

## **Headache Classification Committee of the International Headache Society (IHS)**

# **The International Classification of Headache Disorders, 3rd edition**

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## Preface

On behalf of the Classification Committee of The International Headache Society, I am proud to present the third edition of the *International Classification of Headache Disorders* (ICHD-3).

This follows the publication of ICHD-3 beta in 2013. The idea behind the beta version was to promote more field testing before presentation of the final ICHD-3, and this has worked well. There have been excellent field-testing studies published, in migraine with aura, cluster headache, idiopathic intracranial hypertension and trigeminal neuralgia among others. It was, for example, documented that the Appendix criteria for A1.2 *Migraine with aura* were superior to the criteria for 1.2 *Migraine with aura* in the main body of ICHD-3 beta, better distinguishing this disorder from transient ischaemic attacks. Field testing of the novel associated features in criterion C1 for 3.1. *Cluster headache*, facial flushing and aural fullness, revealed that they did not add to diagnostic discrimination. Consequently, these symptoms are included only in the Appendix of ICHD-3, where they invite further study. These are examples of the evidence-based process of disease classification that now underpins all future changes to the *International Classification of Headache Disorders*.

A contributing reason for the beta version was, as we thought, so that ICHD-3 could when published include the codes of the *International Classification of Diseases, 11th Revision* (ICD-11), from the World Health Organization (WHO). We expected that ICD-11 would be finished in 2016, but unfortunately there have been long and unexpected delays so that the final codes are still not available. We therefore have to publish ICHD-3 without them.

ICHD-3 is published as the first issue of *Cephalalgia* in 2018, exactly 30 years after the first edition of the *International Classification of Headache Disorders*, ICHD-I as we now call it. This first version was based primarily upon the opinions of experts, but proved nevertheless to be largely valid. ICHD-II, published in 2004, included a number of changes prompted partly by new evidence and partly by revised opinions of experts. New scientific evidence played a relatively greater role in the changes made in ICHD-3 beta, and all the further changes included in ICHD-3 are based on such evidence. Thus, headache classification is now and will in the future be driven entirely by research.

A long journey that started in 2010 has ended with the publication of ICHD-3, but the present committee has still much to do for a couple of years. ICHD-3 beta was translated into many languages, and these translations need updating before ICHD-3 can be published in

those languages. Hopefully many additional translations will be published so that ICHD-3 becomes available in all major and even in many minor languages. An electronic version of ICHD-3 beta already developed under the leadership of Professor Hartmut Göbel will be updated to ICHD-3. A case book is planned in a collaboration between Professors Morris Levin and Jes Olesen. Finally, a cross-walk between ICHD-3 and WHO's ICD-11 will be made by Professors Timothy Steiner and Jes Olesen as soon as the codes for ICD-11 become available.

What then is the future of headache classification? Classification must in principle be a conservative discipline. When major changes are made to a classification, all previous studies using those parts of the classification that are changed must be revisited. Drug trials according to previous diagnostic criteria must, for example, be repeated if diagnostic criteria undergo major changes because patients falling under the new diagnosis will be different from those falling under the previous diagnosis. My hope is that the active field testing and scientific analysis that have been done for ICHD-3 will continue, allowing future changes to be entirely evidence-based. Following the tradition, it will be 10–15 years before ICHD-4, but a number of field-testing studies will be produced in the meantime. Modified ICHD-II diagnostic criteria for 1.3 *Chronic migraine* were published in *Cephalalgia*; the Classification Committee endorsed these changes, asking for their immediate use even though they were not integrated into the *International Classification of Headache Disorders* until ICHD-3 beta appeared years later. A future headache classification committee should similarly be able to endorse and support the adoption of new or revised diagnostic criteria before publishing ICHD-4 when they are substantiated by good field-testing studies published in *Cephalalgia*.

ICHD-I took headache classification from being one of the worst-classified neurological diseases to being the best. We have kept this momentum for 30 years, and the superiority of our classification became evident recently during the committee work in Geneva on the neurological section of ICD-11. No other discipline within neurology has such a systematic classification with explicit diagnostic criteria for every disease entity. I sincerely hope that this tradition can be upheld in the future, and that headache can continue to lead the way in the classification of neurological diseases.

**Jes Olesen**  
*Chairman*

Headache Classification Committee  
International Headache Society

## How to use this classification

This extensive document is not intended to be learned 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 types 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: every patient entered into a research project, be it a drug trial or a study of pathophysiology or biochemistry, must fulfil an agreed set of diagnostic criteria.

1. This classification is hierarchical, and you must decide how detailed you wish to make your diagnosis: from the first-digit level only to the fifth. First one forms an impression as to 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, while in specialist practice and headache centres a diagnosis at fourth- or fifth-digit levels is appropriate.
2. For most purposes, patients receive a diagnosis according to the headache phenotypes currently present, or that 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. For example, 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, 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 and when 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 types or 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 while other attacks meet another set. In such cases, two diagnoses exist and both should be given and 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-, fourth- and occasionally fifth-digit levels then demand, as criterion A, fulfilment of the criteria for levels one and/or two and, in criterion B and onwards, state the further specific criteria to be fulfilled.
8. The frequency of primary headache disorders varies widely, from attacks every one to two years to attacks daily. The severity of attacks also varies. ICHD-3 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 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 twofold 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, while similar, do not quite satisfy the criteria. This can be due to 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 the secondary headaches, the most well-known and well-established causes are mentioned and criteria for the consequent headaches are given. However, in many chapters, for example 9. *Headache attributed to infection*, there are an almost endless number of possible infective 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 can 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, while 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 injury to the head* being a good example, persistent headache types or subtypes 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 type (e.g. 5.1 *Acute headache attributed to traumatic injury to the head*) to the persistent type (5.2 *Persistent headache attributed to traumatic injury to the head*) after a specified time interval (three months in this example). Evidence of causation depends upon earlier fulfilment of the criteria for diagnosis of the acute type, and persistence of the same headache. Most such diagnoses are in the Appendix because of insufficient evidence of 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
<b>1.</b>	<b>Migraine</b>
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 (SHM)
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
<b>2.</b>	<b>Tension-type headache (TTH)</b>
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
  - 3.4.1 Hemicrania continua, remitting subtype
  - 3.4.2 Hemicrania continua, unremitting subtype
- 3.5 Probable trigeminal autonomic cephalalgia
  - 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 and/or cervical vascular disorder**
  - 6.1 Headache attributed to cerebral ischaemic event
    - 6.1.1 Headache attributed to ischaemic stroke (cerebral infarction)
      - 6.1.1.1 Acute headache attributed to ischaemic stroke (cerebral infarction)
      - 6.1.1.2 Persistent headache attributed to past 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 Acute headache attributed to non-traumatic intracerebral haemorrhage
    - 6.2.2 Acute headache attributed to non-traumatic subarachnoid haemorrhage (SAH)
    - 6.2.3 Acute headache attributed to non-traumatic acute subdural haemorrhage (ASDH)
    - 6.2.4 Persistent headache attributed to past non-traumatic intracranial haemorrhage
      - 6.2.4.1 Persistent headache attributed to past non-traumatic intracerebral haemorrhage
      - 6.2.4.2 Persistent headache attributed to past non-traumatic subarachnoid haemorrhage
      - 6.2.4.3 Persistent headache attributed to past non-traumatic acute subdural haemorrhage
  - 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
      - 6.5.1.1 Acute headache or facial or neck pain attributed to cervical carotid or vertebral artery dissection
      - 6.5.1.2 Persistent headache or facial or neck pain attributed to past cervical carotid or vertebral artery dissection
    - 6.5.2 Post-endarterectomy headache
    - 6.5.3 Headache attributed to carotid or vertebral angioplasty or stenting
  - 6.6 Headache attributed to cranial venous disorder
    - 6.6.1 Headache attributed to cerebral venous thrombosis (CVT)
    - 6.6.2 Headache attributed to cranial venous sinus stenting
  - 6.7 Headache attributed to other acute intracranial arterial disorder
    - 6.7.1 Headache attributed to an intracranial endarterial procedure
    - 6.7.2 Angiography headache
    - 6.7.3 Headache attributed to reversible cerebral vasoconstriction syndrome (RCVS)
      - 6.7.3.1 Acute headache attributed to reversible cerebral vasoconstriction syndrome (RCVS)

- 6.7.3.2 Acute headache probably attributed to reversible cerebral vasoconstriction syndrome (RCVS)
- 6.7.3.3 Persistent headache attributed to past reversible cerebral vasoconstriction syndrome (RCVS)
- 6.7.4 Headache attributed to intracranial artery dissection
- 6.8 Headache and/or migraine-like aura attributed to chronic intracranial vasculopathy
  - 6.8.1 Headache attributed to Cerebral Autosomal Dominant Arteriopathy with Subcortical Infarcts and Leukoencephalopathy (CADASIL)
  - 6.8.2 Headache attributed to mitochondrial encephalopathy, lactic acidosis and stroke-like episodes (MELAS)
  - 6.8.3 Headache attributed to Moyamoya angiopathy (MMA)
  - 6.8.4 Migraine-like aura attributed to cerebral amyloid angiopathy (CAA)
  - 6.8.5 Headache attributed to syndrome of retinal vasculopathy with cerebral leukoencephalopathy and systemic manifestations (RVCLSM)
  - 6.8.6 Headache attributed to other chronic intracranial vasculopathy
- 6.9 Headache attributed to pituitary apoplexy
- 7. Headache attributed to non-vascular intracranial disorder**
  - 7.1 Headache attributed to increased cerebrospinal fluid (CSF) 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 cause
    - 7.1.3 Headache attributed to intracranial hypertension secondary to chromosomal disorder
    - 7.1.4 Headache attributed to intracranial hypertension secondary to hydrocephalus
  - 7.2 Headache attributed to low cerebrospinal fluid (CSF) pressure
    - 7.2.1 Post-dural puncture headache
    - 7.2.2 Cerebrospinal fluid (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 Ictal epileptic headache
    - 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 Cocaine-induced headache
- 8.1.6 Histamine-induced headache
  - 8.1.6.1 Immediate histamine-induced headache
  - 8.1.6.2 Delayed histamine-induced headache
- 8.1.7 Calcitonin gene-related peptide (CGRP)-induced headache
  - 8.1.7.1 Immediate CGRP-induced headache
  - 8.1.7.2 Delayed CGRP-induced headache
- 8.1.8 Headache attributed to exogenous acute pressor agent
- 8.1.9 Headache attributed to occasional use of non-headache medication
- 8.1.10 Headache attributed to long-term use of non-headache medication
- 8.1.11 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 Non-opioid analgesic-overuse headache
    - 8.2.3.1 Paracetamol (acetaminophen)-overuse headache
    - 8.2.3.2 Non-steroidal anti-inflammatory drug (NSAID)-overuse headache
      - 8.2.3.2.1 Acetylsalicylic acid-overuse headache
    - 8.2.3.3 Other non-opioid analgesic-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 unspecified or 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 localized brain infection
- 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 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
- 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 angle-closure glaucoma
  - 11.3.2 Headache attributed to refractive error
  - 11.3.3 Headache attributed to ocular inflammatory disorder
  - 11.3.4 Trochlear headache
- 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
- 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 lesions of the cranial nerves and other facial pain**
- 13.1 Pain attributed to a lesion or disease of the trigeminal nerve
  - 13.1.1 Trigeminal neuralgia
    - 13.1.1.1 Classical trigeminal neuralgia
      - 13.1.1.1.1 Classical trigeminal neuralgia, purely paroxysmal
      - 13.1.1.1.2 Classical trigeminal neuralgia with concomitant continuous pain
    - 13.1.1.2 Secondary trigeminal neuralgia
      - 13.1.1.2.1 Trigeminal neuralgia attributed to multiple sclerosis
      - 13.1.1.2.2 Trigeminal neuralgia attributed to space-occupying lesion

- 13.1.1.2.3 Trigeminal neuralgia attributed to other cause
- 13.1.1.3 Idiopathic trigeminal neuralgia
  - 13.1.1.3.1 Idiopathic trigeminal neuralgia, purely paroxysmal
  - 13.1.1.3.2 Idiopathic trigeminal neuralgia with concomitant continuous pain
- 13.1.2 Painful trigeminal neuropathy
  - 13.1.2.1 Painful trigeminal neuropathy attributed to herpes zoster
  - 13.1.2.2 Trigeminal post-herpetic neuralgia
  - 13.1.2.3 Painful post-traumatic trigeminal neuropathy
  - 13.1.2.4 Painful trigeminal neuropathy attributed to other disorder
  - 13.1.2.5 Idiopathic painful trigeminal neuropathy
- 13.2 Pain attributed to a lesion or disease of the glossopharyngeal nerve
  - 13.2.1 Glossopharyngeal neuralgia
    - 13.2.1.1 Classical glossopharyngeal neuralgia
    - 13.2.1.2 Secondary glossopharyngeal neuralgia
    - 13.2.1.3 Idiopathic glossopharyngeal neuralgia
  - 13.2.2 Painful glossopharyngeal neuropathy
    - 13.2.2.1 Painful glossopharyngeal neuropathy attributed to a known cause
    - 13.2.2.2 Idiopathic painful glossopharyngeal neuropathy
- 13.3 Pain attributed to a lesion or disease of nervus intermedius
  - 13.3.1 Nervus intermedius neuralgia
    - 13.3.1.1 Classical nervus intermedius neuralgia
    - 13.3.1.2 Secondary nervus intermedius neuralgia
    - 13.3.1.3 Idiopathic nervus intermedius neuralgia
  - 13.3.2 Painful nervus intermedius neuropathy
    - 13.3.2.1 Painful nervus intermedius neuropathy attributed to herpes zoster
    - 13.3.2.2 Post-herpetic neuralgia of nervus intermedius
    - 13.3.2.3 Painful nervus intermedius neuropathy attributed to other disorder
    - 13.3.2.4 Idiopathic painful nervus intermedius neuropathy
- 13.4 Occipital neuralgia
- 13.5 Neck-tongue syndrome
- 13.6 Painful optic neuritis
- 13.7 Headache attributed to ischaemic ocular motor nerve palsy
- 13.8 Tolosa–Hunt syndrome
- 13.9 Paratrigeminal oculosympathetic (Raeder’s) syndrome
- 13.10 Recurrent painful ophthalmoplegic neuropathy
- 13.11 Burning mouth syndrome (BMS)
- 13.12 Persistent idiopathic facial pain (PIFP)
- 13.13 Central neuropathic pain
  - 13.13.1 Central neuropathic pain attributed to multiple sclerosis (MS)
  - 13.13.2 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

A1.2.0.1	Pure menstrual migraine with aura
A1.2.0.2	Menstrually related migraine with aura
A1.2.0.3	Non-menstrual migraine with aura
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
A1.4.6	Visual snow
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
<b>A2.</b>	<b>Tension-type headache (alternative criteria)</b>
A2.1	Infrequent episodic tension-type headache (alternative criteria)
A2.2	Frequent episodic tension-type headache (alternative criteria)
A2.3	Chronic tension-type headache (alternative criteria)
<b>A3.</b>	<b>Trigeminal-autonomic cephalgias (TACs)</b>
A3.1	Cluster headache (alternative criteria)
A3.2	Paroxysmal hemicrania (alternative criteria)
A3.3	Short-lasting unilateral neuralgiform headache attacks (alternative criteria)
A3.4	Hemicrania continua (alternative criteria)
A3.6	Undifferentiated trigeminal autonomic cephalalgia
<b>A4.</b>	<b>Other primary headache disorders</b>
A4.11	Epicrania fugax
<b>A5.</b>	<b>Headache attributed to trauma or injury to the head and/or neck</b>
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
<b>A6.</b>	<b>Headache attributed to cranial and/or cervical vascular disorder</b>
A6.10	Persistent headache attributed to past cranial and/or cervical vascular disorder
<b>A7.</b>	<b>Headache attributed to non-vascular intracranial disorder</b>
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
<b>A8.</b>	<b>Headache attributed to a substance or its withdrawal</b>
A8.4	Persistent headache attributed to past use of or exposure to a substance
<b>A9.</b>	<b>Headache attributed to infection</b>
A9.1	Headache attributed to intracranial infection
A9.1.3.3	Persistent headache attributed to past intracranial fungal or other parasitic infection
A9.3	Headache attributed to human immunodeficiency virus (HIV) infection
<b>A10.</b>	<b>Headache attributed to disorder of homeostasis</b>
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

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A10.8.2	Headache attributed to other metabolic or systemic disorder
A10.9	Persistent headache attributed to past disorder of homoeostasis
<b>A11.</b>	<b>Headache or facial pain attributed to disorder of the cranium, neck, eyes, ears, nose, sinuses, teeth, mouth or other facial or cervical structure</b>
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
A11.3	Headache attributed to disorder of the eyes
A11.3.5	Headache attributed to heterophoria or heterotropia
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
<b>A12.</b>	<b>Headache attributed to psychiatric disorder</b>
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 (PTSD)



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## **Part One**

### **The primary headaches**

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

## 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 (SHM)
  - 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?** Three rules apply to migraine-like headache, according to circumstances.

1. 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.
2. 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 migraine diagnosis (episodic or chronic) and the diagnosis 8.2 *Medication-overuse headache* should be given when medication overuse is present.

3. When *pre-existing migraine* is made *significantly worse* (usually meaning a twofold 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. Many epidemiological studies have documented its high prevalence and socio-economic and personal impacts. In the *Global Burden of Disease Study 2010* (GBD2010), it was ranked as the third most prevalent disorder in the world. In GBD2015, it was ranked the third-highest cause of disability worldwide in both males and females under the age of 50 years.

Migraine has two major types: 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 prodromal phase, occurring hours or days before the headache, and/or a postdromal phase following headache resolution. Prodromal and postdromal 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 type, subtype or subform of migraine, all 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*. However, since the diagnostic criteria for 1.3 *Chronic migraine* subsume attacks of all types, subtypes or subforms, additional coding is unnecessary for episodic subtypes of 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 attacks<sup>1</sup> fulfilling criteria B–D
- B. Headache attacks lasting 4–72 hours (when untreated or unsuccessfully treated)<sup>2,3</sup>
- 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 two 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.

Prodromal symptoms may begin hours or a day or two before the other symptoms of a migraine attack 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. Postdromal symptoms, most commonly

feeling tired or weary, difficulty with concentration and neck stiffness, may follow resolution of the headache, persisting for up to 48 hours; these are less well studied.

Migraine attacks can be associated with cranial autonomic symptoms and symptoms of cutaneous allodynia.

In young children, photophobia and phonophobia may be inferred from their behaviour.

A minority (<10%) of women have attacks of migraine in association with the majority of their menstrual cycles; most such attacks are without aura. Attacks during menstruation tend to be longer and accompanied by more severe nausea than attacks outside the menstrual cycle. ICHD-3 offers criteria for A1.1.1 *Pure menstrual migraine without aura*, A1.1.2 *Menstrually related migraine without aura* and A1.1.3 *Non-menstrual migraine without aura*, but in the Appendix because of uncertainty over whether they should be regarded as separate entities. Criteria are also offered for A1.2.0.1 *Pure menstrual migraine with aura*, A1.2.0.2 *Menstrually related migraine with aura* and A1.2.0.3 *Non-menstrual migraine with aura* to encourage better characterization of these uncommon subforms if they are separate entities.

Very frequent migraine attacks are distinguished as 1.3 *Chronic migraine*. When there is associated medication overuse, both of the 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 1.1 *Migraine without aura*, although blood flow changes in the brainstem may occur, as may cortical changes secondary to pain activation. This contrasts with the pathognomonic spreading oligoemia of 1.2 *Migraine with aura*. While the bulk of the literature suggests that CSD does not occur in 1.1 *Migraine without aura*, some recent studies disagree. Furthermore, it has been suggested that glial waves or other cortical phenomena may be involved in 1.1 *Migraine without aura*. The messenger molecules nitric oxide (NO), serotonin (5-hydroxytryptamine; 5-HT) and calcitonin gene-related peptide (CGRP) are involved. While 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 the last 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, central mesencephalic grey and thalamus, have been recognized. Highly receptor-specific acute medications including 5-HT<sub>1B/D</sub> receptor agonists (triptans), 5-HT<sub>1F</sub> receptor agonists and CGRP receptor antagonists have demonstrated efficacy in the

acute treatment of migraine attacks. Because of their high receptor-specificity, their mechanisms of action provide new insight into migraine mechanisms. It is now clear that 1.1 *Migraine without aura* is a neurobiological disorder, while clinical as well as basic neuroscience studies continue to advance our knowledge of migraine mechanisms.

## 1.2 Migraine with aura

**Previously used terms:** Classic or classical migraine; ophthalmic, hemiparaesthetic, 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 three of the following six characteristics:
  1. at least one aura symptom spreads gradually over  $\geq 5$  minutes
  2. two or more aura symptoms occur in succession
  3. each individual aura symptom lasts 5–60 minutes<sup>1</sup>
  4. at least one aura symptom is unilateral<sup>2</sup>
  5. at least one aura symptom is positive<sup>3</sup>
  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  $3 \times 60$  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.

**Comments:** 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*.

Field testing has compared the diagnostic criteria for 1.2 *Migraine with aura* in the main body of ICHD-3 beta with those for A1.2 *Migraine with aura* in the Appendix. The latter performed better in distinguishing migraine with aura from transient ischaemic attacks. These are now adopted in ICHD-3, which no longer has Appendix criteria for this disorder.

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 headache 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.

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 not recognized in this classification: they are all coded as 1.2.1 *Migraine with typical aura*.

When aura symptoms are multiple, they 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 one hour, but motor symptoms are often longer lasting.

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. When the aura includes motor weakness, the disorder should be coded as 1.2.3 *Hemiplegic migraine* or one of its subforms. 1.2.3 *Hemiplegic migraine* is classified as a separate subtype because of genetic and pathophysiological differences from 1.2.1 *Migraine with typical aura*. Patients with 1.2.3 *Hemiplegic migraine* often have brainstem symptoms in addition.

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.

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

The previously defined syndromes, *migraine with prolonged aura* and *migraine with acute-onset aura*, have been abandoned. It is not rare for aura to last more than one hour but, in most such cases, patients have at least two of the other characteristics of criterion C. Even when most of a patient's attacks do not fulfil criterion C, it is usual that other attacks fulfil criteria for one of the recognized subtypes or subforms of 1.2 *Migraine with aura*, and this should be the diagnosis. The few other cases 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.

Prodromal symptoms may begin hours or a day or two before the other symptoms of a migraine attack with 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 term 'prodrome', which has replaced 'premonitory phase' or 'premonitory symptoms', does not include aura. Postdromal symptoms, most commonly feeling tired or weary, difficulty with concentration and

neck stiffness, may follow resolution of the headache, persisting for up to 48 hours; these are less well studied.

### 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 one hour, a mix of positive and negative features and complete reversibility

#### Diagnostic criteria:

- A. Attacks fulfilling criteria for 1.2 *Migraine with aura* and criterion B below
- B. Aura with both of the following:
  1. fully reversible visual, sensory and/or speech/language symptoms
  2. no motor, brainstem or retinal symptoms.

#### 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. Attacks fulfilling criteria for 1.2.1 *Migraine with typical aura* and criterion B below
- 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. Attacks fulfilling criteria for 1.2.1 *Migraine with typical aura* and criterion B below
- 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. Attacks fulfilling criteria for 1.2 *Migraine with aura* and criterion B below
- B. Aura with both of the following:
  1. at least two of the following fully reversible brainstem symptoms:
    - a. dysarthria<sup>1</sup>
    - b. vertigo<sup>2</sup>
    - c. tinnitus
    - d. hypacusis<sup>3</sup>
    - e. diplopia<sup>4</sup>
    - f. ataxia not attributable to sensory deficit
    - g. decreased level of consciousness (GCS  $\leq 13$ )<sup>5</sup>
  2. no motor<sup>6</sup> or retinal symptoms.

#### Notes:

1. Dysarthria should be distinguished from aphasia.
2. Vertigo does not embrace and should be distinguished from dizziness.
3. This criterion is not fulfilled by sensations of ear fullness.
4. Diplopia does not embrace (or exclude) blurred vision.
5. The Glasgow Coma Scale (GCS) score may have been assessed during admission; alternatively, deficits clearly described by the patient allow GCS estimation.
6. When motor symptoms are present, code as 1.2.3 *Hemiplegic migraine*.

**Comments:** Originally the terms *basilar artery migraine* or *basilar migraine* were used but, since 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 B1 may occur with anxiety and hyperventilation, and are therefore subject to misinterpretation.

### 1.2.3 Hemiplegic<sup>1</sup> migraine

**Description:** Migraine with aura including motor weakness.

#### Diagnostic criteria:

- A. Attacks fulfilling criteria for 1.2 *Migraine with aura* and criterion B below
- B. Aura consisting of both of the following:
  1. fully reversible motor weakness<sup>2</sup>
  2. fully reversible visual, sensory and/or speech/language symptoms.

#### Notes:

1. The term *plegic* means paralysis in most languages, but most attacks are characterized by motor weakness.
2. Motor symptoms generally last less than 72 hours but, in some patients, motor weakness may persist for weeks.

**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. Attacks fulfilling 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* than was previously possible. Specific genetic subforms 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 subform (if discovered) should be specified at the fifth digit.

It has been shown that 1.2.3.1 *Familial hemiplegic migraine* 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 cerebrospinal fluid (CSF) pleocytosis can occur.

1.2.3.1 *Familial hemiplegic migraine* may be mistaken for epilepsy and treated (unsuccessfully) 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. Attacks fulfilling criteria for 1.2.3.1 *Familial hemiplegic migraine*
- B. A mutation on the *CACNA1A* gene has been demonstrated.

#### 1.2.3.1.2 Familial hemiplegic migraine type 2 (FHM2)

##### *Diagnostic criteria:*

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

#### 1.2.3.1.3 Familial hemiplegic migraine type 3 (FHM3)

##### *Diagnostic criteria:*

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

#### 1.2.3.1.4 Familial hemiplegic migraine, other loci

##### *Diagnostic criteria:*

- A. Attacks fulfilling 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 (SHM)

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

##### *Diagnostic criteria:*

- A. Attacks fulfilling 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. Attacks fulfilling criteria for 1.2 *Migraine with aura* and criterion B below
- B. Aura characterized by both of the following:
  1. 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:
    - a. clinical visual field examination
    - b. the patient's drawing of a monocular field defect (made after clear instruction)
  2. at least two of the following:
    - a. spreading gradually over  $\geq 5$  minutes
    - b. symptoms last 5–60 minutes

- c. accompanied, or followed within 60 minutes, by headache
- C. 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 as the underlying aetiology cannot be ascertained.

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

**Description:** Headache occurring on 15 or more days/month for more than three months, which, on at least eight days/month, has the features of migraine headache.

#### Diagnostic criteria:

- A. Headache (migraine-like or tension-type-like<sup>1</sup>) on  $\geq 15$  days/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  $\geq 8$  days/month for  $>3$  months, fulfilling any of the following<sup>2</sup>:
  - 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.<sup>3–5</sup>

#### Notes:

1. The reason for singling out 1.3 *Chronic migraine* from types of 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. Such patients are extremely difficult to keep medication-free in order to observe the natural history of the headache. In this situation, attacks with and those without aura are both counted, as are both migraine-like and tension-type-like headaches (but not secondary headaches).

2. Characterization of frequently recurring headache generally requires a headache diary to record information on pain and associated symptoms day by day for at least one month.
3. Because tension-type-like headache is within the diagnostic criteria for 1.3 *Chronic migraine*, this diagnosis excludes the diagnosis of 2. *Tension-type headache* or its types.
4. 4.10 *New daily persistent headache* may have features suggestive of 1.3 *Chronic migraine*. The latter disorder evolves over time from 1.1 *Migraine without aura* and/or 1.2 *Migraine with aura*; therefore, when these criteria A–C are fulfilled by headache that, unambiguously, is daily and unremitting from  $<24$  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 1.3 *Chronic migraine*.
5. 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 type 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; the diagnosis of 8.2 *Medication-overuse headache* may be inappropriate for these (assuming that chronicity induced by drug overuse is always reversible). For these reasons, and because of the general rule to apply all relevant diagnoses, patients meeting criteria for 1.3 *Chronic migraine* and for 8.2 *Medication-overuse headache* should be coded for both. After drug withdrawal, migraine will either revert to an episodic type or remain chronic, and should be re-diagnosed accordingly; in the latter case, the diagnosis of 8.2 *Medication-overuse headache* may be rescinded.

### 1.4 Complications of migraine

**Comment:** Code separately for both the migraine type, subtype or subform 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 hours<sup>1</sup>
  2. pain and/or associated symptoms are debilitating<sup>2</sup>
- D. Not better accounted for by another ICHD-3 diagnosis.

