights or zigzag lines, often with a scotoma that interferes with reading. Visual auras typically expand over 5–20 minutes and last for less than 60 minutes. They are often, but not always restricted to one hemifield. Other types of migraine aura, for example somatosensory or dysphasic aura, are not included as diagnostic criteria because their phenomenology is less specific and most patients also have visual auras.
8. History and physical examinations do not suggest another vestibular disorder *or* such a disorder has been considered but ruled out by appropriate investigations *or* such a disorder is present as a comorbid or independent condition, but episodes can be clearly differentiated. Migraine attacks may be induced by vestibular stimulation. Therefore, the differential diagnosis should include other vestibular disorders complicated by superimposed migraine attacks.

Comments:

Other symptoms

Transient auditory symptoms, nausea, vomiting, prostration and susceptibility to motion sickness may be associated with A1.6.5 *Vestibular migraine*. However, as they also occur with various other vestibular disorders, they are not included as diagnostic criteria.

Relation to migraine aura and migraine with brainstem aura

Both *migraine aura* and *migraine with brainstem aura* (formerly: *basilar-type migraine*) are terms defined by ICHD-3 beta. Only a minority of patients with A1.6.5 *Vestibular migraine* experience their vertigo in the time frame of 5–60 minutes as defined for an aura symptom. Even fewer have their vertigo immediately before headache starts, as required for 1.2.1.1 *Typical aura with headache*. Therefore, episodes of A1.6.5 *Vestibular migraine* cannot be regarded as migraine auras.

Although vertigo is reported by more than 60% of patients with 1.2.2 *Migraine with brainstem aura*, ICHD-3 beta requires at least two brainstem symptoms in addition to visual, sensory or dysphasic aura symptoms for this diagnosis. Fewer than 10% of patients with A1.6.5 *Vestibular migraine* fulfil these criteria. Therefore, A1.6.5 *Vestibular migraine* and 1.2.2 *Migraine with brainstem aura* are not synonymous, although individual patients may meet the diagnostic criteria for both disorders.

Relation to benign paroxysmal vertigo

Although A1.6.5 *Vestibular migraine* may start at any age, ICHD-3 beta specifically recognizes a childhood disorder, 1.6.2 *Benign paroxysmal vertigo*. The diagnosis requires five episodes of vertigo, occurring without warning and resolving spontaneously after minutes to hours. Between episodes, neurological examination, audiometry, vestibular functions and EEG must be normal. A unilateral throbbing headache may occur during attacks but is not a mandatory criterion. 1.6.2 *Benign paroxysmal vertigo* is regarded as one of the precursor syndromes of migraine. Therefore, previous migraine headaches are not required for diagnosis. As the classification of A1.6.5 *Vestibular migraine* does not involve any age limit, the diagnosis can be applied in children when the respective criteria are met. Only children with different types of vertigo attacks, for example short-duration episodes of less than 5 minutes and longer-lasting ones of more than 5 minutes, should receive both these diagnoses.

Overlap with Menière's disease

1. *Migraine* is more common in patients with Menière's disease than in healthy controls. Many patients with features of both Menière's disease and A1.6.5 *Vestibular migraine* have been reported. In fact, migraine and Menière's disease can be inherited as a symptom cluster. Fluctuating hearing loss, tinnitus and aural pressure may occur in A1.6.5 *Vestibular migraine*, but hearing loss does not progress to profound levels. Similarly, migraine headaches, photophobia and even migraine auras are common during Menière attacks. The pathophysiological relationship between A1.6.5 *Vestibular migraine* and Menière's disease remains uncertain. In the first year after onset of symptoms, differentiation between them may be challenging, as Menière's disease can be mono-symptomatic with only vestibular symptoms in the early stages of the disease.

When the criteria for Menière's disease are met, particularly hearing loss as documented by audiometry, Menière's disease should be diagnosed, even when migraine symptoms occur during the vestibular attacks. Only patients who have two different types of attacks, one fulfilling the criteria for A1.6.5 *Vestibular migraine* and the other for Menière's disease, should be diagnosed with both disorders. A future revision of ICHD may include a vestibular migraine/Menièr's disease overlap syndrome.

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A2. Tension-type headache (alternative criteria)

The following alternative criteria may be applied to A2.1 *Infrequent episodic tension-type headache*, A2.2 *Frequent episodic tension-type headache*, A2.3 *Chronic tension-type headache*. They define a core syndrome of tension-type headache. In other words these criteria are very specific but have low sensitivity.

Alternative diagnostic criteria:

- A. Episodes, or headache, fulfilling criterion A for [whichever of 2.1 *Infrequent episodic tension-type headache*, 2.2 *Frequent episodic tension-type headache* or 2.3 *Chronic tension-type headache*] and criteria B–D below
- B. Episodes, or headache, fulfil criterion B for [whichever of 2.1 *Infrequent episodic tension-type headache*, 2.2 *Frequent episodic tension-type headache* or 2.3 *Chronic tension-type headache*]
- C. Headache has at least three of the following four characteristics:
 1. bilateral location
 2. pressing/tightening (non-pulsating) quality

- 3. mild or moderate intensity
- 4. not aggravated by routine physical activity such as walking or climbing stairs
- D. No nausea, vomiting, photophobia or phonophobia
- E. Not better accounted for by another ICHD-3 diagnosis.