**Notes:**

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

**Comment:** 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 this disorder and the relevant type or subtype of migraine but not for 1.4.1 *Status migrainosus*. When overuse of medication is of shorter duration than three months, code for the appropriate migraine type or subtype(s) only.

**1.4.2 Persistent aura without infarction**

**Description:** Aura symptoms persisting for one 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  $\geq 1$  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 one-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 due to cerebral infarction of other causes. Attacks with prolonged aura lasting less than one 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 occurring in association with an ischaemic brain

lesion in the appropriate territory demonstrated by neuroimaging, with onset during the course of a typical migraine with aura attack.

**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<sup>1</sup>
- C. Neuroimaging demonstrates ischaemic infarction in a relevant area
- D. Not better accounted for by another ICHD-3 diagnosis.

**Note:**

1. There may be additional symptoms attributable to the infarction.

**Comments:** Ischaemic stroke in a migraine sufferer may be categorized as cerebral infarction of other cause coexisting with 1. *Migraine*, cerebral infarction of other cause presenting with symptoms resembling 1.2 *Migraine with aura*, or cerebral infarction occurring during the course of a typical attack of 1.2 *Migraine with aura*. 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 twofold increased risk of ischaemic stroke in patients with 1.2 *Migraine with aura* 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 increased risk and frequency of aura and the nature of aura symptoms denoting the increase in risk are unknown. Most studies have shown a lack of association between 1.1 *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 one hour after an attack of migraine with aura

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

**Comment:** Migraine and epilepsy are prototypical examples of paroxysmal brain disorders. While migraine-like headaches are quite frequently seen in the epileptic post-ictal period, sometimes a seizure occurs during or following a migraine attack. This phenomenon, sometimes referred to as *migrainepathy*, is a rare event, originally described in patients with 1.2 *Migraine with aura*. Evidence of an association with 1.1 *Migraine without aura* is 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 type or 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 subtypes
- 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 for  $\geq 1$  hour, up to 10 days
  - 3. attacks occur  $\geq 1$  week apart
- D. Complete freedom from symptoms between attacks
- E. Not attributed to another disorder.<sup>1</sup>

*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 attacks are predictable.

This disorder was first included as a childhood periodic syndrome 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 1.6.1.1 *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. At least two of the following four associated symptoms or signs:
  - 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.<sup>1</sup>

*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, when 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. Vertigo<sup>1</sup> occurring without warning, maximal at onset and resolving spontaneously after minutes to hours without loss of consciousness
- C. At least one of the following five 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.<sup>2</sup>

**Notes:**

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.
2. In particular, posterior fossa tumours, seizures and vestibular disorders have been excluded.

**Comment:** 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 attacks<sup>1</sup> 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 five associated symptoms or signs:
  1. pallor
  2. irritability
  3. malaise
  4. vomiting
  5. ataxia<sup>2</sup>
- D. Normal neurological examination between attacks
- E. Not attributed to another disorder.<sup>3</sup>

**Notes:**

1. Attacks tend to recur monthly.
2. Ataxia is more likely in older children within the affected age group.
3. 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.

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

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?** Three rules apply to tension-type-like headache, according to circumstances:

1. When a *new headache with the characteristics of tension-type headache* 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.
2. 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.
3. When *pre-existing tension-type headache* is made *significantly worse* (usually meaning a twofold 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 in association with medication overuse, a close temporal relation is often difficult to establish. Both diagnoses, 2.3 *Chronic tension-type headache* and 8.2 *Medication-overuse headache*, should therefore be given in all such cases.

### Introduction

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

While it was previously considered to be primarily psychogenic, a number of studies since ICHD-I strongly suggest a neurobiological basis to 2. *Tension-type headache*, at least for its more severe subtypes.

The division of 2. *Tension-type headache* into *episodic* and *chronic* types, introduced in ICHD-I, has proved extremely useful. In ICHD-II, the episodic type was further divided into an *infrequent* type, with headache episodes less than once per month, and a *frequent* type. 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. 2.3 *Chronic tension-type headache* is a serious disease, causing greatly decreased quality of life and high disability.

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 is the most significant abnormal finding in patients with any type of 2. *Tension-type headache*: it is typically present interictally, is exacerbated during actual headache and increases with the intensity and

frequency of headaches. Increased tenderness is very probably of pathophysiological importance. ICHD-II therefore distinguished patients with and without such disorder of the pericranial muscles, a subdivision maintained in ICHD-3 to stimulate further research in this area.

Pericranial tenderness is easily detected and recorded by manual palpation. Small rotating movements with the index and middle fingers, and firm pressure (preferably aided by use of a palpometer), provide local tenderness scores of 0–3 for frontal, temporal, masseter, pterygoid, sternocleidomastoid, splenius and trapezius muscles. These can be summed to yield a total tenderness score for each patient. These measures are a useful guide for treatment, and add value and credibility to explanations given to the patient.

The diagnostic difficulty most often encountered among the primary headache disorders is in discriminating between 2. *Tension-type headache* and mild forms of 1.1 *Migraine without aura*. This is more so because patients with frequent headaches often suffer from both disorders. Stricter diagnostic criteria have been suggested for 2. *Tension-type headache* in the hope of excluding migraine that phenotypically resembles tension-type headache. Such criteria were proposed in the Appendix of ICHD-II as A2. *Tension-type headache*. However, the increase in specificity of the criteria reduces their sensitivity, resulting in larger proportions of patients whose headaches can be classified only as 2.4 *Probable tension-type headache* or 1.5 *Probable migraine*. With still 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, although photophobia or phonophobia may be present.

#### Diagnostic criteria:

- A. At least 10 episodes of headache occurring on <1 day/month on average (<12 days/year) and fulfilling criteria B–D
- B. Lasting from 30 minutes to seven 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.<sup>1</sup>

#### Note:

1. When headache fulfils criteria for both 1.5 *Probable migraine* and 2.1 *Infrequent episodic tension-type headache*, code as 2.1 *Infrequent episodic tension-type headache* (or as either subtype of it for which the criteria are fulfilled) under the general rule that definite diagnoses always trump probable diagnoses.

#### 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, although photophobia or phonophobia may be present.

**Diagnostic criteria:**

- A. At least 10 episodes of headache occurring on 1–14 days/month on average for >3 months ( $\geq 12$  and <180 days/year) and fulfilling criteria B–D
- B. Lasting from 30 minutes to seven 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.<sup>1</sup>

**Note:**

- 1. When headache fulfils criteria for both 1.5 *Probable migraine* and 2.2 *Frequent episodic tension-type headache*, code as 2.2 *Frequent episodic tension-type headache* (or as either subtype of it for which the criteria are fulfilled) under the general rule that definite diagnoses always trump probable diagnoses.

**Comment:** 2.2 *Frequent episodic tension-type headache* often coexists with 1.1 *Migraine without aura*. Both disorders need to be identified, preferably through use of a diagnostic headache diary, because the treatments of each differ considerably. 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 its adverse consequence of 8.2 *Medication-overuse headache*.

**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  $\geq 15$  days/month on average for >3 months ( $\geq 180$  days/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.<sup>1–3</sup>

**Notes:**

- 1. Both 2.3 *Chronic tension-type headache* and 1.3 *Chronic migraine* require headache on 15 or more days/month. For 2.3 *Chronic tension-type headache*, headache must, on at least 15 days, meet criteria B–D for 2.2 *Frequent episodic tension-type headache*; for 1.3 *Chronic migraine*, headache must, on at least eight days, meet criteria B–D for 1.1 *Migraine without aura*. A patient can therefore fulfil all criteria for both these diagnoses; for example, by having headache on 25 days/month meeting migraine criteria on eight days and tension-type headache criteria on 17 days. In these cases, only the diagnosis 1.3 *Chronic migraine* should be given.
- 2. 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*.

3. In many uncertain cases there is overuse of medication. When this fulfils criterion B for any of the subtypes 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 type. When the disorder remains chronic after withdrawal, the diagnosis of 8.2 *Medication-overuse headache* may be rescinded.

#### 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 type or 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 1.5.1 *Probable migraine without aura*. In such cases, the general rule of hierarchy applies, putting 1. *Migraine* and its types and subtypes before 2. *Tension-type headache* and its types and subtypes.

#### 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
  - 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
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- 3.4 Hemicrania continua
  - 3.4.1 Hemicrania continua, remitting subtype
  - 3.4.2 Hemicrania continua, unremitting subtype
- 3.5 Probable trigeminal autonomic cephalalgia
  - 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

#### General comment

*Primary or secondary headache or both?* Three rules apply to headache with the characteristics of a trigeminal autonomic cephalgia (TAC), according to circumstances.

1. When a *new headache with the characteristics of a 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.
2. 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.
3. When a *pre-existing TAC* is made *significantly worse* (usually meaning a twofold 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 TACs share the clinical features of unilateral headache and, usually, prominent cranial parasympathetic

autonomic features, which are lateralized and ipsilateral to the headache. Experimental and human functional imaging suggests these syndromes activate a normal human trigeminal-parasympathetic reflex, with the 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; erythromelalgia 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)<sup>1</sup>
- 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) miosis and/or ptosis
  2. a sense of restlessness or agitation
- D. Occurring with a frequency between one every other day and eight per day<sup>2</sup>
- E. Not better accounted for by another ICHD-3 diagnosis.

#### Notes:

1. During part, but less than half, of the active time-course of 3.1 *Cluster headache*, attacks

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

**Comments:** Attacks occur in series lasting for weeks or months (so-called cluster periods or bouts) 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*.

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.

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 a single cluster period.

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 13.1.1 *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 seven days to one year, separated by pain-free periods lasting at least three months.

#### Diagnostic criteria:

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

- B. At least two cluster periods lasting from seven days to one year (when untreated) and separated by pain-free remission periods of  $\geq 3$  months.

**Comment:** Cluster periods usually last between two weeks and three months.

### 3.1.2 Chronic cluster headache

**Description:** Cluster headache attacks occurring for one year or longer without remission, or with remission periods lasting less than three months.

#### 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  $< 3$  months, for at least one 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 usually 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. 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) miosis and/or ptosis
- 2. a sense of restlessness or agitation
- D. Occurring with a frequency of >5 per day<sup>1</sup>
- E. Prevented absolutely by therapeutic doses of indomethacin<sup>2</sup>
- F. Not better accounted for by another ICHD-3 diagnosis.

**Notes:**

1. During part, but less than half, of the active time-course of 3.2 *Paroxysmal hemicrania*, attacks may be less frequent.
2. 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 seven days to one year, separated by pain-free periods lasting at least three months.

**Diagnostic criteria:**

- A. Attacks fulfilling criteria for 3.2 *Paroxysmal hemicrania* and occurring in bouts
- B. At least two bouts lasting from seven days to one year (when untreated) and separated by pain-free remission periods of  $\geq 3$  months.

**3.2.2 Chronic paroxysmal hemicrania (CPH)**

**Description:** Attacks of paroxysmal hemicrania occurring for more than one year without remission, or with remission periods lasting less than three months.

**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 <3 months, for at least one year.

**Comment:** Patients who fulfil criteria for both 3.2.2 *Chronic paroxysmal hemicrania (CPH)* and 13.1.1 *Trigeminal neuralgia* (sometimes referred to as *CPH-tic syndrome*) should receive both diagnoses. Their recognition is important, since 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 five 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. miosis and/or ptosis
- D. Occurring with a frequency of at least one a day<sup>1</sup>
- E. Not better accounted for by another ICHD-3 diagnosis.

**Note:**

1. During part, but less than half, of the active time-course of 3.3 *Short-lasting unilateral neuralgiform headache attacks*, attacks may be less frequent.

**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: 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* can usually be triggered without a refractory period. This is in contrast to 13.1.1 *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*, and criterion B below
- B. Both of the following, ipsilateral to the pain:
  1. conjunctival injection
  2. 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 13.1.1 *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 seven days to one year, separated by pain-free periods lasting three months 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 seven days to one year (when untreated) and separated by pain-free remission periods of  $\geq 3$  months.

#### 3.3.1.2 Chronic *SUNCT*

**Description:** Attacks of *SUNCT* occurring for more than one year without remission, or with remission periods lasting less than three months.

#### 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  $< 3$  months, for at least one 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. Not more than one of the following, ipsilateral to the pain:
  1. conjunctival injection
  2. lacrimation (tearing).

#### 3.3.2.1 Episodic *SUNA*

**Description:** Attacks of *SUNA* occurring in periods lasting from seven days to one year, separated by pain-free periods lasting at least three months.

#### 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 seven days to one year (when untreated) and separated by pain-free remission periods of  $\geq 3$  months.

#### 3.3.2.2 Chronic *SUNA*

**Description:** Attacks of *SUNA* occurring for more than one year without remission, or with remission periods lasting less than three months.

#### 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  $< 3$  months, for at least one 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) 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 indomethacin<sup>1</sup>
- 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* is included under 3. *Trigeminal autonomic cephalalgias* in ICHD-3 on the basis that the pain is typically unilateral, as are the cranial autonomic symptoms when present (in ICHD-II it was under 4. *Other primary headache disorders*).

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 24 hours' duration.

**Diagnostic criteria:**

- A. Headache fulfilling criteria for 3.4 *Hemicrania continua*, and criterion B below
- B. Headache is not daily or continuous, but interrupted (without treatment) by remission periods of  $\geq 24$  hours.

**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 for at least one year, without remission periods of at least 24 hours.

**Diagnostic criteria:**

- A. Headache fulfilling criteria for 3.4 *Hemicrania continua*, and criterion B below
- B. Headache is daily and continuous for at least one year, without remission periods of  $\geq 24$  hours.

**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 cephalalgia**

**Description:** Headache attacks that are believed to be a type or subtype of 3. *Trigeminal autonomic cephalalgias*, but which are missing one of the features required to fulfil all criteria for any of the types and 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. only a first bout of cluster headache), or have had a sufficient number 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?* Two rules apply to 4. *Other primary headache disorders*, according to circumstances.

1. When a *new headache with the characteristics of any of the disorders classified here* 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.
2. 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 twofold 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. They are grouped into four categories and coded in sequence in ICHD-3 accordingly.

1. Headaches associated with physical exertion, including 4.1 *Primary cough headache*, 4.2 *Primary exercise headache*, 4.3 *Primary headache associated with sexual activity* and 4.4 *Primary thunderclap headache*.
2. Headaches attributed to direct physical stimuli (considered to be primary headache disorders because they are brought on by physiological [non-damaging] 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).
4. Other miscellaneous primary headache disorders including 4.9 *Hypnic headache* and 4.10 *New daily persistent headache*.

The pathogenesis of these disorders 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 (e.g. 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.

### 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 manoeuvre<sup>1</sup>
- C. Sudden onset<sup>2</sup>
- D. Lasting between one second and two hours<sup>2</sup>
- E. Not better accounted for by another ICHD-3 diagnosis.<sup>3</sup>

**Notes:**

- 1. Headache arises moments after the cough or other stimulus.
- 2. Headache reaches its peak almost immediately, and then subsides over several seconds to a few minutes (although some patients experience mild to moderate headache for two hours).
- 3. 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 spontaneous intracranial hypotension, carotid or vertebral diseases, middle cranial fossa or posterior fossa tumours, midbrain 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. Since 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.

**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* is usually bilateral and posterior, and predominantly affects patients older than 40 years of age. There is a significant correlation between the frequency of the 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*.

While indomethacin (50–200 mg/day) is usually effective in treating 4.1 *Primary cough headache*, a few symptomatic cases have been reported to respond to this treatment.

**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 manoeuvre
- C. Sudden onset
- D. Lasting between one second and two 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 type or 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.<sup>1</sup>

**Note:**

- 1. 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.

**Comments:** 4.2 *Primary exercise headache* occurs particularly in hot weather or at high altitude. Subtypes such as ‘weightlifter’s 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 manoeuvres), 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 five minutes).

There are reports of prevention in some patients by 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 4.2 *Primary exercise headache* have a 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.

#### 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 cephalgia; coital headache; intercourse headache; orgasmic cephalgia; 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 due to 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 one 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.<sup>1,2</sup>

##### Notes:

1. 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, intra- and extracranial arterial dissection and reversible cerebral vasoconstriction syndrome (RCVS).
2. Multiple explosive headaches during sexual activities should be considered as 6.7.3 *Headache attributed to reversible cerebral vasoconstriction syndrome* (qv) until proven otherwise by angiographic studies (including conventional, magnetic resonance (MR) or computed tomography (CT) angiography) or transcranial Doppler ultrasonography. Of note, vasoconstrictions may not be observed in the early stage of RCVS; therefore, follow-up studies may be needed.

**Comments:** Two subtypes (*preorgasmic headache* and *orgasmic headache*) were included in ICHD-I and ICHD-II, but clinical studies have since been unable to distinguish these; therefore, 4.3 *Primary 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 with 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 associated with sexual activity* should always be related to the frequency of sexual activity.

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 one 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  $\geq 5$  minutes
- D. Not better accounted for by another ICHD-3 diagnosis.<sup>1,2</sup>

*Notes:*

- 1. Thunderclap headache is frequently associated with serious vascular intracranial vascular 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, spontaneous intracranial 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.
- 2. 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.

*Comment:* Evidence that thunderclap headache exists as a primary disorder is poor: the search for an underlying cause should be both expedited and exhaustive.

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:* 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 due to external cooling of the head, such as occurs during exposure in very cold weather, when diving into cold water or when 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 upon 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 one hour during sustained external compression of the forehead or scalp
- C. Maximal at the site of external compression
- D. Resolving within one 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 one 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. While 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 one 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 and C
- B. Each stab lasts for up to a few seconds<sup>1</sup>
- C. Stabs recur with irregular frequency, from one to many per day<sup>2</sup>
- D. No cranial autonomic symptoms
- E. Not better accounted for by another ICHD-3 diagnosis.

*Notes:*

- 1. Studies show 80% of stabs last three seconds or less; rarely, stabs last for 10–120 seconds.
- 2. 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 one week.

*Comments:* Field testing has confirmed the validity of these diagnostic criteria for 4.7 *Primary stabbing headache*. They enable the diagnosis of most primary headaches characterized by stabbing pain, which were not classified in ICHD-II.

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 the 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
- B. Two only of the following:
  - 1. each stab lasts for up to a few seconds
  - 2. stabs recur with irregular frequency, from one to many per day
  - 3. no cranial autonomic symptoms
- C. Not fulfilling ICHD-3 criteria for any other headache disorder

D. 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 and 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.<sup>1</sup>

*Note:*

1. Other causes, in particular structural and dermatologic lesions, have been excluded by history, physical examination and appropriate investigations.

*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 three 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.

##### 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 four 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  $\geq 10$  days/month for  $> 3$  months
- D. Lasting from 15 minutes up to four hours after wakening
- E. No cranial autonomic symptoms or restlessness
- F. Not better accounted for by another ICHD-3 diagnosis.<sup>1,2</sup>

*Notes:*

1. Distinction from one of the types or subtypes of 3. *Trigeminal autonomic cephalalgias*, especially 3.1 *Cluster headache*, is necessary for effective management.
2. 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*.

*Comments:* A recent study has suggested these criteria, introduced in ICHD-3 beta, are more sensitive for 4.9 *Hypnic headache* than those of ICHD-II.

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 subtype (on <15 days/month) may occur.

Although it was thought that the features of 4.9 *Hypnic headache* were generally tension-type-like, recent studies found patients could present with migraine-like features and some patients had nausea during attacks.

Onset of 4.9 *Hypnic headache* is not related to sleep stage. A recent magnetic resonance imaging (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.

#### 4.9.1 Probable hypnic headache

##### Diagnostic criteria:

- A. Recurrent headache attacks fulfilling criteria B and C
- B. Developing only during sleep, and causing awakening
- C. Two only of the following:
  1. occurring on  $\geq 10$  days/month for >3 months
  2. lasting from 15 minutes up to four hours after waking
  3. no cranial autonomic symptoms or restlessness
- D. Not fulfilling ICHD-3 criteria for any other headache disorder
- E. 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.<sup>1-4</sup>

##### Notes:

1. 4.10 *New daily persistent headache* 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 associated with or followed by medication overuse.
2. 4.10 *New daily persistent headache* 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* whenever the criteria for this disorder are met. In contrast, when the criteria for both 4.10 *New daily persistent headache* and 3.4 *Hemicrania continua* are met, then the latter is the default diagnosis.
3. Abortive drug use may exceed the limits defined as causative of 8.2 *Medication-overuse headache*. 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* and 8.2 *Medication-overuse headache*, should be given.
4. In all cases, other secondary headaches such as 5.1 *Acute headache attributed to traumatic injury to the head*, 7.1 *Headache attributed to increased cerebrospinal fluid pressure* and 7.2 *Headache attributed to low cerebrospinal fluid pressure* should be ruled out by appropriate investigations.

**Comment:** 4.10 *New daily persistent headache* has two subtypes: a self-limiting subtype that typically resolves within several months without therapy, and a refractory subtype 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  $\leq 3$  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 and/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 the 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 subtypes) – even when the headache phenomenologically appears to be 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 general rules for attribution developed in ICHD-II allowed one or two diagnoses in such circumstances, but relied on judgement. They were modified in ICHD-3 beta to be less open to interpretation, and these modifications are retained.

1. 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).
2. When a *pre-existing* primary headache becomes *chronic* or is made *significantly worse* (usually meaning a twofold 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 adopted in ICHD-3 beta, and this too is retained:

### 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 diagnosed<sup>1</sup>
- C. Evidence of causation demonstrated by at least two of the following<sup>2</sup>:
1. headache has developed in temporal relation to the onset of the presumed causative disorder
  2. either 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 disorder<sup>3</sup>
  4. other evidence exists of causation<sup>4</sup>
- D. Not better accounted for by another ICHD-3 diagnosis.

#### Notes:

1. Since 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, however, diagnostic criteria restrict themselves to information reasonably available to the diagnosing physician in a typical clinical situation.
2. The general criteria require at least 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 upon 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 upon 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 *Acute headache attributed to non-traumatic subarachnoid haemorrhage*. 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*]).

## 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?* The general rules for attribution to another disorder apply to 5. *Headache attributed to trauma or injury to the head and/or neck.*

1. When a *new 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.
2. When a *pre-existing headache* with the characteristics of a primary headache disorder becomes *chronic* or is made *significantly worse* (usually meaning a twofold 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 types or subtypes) should be given, provided that there is good evidence that the disorder can cause headache.

### Introduction

The types 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 three 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 types of 5. *Headache attributed to trauma or injury to the head and/or neck* from other headache disorders; most often these resemble 2. *Tension-type headache* or 1. *Migraine*. Consequently, their diagnosis is largely dependent upon the close temporal relation between the trauma or injury and headache onset. Consistently with those of ICHD-II, the diagnostic criteria of ICHD-3 for all types of 5. *Headache attributed to trauma or injury to the head and/or neck* require that headache must be *reported* to have developed within seven days following trauma or injury, or within seven days after regaining consciousness and/or within seven days after recovering the ability to sense and report pain. Although this seven-day interval is somewhat arbitrary, and 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. Research is encouraged that tests the diagnostic 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* (see Appendix).

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, neuroinflammation, alterations in cerebral haemodynamics, an 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, functional and metabolic abnormalities following minor trauma that are not detectable through conventional diagnostic tests. Post-traumatic sleep disturbances, mood disturbances and psychosocial and other 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 a post-traumatic 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.

It is recognized that some patients develop headache following very minor trauma to the head – so minor that it does not meet criteria even for mild traumatic brain injury. These headaches may begin after a single trauma or following repetitive minor head impacts (e.g. in players of rugby or American football). However, headache due to very minor head trauma has not been adequately studied, so there are insufficient data to support its recognition and inclusion in ICHD-3. Research on headache following very minor trauma to the head, perhaps guided by the diagnostic criteria for A5.8 *Acute headache attributed to other trauma or injury to the head and/or neck* and A5.9 *Persistent headache attributed to other trauma or injury to the head and/or neck*, is encouraged.

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 types 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 due to 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 three months' duration caused by traumatic injury to the head.

**Diagnostic criteria:**

- A. Any headache fulfilling criteria C and D
- B. Traumatic injury to the head<sup>1</sup> has occurred
- C. Headache is reported to have developed within seven 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) impairing ability to sense or report headache following the injury to the head
- D. Either of the following:
  - 1. headache has resolved within three months after its onset
  - 2. headache has not yet resolved but three months have not yet passed since its onset
- 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 upon the head. These include impact between the head and 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 seven days is somewhat arbitrary (see 'Introduction' above). Compared to longer intervals, a seven-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* may be used when the interval between injury and headache onset is greater than seven 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 amnesia lasting >24 hours<sup>1</sup>
  - 4. alteration in level of awareness for >24 hours

5. imaging evidence of a traumatic head injury such as skull fracture, intracranial haemorrhage and/or brain contusion.

**Note:**

1. The duration of post-traumatic amnesia is defined as the time between head injury and resumption of normal continuous recall of events.

**5.1.2 Acute 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<sup>1</sup>
    - d) altered level of awareness for >24 hours
    - e) imaging evidence of a traumatic head injury such as skull fracture, intracranial haemorrhage and/or brain contusion
  2. associated 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 of the following symptoms suggestive of mild traumatic brain injury:
      - i. nausea
      - ii. vomiting
      - iii. visual disturbances
      - iv. dizziness and/or vertigo
      - v. gait and/or postural imbalance
      - vi. impaired memory and/or concentration.

**Note:**

1. The duration of post-traumatic amnesia is defined as the time between head injury and resumption of normal continuous recall of events.

**Comment:** The diagnostic criteria for mild and those for moderate or severe traumatic injury to the head allow

for substantial variability in the severity of the injury classified into 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*. There is insufficient evidence for adding these categories at present, but future studies should investigate the utility of doing so.

**5.2 Persistent headache attributed to traumatic injury to the head**

**Coded elsewhere:** Trauma due to 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 more than three months' duration caused by traumatic injury to the head.

**Diagnostic criteria:**

- A. Any headache fulfilling criteria C and D
- B. Traumatic injury to the head<sup>1</sup> has occurred
- C. Headache is reported to have developed within seven 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) impairing ability to sense or report headache following the injury to the head
- D. Headache persists for >3 months after its onset
- E. Not better accounted for by another ICHD-3 diagnosis.<sup>2</sup>

**Notes:**

1. Traumatic injury to the head is defined as a structural or functional injury resulting from the action of external forces upon the head. These include impact between the head and an object, penetration of the head by a foreign body, forces generated from blasts or explosions, and other forces yet to be defined.
2. When headache following head injury becomes persistent, the possibility of 8.2 *Medication-overuse headache* needs to be considered.

*Comments:* The stipulation that headache must be reported to have developed within seven days is somewhat arbitrary (see ‘Introduction’ above). Compared to longer intervals, a seven-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* may be used when the interval between injury and headache onset is greater than seven 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, three months is the time interval after which headache attributed to trauma or injury to the head 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 amnesia lasting >24 hours<sup>1</sup>
  4. alteration in level of awareness for >24 hours
  5. imaging evidence of a traumatic head injury such as skull fracture, intracranial haemorrhage and/or brain contusion.

*Note:*

1. The duration of post-traumatic amnesia is defined as the time between head injury and resumption of normal continuous recall of events.

**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<sup>1</sup>
    - d) altered level of awareness for >24 hours
    - e) imaging evidence of a traumatic head injury such as skull fracture, intracranial haemorrhage and/or brain contusion
  2. associated 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 of the following symptoms suggestive of mild traumatic brain injury:
      - i. nausea
      - ii. vomiting
      - iii. visual disturbances
      - iv. dizziness and/or vertigo
      - v. gait and/or postural imbalance
      - vi. impaired memory and/or concentration.

*Note:*

1. The duration of post-traumatic amnesia is defined as the time between head injury and resumption of normal continuous recall of events.

**5.3 Acute headache attributed to whiplash<sup>1</sup>**

*Description:* Headache of less than three months’ duration caused by whiplash.

*Diagnostic criteria:*

- A. Any headache fulfilling criteria C and D
- B. Whiplash,<sup>1</sup> associated at the time with neck pain and/or headache, has occurred
- C. Headache has developed within seven days after the whiplash

- D. Either of the following:
1. headache has resolved within three months after its onset
  2. headache has not yet resolved but three months have not yet passed since its onset
- 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 more than three months' duration caused by whiplash.

**Diagnostic criteria:**

- A. Any headache fulfilling criteria C and D
- B. Whiplash,<sup>1</sup> associated at the time with neck pain and/or headache, has occurred
- C. Headache developed within seven days after the whiplash
- D. Headache persists for >3 months after its onset
- E. Not better accounted for by another ICHD-3 diagnosis.<sup>2</sup>

**Notes:**

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.
2. When headache following whiplash 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 three months' duration caused by surgical craniotomy.