A3. Trigeminal-autonomic cephalalgias (TACs)

A3.6 Undifferentiated trigeminal autonomic cephalalgia

Description:

A trigeminal autonomic cephalalgia-like disorder occurring in children and adolescents with characteristics of the disorder not fully developed.

Comments:

Incomplete brain development may alter the presentation of trigeminal autonomic cephalalgias (TACs). Patients coded A3.6 *Undifferentiated trigeminal autonomic cephalalgia* would, typically, be children or adolescents whose headaches have characteristics strongly suggestive of a TAC, but mixed and incomplete; for example, they may have lateralized headache attacks lasting 30 minutes with autonomic features, but without the expected responses to indomethacin, oxygen or triptans.

Longitudinal studies are required to understand these presentations better and in order to propose criteria for their diagnosis.

A4. Other primary headache disorders

A4.11 Epicrania fugax

Description:

Brief paroxysmal head pain, with stabbing quality, describing a linear or zig-zag trajectory across the surface of one hemicranium.

Diagnostic criteria:

- A. Recurrent stabbing head pain attacks lasting 1–10 seconds, fulfilling criterion B
- B. The pain is felt to move across the surface of one hemicranium in a linear or zig-zag trajectory, commencing and terminating in the territories of different nerves
- C. Not better accounted for by another ICHD-3 diagnosis.

Comments:

A structural lesion must be excluded by history, physical examination and, when appropriate, investigation.

Patients with A4.11 *Epicrania fugax* describe their painful experience in terms of the trajectory of the pain between two distant points on the head surface, with motion from onset to termination taking just a few seconds. Such dynamic topography is a distinctive attribute that differentiates A4.11 *Epicrania fugax* from other epicranial headaches and neuralgias. The onset and termination points remain constant in each patient, with the pain strictly unilateral, although some patients have shifting sides. The pain usually moves forward, but backward radiation is also possible. Forward-moving pain starts in a posterior hemicranial area and tends to reach the ipsilateral eye or nose. Backward-moving pain starts in a frontal or periorbital area and tends to reach the occipital region. At the end of the attacks, ipsilateral autonomic signs such as lacrimation, conjunctival injection and/or rhinorrhoea may occur.

Although the attacks are mostly spontaneous, they may occasionally be triggered by touch on the point of onset, which may remain tender in between attacks.

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A5. Headache attributed to trauma or injury to the head and/or neck

A5.1 Acute headache attributed to traumatic injury to the head

Comment:

The current stipulation that headache must begin (or be reported to have begun) within 7 days of head injury (or awareness of the injury) is somewhat arbitrary. Some data suggest that headache may begin after a longer interval. Future studies should continue to

investigate the utility of diagnostic criteria for A5.1 *Acute headache attributed to traumatic injury to the head* that allow for headache to begin up to 30 days after the injury.

A5.1.1.1 Delayed-onset acute headache attributed to moderate or severe traumatic injury to the head

Diagnostic criteria:

- A. Any headache fulfilling criteria C and D
- B. Traumatic injury to the head has occurred, associated with at least one of the following:
 1. loss of consciousness for >30 minutes
 2. Glasgow Coma Scale (GCS) <13
 3. post-traumatic amnesia lasting >24 hours
 4. alteration in level of awareness for >24 hours
 5. imaging evidence of a traumatic head injury such as intracranial haemorrhage and/or brain contusion
- C. Time of onset of headache is uncertain, and/or headache is reported to have developed >7 days after all of the following:
 1. the head injury
 2. regaining of consciousness following the head injury (when applicable)
 3. discontinuation of medication(s) that impair ability to sense or report headache following the head injury (when applicable)
- D. Either of the following:
 1. headache has resolved within 3 months after the head injury
 2. headache has not yet resolved but 3 months have not yet passed since the head injury
- E. Not better accounted for by another ICHD-3 diagnosis.

A5.1.2.1 Delayed-onset acute headache attributed to mild traumatic injury to the head

Diagnostic criteria:

- A. Any headache fulfilling criteria C and D
- B. Traumatic injury to the head has occurred, fulfilling both of the following:
 1. associated with none of the following:
 - a) loss of consciousness for >30 minutes
 - b) Glasgow Coma Scale (GCS) <13
 - c) post-traumatic amnesia lasting >24 hours
 - d) altered level of awareness for >24 hours
 - e) imaging evidence of a traumatic head injury such as intracranial haemorrhage and/or brain contusion

2. associated, immediately following the head injury, with one or more of the following symptoms and/or signs:
 - a) transient confusion, disorientation or impaired consciousness
 - b) loss of memory for events immediately before or after the injury
 - c) two or more other symptoms suggestive of mild traumatic brain injury: nausea, vomiting, visual disturbances, dizziness and/or vertigo, impaired memory and/or concentration
- C. Time of onset of headache is uncertain, and/or headache is reported to have developed >7 days after all of the following:
 1. the head injury
 2. regaining of consciousness following the head injury (when applicable)
 3. discontinuation of medication(s) that impair ability to sense or report headache following the head injury (when applicable)
 - D. Either of the following:
 1. headache has resolved within 3 months after the head injury
 2. headache has not yet resolved but 3 months have not yet passed since the head injury
 - E. Not better accounted for by another ICHD-3 diagnosis.

A5.2 Persistent headache attributed to traumatic injury to the head

Comment:

The current stipulation that headache must begin (or be reported to have begun) within 7 days of head injury (or awareness of the injury) is somewhat arbitrary. Some data suggest that headache may begin after a longer interval. Future studies should continue to investigate the utility of diagnostic criteria for A5.2 *Persistent headache attributed to traumatic injury to the head* that allow for headache to begin up to 30 days after the injury.

A5.2.1.1 Delayed-onset persistent headache attributed to moderate or severe traumatic injury to the head

Diagnostic criteria:

- A. Any headache fulfilling criteria C and D
- B. Traumatic injury to the head has occurred, associated with at least one of the following:
 1. loss of consciousness for >30 minutes
 2. Glasgow Coma Scale (GCS) <13
 3. post-traumatic amnesia lasting >24 hours
 4. alteration in level of awareness for >24 hours

- 5. imaging evidence of a traumatic head injury such as intracranial haemorrhage and/or brain contusion.
- C. Time of onset of headache is uncertain, and/or headache is reported to have developed >7 days after all of the following:
 - 1. the head injury
 - 2. regaining of consciousness following the head injury (when applicable)
 - 3. discontinuation of medication(s) that impair ability to sense or report headache following the head injury (when applicable)
- D. Headache persists for >3 months after the head injury
- E. Not better accounted for by another ICHD-3 diagnosis.

A5.2.2.1 Delayed-onset persistent headache attributed to mild traumatic injury to the head

Diagnostic criteria:

- A. Any headache fulfilling criteria C and D
- B. Traumatic injury to the head has occurred, fulfilling both of the following:
 - 1. associated with none of the following:
 - a) loss of consciousness for >30 minutes
 - b) Glasgow Coma Scale (GCS) <13
 - c) post-traumatic amnesia lasting >24 hours
 - d) altered level of awareness for >24 hours
 - e) imaging evidence of a traumatic head injury such as intracranial haemorrhage and/or brain contusion
 - 2. associated, immediately following the head injury, with one or more of the following symptoms and/or signs:
 - a) transient confusion, disorientation or impaired consciousness
 - b) loss of memory for events immediately before or after the injury
 - c) two or more other symptoms suggestive of mild traumatic brain injury: nausea, vomiting, visual disturbances, dizziness and/or vertigo, impaired memory and/or concentration
- C. Time of onset of headache is uncertain, and/or headache is reported to have developed >7 days after all of the following:
 - 1. the head injury
 - 2. regaining of consciousness following the head injury (when applicable)
 - 3. discontinuation of medication(s) that impair ability to sense or report headache following the head injury (when applicable)

- D. Headache persists for >3 months after the head injury
- E. Not better accounted for by another ICHD-3 diagnosis.

A5.7 Headache attributed to radiosurgery of the brain

Diagnostic criteria:

- A. Any new headache fulfilling criterion C
- B. Radiosurgery of the brain has been performed
- C. Evidence of causation demonstrated by both of the following:
 - 1. headache has developed within 7 days after radiosurgery
 - 2. headache has resolved within 3 months after radiosurgery
- D. Not better accounted for by another ICHD-3 diagnosis.

Comment:

Although de novo headache has been described after radiosurgery, most studies do not provide detailed descriptions of its clinical characteristics, neither is it usually clear whether headache occurring after radiosurgery represents an exacerbation of an underlying headache disorder or a new headache. In cases where a previous history of headache was not present, the headache syndrome was short-lived, occurred more than a year after the procedure, and resembled migraine or thunderclap headache. Therefore, causal relationships between these headaches and the radiosurgical procedures preceding them were highly doubtful. Carefully controlled prospective studies are necessary to determine whether *A5.7 Headache attributed to radiosurgery of the brain* exists as an entity and, if so, how it is related to the type and location of the lesion being irradiated and/or the dosage and radiation field employed.

A5.8 Acute headache attributed to other trauma or injury to the head and/or neck

Diagnostic criteria:

- A. Any headache fulfilling criteria C and D
- B. Trauma or injury to the head and/or neck of a type not described above has occurred
- C. Evidence of causation demonstrated by either or both of the following:
 - 1. headache has developed in close temporal relation to the trauma or injury

- 2. other evidence exists of causation by the trauma or injury
- D. Either of the following:
 - 1. headache has resolved within 3 months after the trauma or injury
 - 2. headache persists but 3 months have not yet passed since the trauma or injury
- E. Not better accounted for by another ICHD-3 diagnosis.

A5.9 Persistent headache attributed to other trauma or injury to the head and/or neck

Diagnostic criteria:

- A. Any headache fulfilling criteria C and D
- B. Trauma or injury to the head and/or neck of a type not described above has occurred
- C. Evidence of causation demonstrated by either or both of the following:
 - 1. headache has developed in close temporal relation to the trauma or injury
 - 2. other evidence exists of causation by the trauma or injury
- D. Headache persists for >3 months after the trauma or injury
- E. Not better accounted for by another ICHD-3 diagnosis.