**Diagnostic criteria:**

- A. Any headache fulfilling criteria C and D
- B. Surgical craniotomy<sup>1</sup> has been performed
- C. Headache is reported to have developed within seven days after one of the following:
  1. the craniotomy
  2. regaining of consciousness following the craniotomy
  3. discontinuation of medication(s) impairing ability to sense or report headache following the craniotomy
- D. Either of the following:
  1. headache has resolved within three months after its onset
  2. headache has not yet resolved but three months have not yet passed since its onset
- E. Not better accounted for by another ICHD-3 diagnosis.<sup>1,2</sup>

**Notes:**

1. When the craniotomy was performed following and because of head injury, code as 5.1.1 *Acute headache attributed to moderate or severe traumatic injury to the head*.
2. 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, consideration should particularly include cervicogenic headache (due to positioning during surgery), headache from cerebrospinal fluid leak, infections, hydrocephalus and intracranial haemorrhage.

**Comments:** 5.5 *Acute headache attributed to craniotomy* occurs in a substantial proportion of patients undergoing surgical craniotomy. In the majority of cases, it begins within the first few days after craniotomy and resolves within the acute postoperative period. It is more common after surgery of the skull base compared to 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.

### 5.6 Persistent headache attributed to craniotomy

**Description:** Headache of more than three months' duration caused by surgical craniotomy.

**Diagnostic criteria:**

- A. Any headache fulfilling criteria C and D
- B. Surgical craniotomy<sup>1</sup> has been performed
- C. Headache is reported to have developed within seven days after one of the following:
  - 1. the craniotomy
  - 2. regaining of consciousness following the craniotomy
  - 3. discontinuation of medication(s) impairing ability to sense or report headache following the craniotomy
- D. Headache persists for >3 months after its onset
- E. Not better accounted for by another ICHD-3 diagnosis.<sup>1,2</sup>

**Notes:**

- 1. When the craniotomy was performed following and because of head injury, code as 5.2.1 *Persistent headache attributed to moderate or severe traumatic injury to the head*.
- 2. When headache following craniotomy becomes persistent, the possibility of 8.2 *Medication-overuse headache* needs to be considered.

**Comment:** 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*.

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## 6. Headache attributed to cranial and/or cervical vascular disorder

- 6.1 Headache attributed to cerebral ischaemic event
  - 6.1.1 Headache attributed to ischaemic stroke (cerebral infarction)
    - 6.1.1.1 Acute headache attributed to ischaemic stroke (cerebral infarction)
    - 6.1.1.2 Persistent headache attributed to past 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 Acute headache attributed to non-traumatic intracerebral haemorrhage
  - 6.2.2 Acute headache attributed to non-traumatic subarachnoid haemorrhage (SAH)
  - 6.2.3 Acute headache attributed to non-traumatic acute subdural haemorrhage (ASDH)
  - 6.2.4 Persistent headache attributed to past non-traumatic intracranial haemorrhage
    - 6.2.4.1 Persistent headache attributed to past non-traumatic intracerebral haemorrhage
    - 6.2.4.2 Persistent headache attributed to past non-traumatic subarachnoid haemorrhage
    - 6.2.4.3 Persistent headache attributed to past non-traumatic acute subdural haemorrhage
- 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
    - 6.5.1.1 Acute headache or facial or neck pain attributed to cervical carotid or vertebral artery dissection
    - 6.5.1.2 Persistent headache or facial or neck pain attributed to past cervical carotid or vertebral artery dissection
  - 6.5.2 Post-endarterectomy headache
  - 6.5.3 Headache attributed to carotid or vertebral angioplasty or stenting
- 6.6 Headache attributed to cranial venous disorder
  - 6.6.1 Headache attributed to cerebral venous thrombosis (CVT)
  - 6.6.2 Headache attributed to cranial venous sinus stenting
- 6.7 Headache attributed to other acute intracranial arterial disorder
  - 6.7.1 Headache attributed to an intracranial endarterial procedure
  - 6.7.2 Angiography headache
  - 6.7.3 Headache attributed to reversible cerebral vasoconstriction syndrome (RCVS)
    - 6.7.3.1 Acute headache attributed to reversible cerebral vasoconstriction syndrome (RCVS)
    - 6.7.3.2 Acute headache probably attributed to reversible cerebral vasoconstriction syndrome (RCVS)
    - 6.7.3.3 Persistent headache attributed to past reversible cerebral vasoconstriction syndrome (RCVS)
- 6.8 Headache and/or migraine-like aura attributed to chronic intracranial vasculopathy
  - 6.8.1 Headache attributed to Cerebral Autosomal Dominant Arteriopathy with Subcortical Infarcts and Leukoencephalopathy (CADASIL)
  - 6.8.2 Headache attributed to mitochondrial encephalopathy, lactic acidosis and stroke-like episodes (MELAS)
  - 6.8.3 Headache attributed to Moyamoya angiopathy (MMA)
  - 6.8.4 Migraine-like aura attributed to cerebral amyloid angiopathy (CAA)
  - 6.8.5 Headache attributed to syndrome of retinal vasculopathy with cerebral leukoencephalopathy and systemic manifestations (RVCLSM)
  - 6.8.6 Headache attributed to other chronic intracranial vasculopathy
- 6.9 Headache attributed to pituitary apoplexy

### General comment

*Primary or secondary headache or both?* The general rules for attribution to another disorder apply to 6. *Headache attributed to cranial and/or cervical vascular disorder.*

1. When a *new 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. This rule applies similarly to *new migraine-aura-like symptoms* occurring for the first time in close temporal relation to a cranial or cervical vascular disorder.
2. When a *pre-existing headache* with the characteristics of a primary headache disorder becomes *chronic*, or is made *significantly worse* (usually meaning a twofold 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 and/or cervical vascular disorder* (or one of its types or 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 and/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 and/or cervical vascular disorder
  2. either or both of the following:
    - a) headache has significantly worsened in parallel with worsening of the cranial and/or cervical vascular disorder
    - b) headache has significantly improved in parallel with improvement of the cranial and/or cervical vascular disorder
  3. headache has characteristics typical for the cranial and/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 cerebral ischaemic event

### 6.1.1 Headache attributed to ischaemic stroke (cerebral infarction)

#### 6.1.1.1 Acute headache attributed to ischaemic stroke (cerebral infarction)

**Description:** New and usually acute-onset headache caused by ischaemic stroke and associated with focal neurological signs of the stroke. It is very rarely the presenting or a prominent feature of ischaemic stroke. It usually has a self-limiting course.

#### **Diagnostic criteria:**

- A. Any new headache fulfilling criteria C and D
- B. Acute ischaemic stroke has been diagnosed
- C. Evidence of causation demonstrated by either or both 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. Either of the following:
  1. headache has resolved within three months<sup>1</sup>
  2. headache has not yet resolved but three months have not yet passed<sup>1</sup>
- E. Not better accounted for by another ICHD-3 diagnosis.

#### **Note:**

1. The three months should be counted from stabilization, spontaneously or through treatment, rather than onset of the ischaemic stroke.

**Comments:** 6.1.1.1 *Acute 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 ipsilateral to the stroke or bilateral. Rarely, an acute ischaemic stroke, notably an embolic cerebellar or supratentorial 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.

Headache is, however, extremely common in acute arterial wall disorders that may lead to ischaemic stroke, such as arterial 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; it is then more correctly coded to the arterial wall disorder.

**6.1.1.2 Persistent headache attributed to past ischaemic stroke (cerebral infarction)**

*Description:* Headache caused by ischaemic stroke and persisting for more than three months after the stroke has stabilized.

*Diagnostic criteria:*

- A. Headache previously diagnosed as 6.1.1.1 *Acute headache attributed to ischaemic stroke (cerebral infarction)*, and fulfilling criterion C
- B. The ischaemic stroke has stabilized, spontaneously or through treatment
- C. Headache has persisted for >3 months after stabilization of the ischaemic stroke
- D. Not better accounted for by another ICHD-3 diagnosis.

*Comments:* A few studies have documented headaches meeting the criteria for 6.1.1.2 *Persistent headache attributed to past ischaemic stroke (cerebral infarction)*. Research is needed to identify risk factors for such persistent headache; previous history of 1. *Migraine* may play a role, as may anxiety/depression.

**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. A 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.<sup>1,2</sup>

*Notes:*

- 1. 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 TIA and more frequently progressive in 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.
- 2. The coincidence of otherwise typical TIA and *severe* headache should prompt the search for some arterial disorders that can directly induce severe headache (arterial dissection, among others).

*Comments:* A transient ischaemic attack (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 one hour.

While more common with basilar- than carotid-territory TIA, headache is very rarely a prominent symptom of TIA.

**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, generally with sudden (even thunderclap) onset. Depending on the type of haemorrhage, it may be isolated or associated with focal neurological deficits.

**6.2.1 Acute headache attributed to non-traumatic intracerebral haemorrhage**

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

**Diagnostic criteria:**

- A. Any new headache fulfilling criteria C and D
- B. Intracerebral haemorrhage (ICH)<sup>1</sup> 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. Either of the following:
  1. headache has resolved within three months<sup>2</sup>
  2. headache has not yet resolved but three months have not yet passed<sup>2</sup>
- E. Not better accounted for by another ICHD-3 diagnosis.

**Notes:**

1. Through usage, the term *intracerebral* is taken in this context to include *intracerebellar*.
2. The three months should be counted from stabilization, spontaneously or through treatment, rather than onset of the intracerebral haemorrhage.

**Comments:** 6.2.1 *Acute headache attributed to non-traumatic intracerebral haemorrhage* is more often due to subarachnoid blood and local compression than to intracranial hypertension. It can occasionally present as thunderclap headache.

Headache is more usual and more severe in haemorrhagic than in ischaemic stroke. When occurring at stroke onset, headache is associated with a higher risk of early mortality in intracerebral haemorrhage but not in ischaemic stroke.

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; this may require emergency surgical decompression.

### 6.2.2 *Acute headache attributed to non-traumatic subarachnoid haemorrhage (SAH)*

**Coded elsewhere:** Non-traumatic subarachnoid haemorrhage (SAH) is distinguished from non-traumatic

convexal subarachnoid haemorrhage (cSAH). The latter disorder can present with highly variable clinical and radiological features according to its various underlying causes, which include reversible cerebral vasoconstriction syndrome (RCVS), cerebral amyloid angiopathy (CAA), endocarditis and cerebral venous thrombosis. Patients with aura-like attacks, cSAH and CAA should be coded as 6.8.4 *Migraine-like aura attributed to cerebral amyloid angiopathy*. Patients with headache, cSAH and RCVS should be coded as 6.7.3 *Headache attributed to reversible cerebral vasoconstriction syndrome*.

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

**Diagnostic criteria:**

- A. Any new headache fulfilling criteria C and D
- 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. Either of the following:
  1. headache has resolved within three months<sup>1</sup>
  2. headache has not yet resolved but three months have not yet passed<sup>1</sup>
- E. Not better accounted for by another ICHD-3 diagnosis.<sup>2,3</sup>

**Notes:**

1. The three months should be counted from stabilization, spontaneously or through treatment, rather than onset of the subarachnoid haemorrhage (SAH).
2. Diagnosis of SAH is confirmed by non-contrast-enhanced CT scan: sensitivity is close to 99% in the first six hours after onset, 98% at 12 hours and 93% at 24 hours (but dropping to 50% at seven days). When CT results are non-diagnostic, lumbar puncture is essential: xanthochromia is present in all cases

with aneurysmal SAH when cerebrospinal fluid (CSF) is collected between 12 hours and two weeks after the onset of symptoms and analysed spectrophotometrically. MRI is not indicated as an initial diagnostic test for SAH; however, fluid-attenuated inversion recovery (FLAIR) and gradient-echo T2-weighted images may be useful when the CT is normal and the CSF abnormal.

3. In the presence of non-traumatic convexal subarachnoid haemorrhage, older age, sensorimotor dysfunction, stereotyped aura-like spells and absence of significant headache suggest cerebral amyloid angiopathy as the underlying cause. Younger age and recurrent thunderclap headache predict reversible cerebral vasoconstriction syndrome.

**Comments:** Non-traumatic subarachnoid haemorrhage (SAH) is one of the most common causes of persistent, intense and incapacitating headache of abrupt onset (thunderclap headache), and is a serious condition (mortality is 40–50%, with 10–20% of patients dying before arriving at hospital; 50% of survivors are left disabled).

6.2.2 *Acute headache attributed to non-traumatic subarachnoid haemorrhage* may nonetheless be moderate and without any associated signs. The abrupt onset is the key feature. Accordingly, any patient with headache of abrupt onset or thunderclap headache should be evaluated for SAH.

Delayed diagnosis often has a catastrophic outcome: SAH is a neurointerventional emergency. However, initial misdiagnosis occurs in one quarter to one half of patients, the most common specific misdiagnosis being migraine. The most common reasons for misdiagnosis are failure to obtain appropriate neuroimaging, or misinterpretation of it, or failure to perform lumbar puncture in cases where this is required.

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). In patients who are initially misdiagnosed and in whom SAH is belatedly recognized when they present again a few days later, there is often no aneurysm and no cause identifiable for the SAH.

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

**Coded elsewhere:** Most cases of acute subdural haemorrhage occur after head trauma; headache in such cases should be coded accordingly.

**Description:** Headache caused by non-traumatic acute subdural haemorrhage, typically severe and sudden, peaking in seconds (thunderclap headache) or minutes.

It is usually accompanied or rapidly followed by focal signs and a decrease in consciousness.

**Diagnostic criteria:**

- A. Any new headache fulfilling criteria C and D
- 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 characteristics:
    - a) sudden or thunderclap onset
    - b) localized in accordance with the site of the haemorrhage
- D. Either of the following:
  1. headache has resolved within three months<sup>1</sup>
  2. headache has not yet resolved but three months have not yet passed<sup>1</sup>
- E. Not better accounted for by another ICHD-3 diagnosis.

**Note:**

1. The three months should be counted from stabilization, spontaneously or through treatment, rather than onset of the acute subdural haemorrhage.

**Comments:** Non-traumatic acute subdural haemorrhage (ASDH) without other intracranial haemorrhage ('pure ASDH') is rare. It represents a life-threatening condition and 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, Moyamoya 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.

#### 6.2.4 Persistent headache attributed to past non-traumatic intracranial haemorrhage

**Description:** Headache caused by non-traumatic intracranial haemorrhage and persisting for more than three months after the haemorrhage has stabilized.

##### Diagnostic criteria:

- A. Headache previously diagnosed as 6.2.1 *Acute headache attributed to non-traumatic intracerebral haemorrhage*, 6.2.2 *Acute headache attributed to non-traumatic subarachnoid haemorrhage* or 6.2.3 *Acute headache attributed to non-traumatic acute subdural haemorrhage* and fulfilling criterion C
- B. The intracranial haemorrhage (of whichever type) has stabilized, spontaneously or through treatment
- C. Headache has persisted for >3 months after stabilization of the intracranial haemorrhage
- D. Not better accounted for by another ICHD-3 diagnosis.

**Comments:** A few studies have documented headaches meeting the criteria for 6.2.4 *Persistent headache attributed to past non-traumatic intracranial haemorrhage*. Research is needed to identify risk factors for such a persistent headache; previous history of 1. *Migraine* may play a role, as may anxiety/depression.

Codable subforms are 6.2.4.1 *Persistent headache attributed to past non-traumatic intracerebral haemorrhage*, 6.2.4.2 *Persistent headache attributed to past non-traumatic subarachnoid haemorrhage* and 6.2.4.3 *Persistent headache attributed to past non-traumatic acute subdural haemorrhage*.

### 6.3 Headache attributed to unruptured vascular malformation

**Coded elsewhere:** New headache attributed to ruptured vascular malformation is coded as 6.2.1 *Acute headache attributed to non-traumatic intracerebral haemorrhage*, 6.2.2 *Acute headache attributed to non-traumatic subarachnoid haemorrhage* or, rarely, 6.2.3 *Acute 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-limiting 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.<sup>1</sup>

##### Note:

1. In particular, 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. Any 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 four weeks prior to diagnosis of aneurysmal rupture. Setting aside the possibility of

memory biases, this suggests these headaches are due to 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 the risks of re-bleeding, patients with sudden severe headaches should undergo complete investigation, including cerebral imaging, CSF study and cerebral angiography (MR 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 growth of the AVM
    - b) headache has significantly improved or resolved in parallel with effective treatment of the AVM
  - 3. headache is localized to the site of the AVM
- D. Not better accounted for by another ICHD-3 diagnosis.<sup>1</sup>

*Note:*

- 1. In particular, intracranial haemorrhage has been excluded by appropriate investigations.

*Comments:* Cases have been reported highlighting the association of arteriovenous malformation (AVM) with different types of 3. *Trigeminal autonomic cephalalgias* including 3.1 *Cluster headache*, 3.2.2 *Chronic paroxysmal hemicrania* and 3.3.1 *Short-lasting unilateral neuralgiform headache attacks 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 migraine-like symptoms much more rarely.

**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 or resolved after effective 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.<sup>1</sup>

*Note:*

- 1. In particular, 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 due to 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 *Acute headache attributed to non-traumatic 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, or led to its discovery
  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.<sup>1</sup>

##### *Note:*

1. In particular, 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 cluster headache-like, 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 *KRIT1* 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.

*Comments:* Sturge Weber syndrome occurs exclusively sporadically, resulting from a somatic mosaic mutation in the *GNAQ* gene (guanine nucleotide-binding protein, Q polypeptide).

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 and/or motor 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 (GCA). Headache may be the sole symptom of GCA, a disease most conspicuously associated with headache. 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 three 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 (GCA) is the disease most conspicuously associated with headache, which is due to 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 very suggestive of GCA and should prompt urgent investigations. The major risk is of blindness due to 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 one 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 (PACNS). 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.<sup>1</sup>

*Note:*

- 1. In particular, central nervous system (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, respectively angiography and histology. 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.

Primary angiitis of the central nervous system (PACNS) can present with angiographic findings quite similar to those of reversible cerebral vasoconstriction syndrome (RCVS), including multifocal stenosis of the intracranial arteries; recurrent thunderclap headaches should suggest a diagnosis of RCVS and not PACNS.

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 (SACNS). 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, respectively angiography and histology. 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 twofold: (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 a cervical artery disorder, or has led to the diagnosis of a 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 one 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 carotid or vertebral artery dissection*

##### 6.5.1.1 *Acute headache or facial or neck pain attributed to cervical carotid or vertebral artery 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 criteria C and D
- 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 the cervical artery dissection, or has led to its diagnosis
  - 2. either or both of the following:
    - a) pain has significantly worsened in parallel with other signs of the cervical artery dissection
    - b) pain has significantly improved or resolved within one 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. Either of the following:
  - 1. headache has resolved within three months<sup>1</sup>
  - 2. headache has not yet resolved but three months have not yet passed<sup>1</sup>
- E. Not better accounted for by another ICHD-3 diagnosis.

**Note:**

- 1. The three months should be counted from stabilization, spontaneously or through treatment, rather than onset of the cervical artery dissection.

**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.

6.5.1.1 *Acute headache or facial or neck pain attributed to cervical carotid or vertebral artery dissection* is usually unilateral (ipsilateral to the dissected artery), severe and prolonged (for a mean of four 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 artery dissection* may be present in addition to 6.5.1.1 *Acute headache or facial or neck pain attributed to cervical carotid or vertebral artery dissection*.

6.5.1.1 *Acute headache or facial or neck pain attributed to cervical carotid or vertebral artery 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, MR and/or CT angiography and, in doubtful cases, conventional angiography. Several of these investigations are commonly needed since 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 three to six months according to the quality of the arterial recovery.

**6.5.1.2 Persistent headache or facial or neck pain attributed to past cervical carotid or vertebral artery dissection**

**Description:** Headache caused by cervical carotid or vertebral artery dissection and persisting for more than three months after the dissection has stabilized.

**Diagnostic criteria:**

- A. Headache previously diagnosed as 6.5.1.1 *Acute headache or facial or neck pain attributed to cervical carotid or vertebral artery dissection* and fulfilling criterion C
- B. The dissection has stabilized, spontaneously or through treatment
- C. Headache has persisted for >3 months after stabilization of the dissection
- D. Not better accounted for by another ICHD-3 diagnosis.

**Comment:** A few studies have documented headaches meeting the criteria for 6.5.1.2 *Persistent headache or facial or neck pain attributed to past cervical carotid or vertebral artery dissection*. Research is needed to identify risk factors for such persistent headache; a previous history of 1. *Migraine* may play a role, as may anxiety/depression.

**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 one week of the carotid endarterectomy
  - 2. headache resolves within one month after the carotid endarterectomy
  - 3. both of the following:
    - a) headache is unilateral, on the side of the carotid endarterectomy
    - b) headache has one of the following three distinct characteristics<sup>1</sup>:
      - i. diffuse mild pain
      - ii. cluster headache-like pain occurring once or twice a day in attacks lasting two to three hours
      - iii. pulsating severe pain
- D. Not better accounted for by another ICHD-3 diagnosis.<sup>2</sup>

**Notes:**

- 1. Three subforms of 6.5.2 *Post-endarterectomy headache* have been described, but are not separately coded:
  - a) a diffuse, mild, isolated headache occurring in the first few days after surgery
  - b) a unilateral cluster headache-like pain with attacks, lasting two to three hours, occurring once or twice a day
  - c) unilateral pulsating and severe pain occurring three days after surgery.
- 2. In particular, arterial dissection has been excluded by appropriate investigations.

**Comment:** Of the three subforms of 6.5.2 *Post-endarterectomy headache*, the first and most frequent (up to 60% of cases) is a benign self-limiting condition, while the second (reported in up to 38% of cases) resolves in about two weeks. The third subform is part of the rare hyperperfusion syndrome, often preceding a rise in blood pressure and the onset of seizures or neurological deficits on or about the seventh day. Urgent treatment is required, since these symptoms can herald cerebral haemorrhage.

### 6.5.3 Headache attributed to carotid or vertebral angioplasty or stenting

**Description:** Headache caused by the endovascular procedures of cervical angioplasty and/or stenting.

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 and/or stenting has been performed
- C. Evidence of causation demonstrated by all of the following:
  - 1. headache has developed within one week of the angioplasty and/or stenting
  - 2. headache has resolved within one month after the angioplasty and/or stenting
  - 3. headache is on the same side as the angioplasty and/or stenting
- D. Not better accounted for by another ICHD-3 diagnosis.<sup>1</sup>

**Note:**

- 1. In particular, arterial dissection has been excluded by appropriate investigations.

**Comments:** Carotid and vertebral angioplasty and/or stenting are performed to treat cervical artery stenosis.

In a series of 64 patients who had carotid stenting, headache occurred in one-third, usually within 10 minutes after the procedure, and was mild, ipsilateral, frontotemporal and pressing in nature; it mostly disappeared within 10 minutes. Otherwise, data on 6.5.3 *Headache attributed to carotid or vertebral angioplasty or stenting* remain scarce. Headache is not mentioned in large trials comparing carotid stenting and endarterectomy.

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

## 6.6 Headache attributed to cranial venous disorder

### 6.6.1 Headache attributed to cerebral venous thrombosis (CVT)

**Description:** Headache caused by cerebral venous thrombosis (CVT). 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.

6.6.1 *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 1.1 *Migraine without aura*, 1.2 *Migraine with aura*, 3.1 *Cluster headache*, 3.4 *Hemicrania continua*, 4.4 *Primary thunderclap headache*, 7.2 *Headache attributed to low cerebrospinal fluid pressure* or 6.2.2 *Acute headache attributed to non-traumatic subarachnoid haemorrhage (SAH)* (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 6.6.1 *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 six months of oral anticoagulation and, whenever indicated, treatment of the underlying cause.

6.6.2 *Headache attributed to cranial venous sinus stenting*

**Description:** Unilateral headache caused by and on the same side as cranial venous sinus stenting.

*Diagnostic criteria:*

- A. New unilateral headache, fulfilling criterion C
- B. Jugular or cranial venous stenting has been performed
- C. Evidence of causation demonstrated by all of the following:
  - 1. headache has developed within one week of the stenting
  - 2. headache has resolved within three months after the stenting
  - 3. headache is ipsilateral to the stenting
- D. Not better accounted for by another ICHD-3 diagnosis.<sup>1</sup>

**Note:**

- 1. In particular, within-stent venous thrombosis has been excluded.

**Comments:** Over the past decade, stenting of lateral sinus stenosis has been used to treat idiopathic intracranial hypertension.

Data about 6.6.2 *Headache attributed to cranial venous sinus stenting* are scarce. In one series of 21 patients stented for idiopathic intracranial hypertension, 10 patients exhibited ‘stent-headaches’ differing from those experienced before treatment, located at the site of the stent, in the mastoid region, and lasting about three weeks.

6.7 *Headache attributed to other acute intracranial arterial disorder*

6.7.1 *Headache attributed to an intracranial endarterial procedure*

**Description:** Unilateral headache caused directly by an intracranial endarterial procedure, ipsilateral to the procedure and lasting less than 24 hours.

*Diagnostic criteria:*

- A. Any new headache fulfilling criterion C
- B. An intracranial endarterial procedure has been performed<sup>1</sup>
- C. Evidence of causation demonstrated by at least three of the following:
  - 1. headache has developed within one week of the procedure
  - 2. headache has resolved within one month after the procedure
  - 3. headache is ipsilateral to the procedure, or bilateral
  - 4. headache has one of the following sets of characteristics<sup>2</sup>:

- a) severe, occurring abruptly within seconds of the procedure and lasting <1 hour
  - b) moderate to severe, developing within hours of the procedure and lasting >24 hours
  - c) occurring in a patient with 1. *Migraine* and 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.<sup>3</sup>

**Notes:**

1. For example, angioplasty, embolization or stent placement.
2. There are three recognized (but not separately coded) subforms of 6.7.1 *Headache attributed to an intracranial endarterial procedure*:
  - a) a very specific subform reported after balloon inflation or embolization of an arteriovenous malformation or aneurysm: severe pain localized according to the artery involved, developing abruptly within a few seconds of the procedure, and disappearing rapidly
  - b) headache developing within hours to one day following the procedure and lasting a few days
  - c) a migraine attack, occurring in a person who has 1. *Migraine* and triggered by the intracranial endarterial procedure; this is sometimes followed by recurrent intermittent headache during several weeks (in these cases, the patient should have both diagnoses: the appropriate type or subtype of 1. *Migraine* and 6.7.1 *Headache attributed to an intracranial endarterial procedure*).
3. In particular, arterial dissection and arterial rupture have been excluded by appropriate investigations.

### 6.7.2 Angiography headache

**Description:** Headache caused directly by cerebral angiography.

**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 or within 24 hours of the angiography
  2. headache has resolved within 72 hours after the angiography

3. headache has one of the following sets of characteristics<sup>1</sup>:

- a) developing during contrast injection and lasting <1 hour
- b) developing a few hours after the angiography and lasting >24 hours
- c) occurring in a patient with 1. *Migraine* and 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.

**Note:**

1. There are three recognized (but not separately coded) subforms of 6.7.2 *Angiography headache*.
  - a) occurring during angiography, and closely related to contrast injection
  - b) occurring later, but within 24 hours (both these subforms are more common in patients with a history of primary headache, but are distinctly different in character from the primary headache)
  - c) a migraine attack, occurring in a person who has 1. *Migraine* and triggered by angiography (in these cases, the patient should have both diagnoses: the appropriate type or subtype of 1. *Migraine* and 6.7.2 *Angiography headache*).

**Comment:** 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)

#### 6.7.3.1 Acute headache attributed to reversible cerebral vasoconstriction syndrome (RCVS)

**Description:** Headache caused by reversible cerebral vasoconstriction syndrome (RCVS), typically thunder-clap headache recurring over one to two weeks, often triggered by sexual activity, exertion, Valsalva manœuvres and/or emotion. Headache can remain the sole symptom of RCVS or be a warning symptom preceding haemorrhagic or ischaemic stroke.

**Diagnostic criteria:**

- A. Any new headache fulfilling criterion C
- B. Reversible cerebral vasoconstriction syndrome (RCVS) has been diagnosed
- C. Evidence of causation demonstrated by either or both of the following:

1. headache, with or without focal deficits and/or seizures, has led to angiography (with ‘string of beads’ appearance) and diagnosis of RCVS
2. headache has one or more of the following characteristics:
  - a) thunderclap onset
  - b) triggered by sexual activity, exertion, Valsalva manœuvres, emotion, bathing and/or showering
  - c) present or recurrent during  $\leq 1$  month after onset, with no new significant headache after  $>1$  month
- D. Either of the following:
  1. headache has resolved within three months of onset
  2. headache has not yet resolved but three months from onset have not yet passed
- E. Not better accounted for by another ICHD-3 diagnosis.<sup>1</sup>

**Note:**

1. In particular, 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 subarachnoid haemorrhage.

RCVS is the most frequent cause of thunderclap headache recurring over a few days or weeks. 6.7.3.1 *Acute headache attributed to reversible cerebral vasoconstriction syndrome* may rarely have other modes of onset: progressing rapidly over hours or more slowly over days.

Large series of patients with confirmed RCVS have shown that up to 75% present with headache as the only symptom, but the condition can be associated with fluctuating focal neurological deficits and sometimes seizures. 6.7.3.1 *Acute headache attributed to reversible cerebral vasoconstriction syndrome* may be a warning symptom preceding haemorrhagic or ischaemic stroke. Headache is absent in a minority of cases of RCVS.

Angiography in RCVS is, by definition, abnormal, with alternating segments of arterial constriction and dilatation (‘string of beads’ or ‘sausage on a string’ 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 6.7.3.2 *Acute headache probably attributed to reversible cerebral vasoconstriction syndrome*. Brain MRI is abnormal in 30% to 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 attributable to exposure to vasoactive substances including illicit drugs, alpha-sympathomimetics and serotonergic drugs. The disease is self-limiting in one to three months, with disappearance of the arterial abnormalities (hence ‘reversible’) and, almost always, resolution of the headache. However, strokes due to RCVS can produce permanent impairment.

**6.7.3.2 Acute headache probably attributed to reversible cerebral vasoconstriction syndrome (RCVS)**

**Description:** Headache typical for reversible cerebral vasoconstriction syndrome (RCVS), namely thunderclap headache, recurring over one to two 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 one month, with all three of the following characteristics:
    - a) thunderclap onset, and peaking in  $<1$  minute
    - b) severe intensity
    - c) lasting  $\geq 5$  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. Either of the following:
  1. headache has resolved within three months of its onset
  2. headache has not yet resolved but three months from its onset have not yet passed
- E. Not better accounted for by another ICHD-3 diagnosis.<sup>1</sup>

**Note:**

1. In particular, aneurysmal subarachnoid haemorrhage has been excluded by appropriate investigations.

**Comment:** ICHD-3 does not generally propose criteria for probable secondary headaches. However, the arterial abnormalities of reversible cerebral vasoconstriction syndrome (RCVS) may be difficult to demonstrate. Some RCVS cases need repeated CT- or MR-angiography during two to three 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 one month but normal initial cerebral angiography, and in whom another cause of the headaches has been excluded by appropriate investigations, a temporary diagnosis of 6.7.3.2 *Headache probably attributed to reversible cerebral vasoconstriction syndrome* can be made.

#### 6.7.3.3 Persistent headache attributed to past reversible cerebral vasoconstriction syndrome (RCVS)

**Description:** Headache caused by reversible cerebral vasoconstriction syndrome (RCVS) and persisting for more than three months after onset.

**Diagnostic criteria:**

- A. Headache previously diagnosed as 6.7.3.1 *Acute headache attributed to reversible cerebral vasoconstriction syndrome (RCVS)* and fulfilling criterion C
- B. Normalization of cerebral arteries, shown by follow-up indirect or direct angiography, within three months of onset of RCVS
- C. Headache has persisted for >3 months after its onset
- D. Not better accounted for by another ICHD-3 diagnosis.

**Comment:** A few studies have documented headaches meeting the criteria for 6.7.3.3 *Persistent headache attributed to past reversible cerebral vasoconstriction syndrome*. Research is needed to identify risk factors for such persistent headache; a previous history of 1. *Migraine* may play a role, as may anxiety/depression.

#### 6.7.4 Headache attributed to intracranial artery 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 subarachnoid haemorrhage or 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 arterial dissection, or has led to its diagnosis
  2. headache resolves within one 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.

**Comments:** Dissection can affect any intracranial artery and may induce subarachnoid haemorrhage, ischaemic infarcts, compression of adjacent structures or, less commonly, intracerebral haemorrhage. In Asians, intracranial arterial dissection is more frequent than cervical artery dissection.

Acute headache is often the presenting symptom and can be the sole symptom of this disorder.

### 6.8 Headache and/or migraine-like aura attributed to chronic intracranial vasculopathy

#### 6.8.1 Headache attributed to cerebral autosomal dominant arteriopathy with subcortical infarcts and leukoencephalopathy (CADASIL)

**Description:** Headache recurring in attacks resembling 1.2 *Migraine with aura*, except for an unusual frequency of prolonged aura, caused by cerebral autosomal dominant arteriopathy with subcortical infarcts and leukoencephalopathy (CADASIL). It is associated with the other clinical features of CADASIL or, often, the first symptom of it.

**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<sup>1</sup>
- 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.

**Note:**

- 1. The diagnosis is made by screening for *NOTCH3* mutations, by a simple skin biopsy with immunostaining of NOTCH3 antibodies, or with electron microscopy to assess for extracellular granular osmiophilic material (GOM) within the arterial media.

**Comments:** Cerebral autosomal dominant arteriopathy with subcortical infarcts and leukoencephalopathy (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 due to mutations of the *NOTCH3* gene.

CADASIL is characterized clinically by recurrent small deep infarcts, subcortical dementia, mood disturbances and, in one-third of cases, by attacks typical of 1.2 *Migraine with aura* except for an unusual frequency of prolonged aura. In such cases, these are 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.

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

**6.8.2 Headache attributed to mitochondrial encephalopathy, lactic acidosis and stroke-like episodes (MELAS)**

**Description:** Headache, which is either recurrent in migraine-like attacks or a presenting symptom of stroke-like episodes, caused by and associated with the other clinical features of mitochondrial encephalopathy, lactic acidosis and stroke-like episodes (MELAS).

**Diagnostic criteria:**

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

**Comments:** Mitochondrial encephalopathy, lactic acidosis and stroke-like episodes (MELAS) is 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). Headache is common 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 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, since migraine attacks, mostly with aura, also occur in other mitochondrial disorders.

**6.8.3 Headache attributed to Moyamoya angiopathy (MMA)**

**Description:** Chronic recurrent headache, which may be migraine-like, caused by and associated with the other clinical features of Moyamoya angiopathy.

**Diagnostic criteria:**

- A. Recurrent headache fulfilling criterion C
- B. Neuroimaging evidence of Moyamoya angiopathy (MMA)
- 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 and/or imaging evidence of MMA, or led to its discovery
  - 2. either or both of the following:
    - a) headache has significantly worsened in parallel with other symptoms and/or clinical and/or radiological signs of worsening of MMA
    - b) headache has significantly improved after revascularization surgery
- D. Not better accounted for by another ICHD-3 diagnosis.

**Comments:** Moyamoya angiopathy (MMA) is characterized by bilateral progressive narrowing and occlusion of the intracranial portion of the internal carotid, middle cerebral and anterior cerebral arteries. Several susceptibility genes have been identified or localized for MMA. In some other patients, MMA is associated with other conditions (sickle cell anaemia, Down syndrome and radiation therapy amongst others) and is referred to as Moyamoya syndrome.

MMA usually presents early in childhood or adolescence with ischaemic or haemorrhagic stroke that can cause acute headache. Outside of these acute vascular events, headache is highly common in both children and adults with MMA, phenotypically most commonly resembling 1.1 *Migraine without aura*, 1.2 *Migraine with aura*, 1.2.3 *Hemiplegic migraine* or 2. *Tension-type headache*; cluster headache-like attacks have been rarely reported.

Revascularization surgery has variable effects on headache in MMA, with improvement in some patients, persistence in others, and postoperative new-onset headache in another subset.

#### 6.8.4 Migraine-like aura attributed to cerebral amyloid angiopathy (CAA)

**Description** Late-onset migraine-like aura attacks without headache or with mild headache, also termed ‘amyloid spells’, caused by and associated with the other clinical features of cerebral amyloid angiopathy, often in the setting of convexal subarachnoid haemorrhage.

##### **Diagnostic criteria:**

- A. New attacks of migraine-like aura, with or without mild headache, fulfilling criterion C
- B. Neuroimaging or brain biopsy evidence of cerebral amyloid angiopathy (CAA)
- C. Evidence of causation demonstrated by one or more of the following:
  1. aura has developed in close temporal relation to other symptoms and/or clinical signs of CAA, or led to its discovery
  2. aura has significantly worsened in parallel with clinical and/or radiological signs of worsening of the CAA
  3. onset after 50 years of age
- D. Not better accounted for by another ICHD-3 diagnosis.<sup>1</sup>

##### **Note:**

1. Blood-sensitive MRI sequences are important in diagnosing cerebral amyloid angiopathy, and should be performed in any patient with late-onset migraine-like aura.

**Comments:** Cerebral amyloid angiopathy (CAA) is a small-vessel disorder associated with progressive amyloid deposition in the walls of cortical and leptomeningeal vessels. Sporadic forms are more common than inherited familial forms.

CAA is a major cause of lobar symptomatic intracerebral haemorrhage, transient focal neurologic

episodes in the elderly and cognitive impairment. Transient focal neurological episodes include both positive migraine aura-like (spreading paraesthesias and/or positive visual phenomena) and negative TIA-like neurological symptoms, and may be caused by superficial cortical siderosis or convexal subarachnoid haemorrhage. These episodes are associated with a high early risk of symptomatic intracerebral haemorrhage.

#### 6.8.5. Headache attributed to syndrome of retinal vasculopathy with cerebral leukoencephalopathy and systemic manifestations (RVCLSM)

**Description:** Headache recurring as migraine-like attacks, mainly without aura, caused by the syndrome of retinal vasculopathy with cerebral leukoencephalopathy and systemic manifestations (RVCLSM). It may be associated with the other clinical features of RVCLSM or be the earliest clinical manifestation of it.

##### **Diagnostic criteria:**

- A. Recurrent migraine-like attacks, with or without aura, fulfilling criterion C
- B. The syndrome of retinal vasculopathy with cerebral leukoencephalopathy and systemic manifestations (RVCLSM) has been demonstrated<sup>1</sup>
- C. Migraine-like attacks are secondary to and part of the clinical manifestations of the syndrome
- D. Not better accounted for by another ICHD-3 diagnosis.

##### **Note:**

1. Diagnosis is made by genetic testing for *TREX1* mutations.

**Comment:** The syndrome of retinal vasculopathy with cerebral leukoencephalopathy and systemic manifestations (RVCLSM) is an autosomal dominant systemic small-vessel disease caused by C-terminal frame-shift mutations in *TREX1*. It is characterized clinically by focal neurological deficits, cognitive impairment, psychiatric disturbances, seizures, various systemic manifestations and, in at least half of cases, by migraine-like attacks. Other clinical manifestations are visual impairment from vascular retinopathy, and neurological decline and premature death due to progressive enhancing cerebral white matter lesions. The clinical spectrum also includes impaired liver and kidney function, anaemia sometimes associated with gastrointestinal bleeding and hypertension. In younger patients, in

whom brain MRI may be normal, the clinical manifestations include mild Raynaud's phenomenon (54%), migraine (mainly without aura: 42%) and psychiatric disturbances (23%). The diagnosis in such cases may be suspected from family history.

**6.8.6 Headache attributed to other chronic intracranial vasculopathy**

*Description:* Migraine-like attacks, with or without aura, caused by and occurring as part of the clinical manifestations of a genetic or non-genetic chronic intracranial vasculopathy other than those described above.

*Diagnostic criteria:*

- A. Recurrent migraine-like attacks, with or without aura, fulfilling criterion C
- B. A genetic or non-genetic chronic intracranial vasculopathy has been demonstrated
- C. Migraine-like attacks are secondary to and part of the clinical manifestations of the chronic intracranial vasculopathy
- D. Not better accounted for by another ICHD-3 diagnosis.

*Comments:* Recurrent migraine-like attacks have been reported as part of the clinical manifestations of the autosomal dominant *hereditary infantile hemiparesis, retinal arterial tortuosity and leucoencephalopathy* (HIHRATL), a condition due to *COL4A1* mutations. Only a few families with this disorder have been reported. Because of the other severe manifestations, these migraine-like attacks have not been systematically investigated in HIHRATL but they appear mainly to resemble 1.2 *Migraine with aura*.

All of the other rare genetic and non-genetic chronic intracranial vasculopathies can, potentially, cause migraine-like attacks.

**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 non-aneurysmal subarachnoid haemorrhage.

It is also one of the causes of thunderclap headache. Most cases occur as the first presentation of rapid enlargement of non-functioning pituitary macroadenomas due to haemorrhage and/or infarction.

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

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## 7. Headache attributed to non-vascular intracranial disorder

- 7.1 Headache attributed to increased cerebrospinal fluid (CSF) 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 cause
  - 7.1.3 Headache attributed to intracranial hypertension secondary to chromosomal disorder
  - 7.1.4 Headache attributed to intracranial hypertension secondary to hydrocephalus
- 7.2 Headache attributed to low cerebrospinal fluid (CSF) pressure
  - 7.2.1 Post-dural puncture headache
  - 7.2.2 Cerebrospinal fluid (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 Ictal epileptic headache
  - 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?* The general rules for attribution to another disorder apply to 7. *Headache attributed to non-vascular intracranial disorder.*

1. When a *new 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.

2. When a *pre-existing headache* with the characteristics of a primary headache disorder becomes *chronic*, or is made *significantly worse* (usually meaning a twofold 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 types or subtypes) should be given, provided that there is good evidence that the disorder can cause headache.

### Introduction

In this chapter are the headaches 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 to 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, or has led to its discovery
  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 one month after successful treatment or spontaneous resolution of the

intracranial disorder usually has other mechanisms. Headache persisting for more than three 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 (CSF) pressure

**Coded elsewhere:** Headache attributed to increased 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.

#### Diagnostic criteria:

- A. New headache, or a significant worsening<sup>1</sup> of a pre-existing headache, fulfilling criterion C
- B. Intracranial hypertension has been diagnosed, with both of the following:
  1. cerebrospinal fluid (CSF) pressure exceeds 250 mm CSF (or 280 mm CSF in obese children)<sup>2</sup>
  2. normal CSF composition
- C. Evidence of causation demonstrated by at least two of the following:
  1. headache has developed in temporal relation to the intracranial hypertension, or led to its discovery
  2. headache is relieved by reducing the intracranial hypertension
  3. papilloedema
- D. Not better accounted for by another ICHD-3 diagnosis.<sup>3</sup>

#### Notes:

1. 'Significant worsening' implies a twofold or greater increase in frequency and/or severity in accordance with the general rule on distinguishing secondary from primary headache.
2. For diagnostic purposes, CSF pressure should be measured in the absence of treatment to lower intracranial pressure. CSF pressure may be measured by lumbar puncture performed in the lateral decubitus position without sedative medications or by epidural or intraventricular monitoring. Because CSF pressure varies during the course of a day, a single measurement may not be indicative of the average CSF

pressure over 24 hours: prolonged lumbar or intraventricular pressure monitoring may be required in cases of diagnostic uncertainty.

3. Intracranial neoplasm has been excluded.

**Comment:** 7.1 *Headache attributed to increased cerebrospinal fluid (CSF) pressure* is a headache type. The diagnosis, when made, should be temporary only, pending determination of the cause of raised CSF pressure; the headache should then be recoded to the appropriate subtype.

#### 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:** New headache, or a significant worsening of a pre-existing headache, caused by and accompanied by other symptoms and/or clinical and/or neuroimaging signs of idiopathic intracranial hypertension (IIH), with typical features suggestive of IIH.

#### Diagnostic criteria:

- A. New headache, or a significant worsening<sup>1</sup> of a pre-existing headache, fulfilling criterion C
- B. Both of the following:
  1. idiopathic intracranial hypertension (IIH) has been diagnosed<sup>2</sup>
  2. cerebrospinal fluid (CSF) pressure exceeds 250 mm CSF (or 280 mm CSF in obese children)<sup>3</sup>
- C. Either or both of the following:
  1. headache has developed or significantly worsened<sup>1</sup> in temporal relation to the IIH, or led to its discovery
  2. headache is accompanied by either or both of the following:
    - a) pulsatile tinnitus
    - b) papilloedema<sup>4</sup>
- D. Not better accounted for by another ICHD-3 diagnosis.<sup>5,6</sup>

#### Notes:

1. 'Significant worsening' implies a twofold or greater increase in frequency and/or severity in accordance with the general rule on distinguishing secondary from primary headache.
2. IIH should be diagnosed with caution in those with altered mental status.

3. For diagnostic purposes, CSF pressure should be measured in the absence of treatment to lower intracranial pressure. CSF pressure may be measured by lumbar puncture performed in the lateral decubitus position without sedative medications or by epidural or intraventricular monitoring. Because CSF pressure varies during the course of a day, a single measurement may not be indicative of the average CSF pressure over 24 hours: prolonged lumbar or intraventricular pressure monitoring may be required in cases of diagnostic uncertainty.
4. Papilloedema must be distinguished from pseudopapilloedema or optic disc oedema. The majority of patients with IIH have papilloedema, and IIH should be diagnosed with caution in patients without this sign.
5. 7.1.1 *Headache attributed to idiopathic intracranial hypertension* may mimic the primary headaches, especially 1.3 *Chronic migraine* and 2.3 *Chronic tension-type headache*; on the other hand, these disorders commonly coexist with IIH.
6. 8.2 *Medication-overuse headache* should be excluded in patients lacking papilloedema, abducens palsy or the characteristic neuroimaging signs of IIH.

**Comments:** Idiopathic intracranial hypertension (IIH) most commonly occurs in obese females of childbearing age (who are also the most likely to be misdiagnosed with IIH).

7.1.1 *Headache attributed to idiopathic intracranial hypertension* lacks specific features, and commonly resembles 1. *Migraine* or 2. *Tension-type headache*. Daily occurrence is not required for diagnosis.

Relief of the headache after cerebrospinal fluid (CSF) removal is supportive of the diagnosis but not on its own diagnostic: it may be seen in patients with other headache types (sensitivity 72% and specificity 77% for 7.1.1 *Headache attributed to idiopathic intracranial hypertension*).

Neuroimaging findings consistent with the diagnosis of IIH include empty sella turcica, distention 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.

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

**Coded elsewhere:** Headache attributed to increased intracranial pressure due to 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 8.1.10 *Headache attributed to long-term use of non-headache medication*.

**Description:** Headache caused by intracranial hypertension secondary to any of a variety of systemic disorders and accompanied by other symptoms and/or clinical and/or neuroimaging signs both of the intracranial hypertension and of the underlying causative disorder. It usually remits with resolution of the systemic disorder.

**Diagnostic criteria:**

- A. Headache fulfilling criteria for 7.1 *Headache attributed to increased cerebrospinal fluid (CSF) pressure* and criterion C below
- B. Intracranial hypertension has been attributed to a metabolic, toxic or hormonal disorder<sup>1</sup>
- C. Evidence of causation demonstrated by either or both of the following:
  1. headache has developed in temporal relation to the increase in CSF pressure, or led to its discovery
  2. either or both of the following:
    - a) headache has significantly worsened in parallel with increasing CSF pressure
    - b) headache has significantly improved in parallel with reduction in CSF pressure
- D. Not better accounted for by another ICHD-3 diagnosis.

**Note:**

1. Potential metabolic, toxic or hormonal causes of intracranial hypertension include acute hepatic failure, renal failure, hypercarbia, acute hypertensive crisis, Reye's hepatocerebral syndrome, cerebral venous sinus thrombosis, right heart failure, a range of substances (including thyroid hormone as replacement in children, all-trans retinoic acid, retinoids, tetracyclines and chlordecone), vitamin A toxicity and corticosteroid withdrawal.

**Comment:** Removal of the inciting agent or treatment of the underlying causative disorder may not be sufficient to normalize the high intracranial pressure; additional treatment is often required to relieve headache and other symptoms and, more importantly, to prevent visual loss.

7.1.3 *Headache attributed to intracranial hypertension secondary to chromosomal disorder*

**Description:** New headache, or a significant worsening of a pre-existing headache, caused by intracranial

hypertension secondary to a chromosomal disorder and accompanied by other symptoms and/or clinical and/or neuroimaging signs both of the intracranial hypertension and of the underlying chromosomal disorder.

*Diagnostic criteria:*

- A. New headache, or a significant worsening<sup>1</sup> of a pre-existing headache, fulfilling criteria for 7.1 *Headache attributed to increased cerebrospinal fluid (CSF) pressure* and criterion C below
- B. Intracranial hypertension has been attributed to a chromosomal disorder<sup>2</sup>
- C. Evidence of causation demonstrated by at least two of the following:
  - 1. headache has developed in temporal relation to the intracranial hypertension, or led to its discovery
  - 2. headache is relieved by reducing the intracranial hypertension
  - 3. papilloedema
- D. Not better accounted for by another ICHD-3 diagnosis.

*Notes:*

- 1. 'Significant worsening' implies a twofold or greater increase in frequency and/or severity in accordance with the general rule on distinguishing secondary from primary headache.
- 2. Chromosomal disorders associated with intracranial hypertension include Turner syndrome and Down syndrome.

**7.1.4 Headache attributed to intracranial hypertension secondary to hydrocephalus**

*Description:* New headache, or a significant worsening of a pre-existing headache, caused by intracranial hypertension secondary to hydrocephalus and accompanied by other symptoms and/or clinical signs of increased cerebrospinal fluid pressure or hydrocephalus.

*Diagnostic criteria:*

- A. New headache, or a significant worsening<sup>1</sup> of a pre-existing headache, fulfilling criteria for 7.1 *Headache attributed to increased cerebrospinal fluid (CSF) pressure* and criterion C below
- B. Intracranial hypertension has been attributed to hydrocephalus

- C. Evidence of causation demonstrated by either or both of the following:
  - 1. headache developed or significantly worsened<sup>1</sup> in temporal relation to development or worsening of the increased CSF pressure, or led to its discovery
  - 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.

*Note:*

- 1. 'Significant worsening' implies a twofold or greater increase in frequency and/or severity in accordance with the general rule on distinguishing secondary from primary headache.

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

**7.2 Headache attributed to low cerebrospinal fluid (CSF) pressure**

*Description:* Orthostatic headache caused by 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<sup>1</sup> fulfilling criterion C
- B. Either or both of the following:
  - 1. low cerebrospinal fluid (CSF) pressure (<60 mm CSF)
  - 2. evidence of CSF leakage on imaging<sup>2</sup>
- C. Headache has developed in temporal relation to the low CSF pressure or CSF leakage, or led to its discovery<sup>3</sup>
- D. Not better accounted for by another ICHD-3 diagnosis.

**Notes:**

1. 7.2 *Headache attributed to low cerebrospinal fluid (CSF) 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 upon as a diagnostic criterion.
2. Brain imaging showing brain sagging or pachymeningeal enhancement, or spine imaging (spine MRI, or MRI, CT or digital subtraction myelography) showing extradural CSF.
3. Evidence of causation may depend upon 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 five 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 two weeks, or after sealing of the leak with autologous epidural lumbar patch.

**Diagnostic criteria:**

- A. Headache fulfilling criteria for 7.2 *Headache attributed to low cerebrospinal fluid (CSF) pressure*, and criterion C below
- B. Dural puncture has been performed
- C. Headache has developed within five 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 Cerebrospinal fluid (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. Headache fulfilling criteria for 7.2 *Headache attributed to low cerebrospinal fluid (CSF) pressure*, and criterion C below
- B. A procedure has been performed, or trauma has occurred, known sometimes to cause persistent CSF leakage (CSF fistula)
- 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; hypoliqorrhoeic 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. Headache fulfilling criteria for 7.2 *Headache attributed to low cerebrospinal fluid (CSF) pressure*, and criterion C below
- B. Absence of a procedure or trauma known to be able to cause CSF leakage<sup>1</sup>
- C. Headache has developed in temporal relation to occurrence of low CSF pressure or CSF leakage, or has led to its discovery<sup>2</sup>
- D. Not better accounted for by another ICHD-3 diagnosis.

**Notes:**

1. 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.
2. Dural puncture to measure CSF pressure directly is not necessary in patients with positive MRI signs of leakage such as dural enhancement with contrast.

**Comments:** Spontaneous cerebrospinal fluid (CSF) leak has been associated with heritable connective tissue disorders. Patients with CSF leaks should be screened for connective tissue and vascular abnormalities.

While 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*. Thus, 7.2.3 *Headache attributed to spontaneous intracranial hypotension* may occur immediately or within seconds of assuming an upright position and resolve quickly (within one minute) after lying horizontally, resembling 7.2.1 *Post-dural puncture headache*, or 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. 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.

In patients with typical orthostatic headache and no apparent cause, and after exclusion of postural orthostatic tachycardia syndrome (POTS), it is reasonable in clinical practice to provide autologous lumbar epidural blood patch (EBP). While 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 targeted (to the site of the leak) and/or non-targeted lumbar EBPs, and surgical intervention may be required.

It is not clear that all patients with 7.2.3 *Headache attributed to spontaneous intracranial hypotension* have an active CSF leak, despite a compelling history or brain imaging signs compatible with CSF leakage. The underlying disorder 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 due to 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 one or more of the following:
  1. headache has developed in temporal relation to the onset of the non-infectious inflammatory disease
  2. headache has significantly worsened in parallel with worsening of the non-infectious inflammatory disease
  3. 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 and associated with other symptoms and signs of neurosarcoidosis.

**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 the neurosarcoidosis
  2. either or both of the following:
    - a) headache has significantly worsened in parallel with worsening of the neurosarcoidosis
    - b) headache has significantly improved in parallel with improvement in the 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 cerebrospinal fluid (CSF) examination<sup>1</sup>
- C. Evidence of causation demonstrated by at least two of the following:
  - 1. headache has developed in temporal relation to the onset of the aseptic meningitis, or led to its discovery
  - 2. either or both of the following:
    - a) headache has significantly worsened in parallel with worsening of the aseptic meningitis
    - b) headache has significantly improved in parallel with improvement in the 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.

**Note:**

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

**Comment:** 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 but not usually a presenting or prominent symptom of any of a variety of autoimmune disorders, and 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<sup>1</sup>
- C. Evidence of causation demonstrated by one or more of the following:

- 1. headache has developed in temporal relation to the onset of the non-infectious inflammatory disease
- 2. headache has significantly worsened in parallel with worsening of the non-infectious inflammatory disease
- 3. headache has significantly improved in parallel with improvement in the non-infectious inflammatory disease
- D. Not better accounted for by another ICHD-3 diagnosis.

**Note:**

- 1. 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 one or more of the following:
  - 1. headache has developed in temporal relation to the onset of the lymphocytic hypophysitis
  - 2. headache has significantly worsened in parallel with worsening of the lymphocytic hypophysitis
  - 3. 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 hypophyseal 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 1–12) accompanied by neurological deficits including hemiparaesthesia, hemiparesis and/or dysphasia, but positive visual symptoms only uncommonly, lasting several hours. There is cerebrospinal fluid lymphocytic pleocytosis. The disorder resolves spontaneously within three months.

*Diagnostic criteria:*

- A. Episodes of migraine-like headache fulfilling criteria B and C<sup>1</sup>
- B. Both of the following:
  1. accompanied or shortly preceded by onset of at least one of the following transient neurological deficits lasting >4 hours
    - a) hemiparaesthesia
    - b) dysphasia
    - c) hemiparesis
  2. associated with cerebrospinal fluid (CSF) lymphocytic pleocytosis (>15 white cells per  $\mu\text{l}$ ), 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 onset or worsening of 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.<sup>2</sup>

*Notes:*

1. Most patients with this syndrome have no prior history of migraine.
2. Other diagnoses that may share some of its clinical features include 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)*. Also to be excluded are neuroborreliosis, neurosyphilis, neurobrucellosis,

mycoplasma, granulomatous and neoplastic arachnoiditis, encephalitis and CNS vasculitis.

*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 three months.

In addition to cerebrospinal fluid (CSF) lymphocytosis (up to 760 cells/ $\mu\text{l}$ ), there are elevations of CSF total protein (up to 250 mg/dl) in more than 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 interictally, but brain imaging during an episode 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. Electroencephalography (EEG) and single-photon emission computed tomography (SPECT) scans may show focally abnormal areas consistent with the focal neurological deficits.

### 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 one or more 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 caused by one or more space-occupying intracranial tumours.

*Diagnostic criteria:*

- A. Any 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 four characteristics:
    - a) progressive
    - b) worse in the morning and/or when lying down
    - c) aggravated by Valsalva-like manoeuvres
    - d) accompanied by nausea and/or vomiting
- D. Not better accounted for by another ICHD-3 diagnosis.