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A6. Headache attributed to cranial or cervical vascular disorder

A6.10 Persistent headache attributed to past cranial or cervical vascular disorder

- A. Headache previously diagnosed as 6. *Headache attributed to cranial or cervical vascular disorder* or one of its subtypes or subforms, and fulfilling criterion C
- B. The cranial or cervical vascular disorder causing the headache has been effectively treated or has spontaneously remitted

- C. Headache has persisted for >3 months after effective treatment or spontaneous remission of the vascular disorder
- D. Not better accounted for by another ICHD-3 diagnosis.

Comment:

Headaches meeting the criteria for A6.10 *Persistent headache attributed to past cranial or cervical vascular disorder*, if they exist, have been poorly documented; research is needed to establish better criteria for causation.

A7. Headache attributed to non-vascular intracranial disorder

A7.6 Headache attributed to epileptic seizure

A7.6.3 *Post-electroconvulsive therapy (ECT) headache*

Diagnostic criteria:

- A. Recurrent headache fulfilling criterion C
- B. A course of electroconvulsive therapy (ECT) has been given
- C. Evidence of causation demonstrated by all of the following:
 - 1. headache has developed after $\geq 50\%$ of ECT sessions
 - 2. each headache has developed within 4 hours after ECT
 - 3. each headache has resolved within 72 hours after ECT
- D. Not better accounted for by another ICHD-3 diagnosis.

Comment:

Clear descriptions of headache associated with electroconvulsive therapy are sparse. Published data are not adequate to define A7.6.3 *Post-electroconvulsive therapy (ECT) headache* operationally.

A7.9 Persistent headache attributed to past non-vascular intracranial disorder

Diagnostic criteria:

- A. Headache previously diagnosed as 7. *Headache attributed to non-vascular intracranial disorder* or one of its subtypes or subforms, and fulfilling criterion C

- B. The non-vascular intracranial disorder causing the headache has been effectively treated or has spontaneously remitted
- C. Headache has persisted for >3 months after effective treatment or spontaneous remission of the vascular disorder
- D. Not better accounted for by another ICHD-3 diagnosis.

Comment:

Headaches meeting the criteria for A7.9 *Persistent headache attributed to past non-vascular intracranial disorder*, if they exist, have been poorly documented; research is needed to establish better criteria for causation.

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A8. Headache attributed to a substance or its withdrawal

A8.4 Persistent headache attributed to past use of or exposure to a substance

Coded elsewhere:

8.2 *Medication-overuse headache.*

Diagnostic criteria:

- A. Headache previously diagnosed as 8.1 *Headache attributed to use of or exposure to a substance* or one of its subtypes, and fulfilling criterion C
- B. Use of or exposure to the substance has ceased

- C. Headache has persisted for >3 months after exposure has ceased
- D. Not better accounted for by another ICHD-3 diagnosis.

A9. Headache attributed to infection

A9.1 Headache attributed to intracranial infection

A9.1.3.3 Persistent headache attributed to past intracranial fungal or other parasitic infection

Diagnostic criteria:

- A. Headache previously fulfilling criteria for 9.1.3 *Headache attributed to intracranial fungal or other parasitic infection*, and fulfilling criterion C
- B. Intracranial fungal or other parasitic infection has resolved
- C. Headache has persisted for >3 months after resolution of the intracranial fungal or other parasitic infection
- D. Not better accounted for by another ICHD-3 diagnosis, and hydrocephalus has been excluded by neuroimaging.

A9.1.6 Headache attributed to other infective space-occupying lesion

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. A space-occupying lesion of infective nature, other than brain abscess or subdural empyema, has been demonstrated
- C. Evidence of causation demonstrated by at least two of the following:
 1. headache has developed in temporal relation to development of the infective space-occupying lesion, or led to its discovery
 2. headache has significantly worsened in parallel with deterioration of the infective space-occupying lesion, shown by any of the following:
 - a) worsening of other symptoms and/or clinical signs arising from the infective space-occupying lesion
 - b) evidence of enlargement of the infective space-occupying lesion
 - c) evidence of rupture of the infective space-occupying lesion
 3. headache has significantly improved in parallel with improvement in the infective space-occupying lesion

4. headache has at least one of the following three characteristics:
 - a) intensity increasing gradually to moderate or severe
 - b) aggravated by straining or other Valsalva manoeuvre
 - c) accompanied by nausea
- D. Not better accounted for by another ICHD-3 diagnosis.

A9.3 Headache attributed to human immunodeficiency virus (HIV) infection

Coded elsewhere:

Headache occurring in patients with HIV infection but caused by a specific opportunistic infection should be coded to that infection. Headache caused by use of antiretroviral drugs should be coded to 8.1.11 *Headache attributed to long-term use of non-headache medication*.

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. Both of the following:
 1. systemic HIV infection has been demonstrated
 2. other ongoing systemic and/or intracranial infection has been excluded
- C. Evidence of causation demonstrated by at least two of the following:
 1. headache has developed in temporal relation to the onset of HIV infection
 2. headache has developed or significantly worsened in temporal relation to worsening of HIV infection as indicated by CD4 cell count and/or viral load
 3. headache has significantly improved in parallel with improvement in HIV infection as indicated by CD4 cell count and/or viral load
- D. Not better accounted for by another ICHD-3 diagnosis.

Comments:

Headache is reported by more than half of people infected by human immunodeficiency virus (HIV)/acquired immune deficiency syndrome (AIDS), and may be a part of the symptomatology of both acute and chronic HIV infection (through aseptic meningitis and similar mechanisms). In most cases, headache is dull and bilateral, or has the features of a primary headache disorder (1. *Migraine* or 2. *Tension-type headache*). Headache severity, frequency and disability seem associated with severity of HIV infection as indicated by CD4 cell count and/or viral load, but not with the

duration of HIV infection or the number of prescribed antiretroviral medications. Only a minority of HIV patients have headache attributable to opportunistic infections, probably as a consequence of the availability of highly active antiretroviral therapy.

The rationale for separating A9.3 *Headache attributed to human immunodeficiency virus (HIV) infection* from headaches attributed to other infections is three-fold:

- a) HIV infection is always both systemic and within the central nervous system;
- b) the central nervous system infection may progress independently of the systemic infection;
- c) HIV infection is still not curable.

The positioning of A9.3 *Headache attributed to human immunodeficiency virus (HIV) infection* within the Appendix has been deemed necessary because it is extremely difficult to distinguish headache attributed purely to HIV infection from the primary-like headaches reported by most HIV patients. Application of these criteria in prospective studies may provide more conclusive evidence.

During HIV infection, secondary meningitis and/or encephalitis associated with opportunistic infections or neoplasms can develop. The most common intracranial infections associated with HIV infection and causing headache are toxoplasmosis and cryptococcal meningitis. Headache occurring in patients with HIV infection but attributed to a specific opportunistic infection should be coded to that infection.

Antiretroviral drugs can also cause headache. In these cases, the headache should be coded as 8.1.11 *Headache attributed to long-term use of non-headache medication*.

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A10. Headache attributed to disorder of homoeostasis

A10.7 Head and/or neck pain attributed to orthostatic (postural) hypotension

Description:

Pain, mostly in the back of the neck but sometimes spreading upwards to the occipital region (‘coathanger’ distribution), attributed to postural hypotension and developing only in upright posture.

Diagnostic criteria:

- A. Headache fulfilling criterion C
- B. Orthostatic (postural) hypotension has been demonstrated
- C. Evidence of causation demonstrated by two of the following:
 - 1. headache develops exclusively during upright posture
 - 2. headache spontaneously improves in horizontal posture
 - 3. headache is mostly in the back of the neck, sometimes spreading upwards to the occipital region (‘coathanger’ distribution)
- D. Not better accounted for by another ICHD-3 diagnosis.

Comment:

When specifically asked, 75% of patients with orthostatic hypotension reported neck pain.

A10.8 Headache attributed to other disorder of homoeostasis

A10.8.1 Headache attributed to travel in space

Description:

Non-specific headache caused by travel in space. The majority of headache episodes are not associated with symptoms of space motion sickness.

Diagnostic criteria:

- A. Any new headache fulfilling criterion C
- B. The subject is travelling through space
- C. Evidence of causation demonstrated by both of the following:
 - 1. headache has occurred exclusively during space travel
 - 2. headache has spontaneously improved on return to earth
- D. Not better accounted for by another ICHD-3 diagnosis.

Comment:

Of the 16 male and one female astronauts who participated in a survey, 12 (71%) reported at least one headache episode experienced while in space, whereas they had not suffered from headache when on earth.

A10.8.2 Headache attributed to other metabolic or systemic disorder

Headaches attributed to the following disorders may occur, but are not sufficiently validated:

anaemia, adrenocortical insufficiency, mineralocorticoid deficiency, hyperaldosteronism, polycythaemia, hyperviscosity syndrome, thrombotic thrombocytopenic purpura, plasmapheresis, anticardiolipin antibody syndrome, Cushing’s disease, hyponatraemia, hyperthyroidism, hyperglycaemia, hypercalcaemia, systemic lupus erythematosus, chronic fatigue syndrome, fibromyalgia.

Well-controlled, prospective studies are needed to define more clearly the incidence and characteristics of headaches that occur in association with these disorders. In each case, only those patients who meet well-established diagnostic criteria for the disorders themselves should be evaluated.

A10.9 Persistent headache attributed to past disorder of homoeostasis

Diagnostic criteria:

- A. Headache previously diagnosed as 10. *Headache attributed to disorder of homoeostasis*, and fulfilling criterion C
- B. The disorder of homoeostasis causing the headache has been effectively treated or has spontaneously remitted
- C. Headache has persisted for >3 months after effective treatment or spontaneous remission of the disorder of homoeostasis
- D. Not better accounted for by another ICHD-3 diagnosis.

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A11. Headache or facial pain attributed to disorder of the cranium, neck, eyes, ears, nose, sinuses, teeth, mouth or other facial or cervical structure

A11.2 Headache attributed to disorder of the neck

A11.2.4 Headache attributed to upper cervical radiculopathy

Diagnostic criteria:

- A. Head and/or neck pain fulfilling criterion C
- B. Clinical or radiological evidence of a C2 or C3 radiculopathy
- C. Evidence of causation demonstrated by both of the following:
 1. at least two of the following:
 - a) pain has developed in temporal relation to onset of the radiculopathy, or led to its discovery
 - b) pain has significantly improved or significantly worsened in parallel with improvement in or worsening of the radiculopathy
 - c) pain is temporarily abolished by local anaesthesia of the relevant nerve root
 2. headache is ipsilateral to the radiculopathy
- D. Not better accounted for by another ICHD-3 diagnosis.