*Comments:* The prevalence of headache in patients with intracranial tumours ranges from 32% to 71%. The likelihood of headache is greater in young patients (including children), in patients with a history of primary headache, and with rapid growth of the tumour or posterior fossa or midline localization. A low threshold for investigation is suggested in any patient with a current or prior history of cancer.

There are no pathognomonic features of 7.4.1 *Headache attributed to intracranial neoplasm*, although progression or deterioration is a key feature. The other suggestive symptoms (severe, worse in the morning and associated with nausea and vomiting) are not a classical

triad; they are more likely in the context of intracranial hypertension and with posterior fossa tumours.

The headache is not necessarily ipsilateral to the tumour. Masses adjacent to the skull or dura mater tend to be more associated with ipsilateral headaches but intracranial hypertension produces a more diffuse headache. The headache caused by a brain tumour rarely remains the only symptom: isolated headache occurs in 2–16% of patients but neurological deficits and seizures are common.

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, often triggered by postural change or Valsalva-like manoeuvre, and associated with 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.

*Comments:* 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 associated with pituitary adenoma has been demonstrated<sup>1</sup>
- 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.

**Note:**

1. Including prolactin, growth hormone (GH) and/or adrenocorticotrophic hormone (ACTH) hypersecretion.

#### 7.5 Headache attributed to intrathecal injection

**Description:** Headache experienced in both upright and recumbent postures, caused by and occurring within four 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 four days of the intrathecal injection<sup>1</sup>
  2. headache has significantly improved within 14 days after the intrathecal injection<sup>2</sup>
  3. signs of meningeal irritation
- D. Not better accounted for by another ICHD-3 diagnosis.

**Notes:**

1. Headache usually develops within four days after intrathecal injection, and is present in both upright and recumbent postures.
2. When headache persists beyond 14 days, alternative diagnoses should be considered, such as 7.2.2 *Cerebrospinal fluid (CSF) fistula headache*, meningitis or leptomenigeal disease.

### 7.6 Headache attributed to epileptic seizure

**Coded elsewhere:** 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, 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 three 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 or soon after onset of the seizure
  - 2. headache has resolved spontaneously after the seizure has terminated
- D. Not better accounted for by another ICHD-3 diagnosis.

**Comments:** Well-documented reports support recognition of the subtypes 7.6.1 *Ictal epileptic headache* and 7.6.2 *Post-ictal headache*, according to their temporal association with the epileptic seizure.

Pre-ictal headache has also been described. In a small study of 11 patients with intractable focal epilepsy, frontotemporal headache was 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. Pre-ictal headache must also be distinguished from 1.4.4 *Migraine aura-triggered seizure*.

#### 7.6.1 Ictal epileptic headache

**Previously used term:** Ictal headache.

**Description:** Headache caused by and 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 is ipsilateral to the ictal discharge
    - b) headache significantly improves or remits immediately after the partial seizure has terminated
- D. Not better accounted for by another ICHD-3 diagnosis.

**Comments:** 7.6.1 *Ictal epileptic headache* may be followed by other epileptic manifestations (motor, sensory or autonomic).

This condition should be differentiated from ‘pure’ or ‘isolated’ ictal epileptic headache occurring as the sole epileptic manifestation and requiring differential diagnosis from other headache types.

‘Hemicrania epileptica’ (if confirmed to exist) is a very rare variant of 7.6.1 *Ictal epileptic headache* characterized by ipsilateral location of headache and ictal EEG paroxysms.

#### 7.6.2 Post-ictal headache

**Description:** Headache caused by and occurring within three 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 three 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 (CM1)

**Description:** Headache caused by Chiari type I malformation, usually occipital or suboccipital, of short duration (less than five minutes) and provoked by cough or other Valsalva-like manoeuvres. 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 demonstrated<sup>1</sup>
- 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 or led to its discovery
    - b) headache has resolved within three months after successful treatment of the CM1
  2. headache has one or more of the following three characteristics:
    - a) precipitated by cough or other Valsalva-like manoeuvre
    - 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<sup>2</sup>
- D. Not better accounted for by another ICHD-3 diagnosis.<sup>3</sup>

**Notes:**

1. Diagnosis of Chiari malformation type I (CM1) 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 cerebrospinal fluid (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. Almost all (95%) patients with CM1 report a constellation of five or more distinct symptoms.
3. Patients with altered CSF pressure, either increased as in idiopathic intracranial hypertension (IIH) or decreased as in 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 either as 7.1.1 *Headache attributed to idiopathic intracranial hypertension* or as 7.2.3 *Headache attributed to spontaneous intracranial hypotension*. Therefore, in all patients presenting with headache and CM1, abnormal CSF pressure 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).

Prevalence studies show tonsillar herniation of at least 5 mm in 0.24–3.6% of the population, with prevalence decreasing in older age.

*The clinical context of CM1 is important as many of these patients can be asymptomatic.* There are conflicting data regarding the degree of herniation and the severity of associated headache and level of disability: patients can exhibit ‘Chiari-like’ symptoms with minimal cerebellar tonsillar herniation, while others may be asymptomatic with large herniations.

These criteria for 7.7 *Headache attributed to Chiari malformation type I (CM1)* require validation: prospective studies with long-term non-surgical and surgical outcomes are needed. Meanwhile, rigid adherence to both clinical and radiological criteria is recommended in considering surgical intervention to avoid an unnecessary procedure with significant potential for surgical morbidity. Current data suggest that, in carefully selected patients, cough headaches more than headaches without Valsalva-like precipitants, and occipital headaches more than non-occipital, are responsive to surgical intervention.

Emerging data suggest a relationship between obesity and likelihood of headache in CM1; this finding warrants further research, particularly from a treatment viewpoint.

### 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 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 Cocaine-induced headache
  - 8.1.6 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
  - 8.1.8 Headache attributed to exogenous acute pressor agent
  - 8.1.9 Headache attributed to occasional use of non-headache medication
  - 8.1.10 Headache attributed to long-term use of non-headache medication
  - 8.1.11 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 Non-opioid analgesic-overuse headache
    - 8.2.3.1 Paracetamol (acetaminophen)-overuse headache
    - 8.2.3.2 Non-steroidal anti-inflammatory drug (NSAID)-overuse headache
      - 8.2.3.2.1 Acetylsalicylic acid-overuse headache
    - 8.2.3.3 Other non-opioid analgesic-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 unspecified or 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?* The general rules for attribution to another disorder apply with some adaptation to 8. *Headache attributed to a substance or its withdrawal.*

1. When a *new 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.
2. When a *pre-existing headache* with the characteristics of a primary headache disorder becomes *chronic*, or is made *significantly worse* (usually meaning a twofold 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 types or subtypes) should be given, provided that there is good evidence that exposure to or withdrawal from that substance can cause headache.
3. Certain subforms of headache attributed to exposure to a pharmacologically active substance occur some hours after the exposure and only in a patient with a primary headache disorder, phenomenologically resembling the primary headache type. They are presumed to be mechanistically distinct, responding to a non-physiological stimulus, and therefore are regarded as secondary. Diagnoses should be given for both the primary headache disorder and the appropriate subform of 8.1 *Headache attributed to use of or exposure to a substance.*

### 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.

Associations between headache and substances are often anecdotal, many based on reports of adverse drug reactions. The fact of association with headache does not prove causation, or eliminate the need to consider other aetiologies. Because common events happen commonly, an association between headache and an

exposure to a substance may be mere coincidence. Headache can occur by chance. Headache may be a symptom of a systemic disease, and drugs given to treat such a condition will be associated with headache. In trials of drugs for acute migraine, in particular, headache as well as associated symptoms are listed as adverse drug reactions despite being symptoms of the treated disorder rather than an outcome 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, exposure to or withdrawal from a substance known to be able to cause headache has occurred
- C. Evidence of causation demonstrated by at least two of the following:
  1. headache has developed in temporal relation to use of, exposure to or withdrawal from the substance
  2. either of the following:
    - a) headache has significantly improved or resolved in close temporal relation to cessation of use of or exposure to the substance
    - b) headache has significantly improved or resolved within a defined period after withdrawal from the substance
  3. headache has characteristics typical for use of, exposure to or withdrawal from 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 an unwanted effect of a substance in normal therapeutic use or in experimental studies, or caused by a toxic substance.

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 may 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 intake 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. 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. It resolves spontaneously.

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

GTN induces immediate headache in most normal people, but can also cause a delayed headache in people with 1. *Migraine* 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, five to six hours after exposure. People with 3.1 *Cluster headache* develop delayed headache only during cluster

periods: GTN usually induces a cluster headache attack one to two 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 this 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 one hour after absorption of the NO donor
  2. headache has resolved within one 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.<sup>1</sup>

*Note:*

1. Phenomenologically, 8.1.1.2 *Delayed NO donor-induced headache* resembles the patient's primary headache type, but is regarded as secondary, attributed to the drug. The patient should be coded both for the primary headache and 8.1.1.2 *Delayed NO donor-induced headache*.

*Comment:* While 8.1.1.2 *Delayed NO donor-induced headache* occurs only in a person affected by a primary headache disorder and phenomenologically resembles that headache type, it is presumed that it is mechanistically distinct.

#### 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 phosphodiesterase (PDE) inhibitor has been taken
- C. Evidence of causation demonstrated by all of the following:
  1. headache has developed within five 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 is usually tension-type-like, but in people with 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). It resolves spontaneously.

**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 three 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:** 8.1.4.1 *Immediate alcohol-induced headache* is much rarer than 8.1.4.2 *Delayed alcohol-induced headache*. The effective dose of alcohol to cause the former is variable: in people with 1. Migraine it can sometimes be very small while at other times they may tolerate alcohol at the same level as non-migraineurs.

**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 secondary headaches. 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 Cocaine-induced headache**

**Description:** Headache developing within one hour of, and caused by, administration of cocaine by any route. It resolves spontaneously within 72 hours.

**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 one 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).

#### 8.1.6 Histamine-induced headache

**Description:** Headache caused immediately, or after a delay, by acute exposure to histamine. It resolves spontaneously.

**Comments:** Histamine has a 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 people with 1. *Migraine*, 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, five to six hours after exposure. People with 3.1 *Cluster headache* develop delayed headache with the characteristics of that disorder only during cluster periods, usually one to two hours after exposure.

##### 8.1.6.1 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 one hour of histamine absorption
  2. headache has resolved within one 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.

##### 8.1.6.2 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.<sup>1</sup>

**Note:**

1. Phenomenologically, 8.1.6.2 *Delayed histamine-induced headache* resembles the patient's primary headache type, but is regarded as secondary, attributed to the drug. The patient should be coded both for the primary headache and 8.1.6.2 *Delayed histamine-induced headache*.

**Comment:** While 8.1.6.2 *Delayed histamine-induced headache* occurs only in a person affected by a primary headache disorder and phenomenologically resembles that headache type, it is presumed that it is mechanistically distinct.

##### 8.1.7 Calcitonin gene-related peptide (CGRP)-induced headache

**Description:** Headache caused immediately, or after a delay, by acute exposure to calcitonin gene-related peptide (CGRP). It resolves spontaneously.

**Comments:** Calcitonin gene-related peptide (CGRP), administered by infusion, causes an immediate headache. It can also cause a delayed headache in people with 1. *Migraine*, on average five to six hours after exposure, which fulfils the diagnostic criteria for 1.1 *Migraine without aura*.

An increasing number of CGRP-receptor antagonists have been found effective in the acute treatment of migraine.

##### 8.1.7.1 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 one hour of CGRP absorption
  2. headache has resolved within one 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.

#### 8.1.7.2 Delayed CGRP-induced headache

##### *Diagnostic criteria:*

- A. Headache, in a person affected by 1. *Migraine*, and with the characteristics of this headache, 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.<sup>1</sup>

##### *Note:*

1. Phenomenologically, 8.1.7.2 *Delayed CGRP-induced headache* resembles migraine, but is regarded as secondary, attributed to the drug. The patient should be coded both for the appropriate type or subtype of 1. *Migraine* and 8.1.7.2 *Delayed CGRP-induced headache*.

*Comment:* While 8.1.7.2 *Delayed CGRP-induced headache* occurs only in a person affected by 1. *Migraine* and phenomenologically resembles this headache type, it is presumed that it is mechanistically distinct.

#### 8.1.8 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 one 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.

#### 8.1.9 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:* 8.1.9 *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, sildenafil.

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.

#### 8.1.10 Headache attributed to long-term use of non-headache medication

*Coded elsewhere:* 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.

Headache occurring during the pill-free interval of combined oral contraception is coded as 8.3.3 *Oestrogen-withdrawal headache*.

**Description:** Headache developing as an adverse event during long-term use of a medication for purposes other than the treatment of headache. It is not necessarily reversible.

**Diagnostic criteria:**

- A. Headache present on  $\geq 15$  days/month and fulfilling criterion C
- B. Long-term use of a medication has occurred 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 long-term use or exposure that may result in headache varies from medication to medication. Similarly, the time required for resolution varies – if the effect is reversible.

Exogenous hormones, taken usually for contraception or as hormone replacement therapy, are non-headache medication; therefore, 8.1.10 *Headache attributed to long-term use of non-headache medication* now subsumes headache developing as an adverse event during hormone therapy (previously coded as 8.1.12 *Headache attributed to exogenous hormone*). Regular use of exogenous hormones can be associated with an increase in frequency or new development of migraine-like 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 8.1.10 *Headache attributed to long-term use of non-headache*

*medication*. When a pre-existing headache with the characteristics of a primary headache disorder becomes *chronic*, or is made *significantly worse* (usually meaning a twofold 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 8.1.10 *Headache attributed to long-term use of non-headache medication* should be given. However, headache occurring only during the pill-free interval of combined oral contraception is coded as 8.3.3 *Oestrogen-withdrawal headache*.

Otherwise, 8.1.10 *Headache attributed to long-term use of non-headache medication* can be due to a 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.

**8.1.11 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 has occurred 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:** 8.1.11 *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. It 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, chlordecone, EDTA, heptachlor, hydrogen sulfide, kerosene, long-chain alcohols, methyl alcohol, methyl bromide, methyl chloride, methyl iodine, naphthalene, organophosphorous compounds (parathion, pyrethrum).

The characteristics of 8.1.11 *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:* Drug-induced headache; medication-misuse headache; rebound headache.

*Coded elsewhere:* Patients with a pre-existing primary headache who, in association with medication overuse, develop a new type of headache or a significant 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/month in a patient with a pre-existing primary headache and developing as a consequence of regular overuse of acute or symptomatic headache medication (on 10 or more or 15 or more days/month, depending on the medication) for more than three months. It usually, but not invariably, resolves after the overuse is stopped.

*Diagnostic criteria:*

- A. Headache occurring on  $\geq 15$  days/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 headache<sup>1-3</sup>
- C. Not better accounted for by another ICHD-3 diagnosis.

*Notes:*

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 *Non-opioid 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.
2. 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*.
3. 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 unspecified or 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 disorders such as 3.1.2 *Chronic cluster headache* or 4.10 *New daily persistent headache*.

The diagnosis of 8.2 *Medication-overuse headache* is extremely important clinically. Epidemiological evidence from many countries indicates that more than half of people with headache on 15 or more days/month have 8.2 *Medication-overuse headache*. Clinical 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 and can be provided with success in primary care. 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.

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.

In the criteria 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.

It is recognized that cross-sectional population-based studies estimating the prevalence of 8.2 *Medication-overuse headache* can record the coexistence in participants of headache on  $\geq 15$  days/month and overuse of drugs for acute and/or symptomatic treatment of headache but are rarely able to collect information on earlier headache, on duration of the current headache or medication overuse, and/or that might support a presumption of causation. In consequence, either or both of criteria A and B may not be entirely fulfilled. Provided that criteria are not fulfilled for another ICHD-3 diagnosis, such cases should be reported as *probable medication-overuse headache* (pMOH), although ICHD-3 does not provide a coding for this.

#### 8.2.1 Ergotamine-overuse headache

**Description:** Headache occurring on 15 or more days/month in a patient with a pre-existing primary headache and developing as a consequence of regular use of ergotamine on 10 or more days/month for more than three months. It usually, but not invariably, resolves after the overuse is stopped.

##### Diagnostic criteria:

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

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

#### 8.2.2 Triptan-overuse headache

**Description:** Headache occurring on 15 or more days/month in a patient with a pre-existing primary headache and developing as a consequence of regular use of one or more triptans on 10 or more days/month for more than three months. It usually, but not invariably, resolves after the overuse is stopped.

##### Diagnostic criteria:

- A. Headache fulfilling criteria for 8.2 *Medication-overuse headache*
- B. Regular intake of one or more triptans,<sup>1</sup> in any formulation, on  $\geq 10$  days/month for  $>3$  months.

##### Note:

1. The triptan(s) will usually be specified in parenthesis.

**Comment:** Triptan overuse by people with 1.1 *Migraine without aura* or 1.2 *Migraine with aura* may increase attack frequency to that of 1.3 *Chronic migraine*. Evidence suggests that this occurs sooner with triptan overuse than with ergotamine overuse.

#### 8.2.3 Non-opioid analgesic-overuse headache

**Description:** Headache occurring on 15 or more days/month in a patient with a pre-existing primary headache and developing as a consequence of regular use of one or more non-opioid analgesics on 15 or more days/month for more than three months. It usually, but not invariably, resolves after the overuse is stopped.

**Comments:** A patient who fulfils criteria for more than one of the subforms of 8.2.3 *Non-opioid analgesic-overuse headache* should be given all applicable codes.

Many patients use more than one non-opioid analgesic: a common example is paracetamol (acetaminophen) and a non-steroidal anti-inflammatory drug (NSAID). For the purposes of ICHD-3, all non-opioid analgesics are regarded as a single class; therefore, a patient who uses more than one non-opioid analgesic cumulatively, but not any single drug, on 15 or more days/month is coded 8.2.3 *Non-opioid analgesic-overuse headache* (with the individual drugs specified in parenthesis) and not 8.2.6 *Medication-overuse headache attributed to multiple drug classes not individually overused*.

##### 8.2.3.1 Paracetamol (acetaminophen)-overuse headache

##### Diagnostic criteria:

- A. Headache fulfilling criteria for 8.2 *Medication-overuse headache*
- B. Regular intake of paracetamol on  $\geq 15$  days/month for  $>3$  months.

### 8.2.3.2 Non-steroidal anti-inflammatory drug (NSAID)-overuse headache

*Coded elsewhere:* Acetylsalicylic acid is a non-steroidal anti-inflammatory drug (NSAID) but has other, unique activity. Consequently, 8.2.3.2.1 *Acetylsalicylic acid-overuse headache* is coded as a separate subform.

#### Diagnostic criteria:

- A. Headache fulfilling criteria for 8.2 *Medication-overuse headache*
- B. Regular intake of one or more non-steroidal anti-inflammatory drugs (NSAIDs)<sup>1</sup> (other than acetylsalicylic acid) on  $\geq 15$  days/month for  $>3$  months.

#### Note:

1. The NSAID(s) should be specified in parenthesis.

### 8.2.3.2.1 Acetylsalicylic acid-overuse headache

#### Diagnostic criteria:

- A. Headache fulfilling criteria for 8.2 *Medication-overuse headache*
- B. Regular intake of acetylsalicylic acid on  $\geq 15$  days/month for  $>3$  months.

*Comment:* Acetylsalicylic acid is a non-steroidal anti-inflammatory drug (NSAID) but has other, unique activity. Consequently, 8.2.3.2.1 *Acetylsalicylic acid-overuse headache* is coded as a separate subform.

### 8.2.3.3 Other non-opioid analgesic-overuse headache

#### Diagnostic criteria:

- A. Headache fulfilling criteria for 8.2 *Medication-overuse headache*
- B. Regular intake of a non-opioid analgesic other than paracetamol or non-steroidal anti-inflammatory drugs (including acetylsalicylic acid) on  $\geq 15$  days/month for  $>3$  months.

### 8.2.4 Opioid-overuse headache

#### Diagnostic criteria:

- A. Headache fulfilling criteria for 8.2 *Medication-overuse headache*
- B. Regular intake of one or more opioids<sup>1</sup> on  $\geq 10$  days/month for  $>3$  months.

#### Note:

1. The opioid(s) should be specified in parenthesis.

*Comment:* Prospective studies indicate that patients overusing opioids have the highest relapse rate after withdrawal treatment.

### 8.2.5 Combination-analgesic-overuse<sup>1</sup> headache

#### Diagnostic criteria:

- A. Headache fulfilling criteria for 8.2 *Medication-overuse headache*
- B. Regular intake of one or more combination-analgesic medications<sup>1,2</sup> on  $\geq 10$  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 (e.g. paracetamol and codeine) or acting as adjuvants (e.g. caffeine). Drugs that combine only two non-opioid analgesics (such as acetylsalicylic acid and paracetamol), without an adjuvant, are not considered combination-analgesics since, for the purposes of ICHD-3, both drugs are in the same class.
2. The combination-analgesic(s) should 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 8.2 *Medication-overuse headache*. For this reason, 8.2.5 *Combination-analgesic-overuse headache* has a separate coding.

The most commonly overused combination-analgesics combine non-opioid 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, non-opioid analgesics and/or opioids<sup>1</sup> on a total of  $\geq 10$  days/month for  $>3$  months without overuse of any single drug or drug class alone.<sup>2</sup>

Notes:

1. The drugs or drug classes should be specified in parenthesis.
2. *Without overuse of any single drug or drug class alone* means that criterion B has not been fulfilled for any of the specific subtypes 8.2.1–8.2.5.

**8.2.7 Medication-overuse headache attributed to unspecified or 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, non-opioid analgesics and/or opioids on  $\geq 10$  days/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. While 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  $\geq 10$  days/month for  $>3$  months, of one or more medications other than those described above,<sup>1</sup> taken for acute or symptomatic treatment of headache.

Note:

1. The medication(s) should be specified in parenthesis.

**8.3 Headache attributed to substance withdrawal**

*Description:* Headache following and caused by interruption in use of or exposure to a medication or other substance that has lasted for weeks or months.

**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 two weeks, which has been interrupted. It resolves spontaneously within seven 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 one hour by intake of caffeine 100 mg
    - b) headache has resolved within seven 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 three months, which has been interrupted. It resolves spontaneously within seven 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 seven 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 five days after daily consumption of exogenous oestrogen for three 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 three days in the absence of further consumption.

**Diagnostic criteria:**

- A. Headache or migraine fulfilling criterion C
- B. Daily use of exogenous oestrogen for  $\geq 3$  weeks, which has been interrupted
- C. Evidence of causation demonstrated by both of the following:
  1. headache or migraine has developed within five days after the last use of oestrogen
  2. headache or migraine has resolved within three days of its onset
- D. Not better accounted for by another ICHD-3 diagnosis.

**Comment:** Oestrogen-withdrawal following cessation of a course of exogenous oestrogens (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, interruption in 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 three months after total withdrawal from use of the substance
- D. Not better accounted for by another ICHD-3 diagnosis.

**Comments:** 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).

There may be other substances not yet recognized.

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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 localized brain infection
- 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 types or 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 comments

The triad of headache, fever and nausea/vomiting is highly suggestive of 9. *Headache attributed to infection.* The probability is increased when lethargy or convulsions are also part of the clinical picture.

*Primary or secondary headache or both?* The general rules for attribution to another disorder apply to 9. *Headache attributed to infection.*

1. When a *new 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.
2. When a *pre-existing headache* with the characteristics of a primary headache disorder becomes *chronic*, or is made *significantly worse* (usually meaning a twofold 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 types or subtypes) should be given, provided that there is good evidence that the disorder can cause headache.

*Acute, chronic or persistent?* 9. *Headache attributed to infection* is usually the consequence of active infection, resolving within three 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; after three months, both the infection and the headache are referred to as *chronic*.

In other, rarer cases, the infection resolves or is eradicated but the headache does not remit; after three 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 that is diffuse

and associated with focal neurological signs and/or altered mental state and 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; when evidence is lacking, the diagnostic criteria for some of the subtypes of 9.1 *Headache attributed to intracranial infection* are at least partly reliant upon expert consensus, including the views of experts in neuroinfection.

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 are included in a single entity of 9.1.1 *Headache attributed to bacterial meningitis or meningoencephalitis*.

A variety of bacteria may cause meningitis and/or encephalitis, including *Streptococcus pneumoniae*, *Neisseria meningitidis* and *Listeria monocytogenes*. The immunologic background is very important because immunosuppression (due to HIV or post-transplant or other chronic immunodepressant treatments) influences susceptibility and clinical and biological profiles.

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 three months *after* resolution of the causative infection. Three separate subforms of 9.1.1 *Headache attributed to bacterial meningitis or meningoencephalitis* are therefore described below 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 B below
- B. 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<sup>1</sup> or has resolved within the last three months
- C. Headache has been present for >3 months.

*Note:*

- 1. Demonstrated by MRI focal or multifocal contrast enhancement and/or persistence of cerebrospinal fluid (CSF) pleocytosis with or without evidence of blood–brain barrier damage.

**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, neck stiffness, light sensitivity and nausea and/or vomiting.

While enteroviruses account for most cases of 9.1.2 *Headache attributed to viral meningitis or encephalitis*, a variety of other viral agents may also be responsible: arbovirus, poliovirus, echovirus, coxsackievirus, herpes simplex, varicella zoster, adenovirus, mumps and others. Cerebrospinal fluid (CSF) polymerase chain reaction (PCR) gives the specific diagnosis in the majority of cases. A positive CSF PCR for herpes simplex virus (HSV) types 1 or 2 and serology for HSV-1 and -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 one week after the onset of the symptoms, causing false negatives. When PCR performed after one week is negative, the diagnosis can be made on the basis of an altered CSF/blood antibody ratio.

As with intracranial bacterial infection, it may be difficult in a viral infection 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 exclusively.

#### 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 or multifocal brain oedema<sup>1</sup>
  2. at least one of the following:
    - a) altered mental state
    - b) focal neurological deficits
    - c) seizures.

##### Note:

1. There may also be associated leptomeningeal enhancement.

**Comments:** Pain is usually diffuse, with the focus in frontal and/or retro-orbital areas, severe or extremely severe and throbbing or pressing in nature.

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. 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; 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,<sup>1</sup> 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.<sup>2</sup>

##### Notes:

1. The clinical symptoms tend to evolve over weeks, in parallel with the level of immunosuppression.
2. Early diagnosis is best made by CT or MRI.

**Comments:** 9.1.3 *Headache attributed to intracranial fungal or other parasitic infection* should be suspected

whenever headache in a person who is immunocompromised is associated with fever, a 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.

Fungi that may cause meningitis and/or encephalitis include *Candida*, *Aspergillus* and *Cryptococcus neoformans*; parasites include toxoplasma. Besides cerebrospinal fluid (CSF) culture and CSF PCR investigations, other tests on CSF and blood include direct (cytological detection, microscopic visualization, culture and identification of fungal elements in the biological materials under observation) and indirect detection of the pathogen (identification of an antigen or another element of the capsule). In the case of aspergillosis, the galactomannan 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.

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 at risk:

- 1) people with significant neutropaenia (<500 neutrophils/mm<sup>3</sup>)
- 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 three 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* occurs but is not well documented in the literature; 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 B below
- B. 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 B below
- B. Headache has been present for >3 months.

#### 9.1.4 Headache attributed to localized brain infection

**Description:** Headache caused by brain abscess, subdural empyema, infectious granuloma or other localized infective lesion, 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 localized brain infection has been demonstrated by neuroimaging and/or specimen analysis
- C. Evidence of causation demonstrated by at least two of the following:
  1. headache has developed in temporal relation to development of the localized brain infection, or led to its discovery
  2. headache has significantly worsened in parallel with deterioration of the localized brain infection shown by either of the following:
    - a) worsening of other symptoms and/or clinical signs arising from the localized brain infection
    - b) evidence of enlargement (or rupture, in the case of brain abscess) of the localized brain infection
  3. headache has significantly improved in parallel with improvement in the localized brain infection
  4. headache has at least one of the following four 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 fever, nausea and/or vomiting
    - d) unilateral, and ipsilateral to the localized brain infection
- D. Not better accounted for by another ICHD-3 diagnosis.

**Comments:** Brain abscesses are usually caused by anaerobic or, sometimes, mixed bacteria, often including anaerobic streptococci or bacteroides. Staphylococci are common after cranial trauma, neurosurgery, or endocarditis. Enterobacteria are common in chronic ear infections. Fungi (e.g. *Aspergillus*) and protozoa (e.g. *Toxoplasma gondii*, particularly in HIV-infected patients) can cause abscesses.

Subdural empyema is often secondary to sinusitis or otitis media. It may also be a complication of meningitis.

Brain granulomas have been associated with cystercosis, sarcoidosis, toxoplasmosis and aspergillosis.

The mechanisms causing 9.1.4 *Headache attributed to localized brain infection* include direct compression, irritation of the meningeal and/or arterial structures, increased intracranial pressure and fever. Headache attributed to subdural empyema is particularly associated with fever and symptoms and/or clinical signs of meningeal irritation and increased intracranial pressure.

## 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 or subform 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. 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 B below
- B. 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 B below
- B. 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. 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 B below
- B. 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 three 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. 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.

**Comments:** 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 B below
- B. 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 B below
- B. Headache has been present for >3 months.

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### 9.2 Headache attributed to systemic infection

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## 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 phaeochromocytoma
  - 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 homeostasis

### 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?* The general rules for attribution to another disorder apply to 10. *Headache attributed to disorder of homeostasis.*

1. When a *new headache* occurs for the first time in close temporal relation to a disorder of homeostasis, 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.
2. When a *pre-existing headache* with the characteristics of a primary headache disorder becomes *chronic*, or is made *significantly worse* (usually meaning a twofold or greater increase in frequency and/or severity), in close temporal relation to a disorder of homeostasis, both the initial headache diagnosis and a diagnosis of 10. *Headache attributed to disorder of homeostasis* (or one of its types or subtypes) should be given, provided that there is good evidence that that disorder can cause headache.

### Introduction

The mechanisms behind causation of the different types of 10. *Headache attributed to disorder of homeostasis* 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 homeostasis 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 homeostasis
  2. either or both of the following:
    - a) headache has significantly worsened in parallel with worsening of the disorder of homeostasis
    - b) headache has significantly improved after resolution of the disorder of homeostasis
  3. headache has characteristics typical for the disorder of homeostasis
- 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 either 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 either or both 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 metres has occurred
- 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 metres
  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, restrictions in venous outflow and low fluid intake (<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 two days of acclimatization prior to engaging in strenuous exercise at high altitudes, liberal fluid intake and avoidance of alcohol.

Dwelling at altitudes above 1000 metres increases not only prevalence but also the severity of the symptoms of 1. *Migraine*. The mechanisms are unknown, and probably unrelated to those of 10.1.1 *High-altitude headache*.

### 10.1.2 Headache attributed to aeroplane travel

**Description:** Headache, often severe, usually unilateral and periocular 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 during the aeroplane flight
  2. either or both of the following:
    - a) headache has worsened in temporal relation to ascent following 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<sup>1</sup>
    - b) orbitofrontal location<sup>2</sup>
    - c) jabbing or stabbing quality<sup>3</sup>
- D. Not better accounted for by another ICHD-3 diagnosis.<sup>4</sup>

**Notes:**

1. Side-shift between different flights occurs in around 10% of cases.
2. Parietal spread may occur.
3. Pulsation (throbbing) may also be noted.
4. In particular, sinus disorder should be excluded.

**Comments:** A recent Scandinavian survey has indicated that up to 8.3% of air-travellers experience 10.1.2 *Headache attributed to aeroplane travel*. It occurs during landing in more than 90% of cases.

Accompanying symptoms are reported in up to 30% of cases. Most frequent are restlessness and unilateral tearing; other localized parasympathetic symptoms, nausea or photo/phonophobia have been described in fewer than 5% of cases.

A proportion of subjects experiencing 10.1.2 *Headache attributed to aeroplane travel* report similar

headache during free snorkelling and/or rapid descent from mountains, suggesting these headaches are due to imbalance between intracranial and external air pressures.

### 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.

Diving has been known to cause cervical carotid or vertebral artery dissection. Headache occurring as a result should be coded to 6.5.1.1. *Acute headache or facial or neck pain attributed to cervical carotid or vertebral artery dissection*.

**Description:** Headache caused by diving to a depth greater than 10 metres, occurring during the dive but often intensified upon resurfacing, in the absence of decompression illness. It is usually accompanied by symptoms of carbon dioxide (CO<sub>2</sub>) intoxication. It remits quickly with oxygen or, if this is not given, spontaneously within three 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 >10 metres
  2. 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 three days of completion of the dive
      - ii. headache has remitted within one hour after treatment with 100% oxygen
  3. at least one of the following symptoms of CO<sub>2</sub> 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.

**Comments:** There is evidence that hypercapnia in the absence of hypoxia is associated with headache. Hypercapnia (arterial pCO<sub>2</sub> >50 mmHg) is known to cause relaxation of cerebrovascular smooth muscle, leading to intracranial vasodilatation and increased intracranial pressure. 10.1.3 *Diving headache* is the best clinical example of headache attributed to hypercapnia. Carbon dioxide (CO<sub>2</sub>) 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 CO<sub>2</sub> production more than 10-fold, resulting in a transient elevation of pCO<sub>2</sub> to >60 mmHg.

10.1.3 *Diving headache* usually intensifies during the decompression phase of the dive or upon resurfacing.

### 10.1.4 Sleep apnoea headache

**Description:** Morning headache, usually bilateral and with a duration of less than four 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 with apnoea-hypopnoea index  $\geq 5$  has been diagnosed<sup>1</sup>
- 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) recurring on  $\geq 15$  days/month
    - b) all of the following:
      - i. bilateral location
      - ii. pressing quality

- iii. not accompanied by nausea, photophobia or phonophobia
- c) resolving within four hours
- D. Not better accounted for by another ICHD-3 diagnosis.<sup>2</sup>

**Notes:**

1. The apnoea-hypopnoea index is calculated by dividing the number of apnoeic events by the number of hours of sleep (5–15/hour = mild; 15–30/hour = moderate; >30/hour = severe).
2. A definitive diagnosis requires overnight polysomnography.

*Comments:* 10.1.4 *Sleep apnoea headache* seems to be less frequent and of longer duration than previously assumed. Although morning headache is significantly more common in patients with sleep apnoea than in the general population, headache present upon awakening is a non-specific symptom 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.<sup>1</sup>

**Note:**

1. Caffeine is rapidly removed by dialysis: 8.3.1 *Caffeine-withdrawal headache* should be considered in patients who consume large quantities of caffeine.

*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.

Variations in urea, sodium and magnesium levels, and in blood pressure and body weight, may be risk factors for developing 10.2 *Dialysis headache*.

### 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  $\geq 180$  mmHg) and/or diastolic (to  $\geq 120$  mmHg) blood pressure. It remits after normalization of blood pressure.

**Diagnostic criteria:**

- A. Any headache fulfilling criterion C
- B. Hypertension, with systolic pressure  $\geq 180$  mmHg and/or diastolic pressure  $\geq 120$  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 do not appear to *cause* headache. Whether moderate hypertension *predisposes* to headache 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 phaeochromocytoma

**Coded elsewhere:** When hypertensive encephalopathy is present, headache is coded as 10.3.3 *Headache attributed to hypertensive encephalopathy*. When the diagnosis of phaeochromocytoma 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 one hour) and accompanied by sweating, palpitations, pallor and/or anxiety, caused by phaeochromocytoma.

#### Diagnostic criteria:

- A. Recurrent discrete short-lasting headache episodes fulfilling criterion C
- B. Phaeochromocytoma 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 phaeochromocytoma, 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.<sup>1</sup>

#### Note:

1. The diagnosis of phaeochromocytoma is established by 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.

**Comments:** 10.3.1 *Headache attributed to phaeochromocytoma* occurs as a paroxysmal headache in 51–80% of patients with phaeochromocytoma.

10.3.1 *Headache attributed to phaeochromocytoma* is often severe, frontal or occipital and usually described as either pulsating or constant in quality. An important feature is its short duration: less than 15 minutes in 50% and less than one 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.

### 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  $\geq 180$  mmHg and/or diastolic  $\geq 120$  mmHg). It remits after normalization of blood pressure.

#### Diagnostic criteria:

- A. Headache fulfilling criterion C
- B. Both of the following:
  1. a hypertensive crisis<sup>1</sup> is occurring
  2. 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.
  - b) pulsating quality
  - c) aggravated by physical activity

**Note:**

1. A hypertensive crisis is defined as a paroxysmal rise in systolic (to  $\geq 180$  mmHg) and/or diastolic (to  $\geq 120$  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

**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 four 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 multisystem disorders with various forms. Their diagnosis requires hypertension (>140/90 mmHg) documented on two blood pressure readings at least four hours apart, or a rise in diastolic pressure of  $\geq 15$  mmHg or in systolic pressure of  $\geq 30$  mmHg, coupled with urinary protein excretion >0.3 g/24 hours. In addition, tissue oedema, thrombocytopaenia 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  $\geq 30$  mmHg and/or diastolic pressure of  $\geq 20$  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 four 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 sometimes somatic (pressure ulcers, ingrown toenail, burns, trauma or surgical or invasive diagnostic procedures).

#### 10.4 Headache attributed to hypothyroidism

**Coded elsewhere:** In the presence of hypothyroidism, headache can also be a manifestation of pituitary adenoma, coded as 7.4.3 *Headache attributed to hypothalamic or pituitary hyper- or hyposecretion*.

**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 the 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 either or both of the following characteristics:



- a) bilateral location
- b) 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.

While 10.4 *Headache attributed to hypothyroidism* is not understood to be associated with nausea or vomiting, a recent study found that patients with hypothyroidism may present with unilateral, episodic, pulsating headache accompanied by nausea and/or vomiting. Half of the patients studied had a history of 1. *Migraine*, so the significance of these results is unclear and they require confirmation in future studies.

### 10.5 Headache attributed to fasting

**Coded elsewhere:** An episode of migraine triggered by fasting is coded as 1. *Migraine* or one of its types.

**Description:** Diffuse non-pulsating headache, usually mild to moderate, occurring during and caused by fasting for at least eight hours. It is relieved after eating.

**Diagnostic criteria:**

- A. Diffuse headache not fulfilling the criteria for 1. *Migraine* or any of its types but fulfilling criterion C below
- B. The patient has fasted for  $\geq 8$  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.<sup>1</sup>

**Note:**

- 1. 10.5 *Headache attributed to fasting* is typically diffuse, non-pulsating and mild to moderate in intensity. In patients with a prior history of 1. *Migraine*, headache during fasting may resemble 1.1 *Migraine without aura*, and should be coded accordingly (fasting being a precipitating factor) when the criteria for this disorder are met.

**Comments:** 10.5 *Headache attributed to fasting* is significantly more common in people who have a prior history of a primary headache disorder.

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 cephalgia

**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 the 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 cephalgia* 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 since 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 at least one of the following:
  1. headache has developed in temporal relation to the onset of the disorder of homoeostasis
  2. headache has significantly worsened in parallel with worsening of the disorder of homoeostasis
  3. 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 angle-closure glaucoma
  - 11.3.2 Headache attributed to refractive error
  - 11.3.3 Headache attributed to ocular inflammatory disorder
  - 11.3.4 Trochlear headache
- 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
- 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 lesions of the cranial nerves and other facial pain*.

### General comment

**Primary or secondary headache or both?** The general rules for attribution to another disorder apply to 11. *Headache or facial pain attributed to disorder of the cranium, neck, eyes, ears, nose, sinuses, teeth, mouth or other facial or cervical structure*.

1. When a *new headache* occurs for the first time in close temporal relation to a cranial, cervical, facial, 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.
2. When a *pre-existing headache* with the characteristics of a primary headache disorder becomes *chronic*, or is made *significantly worse* (usually meaning a twofold or greater increase in frequency and/or severity), in close temporal relation to a cranial, cervical, facial, 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 types or 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 been regarded as common causes of headache, since 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 conclusively the explanation of associated 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, since these manifestations are not unique. The purpose of the criteria in this chapter is not to describe headaches in all their possible subtypes and 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 criteria have not been investigated. Instead, the aim 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

**Coded elsewhere:** Headache caused by trauma to the cranium is classified under 5. *Headache attributed to trauma or injury to the head and/or neck* or one of its types.

**Description:** Headache caused by a non-traumatic 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 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 types.

**Description:** Headache caused by a non-traumatic 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, when 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*. A11.2.5 *Headache attributed to cervical myofascial pain* is an Appendix diagnosis awaiting 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 which 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 and/or imaging evidence<sup>1</sup> of a disorder or lesion within the cervical spine or soft tissues of the neck, known to be able to cause headache<sup>2</sup>
- 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.<sup>3-5</sup>

**Notes:**

1. Imaging findings in the upper cervical spine are common in patients without headache; they are suggestive but not firm evidence of causation.
2. Tumours, fractures, infections and rheumatoid arthritis of the upper cervical spine have not been formally validated as causes of headache, but are accepted to fulfil criterion B in individual cases. Cervical spondylosis and osteochondritis may or may not be valid causes fulfilling criterion B, again depending on the individual case.
3. 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 in the Appendix.
4. 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 in the Appendix as A11.2.4 *Headache attributed to upper cervical radiculopathy*.
5. 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, while 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*.

**11.2.2 Headache attributed to retropharyngeal tendonitis**

**Description:** Headache caused by inflammation or calcification in the retropharyngeal soft tissues, 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, or led to its discovery
  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<sup>1</sup>
  4. there is tenderness over the spinous processes of the upper three cervical vertebrae<sup>2</sup>
- D. Not better accounted for by another ICHD-3 diagnosis.<sup>3</sup>

**Notes:**

1. Although retroflexion of the neck most consistently aggravates pain, the same usually occurs also with rotation of the head and swallowing.
2. Tissues over the transverse processes of the upper three vertebrae are usually tender to palpation.
3. Upper carotid artery 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.

**Comments:** Body temperature and erythrocyte sedimentation rate (ESR) are usually elevated in retropharyngeal tendonitis.

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.

### 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 and/or head due to 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 and/or head due to muscular hyperactivity
- C. Evidence of causation demonstrated by at least two of the following:
  1. pain has developed in temporal relation to the onset of craniocervical dystonia
  2. pain has significantly worsened in parallel with progression of the craniocervical dystonia
  3. pain has significantly improved or resolved in parallel with improvement in or resolution of the craniocervical dystonia
  4. pain 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 angle-closure glaucoma

**Description:** Headache, usually unilateral, caused by acute angle-closure glaucoma and associated with other symptoms and clinical signs of this disorder.

**Diagnostic criteria:**

- A. Any headache fulfilling criterion C
- B. Acute angle-closure glaucoma has been diagnosed, with proof of increased intraocular pressure

- C. Evidence of causation demonstrated by at least two of the following:
  1. headache has developed in temporal relation to the onset of the glaucoma
  2. headache has significantly worsened in parallel with progression of the glaucoma
  3. headache has significantly improved or resolved in parallel with improvement in or resolution of the glaucoma
  4. pain location includes the affected eye
- D. Not better accounted for by another ICHD-3 diagnosis.

**Comments:** Acute angle-closure glaucoma generally causes eye and/or periorbital pain, visual acuity loss (blurring), conjunctival injection and oedema, 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.

**Comments:** Most patients with 11.3.2 *Headache attributed to refractive error* will seek advice from an ophthalmologist.

While 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 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 an ocular inflammatory disease known to be able to cause headache<sup>1</sup>
- 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 inflammatory disease
    - b) headache has significantly improved or resolved in parallel with improvement in or resolution of the ocular inflammatory disease
  - 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 ocular inflammatory disease, headache is localized and ipsilateral to it<sup>2</sup>
- D. Not better accounted for by another ICHD-3 diagnosis.

**Notes:**

- 1. Ocular inflammatory diseases known to cause headache include iritis, uveitis, cyclitis, scleritis, choroiditis, conjunctivitis and corneal inflammation.
- 2. 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, when the ocular inflammatory disease is unilateral, headache is likely to be localized and ipsilateral.

**Comment:** 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).

**11.3.4 Trochlear headache**

**Previously used terms:** Headache attributed to trochleitis.

A non-inflammatory disorder associated with trochlear dysfunction, termed *primary trochlear headache*, produces pain in the trochlear and temporoparietal regions that worsens with supraduction of the eye. It is diagnosed and treated similarly to trochleitis, and therefore included within 11.3.4 *Trochlear headache*.

**Description:** Headache, usually frontal and/or periorbital in location, with or without eye pain, caused by peritrochlear inflammation or dysfunction. It is often exacerbated by movements of the eye.

**Diagnostic criteria:**

- A. Periorbital and/or frontal headache fulfilling criterion C
- B. Clinical and/or imaging evidence of trochlear inflammation or dysfunction including tenderness upon palpation of the trochlea in the superomedial orbit
- C. Evidence of causation demonstrated by at least two of the following:
  - 1. unilateral ocular pain
  - 2. headache is exacerbated by movements of the eye<sup>1</sup>
  - 3. headache is significantly improved by injection of local anaesthetic or steroid agent into the peritrochlear region
  - 4. headache is localized and ipsilateral to the implicated trochlea
- D. Not better accounted for by another ICHD-3 diagnosis.

**Note:**

- 1. Particularly vertical movements.

**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. While not common, it is not rare, and must be considered when evaluating unilateral periorbital head pain.

Trochleitis can also trigger an episode of migraine in patients with 1. *Migraine*, which should be coded according to its type or subtype.

11.3.4 *Trochlear headache* can be provoked by reading.

#### 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 and 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

headache disorders and to 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 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 and ipsilateral to it
- D. Not better accounted for by another ICHD-3 diagnosis.<sup>1</sup>

##### **Note:**

1. 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.

**Comments:** Pain due to 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.

An episode of 1. *Migraine* may be triggered or exacerbated by nasal or sinus pathology.

**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 and other symptoms of the chronic rhinosinusitis
  3. headache is exacerbated by pressure applied over the paranasal sinuses
  4. in the case of a unilateral rhinosinusitis, headache is localized and ipsilateral to it
- D. Not better accounted for by another ICHD-3 diagnosis.

**Comment:** It has been questioned whether chronic sinus pathology can produce persistent headache. Recent studies seem to support such causation. However, pathological changes seen on imaging or endoscopy correlating with the patient's pain description are not on their own enough to secure the diagnosis of 11.5.2 *Headache attributed to chronic or recurring rhinosinusitis*.

**11.6 Headache attributed to disorder of the teeth**

**Description:** Headache caused by a disorder involving the teeth.

**Diagnostic criteria:**

- A. Any headache fulfilling criterion C

- B. Clinical and/or imaging evidence of a disorder or lesion of one or more teeth, 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 palpation, probing or pressure applied to the affected tooth or teeth
  4. in the case of a unilateral disorder or lesion, headache is localized and 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, but may refer pain to the head. The most common causes of 11.6 *Headache attributed to disorder of the teeth* are an endodontic or periodontal infection or abscess, or traumatic irritation such as pericoronitis around a partially erupted lower wisdom tooth.

**11.7 Headache attributed to temporomandibular disorder (TMD)**

**Coded elsewhere:** Jaw disease other than temporomandibular disorder, such as jaw malignancy, osteomyelitis or fracture, produces localized pain which can radiate to the face and head but rarely headache alone. When headache occurs in such cases, code as 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 caused by a disorder involving structures in the temporomandibular region.

**Diagnostic criteria:**

- A. Any headache<sup>1</sup> fulfilling criterion C
- B. Clinical evidence of a painful pathological process affecting elements of the temporomandibular joint(s), muscles of mastication and/or associated structures on one or both sides
- C. Evidence of causation demonstrated by at least two of the following:

1. the headache has developed in temporal relation to the onset of the temporomandibular disorder, or led to its discovery
  2. the headache is aggravated by jaw motion, jaw function (e.g. chewing) and/or jaw parafunction (e.g. bruxism)
  3. the headache is provoked on physical examination by temporalis muscle palpation and/or passive movement of the jaw
- D. Not better accounted for by another ICHD-3 diagnosis.<sup>2</sup>

**Notes:**

1. Usually temporally located, on one or both sides.
2. There is some overlap between 11.7 *Headache attributed to temporomandibular disorder (TMD)* arising from 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 types or subtypes (presumably with pericranial muscle tenderness).

**Comments:** 11.7 *Headache attributed to temporomandibular disorder (TMD)* is usually most prominent in the temporal region(s), preauricular area(s) of the face and/or masseter muscle(s). It may be unilateral, but is likely to be bilateral when the underlying pathology involves both temporomandibular regions. Pain referral to the face is common; after tooth pain, TMD is the most common cause of facial pain.

Pain generators include disc displacements, joint osteoarthritis, degenerative disease and/or hypermobility, and regional myofascial pain.

Diagnosis of TMD can be difficult, with some controversy regarding the relative importance of clinical and radiographic evidence. Use of the diagnostic criteria evolved by the International RDC/TMD Consortium Network and Orofacial Pain Special Interest Group is recommended.

### **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<sup>1</sup>
- 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 injection of local anaesthetic agent into 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.

**Note:**

1. 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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## 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 as types or subtypes of 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 comorbidity by chance alone is expected. Nevertheless, a causal relationship may exist between a new or significantly worsening headache and psychiatric disorder. The general rules for attribution to another disorder apply to 12. *Headache attributed to psychiatric disorder* with some adaptation.

1. When a *new headache* occurs for the first time in close temporal relation to a psychiatric disorder, and causation is confirmed, the headache 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.
2. When a *pre-existing headache* with the characteristics of a primary headache disorder is made *significantly worse* (usually meaning a twofold or greater increase in frequency and/or severity) in close temporal relation to a psychiatric disorder, and causation is confirmed, both the initial headache diagnosis and a diagnosis of 12. *Headache attributed to psychiatric disorder* (or one of its types) should be given, provided that there is good evidence that that disorder can cause headache.
3. When in either case 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. Therefore, 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 sufficiently low 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 as a single episode or recurrent, and 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 and adjustment disorders). In such cases, when there is no evidence of a causal relationship, both a headache diagnosis and a separate psychiatric diagnosis should be made.

Epidemiological data nonetheless show that headache and psychiatric disorders are comorbid more frequently than would be expected by chance. Common underlying factors may cause or predispose to both types of disorder; alternatively, or also, confounding factors may lead to comorbidity being overestimated (e.g. patients who have one diagnosis are more likely to be diagnosed with other conditions simply because they receive more medical scrutiny). Genuine causal associations also are possible, with the headache causing the psychiatric disorder, the psychiatric disorder causing the headache, or a reciprocal (bidirectional) influence between the headache and the psychiatric disorder.

In this context, 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, may be attributed to these disorders, uncertainties persist because of relative lack of evidence of causation. Criteria for headaches attributed to these and all but two other psychiatric disorders therefore remain in the Appendix. Further clarification of the mechanisms underlying these causal associations is necessary for sturdy conclusions.



Evidence suggests that a comorbid psychiatric disorder tends to worsen the course of 1. *Migraine* and of 2. *Tension-type headache*, increasing the frequency and severity of the headache and/or making it less responsive to treatment. Therefore, identification and treatment of any comorbid psychiatric condition is important for the proper management of these headaches. In children and adolescents, primary headache disorders (1. *Migraine*, 2.2 *Frequent episodic tension-type headache* and, especially, 2.3 *Chronic tension-type headache*) are often comorbid with psychiatric disorder. Sleep disorders, post-traumatic stress disorder (PTSD), 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 the disability and prognosis of paediatric headache.

To ascertain whether or not a headache should be attributed to a psychiatric disorder, it is necessary first to determine whether or not there is a concomitant psychiatric disorder. It is recommended to enquire in all headache patients about symptoms of commonly comorbid psychiatric disorders, such as depressive and anxiety disorders. When a psychiatric disorder is suspected to be a possible cause of the headache disorder, evaluation by an experienced psychiatrist or psychologist is recommended.

### 12.1 Headache attributed to somatization disorder<sup>1</sup>

**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<sup>1</sup> 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 has been 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 the somatization disorder
    - 2. constant or remitting headache parallels in time the fluctuation of other somatic symptoms attributed to the somatization disorder
    - 3. headache has remitted in parallel with remission of the other somatic symptoms attributed to the somatization disorder
  - D. Not better accounted for by another ICHD-3 diagnosis.

**Note:**

- 1. 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, published in 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, a persistently high level of anxiety about health or symptoms, and/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. Therefore, ICHD-3 continues to refer to the DSM-IV definition of somatization disorder.

**Comment:** 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.

### 1.2.2 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<sup>1</sup>
- C. Evidence of causation demonstrated by either or both of the following:
  1. headache has developed with or after the onset of the delusion, or led to its diagnosis
  2. headache has remitted after remission of the delusion
- D. Not better accounted for by another ICHD-3 diagnosis.<sup>2</sup>

#### Notes:

1. For example: 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.

2. When a patient first develops a headache (e.g. one of the primary headache disorders classified in Part One of ICHD-3) and *then* develops a delusional explanation for the headache, such as its being due to 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*.

**Comment:** 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 causing 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 the patient's head and causing the headache.

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**Part Three***Painful cranial neuropathies, other facial pain and other headaches*

13. Painful lesions of the cranial nerves and other facial pain
14. Other headache disorders

### 13. Painful lesions of the cranial nerves and other facial pain

- 13.1 Pain attributed to a lesion or disease of the trigeminal nerve
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#### Introduction

This chapter sets out a classification system for painful lesions of the cranial nerves and other facial pains based on a consensus between the International Headache Society (IHS) and the International Association for the Study of Pain (IASP).

The existing nosology of cranial nerve pains does not fully portray the subtle differences between various conditions. However, rather than abandoning many long-established diagnostic terms, this classification retains them, providing detailed definitions for differential diagnoses and their types, subtypes and subforms.

Afferent fibres in the trigeminal, intermedius, glossopharyngeal and vagus nerves, in addition to the upper cervical roots via the occipital nerves, convey nociceptive input to central pathways in the brainstem and to the brain areas that process nociception and pain in the head and neck. The brain perceives pain in the innervated area.

The pain may manifest in any of many distinct forms that are believed to reflect differences in neural pathophysiology, even if the details are not well known. What is known is that neuropathic facial pains can be classified on the basis of their distinct clinical characteristics and aetiology. Central to this concept is initial determination clinically of the main diagnostic group into which the patient’s pain best fits, to be followed by aetiological investigations for diagnostic types and subtypes and therapeutic decision-making.

There are several axes of classification.

#### a) *Syndromology: neuralgia or neuropathy*

The division between, for example, trigeminal neuralgia and trigeminal neuropathy should be viewed as a pragmatic way of distinguishing conditions in which clinical presentations and treatment approaches differ while the two conditions cannot be classified on the basis of currently known pathology or

pathophysiology. The same applies to painful conditions associated with the glossopharyngeal and intermedus nerves.

An important cause of cranial nerve pain is herpes zoster. Despite the fact that trigeminal pain following herpes zoster probably leads to different types of pathological change in trigeminal pathways (i.e. 'irritable nociceptor' versus 'deafferentation' type), available data are too limited to classify them as neuralgia versus neuropathy. Therefore, the well-established term *post-herpetic neuralgia* is maintained.

**b) Location: central or peripheral neuropathic pain**

A lesion or undue activation of these nerves (peripheral neuropathic pain), or of their central pathways (central neuropathic pain), causes neuropathic pain in the face.

**c) Aetiology: classical, idiopathic or secondary**

The cause of a neuropathic pain may be clear, such as infection by varicella-zoster virus or a structural abnormality (e.g. multiple sclerosis plaque) demonstrated by imaging: such pain is termed *secondary*, and attributed to the cause. In other cases no cause is apparent (termed *idiopathic*).

For the trigeminal, glossopharyngeal and intermedus neuralgias, the term *classical* is reserved for cases where imaging or surgery has revealed vascular compression of the respective nerve. Strictly speaking, classical neuralgias are *secondary* (to the neurovascular compression), but it is beneficial to separate them from other causes on the basis of the wider therapeutic options and potentially different nerve pathophysiology.

### 13.1 Pain attributed to a lesion or disease of the trigeminal nerve

#### 13.1.1 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. Additionally, there may be concomitant continuous pain of moderate intensity within the distribution(s) of the affected nerve division(s).

**Previously used terms:** Tic douloureux, primary trigeminal neuralgia.

**Diagnostic criteria:**

- A. Recurrent paroxysms of unilateral facial pain in the distribution(s) of one or more divisions of

the trigeminal nerve, with no radiation beyond,<sup>1</sup> and fulfilling criteria B and C

- B. Pain has all of the following characteristics:
1. lasting from a fraction of a second to two minutes<sup>2</sup>
  2. severe intensity<sup>3</sup>
  3. electric shock-like, shooting, stabbing or sharp in quality
- C. Precipitated by innocuous stimuli within the affected trigeminal distribution<sup>4</sup>
- D. Not better accounted for by another ICHD-3 diagnosis.

**Notes:**

1. In a few patients, pain may radiate to another division, but it remains within the trigeminal dermatomes.
2. Duration can change over time, with paroxysms becoming more prolonged. A minority of patients will report attacks predominantly lasting for >2 minutes.
3. Pain may become more severe over time.
4. Some attacks may be, or appear to be, spontaneous, but there must be a history or finding of pain provoked by innocuous stimuli to meet this criterion. Ideally, the examining clinician should attempt to confirm the history by replicating the triggering phenomenon. However, this may not always be possible because of the patient's refusal, awkward anatomical location of the trigger and/or other factors.