Comment:

Pain is usually posterior but may radiate to more anterior regions. Often there are lancinations of pain in one of the areas subserved by the upper cervical roots on one or both sides, generally in the occipital, retroauricular or upper posterior cervical regions.

A11.2.5 Headache attributed to cervical myofascial pain

Diagnostic criteria:

- A. Head and/or neck pain fulfilling criterion C
- B. A source of myofascial pain in the muscles of the neck, including reproducible trigger points, has been demonstrated

- C. Evidence of causation demonstrated by at least two of the following:
 1. either or both of the following:
 - a) pain has developed in temporal relation to onset of the cervical myofascial pain disorder
 - b) pain has significantly improved in parallel with improvement in the cervical myofascial pain disorder
 2. significant pressure-tenderness is elicited in cervical muscles corresponding to the pain perceived by the patient
 3. pain is temporarily abolished by local anaesthetic injections into trigger points, or by trigger point massage
- D. Not better accounted for by another ICHD-3 diagnosis.

Comment:

Myofascial pain and its relation to so-called ‘trigger points’ is controversial. It has been difficult consistently to demonstrate supposed trigger points, and response to treatment varies.

A11.5 Headache attributed to disorder of the nose or paranasal sinuses

A11.5.3 Headache attributed to disorder of the nasal mucosa, turbinates or septum

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. Clinical, nasal endoscopic and/or imaging evidence of a hypertrophic or inflammatory process within the nasal cavity¹
- C. Evidence of causation demonstrated by at least two of the following:
 1. headache has developed in temporal relation to the onset of the intranasal lesion
 2. headache has significantly improved or significantly worsened in parallel with improvement in (with or without treatment) or worsening of the nasal lesion
 3. headache has significantly improved following local anaesthesia of the mucosa in the region of the lesion
 4. headache is ipsilateral to the site of the lesion
- D. Not better accounted for by another ICHD-3 diagnosis.

Note:

Examples are concha bullosa and nasal septal spur.

A12. Headache attributed to psychiatric disorder

Introduction

Headaches are commonly associated with various psychiatric disorders, but evidence of a causal relationship is lacking for most. The following are offered as candidate criterion sets to facilitate research into the possible causal relationships between certain psychiatric disorders and headache. It is not recommended that they be used routinely in clinical practice to describe the association between comorbid headache and psychiatric disorders. In the vast majority of cases, headache associated with these disorders most probably reflects common underlying risk factors or aetiologies.

In order to make any of the diagnoses listed below, it is crucial to establish a causal relationship between the headache and the psychiatric disorder in question. Thus, either the headache onset occurs simultaneously with the psychiatric disorder or the headache clearly worsens after the psychiatric disorder becomes evident. Definite biomarkers and clinical proof of headache causation are difficult to obtain, and the diagnosis should be based on high levels of clinical suspicion. Thus, for example, in a child with separation anxiety disorder, headache should be attributed to this disorder only in those cases where it occurs solely in the context of actual or threatened separation, without any better explanation. Similarly, in an adult with panic disorder, headache should be attributed to the disorder only in those cases where it occurs solely as one of the symptoms of a panic attack.

A12.3 Headache attributed to depressive disorder

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. Major depressive disorder (single episode or recurrent) or persistent depressive disorder has been diagnosed according to DSM-5 criteria
- C. Headache occurs exclusively during depressive episodes
- D. Not better accounted for by another ICHD-3 diagnosis.

Comment:

Many antidepressants, especially tricyclic antidepressants, are effective against headache disorders even when depression is not present. This makes it difficult to determine whether remission of or improvement in a headache disorder associated with depression and treated with a tricyclic antidepressant is, in fact, evidence of

causation. Remission of headache is more suggestive of a psychiatric cause when a major depressive disorder improves under treatment with other type of antidepressants shown to be less effective in headache treatment.

A12.4 Headache attributed to separation anxiety disorder

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. Separation anxiety disorder has been diagnosed according to DSM-5 criteria
- C. Headache occurs exclusively in the context of actual or threatened separation from home or from major attachment figures
- D. Not better accounted for by another ICHD-3 diagnosis.

Comment:

Separation anxiety disorder is persistent, typically lasting at least 6 months, although a shorter duration may meet diagnostic criteria in cases of acute onset or exacerbation of severe symptoms (e.g. school refusal, or complete inability to separate from home or attachment figures). The disorder causes clinically significant distress and/or impairment in social, academic, occupational and/or other important areas of functioning.

A12.5 Headache attributed to panic disorder

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. Recurrent unexpected panic attacks fulfilling DSM-5 criteria for panic disorder
- C. Headache occurs exclusively during panic attacks
- D. Not better accounted for by another ICHD-3 diagnosis.

A12.6 Headache attributed to specific phobia

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. Specific phobia has been diagnosed according to DSM-5 criteria
- C. Headache occurs exclusively when the patient is exposed or anticipating exposure to the phobic stimulus
- D. Not better accounted for by another ICHD-3 diagnosis.

Comment:

Specific phobias typically last 6 or more months, causing clinically significant distress and/or impairment in social, occupational and/or other important areas of functioning.

A12.7 Headache attributed to social anxiety disorder (social phobia)

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. Social anxiety disorder (social phobia) has been diagnosed according to DSM-5 criteria
- C. Headache occurs solely when the patient is exposed or anticipating exposure to social situations
- D. Not better accounted for by another ICHD-3 diagnosis.

Comment:

In social anxiety disorder (social phobia), there is marked fear or anxiety about one or more social situations in which the individual is exposed to possible scrutiny by others. Examples include social interactions (e.g. having a conversation), being observed (e.g. eating or drinking) or performing in front of others (e.g. giving a speech). The person fears that he or she will act in a way or show anxiety symptoms that will cause him or her to be negatively evaluated (e.g. be humiliated, embarrassed or rejected) or that will offend others. In children, the fear or anxiety may be expressed by crying, tantrums, freezing, clinging, shrinking or failure to speak in social situations. The fear or anxiety is out of proportion to the actual threat posed by the social situation. The disorder is persistent, typically lasting 6 or more months.

A12.8 Headache attributed to generalized anxiety disorder

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. Generalized anxiety disorder has been diagnosed according to DSM-5 criteria
- C. Headache occurs solely during periods of anxiety
- D. Not better accounted for by another ICHD-3 diagnosis.

Comment:

Patients with generalized anxiety disorder present excessive anxiety and worry (apprehensive expectation)

about two (or more) domains of activities or events (e.g. family, health, finances, school/work difficulties), on more days than not, for 3 months or more. Symptoms may include restlessness or feeling keyed up or on edge, and muscle tension. Behaviours associated with this disorder include avoidance of activities or events with possible negative outcomes, marked investment of time and effort in preparing for activities or events with possible negative outcomes, marked procrastination in behaviour or decision-making because of worries, and repeatedly seeking reassurance because of worries.

A12.9 Headache attributed to post-traumatic stress disorder

Diagnostic criteria:

- A. Any headache fulfilling criterion C
- B. Post-traumatic stress disorder (PTSD) has been diagnosed according to DSM-5 criteria
- C. The headache first developed after exposure to the trauma stressor and occurs solely in the context of other symptoms of PTSD¹
- D. Not better accounted for by another ICHD-3 diagnosis.

Note:

1. For example, headache occurs on exposure to reminders of the trauma.

Comments:

Exposure to actual or threatened death, serious injury or sexual violation may occur directly by experiencing the event, or it may occur indirectly: by witnessing the event; by learning that the event occurred to a close family member or friend; by experiencing repeated or extreme exposure to aversive details of the event (e.g. first responders collecting human remains; police officers repeatedly exposed to details of child abuse). This is not true of exposure through electronic media, television, movies or pictures, unless this exposure is work-related.

Given the high rate of comorbid depression with post-traumatic stress disorder (PTSD), the diagnosis of A12.9 *Headache attributed to post-traumatic stress disorder* should be reserved for those patients whose headache is not explained by comorbid depression (i.e. cases of headache attributed to PTSD in patients *without* comorbid depression).

Definition of terms

Accompanying symptoms: Symptoms that typically accompany rather than precede or follow headache. In migraine, for example, the most frequent are nausea, vomiting, photophobia and phonophobia; osmophobia, diarrhoea and other symptoms occur more rarely.

Anorexia: Lack of appetite and dislike for food to a mild degree.

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

Aura: Early symptoms of an attack of migraine with aura, being the manifestations of focal cerebral dysfunction. The aura typically lasts 20–30 minutes and precedes the headache. See also: *Focal symptoms*, *Prodrome*, *Premonitory symptoms*, *Warning symptoms* and *Neurological symptoms*.

Central neuropathic pain: Pain (qv) caused by a lesion or disease of the central somatosensory nervous system (see also *Neuropathic pain*).

Chronic: In pain terminology, *chronic* denotes persistence over a period longer than three months. In headache terminology, it retains this meaning for secondary headache disorders. For primary headache disorders that are more usually episodic (qv), *chronic* is used whenever attacks of headache (qv) occur on more days than not over a period longer than 3 months. The trigeminal autonomic cephalalgias are the exception: in these disorders, *chronic* is not used until the disorder has been unremitting for more than one year.

Close temporal relation: This term is used to describe the relation between an organic disorder and headache. Specific temporal relations may be known for disorders of acute onset where causation is likely, but have often not been studied sufficiently. For chronic disorders the temporal relation as well as causation are often very difficult to ascertain.

Cluster headache attack: One episode of continuous pain lasting 15–180 minutes.

Cluster period: The time during which cluster headache attacks occur regularly and at least once every other day.

Cluster remission period: The time during which attacks cease to occur spontaneously and cannot be induced with alcohol or nitroglycerine. To be considered a remission, the attack-free period must exceed one month.

Duration of attack: Time from onset until termination of an attack of headache (or pain) (qv) meeting

criteria for a particular headache type or subtype. After migraine or cluster headache, a low-grade non-pulsating headache without accompanying symptoms may persist, but this is not part of the attack and is not included in duration. If the patient falls asleep during an attack and wakes up relieved, duration is until time of awakening. If an attack of migraine is successfully relieved by medication but symptoms recur within 48 hours, these may represent a relapse of the same attack or a new attack. Judgement is required to make the distinction (see also *Frequency of attacks*).