**Comments:** The diagnosis of 13.1.1 *Trigeminal neuralgia* must be established clinically. Investigations are designed to identify a likely cause.

Other than the triggering phenomenon, most patients with 13.1.1 *Trigeminal neuralgia* fail to show sensory abnormalities within the trigeminal distribution unless advanced methods are employed (e.g. quantitative sensory testing). However, in some, clinical neurological examination may show sensory deficits, which should prompt neuroimaging investigations to explore possible cause. Diagnosis of subforms such as 13.1.1.1 *Classical trigeminal neuralgia*, 13.1.1.2 *Secondary trigeminal neuralgia* or 13.1.1.3 *Idiopathic trigeminal neuralgia* is then possible.

When very severe, the pain often evokes contraction of the muscles of the face on the affected side (*tic douloureux*).

Mild autonomic symptoms such as lacrimation and/or redness of the ipsilateral eye may be present.

Following a painful paroxysm there is usually a refractory period during which pain cannot be triggered.

13.1.1.1 Classical trigeminal neuralgia

**Description:** Trigeminal neuralgia developing without apparent cause other than neurovascular compression.

**Diagnostic criteria:**

- A. Recurrent paroxysms of unilateral facial pain fulfilling criteria for 13.1.1 *Trigeminal neuralgia*
- B. Demonstration on MRI or during surgery of neurovascular compression (not simply contact), with morphological changes<sup>1</sup> in the trigeminal nerve root.

**Note:**

- 1. Typically atrophy or displacement.

**Comments:** Nerve root atrophy and/or displacement due to neurovascular compression are independently associated with the signs and symptoms of 13.1.1 *Trigeminal neuralgia*. When these anatomical changes are present, the condition is diagnosed as 13.1.1.1 *Classical trigeminal neuralgia*.

The common site of neurovascular compression is at the root entry zone, with compression by an artery more clearly associated with symptoms than compression by a vein. MRI techniques to measure volume and cross-sectional area of the root are available. Atrophic changes may include demyelination, neuronal loss, changes in microvasculature and other morphological changes. While the exact mechanisms of how atrophic changes in the trigeminal nerve contribute to the generation of pain, some evidence suggests that, when present preoperatively, they predict a good outcome following microvascular decompression.

Many patients with 13.1.1.1 *Classical trigeminal neuralgia* have a memorable onset of pain.

13.1.1.1 *Classical trigeminal neuralgia* usually appears in the second or third divisions. The pain rarely occurs bilaterally (sequentially rather than concomitantly).

13.1.1.1 *Classical trigeminal neuralgia* may be preceded by a period of atypical continuous pain termed *pre-trigeminal neuralgia* in the literature.

Between paroxysms, most patients are asymptomatic. In the subform 13.1.1.1.2 *Classical trigeminal neuralgia with concomitant continuous pain*, there is prolonged background pain in the affected area.

13.1.1.1.1 Classical trigeminal neuralgia, purely paroxysmal

**Description:** Classical trigeminal neuralgia without persistent background facial pain.

**Diagnostic criteria:**

- A. Recurrent paroxysms of unilateral facial pain fulfilling criteria for 13.1.1.1 *Classical trigeminal neuralgia*
- B. Pain-free between attacks in the affected trigeminal distribution.

**Comment:** 13.1.1.1.1 *Classical trigeminal neuralgia, purely paroxysmal* is usually responsive, at least initially, to pharmacotherapy (especially carbamazepine or oxcarbazepine).

13.1.1.1.2 Classical trigeminal neuralgia with concomitant continuous pain

**Previously used terms:** Atypical trigeminal neuralgia; trigeminal neuralgia type 2.

**Description:** Classical trigeminal neuralgia with persistent background facial pain.

**Diagnostic criteria:**

- A. Recurrent paroxysms of unilateral facial pain fulfilling criteria for 13.1.1.1 *Classical trigeminal neuralgia*
- B. Concomitant continuous or near-continuous pain between attacks in the affected trigeminal distribution.

**Comment:** Peripheral or central sensitization may account for the continuous pain.

13.1.1.2 Secondary trigeminal neuralgia

**Description:** Trigeminal neuralgia caused by an underlying disease. Clinical examination shows sensory changes in a significant proportion of these patients.

**Diagnostic criteria:**

- A. Recurrent paroxysms of unilateral facial pain fulfilling criteria for 13.1.1 *Trigeminal neuralgia*, either purely paroxysmal or associated with concomitant continuous or near-continuous pain
- B. An underlying disease has been demonstrated known that is to be able to cause, and explaining, the neuralgia<sup>1</sup>
- C. Not better accounted for by another ICHD-3 diagnosis.<sup>2</sup>

**Notes:**

- 1. Recognized causes are tumour in the cerebellopontine angle, arteriovenous malformation and multiple sclerosis.

2. MRI is best equipped to detect an underlying cause for 13.1.1.2 *Secondary trigeminal neuralgia*. Other investigations may include neurophysiological recording of trigeminal reflexes and trigeminal evoked potentials, suitable for patients who cannot undergo MRI.

#### 13.1.1.2.1 Trigeminal neuralgia attributed to multiple sclerosis

*Coded elsewhere:* 13.13.1 *Central neuropathic pain attributed to multiple sclerosis*.

*Description:* Trigeminal neuralgia caused by a multiple sclerosis (MS) plaque or plaques in the pons or trigeminal nerve root entry zone, and associated with other symptoms and/or clinical signs or laboratory findings of MS.

*Diagnostic criteria:*

- A. Recurrent paroxysms of unilateral facial pain fulfilling criteria for 13.1.1 *Trigeminal neuralgia*
- B. Both of the following:
  1. multiple sclerosis (MS) has been diagnosed
  2. an MS plaque at the trigeminal root entry zone or in the pons affecting the intrapontine primary afferents has been demonstrated by MRI, or its presence is suggested by routine electrophysiological studies<sup>1</sup> showing impairment of the trigeminal pathways
- C. Not better accounted for by another ICHD-3 diagnosis.

*Note:*

1. Blink reflex or trigeminal evoked potentials.

*Comments:* 13.1.1.2.1 *Trigeminal neuralgia attributed to multiple sclerosis* occurs in 2–5% of patients with multiple sclerosis (MS), sometimes bilaterally. Conversely, MS is detected in only 2–4% of cases of 13.1.1 *Trigeminal neuralgia*. Symptoms of trigeminal neuralgia are rarely a presenting feature of MS.

The lesion in the pons affects the intrapontine central terminals of the trigeminal afferents projecting to the trigeminal brainstem nuclei. Pontine lesions affecting the second order neurones of the trigeminothalamic tract usually lead to non-paroxysmal pain and/or dysaesthesias and should be classified as 13.13.1 *Central neuropathic pain attributed to multiple sclerosis*.

Some patients with MS are found to have neurovascular compression of the trigeminal root. It is thought that MS increases the susceptibility of the nerve root to

the effects of compression, leading more readily to painful paroxysms.

Patients with 13.1.1.2.1 *Trigeminal neuralgia attributed to multiple sclerosis* benefit less from pharmacological and surgical interventions than those with 13.1.1.1 *Classical trigeminal neuralgia*.

#### 13.1.1.2.2 Trigeminal neuralgia attributed to space-occupying lesion

*Description:* Trigeminal neuralgia caused by contact between the affected trigeminal nerve and a space-occupying lesion.

*Diagnostic criteria:*

- A. Recurrent paroxysms of unilateral facial pain fulfilling criteria for 13.1.1 *Trigeminal neuralgia*
- B. Both of the following:
  1. a space-occupying lesion in contact with the affected trigeminal nerve has been demonstrated
  2. pain has developed after identification of the lesion, or led to its discovery
- C. Not better accounted for by another ICHD-3 diagnosis.

*Comment:* Patients with 13.1.1.2.2 *Trigeminal neuralgia attributed to space-occupying lesion* may or may not have clinically detectable sensory signs, while electrophysiological tests such as trigeminal brainstem reflexes show abnormalities in nearly all cases.

#### 13.1.1.2.3 Trigeminal neuralgia attributed to other cause

*Description:* Trigeminal neuralgia caused by an underlying disease other than those described above.

*Diagnostic criteria:*

- A. Recurrent paroxysms of unilateral facial pain fulfilling criteria for 13.1.1 *Trigeminal neuralgia*, either purely paroxysmal or associated with concomitant continuous or near-continuous pain, but not necessarily unilateral
- B. Both of the following:
  1. a disorder other than those described above, but known to be able to cause trigeminal neuralgia, has been diagnosed<sup>1</sup>
  2. pain has developed after onset of the disorder, or led to its discovery
- C. Not better accounted for by another ICHD-3 diagnosis.



**Note:**

1. Recognized causes are skull-base bone deformity, connective tissue disease, arteriovenous malformation, dural arteriovenous fistula and genetic causes of neuropathy or nerve hyperexcitability.

**13.1.1.3 Idiopathic trigeminal neuralgia**

**Description:** Trigeminal neuralgia with neither electrophysiological tests nor MRI showing significant abnormalities.

**Diagnostic criteria:**

- A. Recurrent paroxysms of unilateral facial pain fulfilling criteria for 13.1.1 *Trigeminal neuralgia*, either purely paroxysmal or associated with concomitant continuous or near-continuous pain
- B. Neither 13.1.1.1 *Classical trigeminal neuralgia* nor 13.1.1.2 *Secondary trigeminal neuralgia* has been confirmed by adequate investigation including electrophysiological tests and MRI<sup>1</sup>
- C. Not better accounted for another ICHD-3 diagnosis.

**Note:**

1. A contact between a blood vessel and the trigeminal nerve and/or nerve root is a common finding on neuroimaging in healthy subjects. When such a contact is found in the presence of 13.1.1 *Trigeminal neuralgia* but without evidence of morphological changes (e.g. atrophy or displacement) in the nerve root, the criteria for 13.1.1.1 *Classical trigeminal neuralgia* are not fulfilled and the condition is considered idiopathic.

**13.1.1.3.1 Idiopathic trigeminal neuralgia, purely paroxysmal**

**Diagnostic criteria:**

- A. Recurrent paroxysms of unilateral facial pain fulfilling criteria for 13.1.1.3 *Idiopathic trigeminal neuralgia*
- B. Pain-free between attacks in the affected trigeminal distribution.

**13.1.1.3.2 Idiopathic trigeminal neuralgia with concomitant continuous pain**

**Diagnostic criteria:**

- A. Recurrent paroxysms of unilateral facial pain fulfilling criteria for 13.1.1.3 *Idiopathic trigeminal neuralgia*
- B. Concomitant continuous or near-continuous pain between attacks in the affected trigeminal distribution.

**13.1.2 Painful trigeminal neuropathy**

**Description:** Facial pain in the distribution(s) of one or more branches of the trigeminal nerve caused by another disorder and indicative of neural damage. The primary pain is usually continuous or near-continuous, and commonly described as burning or squeezing, or likened to pins and needles. Superimposed brief pain paroxysms may occur, but these are not the predominant pain type. This combination distinguishes painful trigeminal neuropathy from the subtypes of trigeminal neuralgia. There are clinically detectable sensory deficits within the trigeminal distribution, and mechanical allodynia and cold hyperalgesia are common, fulfilling IASP criteria for neuropathic pain. As a rule, allodynic areas are much larger than the punctate trigger zones present in trigeminal neuralgia.

**13.1.2.1 Painful trigeminal neuropathy attributed to herpes zoster**

**Description:** Unilateral facial pain of less than three months' duration in the distribution(s) 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 facial pain in the distribution(s) of a trigeminal nerve branch or branches, lasting <3 months
- B. One or more of the following:
  1. herpetic eruption has occurred in the same trigeminal distribution
  2. varicella-zoster virus (VZV) has been detected in the cerebrospinal fluid (CSF) by polymerase chain reaction (PCR)
  3. direct immunofluorescence assay for VZV antigen or PCR assay for VZV DNA is positive in cells obtained from the base of lesions

- C. 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 herpate*). 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 herpes zoster* is usually burning, stabbing/shooting, tingling or aching, and accompanied by cutaneous allodynia.

Ophthalmic herpes may be associated with IIIrd, IVth and/or 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 Trigeminal post-herpetic neuralgia

**Previously used term:** Post-herpetic trigeminal neuropathy.

**Description:** Unilateral facial pain persisting or recurring for at least three months in the distribution(s) of one or more branches of the trigeminal nerve, with variable sensory changes, caused by herpes zoster.

**Diagnostic criteria:**

- A. Unilateral facial pain in the distribution(s) of a trigeminal nerve branch or branches, persisting or recurring for >3 months and fulfilling criterion C
- B. Herpes zoster has affected the same trigeminal nerve branch or branches
- C. Pain developed in temporal relation to the herpes zoster infection<sup>1</sup>
- D. Not better accounted for by another ICHD-3 diagnosis.

**Note:**

1. Usually, pain will have developed while the rash was still active, but on occasion later, after rash has healed. In such cases, pale or light purple scars may be present as sequelae of the herpetic eruption.

**Comments:** Despite its long-preferred name, *post-herpetic neuralgia* is actually a neuropathy or neuronopathy: significant pathoanatomical changes have been shown in the nerve, ganglion and nerve root. In 13.1.2.2 *Trigeminal post-herpetic neuralgia*, there is also evidence of the inflammation extending into the trigeminal brainstem complex.

Following acute herpes zoster, post-herpetic neuralgia is more likely in the elderly.

The first division of the trigeminal nerve is most commonly affected in 13.1.2.2 *Trigeminal post-herpetic neuralgia*, but the second and third divisions can be involved also.

Typically, the pain of post-herpetic neuralgia is burning and itching – the latter sometimes very prominent and extremely bothersome. Also typically, patients with post-herpetic neuralgia show a clear sensory deficit and brush-evoked mechanical allodynia in the trigeminal distribution involved. Many patients, however, show little sensory loss, and instead demonstrate heightened responses to thermal and/or punctate stimuli.

#### 13.1.2.3 Painful post-traumatic trigeminal neuropathy

**Previously used term:** Anaesthesia dolorosa.

**Description:** Unilateral or bilateral facial or oral pain following and caused by trauma to the trigeminal nerve(s), with other symptoms and/or clinical signs of trigeminal nerve dysfunction.

**Diagnostic criteria:**

- A. Facial and/or oral pain in the distribution(s) of one or both trigeminal nerve(s) and fulfilling criterion C
- B. History of an identifiable traumatic event<sup>1</sup> to the trigeminal nerve(s), with clinically evident positive (hyperalgesia, allodynia) and/or negative (hypoesthesia, hypalgesia) signs of trigeminal nerve dysfunction
- C. Evidence of causation demonstrated by both of the following:
  1. pain is localized to the distribution(s) of the trigeminal nerve(s) affected by the traumatic event
  2. pain has developed <6 months after 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. Neuroablative procedures for trigeminal neuralgia, aimed at the trigeminal ganglion or nerve root, may result in neuropathic pain involving one or more trigeminal divisions; this should be considered as post-traumatic and coded here.

**Comments:** Pain duration ranges widely from paroxysmal to constant, and may be mixed.

Specifically following radiation-induced postganglionic injury, neuropathy may appear after more than three months.

13.1.2.3 *Painful post-traumatic trigeminal neuropathy* following neuroablative procedures aimed at the trigeminal ganglion or nerve root may coexist with 13.1.1 *Trigeminal neuralgia* if the latter recurs.

**13.1.2.4 Painful trigeminal neuropathy attributed to other disorder**

**Description:** Unilateral or bilateral facial or oral pain in the distribution(s) of one or more branches of the trigeminal nerve, caused by a disorder other than those described above, with other symptoms and/or clinical signs of trigeminal nerve dysfunction.

**Diagnostic criteria:**

- A. Unilateral or bilateral facial pain in the distribution(s) of one or both trigeminal nerve(s) and fulfilling criterion C
- B. A disorder, other than those described above but known to be able to cause painful trigeminal neuropathy with clinically evident positive (hyperalgesia, allodynia) and/or negative (hypoesthesia, hypalgesia) signs of trigeminal nerve dysfunction, and affecting one or both trigeminal nerves, has been diagnosed
- C. Evidence of causation demonstrated by both of the following:
  - 1. pain is localized to the distribution(s) of the trigeminal nerve(s) affected by the disorder
  - 2. pain developed after onset of the disorder, or led to its discovery
- D. Not better accounted for by another ICHD-3 diagnosis.

**Comments:** Painful trigeminal neuropathy may develop secondary to multiple sclerosis, space-occupying lesion or systemic disease, with only the clinical characteristics (quality of spontaneous pain, evoked pain and presence of sensory deficits) distinguishing between 13.1.1.2 *Secondary trigeminal neuralgia* and 13.1.2 *Painful trigeminal neuropathy*.

13.1.2 *Painful trigeminal neuropathy* caused by a connective tissue disease or hereditary disorders is usually bilateral but may begin asymmetrically and occasionally present with paroxysmal pain superimposed on the background pain. Patients will eventually develop bilateral sensory deficits and continuous pain, which clarify the diagnosis. MRI is

normal, but trigeminal reflexes are invariably delayed or absent.

**13.1.2.5 Idiopathic painful trigeminal neuropathy**

**Description:** Unilateral or bilateral pain in the distribution of one or more branches of the trigeminal nerve(s), indicative of neural damage but of unknown aetiology.

**Diagnostic criteria:**

- A. Unilateral or bilateral facial pain in the distribution(s) of one or both trigeminal nerve(s) and fulfilling criterion B
- B. Clinically evident positive (hyperalgesia, allodynia) and/or negative (hypoesthesia, hypalgesia) signs of trigeminal nerve dysfunction
- C. No cause has been identified
- D. Not better accounted for by another ICHD-3 diagnosis.

**13.2 Pain attributed to a lesion or disease of the glossopharyngeal nerve**

**13.2.1 Glossopharyngeal neuralgia**

**Previously used term:** Vaguglossopharyngeal neuralgia.

**Description:** A disorder characterized by unilateral brief stabbing pain, abrupt in onset and termination, in the distributions not only of the glossopharyngeal nerve but also of the auricular and pharyngeal branches of the vagus nerve. Pain is 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 or coughing and may remit and relapse in the fashion of trigeminal neuralgia.

**Diagnostic criteria:**

- A. Recurring paroxysmal attacks of unilateral pain in the distribution of the glossopharyngeal nerve<sup>1</sup> and fulfilling criterion B
- B. Pain has all of the following characteristics:
  - 1. lasting from a few seconds to two minutes
  - 2. severe intensity
  - 3. electric shock-like, shooting, stabbing or sharp in quality
  - 4. precipitated by swallowing, coughing, talking or yawning
- C. Not better accounted for by another ICHD-3 diagnosis.

**Note:**

1. Within the posterior part of the tongue, tonsillar fossa, pharynx or angle of the lower jaw and/or in the ear.

**Comments:** 13.2.1 *Glossopharyngeal neuralgia* can occur together with 13.1.1 *Trigeminal neuralgia*.

The superior laryngeal nerve is a branch of the vagus. Neuralgia of the superior laryngeal nerve presents similarly to 13.2.1 *Glossopharyngeal neuralgia* in its location and clinically can be difficult to distinguish from it.

Imaging may show neurovascular compression of the glossopharyngeal nerve.

Prior to development of 13.2.1 *Glossopharyngeal neuralgia*, unpleasant sensations may be felt in affected areas for weeks to several months.

The pain of 13.2.1 *Glossopharyngeal neuralgia* may radiate to involve the eye, nose, chin or shoulder. It can be severe enough for patients to lose weight. In rare cases, attacks of pain are associated with vagal symptoms such as cough, hoarseness, syncope and/or bradycardia. Some authors propose distinguishing between pharyngeal, otalgic and vagal subforms of neuralgia, and have suggested using the term *vagoglossopharyngeal neuralgia* when pain is accompanied by asystole, convulsions and syncope.

Clinical examination usually fails to show sensory changes in the nerve distribution but, if mild sensory deficits are encountered, they do not invalidate the diagnosis. Major changes or a reduced/missing gag reflex should prompt aetiological investigations.

13.2.1 *Glossopharyngeal neuralgia* is usually responsive, at least initially, to pharmacotherapy (especially carbamazepine or oxcarbazepine). It has been suggested that application of local anaesthetic to the tonsil and pharyngeal wall can prevent attacks for a few hours.

#### 13.2.1.1 Classical glossopharyngeal neuralgia

**Description:** Glossopharyngeal neuralgia developing without apparent cause other than neurovascular compression.

**Diagnostic criteria:**

- A. Recurrent paroxysms of unilateral pain fulfilling criteria for 13.2.1 *Glossopharyngeal neuralgia*
- B. Demonstration on MRI or during surgery of neurovascular compression of the glossopharyngeal nerve root.

#### 13.2.1.2 Secondary glossopharyngeal neuralgia

**Description:** Glossopharyngeal neuralgia caused by an underlying disease.

**Diagnostic criteria:**

- A. Recurrent paroxysms of unilateral pain fulfilling criteria for 13.2.1 *Glossopharyngeal neuralgia*
- B. An underlying disease has been demonstrated known to be able to cause, and explaining, the neuralgia.<sup>1</sup>

**Note:**

1. There are single reports of 13.2.1.2 *Secondary glossopharyngeal neuralgia* caused by neck trauma, multiple sclerosis, tonsillar or regional tumours, cerebellopontine angle tumours and Arnold–Chiari malformation.

#### 13.2.1.3 Idiopathic glossopharyngeal neuralgia

**Description:** Glossopharyngeal neuralgia with no evidence either of neurovascular compression or of causative underlying disease.

**Diagnostic criteria:**

- A. Recurrent paroxysms of unilateral pain fulfilling criteria for 13.2.1 *Glossopharyngeal neuralgia*
- B. Investigations have found neither neurovascular compression nor an underlying disease known to be able to cause 13.2.1.2 *Secondary glossopharyngeal neuralgia*
- C. Not better accounted for another ICHD-3 diagnosis.

#### 13.2.2 Painful glossopharyngeal neuropathy

**Description:** Pain within the distribution of the glossopharyngeal nerve (posterior part of the tongue, tonsillar fossa, pharynx and/or beneath the angle of the lower jaw). In addition, pain is commonly perceived in the ipsilateral ear. The primary pain is usually continuous or near-continuous, and commonly described as burning or squeezing, or likened to pins and needles. Brief paroxysms may be superimposed, but they are not the predominant pain type. This combination distinguishes painful glossopharyngeal neuropathy from the subforms of 13.2.1 *Glossopharyngeal neuralgia*. Sensory deficits may be present in the ipsilateral posterior part of the tongue and tonsillar fossa, and the gag reflex may be weak or missing.

### 13.2.2.1 Painful glossopharyngeal neuropathy attributed to a known cause

**Description:** Unilateral continuous or near-continuous pain, with or without superimposed brief paroxysms, in the distribution of the glossopharyngeal nerve and caused by another identified disorder.

**Diagnostic criteria:**

- A. Unilateral continuous or near-continuous pain<sup>1</sup> in the distribution of the glossopharyngeal nerve and fulfilling criterion C
- B. A disorder known to be able to cause painful glossopharyngeal neuropathy has been diagnosed<sup>2</sup>
- C. Evidence of causation demonstrated by both of the following:
  1. pain is ipsilateral to the glossopharyngeal nerve affected by the disorder
  2. pain has developed after onset of the disorder, or led to its discovery
- D. Not better accounted for by another ICHD-3 diagnosis.

**Notes:**

1. Brief paroxysms may be superimposed, but are not the predominant pain type.
2. Tumours at the cerebellopontine angle and iatrogenic injury during procedures have been reported to cause painful glossopharyngeal neuropathy.

### 13.2.2.2 Idiopathic painful glossopharyngeal neuropathy

**Description:** Unilateral continuous or near-continuous pain, with or without superimposed brief paroxysms, in the distribution(s) of the glossopharyngeal nerve and of unknown aetiology.

**Diagnostic criteria:**

- A. Unilateral continuous or near-continuous pain<sup>1</sup> in the distribution of the glossopharyngeal nerve
- B. No cause has been identified
- C. Not better accounted for by another ICHD-3 diagnosis.

**Note:**

1. Brief paroxysms may be superimposed, but are not the predominant pain type.

## 13.3 Pain attributed to a lesion or disease of nervus intermedius

### 13.3.1 Nervus intermedius neuralgia

**Previously used term:** Geniculate neuralgia.

**Description:** A rare disorder characterized by brief paroxysms of pain felt deeply in the auditory canal, sometimes radiating to the parieto-occipital region. In the vast majority of cases, vascular compression is found at operation, occasionally with a thickened arachnoid, but it may develop without apparent cause or as a complication of herpes zoster or, very rarely, multiple sclerosis or tumour. It is provoked by stimulation of a trigger area in the posterior wall of the auditory canal and/or periauricular region.

**Diagnostic criteria:**

- A. Paroxysmal attacks of unilateral pain in the distribution of nervus intermedius<sup>1</sup> and fulfilling criterion B
- B. Pain has all of the following characteristics:
  1. lasting from a few seconds to minutes
  2. severe in 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
- C. Not better accounted for by another ICHD-3 diagnosis.<sup>2</sup>

**Notes:**

1. Pain is located in the auditory canal, auricle, in the region of the mastoid process and occasionally the soft palate, and may sometimes radiate to the temporal region or the angle of the mandible.
2. In view of the complex and overlapping innervation of the external ear, deriving from trigeminal (auriculotemporal), facial (nervus intermedius), glossopharyngeal, vagus and second cranial nerves, attribution of neuralgias to a single nerve may not be easy in this body region when a specific neurovascular contact cannot be visualized.

**Comment:** Disorders of lacrimation, salivation and/or taste sometimes accompany the pain of 13.3.1 *Nervus intermedius neuralgia*.

### 13.3.1.1 Classical nervus intermedius neuralgia

**Description:** Nervus intermedius neuralgia developing without apparent cause other than neurovascular compression.

**Diagnostic criteria:**

- A. Recurrent paroxysms of unilateral pain fulfilling criteria for 13.3.1 *Nervus intermedius neuralgia*
- B. Demonstration on MRI or during surgery of neurovascular compression of the nervus intermedius nerve root.

### 13.3.1.2 Secondary nervus intermedius neuralgia

**Description:** Nervus intermedius neuralgia caused by an underlying disease.

**Diagnostic criteria:**

- A. Recurrent paroxysms of unilateral pain fulfilling criteria for 13.3.1 *Nervus intermedius neuralgia*
- B. An underlying disease has been demonstrated known to be able to cause, and explaining, the neuralgia.<sup>1</sup>

**Note:**

1. There are single reports of 13.3.1.2 *Secondary nervus intermedius neuralgia* caused by multiple sclerosis or tumour. In the latter case, neurological deficits arising from damage to other nerves in close proximity tend to dominate the clinical presentation. Herpes zoster typically usually leads to 13.3.2.1 *Painful nervus intermedius neuropathy attributed to herpes zoster* rather than 13.3.1.2 *Secondary nervus intermedius neuralgia*.

### 13.3.1.3 Idiopathic nervus intermedius neuralgia

**Description:** Nervus intermedius neuralgia with no evidence either of neurovascular compression or of causative underlying disease.

**Diagnostic criteria:**

- A. Recurrent paroxysms of unilateral pain fulfilling criteria for 13.3.1 *Nervus intermedius neuralgia*
- B. Investigations have found neither neurovascular compression nor an underlying disease known to

be able to cause 13.3.1.2 *Secondary nervus intermedius neuralgia*

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

### 13.3.2 Painful nervus intermedius neuropathy

**Description:** Pain within the distribution(s) of the intermediate nerve(s) (auditory canal, auricle or region of the mastoid process), usually described by the patient as dull, deep in the ear and continuous or near-continuous. Brief paroxysms may be superimposed, but they are not the predominant pain type. This combination distinguishes painful nervus intermedius neuropathy from the subforms of 13.3.1 *Nervus intermedius neuralgia*. Sensory deficits, usually slight, may be present in the ear canal, auricle or skin overlying the mastoid process.

#### 13.3.2.1 Painful nervus intermedius neuropathy attributed to herpes zoster

**Previously used term:** 13.3.2.1 *Painful nervus intermedius neuropathy attributed to herpes zoster* associated with facial paresis is known as Ramsay Hunt syndrome.

**Description:** Unilateral continuous or near-continuous pain, with or without brief paroxysms superimposed, in the distribution of nervus intermedius and felt deeply in the auditory canal, caused by nervus intermedius herpes zoster infection and commonly associated with facial paresis and other symptoms and/or clinical signs of the infection or its aftermath.