Episodic: Recurring and remitting in a regular or irregular pattern of attacks of headache (or pain) (qv) of constant or variable duration. Through long usage the term has acquired special meaning in the context of *episodic cluster headache*, referring to the occurrence of cluster periods separated by cluster remission periods (qv) rather than to attacks. Similar usage has been adopted in paroxysmal hemicrania.

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

Focal symptoms: Symptoms of focal brain (usually cerebral) disturbance such as occur in migraine aura.

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

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

Headache: Pain (qv) located above the orbitomeatal line.

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

Heterophoria: Latent strabismus.

Heterotropia: Manifest strabismus.

Intensity of pain: Degree of pain (qv) usually expressed in terms of its functional consequence and scored on a verbal four-point scale: 0, no pain; 1, mild pain, does not interfere with usual activities; 2, moderate pain, inhibits but does not wholly prevent usual

- activities; 3, severe pain, prevents all activities. It may also be expressed on a visual analogue scale.
- Lancinating*: Brief, electric, shock-like along a root or nerve.
- Neuralgia*: Pain (qv) in the distribution of a nerve or nerves. (Common usage, especially in Europe, often implies a paroxysmal or lancinating (qv) quality, but the term *neuralgia* should not be reserved for paroxysmal pains.)
- Neuritis*: A special case of neuropathy (qv); the term is now reserved for inflammatory processes affecting nerves.
- Neuroimaging*: CT, MRI, PET, SPECT or scintigraphy of the brain.
- Neuropathic pain*: Pain (qv) caused by a lesion or disease of the somatosensory nervous system.
- Neuropathy*: A disturbance of function or pathological change in a nerve or nerves (in one nerve: mononeuropathy; in several nerves: mononeuropathy multiplex; when diffuse and bilateral: polyneuropathy). The term *neuropathy* is not intended to cover neuropathic pain, neurotmesis, section of a nerve, disturbances of a nerve due to transient impact such as a blow, stretching or epileptic discharge (the term *neurogenic* applies to pain attributed to such temporary perturbations).
- New headache*: Any type, subtype or subform of headache (qv) from which the patient was not previously suffering.
- Not sufficiently validated*: Of doubtful validity as a diagnostic entity judged from the experience of the subcommittee and/or controversy in the literature.
- Nuchal region*: Dorsal (posterior) aspect of upper neck including the region of insertion of neck muscles on the cranium.
- Pain*: An unpleasant sensory and emotional experience associated with actual or potential tissue damage, or described in terms of such damage (see also: *Neuropathic pain*, *Central neuropathic pain* and *Peripheral neuropathic pain*).
- Pericranial muscles*: Neck muscles, muscles of mastication, facial muscles of expression and speech and muscles of the inner ear (tensor tympani, stapedius).
- Peripheral neuropathic pain*: Pain (qv) caused by a lesion or disease of the peripheral somatosensory nervous system (see also *Neuropathic pain*).
- Phonophobia*: Hypersensitivity to sound, usually causing avoidance.
- Photophobia*: Hypersensitivity to light, usually causing avoidance.
- Premonitory symptoms*: Symptoms preceding and forewarning of a migraine attack by 2–48 hours, occurring before the aura in migraine with aura and before the onset of pain in migraine without aura. Among the common premonitory symptoms are: fatigue, elation, depression, unusual hunger, craving for certain foods.
- Pressing/tightening*: Pain of a constant quality often compared to an iron band around the head.
- Previously used term*: A diagnostic term that has been used previously with a similar or identical meaning to the classified term or is subsumed within it. Previously used terms are often ambiguous and/or have been used differently in different countries.
- Prodrome*: This term has been used with different meanings, most often synonymously with premonitory symptoms. It should be avoided in the future.
- Pulsating*: Varying with the heart beat; throbbing.
- Referred pain*: Pain (qv) perceived in another area than the one where nociception arises.
- Refraction error*: Myopia, hypermetropia or astigmatism.
- Scintillation*: Visual hallucinations that are bright and fluctuate in intensity, often at approximately 8–10 cycles/second. They are typical of migraine aura.
- Scotoma*: Loss of part(s) of the visual field of one or both eyes. Scotoma may be absolute (no vision) or relative (obscured or reduced vision).
- Stab of pain*: Sudden pain (qv) lasting a minute or less (usually a second or less).
- Substance*: Organic or inorganic chemical, food or additive, alcoholic beverage, gas or vapour, drug or medication, herbal, animal or other substance given with medicinal intent although not licensed as medicinal products, etc.
- Teichopsia*: Synonym for fortification spectrum (qv).
- Tenderness*: A feeling of discomfort or pain caused by pressure that would not normally be sufficient to cause such sensations.
- Throbbing*: Synonym for pulsating (qv).
- Unilateral*: On either the right or the left side, not crossing the mid line. Unilateral headache does not necessarily involve all of the right or left side of the head, but may be frontal, temporal or occipital only. When used for sensory or motor disturbances of migraine aura it includes complete or partial hemidistribution.
- Vasospasm*: Constriction of artery or arterioles to such a degree that tissue perfusion is reduced.
- Warning symptoms*: Previously used term for either aura or premonitory symptoms and therefore ambiguous. It should not be used.
- Zigzag line*: Synonym for fortification spectrum (qv).