**Diagnostic criteria:**

- A. Unilateral continuous or near-continuous pain<sup>1</sup> in the distribution of nervus intermedius<sup>2</sup> and fulfilling criterion C
- B. One or more of the following:
  1. herpetic eruption has occurred in the distribution of nervus intermedius<sup>3</sup>
  2. varicella-zoster virus (VZV) has been detected in the cerebrospinal fluid (CSF) by polymerase chain reaction (PCR)
  3. direct immunofluorescence assay for VZV antigen or PCR assay for VZV DNA is positive in cells obtained from the base of lesions
- C. Pain developed in temporal relation to the herpes zoster<sup>4</sup>
- D. Not better accounted for by another ICHD-3 diagnosis.<sup>5</sup>

**Notes:**

1. Brief paroxysms may be superimposed, but are not the predominant pain type.
2. In the auditory canal, auricle and/or region of the mastoid process.
3. Owing to viral spread, other cranial nerves may become affected.
4. Pain can precede the herpetic eruption.
5. The diagnosis is confirmed clinically in the acute stages by detection of vesicles on the tympanic membrane, auditory canal, auricle and/or skin overlying the mastoid process. They may also be seen in the anterior third of the tongue, which the virus has reached via chorda tympani, or on the hard palate, supplied by a vestigial remnant branch of the facial nerve.

**Comments:** Other cranial nerves (VIII, IX, X, XI) may also be affected, leading to tinnitus, hearing loss, vertigo, nausea, hoarseness and dysphagia.

While little is known of the natural course of 13.3.2.1 *Painful nervus intermedius neuropathy attributed to herpes zoster*, the pain may continue for more than three months; it should then be classified as 13.3.2.2 *Post-herpetic neuralgia of nervus intermedius*.

**13.3.2.2 Post-herpetic neuralgia of nervus intermedius**

**Description:** Unilateral pain persisting or recurring for at least three months in the distribution of nervus intermedius, felt deeply in the auditory canal, caused by nervus intermedius herpes zoster infection.

**Diagnostic criteria:**

- A. Unilateral pain in the distribution of nervus intermedius,<sup>1</sup> persisting or recurring for >3 months and fulfilling criterion C
- B. Nervus intermedius herpes zoster infection has occurred
- C. Pain developed in temporal relation to the herpes zoster infection<sup>2</sup>
- D. Not better accounted for by another ICHD-3 diagnosis.

**Notes:**

1. In the auditory canal, auricle and/or region of the mastoid process.
2. Usually, pain will have developed while the infection was still active, but on occasion later.

**13.3.2.3 Painful nervus intermedius neuropathy attributed to other disorder**

**Description:** Unilateral continuous or near-continuous pain, with or without superimposed brief paroxysms, in the distribution of nervus intermedius and caused by a disorder other than herpes zoster infection. There may be other symptoms and/or clinical signs of the causative disorder.

**Diagnostic criteria:**

- A. Unilateral continuous or near-continuous pain<sup>1</sup> in the distribution of nervus intermedius,<sup>2</sup> fulfilling criterion C
- B. A disorder affecting nervus intermedius, other than herpes zoster infection but known to be able to cause painful nervus intermedius neuropathy, has been diagnosed<sup>3</sup>
- C. Pain has developed after onset of the disorder, or led to its discovery
- D. Not better accounted for by another ICHD-3 diagnosis.

**Notes:**

1. Brief paroxysms may be superimposed, but are not the predominant pain type.
2. In the auditory canal, auricle and/or region of the mastoid process.
3. 13.3.2 *Painful nervus intermedius neuropathy* has been rarely described in patients with facial tumours or injury to the geniculate ganglion.

**13.3.2.4 Idiopathic painful nervus intermedius neuropathy**

**Description:** Unilateral continuous or near-continuous pain, with or without superimposed brief paroxysms, in the distribution(s) of nervus intermedius and of unknown aetiology.

**Diagnostic criteria:**

- A. Pain in the distribution(s) of nervus intermedius<sup>1</sup> on one or both sides
- B. No cause has been identified
- C. Not better accounted for by another ICHD-3 diagnosis.

**Note:**

1. In the auditory canal, auricle and/or region of the mastoid process.

### 13.4 Occipital neuralgia

**Description:** Unilateral or bilateral paroxysmal, shooting or stabbing pain in the posterior part of the scalp, in the distribution(s) of the greater, lesser and/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 in the distribution(s) of the greater, lesser and/or third occipital nerves and fulfilling criteria B–D
- B. Pain has at least two of the following three characteristics:
  1. recurring in paroxysmal attacks lasting from a few seconds to minutes
  2. severe in intensity
  3. shooting, stabbing or sharp in quality
- C. 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 distribution of C2
- D. Pain is eased temporarily by local anaesthetic block of the affected nerve(s)
- E. 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 interneuronal 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 Neck-tongue syndrome

**Description:** Immediate-onset, unilateral, sharp or stabbing and usually severe occipital and/or upper neck pain brought on by sudden rotatory head movement, accompanied by abnormal sensation and/or posture of the ipsilateral tongue.

**Diagnostic criteria:**

- A. At least two episodes fulfilling criteria B–D

- B. Sharp or stabbing unilateral pain<sup>1</sup> in the upper neck and/or occipital region with concurrent abnormal sensation and/or posture of the ipsilateral tongue
- C. Precipitated by sudden turning of the neck
- D. Lasting from seconds to several minutes
- E. Not better accounted for by another ICHD-3 diagnosis.

**Note:**

1. There may or may not be simultaneous dysaesthesia.

**Comment:** A recent study has described this condition in detail, warranting its promotion from the Appendix (where it appeared in ICHD-3 beta).

### 13.6 Painful 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 retro-orbital, orbital, frontal and/or temporal pain fulfilling criterion C
- B. Clinical, electrophysiological, imaging and/or laboratory evidence confirming optic neuritis<sup>1</sup>
- C. Evidence of causation demonstrated by both of the following:
  1. pain has developed in temporal relation to the optic neuritis
  2. pain is aggravated by eye movement
- D. Not better accounted for by another ICHD-3 diagnosis.

**Note:**

1. Gadolinium-enhanced MRI shows optic nerve enhancement in 90% of cases of 13.6 *Painful optic neuritis*.

**Comments:** Clinical series report the prevalence of pain in optic neuritis to be about 90%. Pain may precede impairment of vision.

13.6 *Painful optic neuritis* is often a manifestation of multiple sclerosis.



### 13.7 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 and/or VIth cranial nerve(s).

**Diagnostic criteria:**

- A. Unilateral frontal and/or periorbital headache fulfilling criterion C
- B. Clinical and imaging evidence confirming an ischaemic ocular motor nerve palsy<sup>1</sup>
- C. Evidence of causation demonstrated by both of the following:
  - 1. headache is ipsilateral to the motor nerve palsy
  - 2. headache has developed in temporal relation to the motor nerve palsy
- D. Not better accounted for by another ICHD-3 diagnosis.

**Note:**

- 1. 13.7 *Headache attributed to ischaemic ocular motor nerve palsy* can occur prior to or concurrently with the onset of diplopia.

**Comment:** The majority of ocular motor nerve palsies are painful, regardless of the presence or absence of diabetes. Pain is most frequent in patients with IIIrd nerve palsies, less so in those with VIth nerve paresis and least frequent in cases of IVth nerve paresis.

### 13.8 Tolosa–Hunt syndrome

**Description:** Unilateral orbital or periorbital 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 orbital or periorbital 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 is ipsilateral to the granulomatous inflammation
- 2. headache has preceded paresis of the IIIrd, IVth and/or VIth nerves by  $\leq 2$  weeks, or developed with it
- D. Not better accounted for by another ICHD-3 diagnosis.

**Comments:** Some reported cases of 13.8 *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.

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 13.8 *Tolosa–Hunt syndrome* resolve when it is treated adequately with corticosteroids.

### 13.9 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 ipsilateral 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. Ipsilateral Horner’s syndrome, with imaging evidence of underlying disease of either the middle cranial fossa or 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, or led to its discovery
  - 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. Not better accounted for by another ICHD-3 diagnosis.

**Comment:** The original description of 13.9 *Paratrigeminal oculosympathetic (Raeder’s) syndrome* is regarded as a classical example of clinico-anatomical methodology in the early 20th century, and was useful because the involvement of oculosympathetic

fibres indicated a lesion of the middle cranial fossa. 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.

### 13.10 Recurrent painful ophthalmoplegic neuropathy

**Previously used term:** Ophthalmoplegic migraine (this old and inappropriate term was rejected because this syndrome is not migrainous but rather a recurrent painful neuropathy).

**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. Both of the following:
  - 1. unilateral headache
  - 2. ipsilateral paresis of one, two or all three ocular motor nerves<sup>1</sup>
- C. Orbital, parasellar or posterior fossa lesion has been excluded by appropriate investigation
- D. Not better accounted for by another ICHD-3 diagnosis.

#### Note:

- 1. Some data suggest that headache can develop up to 14 days prior to ocular motor paresis.

**Comments:** Gadolinium enhancement or nerve thickening can be demonstrated using MRI.

Treatment with corticosteroids is beneficial in some patients.

### 13.11 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 two hours/day over more than three months, without clinically evident causative lesions.

#### Diagnostic criteria:

- A. Oral pain<sup>1</sup> fulfilling criteria B and C
- B. Recurring daily for >2 hours/day for >3 months

- C. Pain has both of the following characteristics:
  - 1. burning quality<sup>2</sup>
  - 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.

#### Notes:

- 1. The pain is usually bilateral; the most common site is the tip of the tongue.
- 2. Pain intensity fluctuates.

**Comments:** 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. Laboratory investigations and brain imaging 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.

### 13.12 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 two hours/day over more than three 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/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 by patients to describe the character of 13.12 *Persistent idiopathic facial pain* but it is most often depicted as dull, nagging or aching, either deep or superficial. It can have sharp exacerbations, and is aggravated by stress. With time, it may spread to a wider area of the craniocervical region.

Patients with 13.12 *Persistent idiopathic facial pain* are predominantly female.

13.12 *Persistent idiopathic facial pain* 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.

13.12 *Persistent idiopathic facial pain* may originate from a minor operation or injury to the face, maxillae, teeth or gums, but persists after healing of the initial noxious event without any demonstrable local cause. However, psychophysical or neurophysiological tests may demonstrate sensory abnormalities. A continuum seems to exist from 13.12 *Persistent idiopathic facial pain* induced by insignificant trauma to 13.1.2.3 *Painful post-traumatic trigeminal neuropathy* caused obviously by significant insult to the peripheral nerves.

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 subtype of 13.12 *Persistent idiopathic facial pain* 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 subtypes/forms, if they exist, have not been sufficiently studied to propose diagnostic criteria.

### 13.13 Central neuropathic pain

**Description:** Unilateral or bilateral craniocervical pain of central origin, with variable presentation and with or without sensory changes. Depending on the cause, it may be constant or remitting and relapsing.

#### 13.13.1 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<sup>1</sup>

- B. Multiple sclerosis 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.

**Note:**

- 1. Pain may be paroxysmal or continuous.

**Comment:** Non-painful sensory abnormalities (usually dysaesthesia but also hypaesthesia, anaesthesia, hypalgesia, paraesthesia, etc) may coexist with pain in 13.13.1 *Central neuropathic pain attributed to multiple sclerosis*.

#### 13.13.2 Central post-stroke pain (CPSP)

**Description:** Usually unilateral facial and/or head pain, with varying presentations involving parts or all of the craniocervical region and associated with impaired sensation, occurring within six 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 six months after the stroke
  - 2. imaging<sup>1</sup> has demonstrated a vascular lesion in an appropriate site
- D. Not better accounted for by another ICHD-3 diagnosis.

**Note:**

- 1. Usually MRI.

**Comments:** 13.13.2 *Central post-stroke pain* is attributed to a lesion of the ascending projections of the trigeminal nuclei. Cervical spinothalamic pathways and cortical processing may also play significant roles.

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. Headache 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.

## A. 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 and/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
  - A1.2.0.1 Pure menstrual migraine with aura
  - A1.2.0.2 Menstrually related migraine with aura
  - A1.2.0.3 Non-menstrual migraine with aura
- 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
  - A1.4.6 Visual snow
- 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)**

- A2.1 Infrequent episodic tension-type headache (alternative criteria)
- A2.2 Frequent episodic tension-type headache (alternative criteria)
- A2.3 Chronic tension-type headache (alternative criteria)

**A3. Trigeminal-autonomic cephalalgias (TACs)**

- A3.1 Cluster headache (alternative criteria)
- A3.2 Paroxysmal hemicrania (alternative criteria)
- A3.3 Short-lasting unilateral neuralgiform headache attacks (alternative criteria)
- A3.4 Hemicrania continua (alternative criteria)
- 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 and/or cervical vascular disorder**

- A6.10 Persistent headache attributed to past cranial and/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
- A9.3 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

A11.3 Headache attributed to disorder of the eyes

A11.3.5 Headache attributed to heterophoria or heterotropia

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 (PTSD)

## 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.

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 has happened between ICHD-II, ICHD-3 beta and ICHD-3, 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,<sup>1</sup> fulfilling criteria for 1.1 *Migraine without aura* and criterion B below
- B. Occurring exclusively on day  $1 \pm 2$  (i.e. days  $-2$  to  $+3$ )<sup>2</sup> of menstruation<sup>1</sup> in at least two out of three menstrual cycles and at no other times of the cycle.<sup>3</sup>

##### Notes:

1. For the purposes of ICHD-3, menstruation is considered to be endometrial bleeding resulting either from 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.
3. For research purposes a prospective diary is recommended, but this is not mandatory for clinical diagnosis of A1.1.1 *Pure menstrual migraine without aura*.

#### A1.1.2 Menstrually related migraine without aura

##### Diagnostic criteria:

- A. Attacks, in a menstruating woman,<sup>1</sup> fulfilling criteria for 1.1 *Migraine without aura* and criterion B below
- B. Occurring on day  $1 \pm 2$  (i.e. days  $-2$  to  $+3$ )<sup>2</sup> of menstruation<sup>1</sup> in at least two out of three menstrual cycles, and additionally at other times of the cycle.<sup>3</sup>

##### Notes:

1. For the purposes of ICHD-3, menstruation is considered to be endometrial bleeding resulting either from 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.
3. For research purposes a prospective diary is recommended, but this is not mandatory for clinical diagnosis of A1.1.2 *Menstrually related migraine without aura*.

#### A1.1.3 Non-menstrual migraine without aura

##### Diagnostic criteria:

- A. Attacks, in a menstruating woman,<sup>1</sup> fulfilling criteria for 1.1 *Migraine without aura* and criterion B below
- B. Not fulfilling 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, 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.

Menstrual migraine attacks are mostly without aura. 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.

Many women over-report an association between attacks and menstruation; for research purposes, diary-documented, prospectively recorded evidence over a minimum of three cycles is necessary to confirm the diagnosis.

The mechanism(s) of migraine may be different with endometrial bleeding resulting from the normal menstrual cycle and bleeding due to 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 distinct subpopulations even though the diagnostic criteria do not. Management strategies may also differ for these 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

### A1.2.0.1 Pure menstrual migraine with aura

*Diagnostic criteria:*

- A. Attacks, in a menstruating woman,<sup>1</sup> fulfilling criteria for 1.2 *Migraine with aura* and criterion B below
- B. Occurring exclusively on day  $1 \pm 2$  (i.e. days  $-2$  to  $+3$ )<sup>2</sup> of menstruation<sup>1</sup> in at least two out of three menstrual cycles and at no other times of the cycle.<sup>3</sup>

*Notes:*

1. For the purposes of ICHD-3, menstruation is considered to be endometrial bleeding resulting either from 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.
3. For research purposes a prospective diary is recommended, but this is not mandatory for clinical diagnosis of A1.2.0.1 *Pure menstrual migraine with aura*.

### A1.2.0.2 Menstrually related migraine with aura

*Diagnostic criteria:*

- A. Attacks, in a menstruating woman,<sup>1</sup> fulfilling criteria for 1.2 *Migraine without aura* and criterion B below
- B. Occurring on day  $1 \pm 2$  (i.e. days  $-2$  to  $+3$ )<sup>2</sup> of menstruation<sup>1</sup> in at least two out of three menstrual cycles, and additionally at other times of the cycle.<sup>3</sup>

*Notes:*

1. For the purposes of ICHD-3, menstruation is considered to be endometrial bleeding resulting either from 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.
3. For research purposes a prospective diary is recommended, but this is not mandatory for clinical diagnosis of A1.2.0.2 *Menstrually related migraine with aura*.

### A1.2.0.3 Non-menstrual migraine with aura

*Diagnostic criteria:*

- A. Attacks, in a menstruating woman,<sup>1</sup> fulfilling criteria for 1.2 *Migraine with aura* and criterion B below
- B. Not fulfilling criterion B for A1.2.0.1 *Pure menstrual migraine with aura* or A1.2.0.2 *Menstrually related migraine with aura*.

*Note:*

1. For the purposes of ICHD-3, 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 1.2 *Migraine with aura* is clearly applicable only to menstruating women as defined above.

Menstrual attacks of migraine are mostly without aura. These criteria for A1.2.0.1 *Pure menstrual migraine with aura* and A1.2.0.2 *Menstrually related migraine with aura* are included to allow these uncommon subforms to be better characterized. Criteria for A1.2.0.3 *Non-menstrual migraine with aura* are included for completeness.

Many women over-report an association between attacks and menstruation; for research purposes, diary-documented, prospectively recorded evidence over a minimum of three cycles is necessary to confirm the diagnosis.

The mechanism(s) of migraine may be different with endometrial bleeding resulting from the normal menstrual cycle and bleeding due to 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 distinct subpopulations even though the diagnostic criteria do not.

## A1.3 Chronic migraine (alternative criteria)

*Alternative diagnostic criteria:*

- A. Headache (migraine-like or tension-type-like) on  $\geq 15$  days/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  $\geq 8$  days/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.

- B. Additional visual symptoms of at least two of the following four types:
  1. palinopsia<sup>2</sup>
  2. enhanced entoptic phenomena<sup>3</sup>
  3. photophobia
  4. impaired night vision (nyctalopia)
- C. Symptoms are not consistent with typical migraine visual aura<sup>4</sup>
- D. Symptoms are not better accounted for by another disorder<sup>5</sup>

#### 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  $\geq 5$  days/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  $\geq 5$  days/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 three auras occur over a period of three days.

**Comment:** Other neurological disorders including reversible cerebral vasoconstriction syndrome, posterior reversible encephalopathy syndrome and arterial dissection should be excluded by appropriate investigation.

#### A1.4.6 Visual snow

##### Diagnostic criteria:

- A. Dynamic, continuous, tiny dots across the entire visual field,<sup>1</sup> persisting for  $>3$  months

##### Notes:

1. Patients compare visual snow to television static ('television snow'). The dots are usually black or grey on a white background and grey or white on a black background, but also reported are transparent dots, white flashing dots and coloured dots.
2. Palinopsia may be visual after-images and/or trailing of moving objects. Visual after-images are different from retinal after-images, which occur only after staring at a high-contrast image and are in complementary colour.
3. These phenomena, arising from the structure of the visual system itself, include excessive floaters in both eyes, excessive blue field entoptic phenomenon (uncountable little grey/white/black dots or rings shooting over the visual field of both eyes when looking at homogeneous bright surfaces such as the blue sky), self-lighting of the eye (coloured waves or clouds perceived when closing the eyes in the dark) and spontaneous photopsia (bright flashes of light).
4. As described under 1.2.1 *Migraine with typical aura*.
5. Normal ophthalmology tests (corrected visual acuity, dilated-pupil funduscopy, visual field examination and electroretinography) and no intake of psychotropic drugs.

**Comments:** A1.4.6 *Visual snow* is newly included in the Appendix of ICHD-3. It may not per se be part of the migraine spectrum but appears to be epidemiologically associated with 1.2 *Migraine with aura*. Further research is needed into whether these disorders share pathophysiological mechanisms causing visual symptoms but, meanwhile, it is hypothesized that cortical hyperexcitability plays a role in both. Patients with 1. *Migraine* have an increased prevalence of palinopsia and heightened visual sensitivity outside attacks: A1.4.6 *Visual snow* features both palinopsia and photophobia. Patients with A1.4.6 *Visual snow* and comorbid 1. *Migraine* more often have palinopsia, spontaneous photopsia, photophobia, nyctalopia and tinnitus than those without comorbid migraine.



Two other reasons support inclusion of A1.4.6 *Visual snow* in ICHD-3. First, it creates awareness of this condition, and aids physicians in recognizing it. Patients complaining of visual snow as a symptom often have (a history of) 1. *Migraine*; physicians unaware of A1.4.6 *Visual snow* may misinterpret its symptoms as persistent visual aura. Second, in a similar argument applied to research, future studies on persistent visual symptoms need homogeneous study groups; inclusion of criteria for A1.4.6 *Visual snow* makes it clear to researchers how this disorder is currently defined.

### 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 four months of age, fulfilling criterion B
- B. Both of the following:
  - 1. episodes last for  $\geq 3$  hours/day
  - 2. episodes occur on  $\geq 3$  days/week for  $\geq 3$  weeks
- C. Not attributed to another disorder.<sup>1</sup>

**Note:**

- 1. In particular, failure to thrive has been excluded.

**Comments:** Infantile colic affects one baby in five.

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 increases twofold.

#### 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 criterion B

- B. Onset before the age of 18 months
- C. At least one other paroxysmal phenomenon<sup>1</sup> associated with the bouts of hemiplegia or occurring independently
- D. Evidence of mental and/or neurological deficit(s)
- E. Not attributed to another disorder.

**Note:**

- 1. Such as tonic spells, dystonic posturing, choreoathetoid movements, nystagmus or other ocular motor abnormalities and/or autonomic disturbances.

**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 *ATPIA3* gene (encoding the sodium-potassium [Na<sup>+</sup>/K<sup>+</sup>] ATPase  $\alpha 3$  subunit) are likely to be responsible for at least 70% of cases.

#### A1.6.6 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*<sup>1</sup>
- C. Vestibular symptoms<sup>2</sup> of moderate or severe intensity,<sup>3</sup> lasting between five minutes and 72 hours<sup>4</sup>
- D. At least half of episodes are associated with at least one of the following three migrainous features<sup>5</sup>:
  - 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 phonophobia
  - 3. visual aura
- E. Not better accounted for by another ICHD-3 diagnosis or by another vestibular disorder.<sup>6</sup>

**Notes:**

- 1. Code also for the underlying migraine diagnosis.
- 2. Vestibular symptoms, as defined by the Bárány Society's *International Classification of Vestibular*

*Disorders* and qualifying for a diagnosis of A1.6.6 *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 four 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. 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 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:** A surprisingly high prevalence of A1.6.6 *Vestibular migraine* of 10.3% was recently described among migraine patients in Chinese neurological departments.

#### **Other symptoms**

Transient auditory symptoms, nausea, vomiting, prostration and susceptibility to motion sickness may

be associated with A1.6.6 *Vestibular migraine*. However, since 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. Only a minority of patients with A1.6.6 *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 A1.6.6 *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 requires at least two brainstem symptoms in addition to visual, sensory or dysphasic aura symptoms for this diagnosis. Fewer than 10% of patients with A1.6.6 *Vestibular migraine* fulfil these criteria. Therefore, A1.6.6 *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**

While A1.6.6 *Vestibular migraine* may start at any age, ICHD-3 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. Since the classification of A1.6.6 *Vestibular migraine* does not involve any age limit, the diagnosis can be applied in children when the respective criteria are met, but only children with different types of vertigo attacks (e.g. short-duration episodes of less than five minutes and longer-lasting ones of more than five 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 A1.6.6 *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 A1.6.6 *Vestibular migraine*, but hearing loss does not progress to profound levels. Similarly, migraine headaches,

photophobia and even migraine auras are common during Ménière attacks. The pathophysiological relationship between A1.6.6 *Vestibular migraine* and Ménière's disease remains uncertain. In the first year after onset of symptoms, differentiation between them may be challenging, since Ménière's disease can be monosymptomatic with only vestibular symptoms in the early stages of the disease.

When the criteria for Ménière's disease are met, particularly hearing loss as documented by audiometry, Ménière'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 A1.6.6 *Vestibular migraine* and the other for Ménière's disease, should be diagnosed with both disorders. A future revision of ICHD may include a vestibular migraine/Ménière'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* and 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.

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## A3. Trigeminal-autonomic cephalalgias (TACs)

### A3.1 Cluster headache (alternative criteria)

#### Alternative 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)<sup>1</sup>

- 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. Occurring with a frequency between one every other day and eight per day<sup>2</sup>
- E. Not better accounted for by another ICHD-3 diagnosis.

#### Notes:

1. During part, but less than half, of the active time-course of A3.1 *Cluster headache*, attacks may be less severe and/or of shorter or longer duration.
2. During part, but less than half, of the active time-course of A3.1 *Cluster headache*, attacks may be less frequent.

*Comment:* Opinion is divided on inclusion of (e) and (f) in criterion C1. Experts in the working group believe it improves sensitivity without significant loss of specificity, but formal field testing has not confirmed this.

### A3.2 Paroxysmal hemicrania (alternative criteria)

#### Alternative 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. 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. Occurring with a frequency of >5 per day<sup>1</sup>
- E. Prevented absolutely by therapeutic doses of indomethacin<sup>2</sup>
- F. Not better accounted for by another ICHD-3 diagnosis.

Notes:

1. During part, but less than half, of the active time-course of A3.2 *Paroxysmal hemicrania*, attacks may be less frequent.
2. 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:** Opinion is divided on inclusion of (e) and (f) in criterion C1. Experts in the working group believe it improves sensitivity without significant loss of specificity, but formal field testing has not been performed to support the change in criteria.

**A3.3 Short-lasting unilateral neuralgiform headache attacks (alternative criteria)**

*Alternative 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. Occurring with a frequency of at least one a day<sup>1</sup>
- E. Not better accounted for by another ICHD-3 diagnosis.

Note:

1. During part, but less than half, of the active time-course of A3.3 *Short-lasting unilateral neuralgiform headache attacks*, attacks may be less frequent.

**Comment:** Opinion is divided on inclusion of C5 and C6. Experts in the working group believe it improves sensitivity without significant loss of specificity, but formal field testing has not been performed to support the change in criteria.

**A3.4 Hemicrania continua (alternative criteria)**

*Alternative 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 indomethacin<sup>1</sup>
- 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.

**Comment:** Opinion is divided on inclusion of (e) and (f) in criterion C1. Experts in the working group believe it improves sensitivity without significant loss of specificity, but formal field testing has not been performed to support the change in criteria.

**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.

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## A4. Other primary headache disorders

### A4.11 *Epicrania fugax*

**Description:** Brief paroxysmal head pain, with stabbing quality, describing a linear or zigzag trajectory across the surface of one hemicranium.

#### Diagnostic criteria:

- A. Recurrent stabbing head pain attacks lasting 1–10 seconds and fulfilling criterion B
- B. Pain moving with a linear or zigzag trajectory across the surface of one hemicranium, commencing and terminating in the distributions of different nerves
- C. Not better accounted for by another ICHD-3 diagnosis.<sup>1</sup>

#### Note:

1. A structural lesion must be excluded by history, physical examination and, when appropriate, investigation.

**Comments:** Patients with A4.11 *Epicrania fugax* describe their pain in terms of its trajectory 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 pain usually moving forward from a posterior hemicranial area towards the ipsilateral eye or nose, but backward radiation is also possible from a frontal or periorbital area towards the occipital region. In all cases, pain is strictly unilateral, although some patients have shifting sides.

At the end of attacks, ipsilateral autonomic signs such as lacrimation, conjunctival injection and/or rhinorrhoea may occur.

Although attacks are mostly spontaneous, they may occasionally be triggered by touch on the point of onset, which may remain tender 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 seven days of head injury (or awareness of the injury) is somewhat arbitrary. Some data suggest that headache may begin after a longer interval. In the following suggested diagnostic criteria, the maximal time interval between the head injury and headache onset is set at three months, but it is presumed that headaches that begin in closer temporal proximity to the injury are more likely to be accurately attributed to the injury. Future studies should continue to investigate the utility of these and alternative diagnostic criteria for A5.1 *Acute headache attributed to traumatic injury to the head* that allow for headache to begin beyond seven days and up to three months 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 skull fracture, intracranial haemorrhage and/or brain contusion
- C. Headache is reported to have developed between seven days and three months after all of the following:
  - 1. the head injury
  - 2. regaining of consciousness following the head injury (when applicable)
  - 3. discontinuation of medication(s) impairing ability to sense or report headache following the head injury (when applicable)
- D. Either of the following:
  - 1. headache has resolved within three months after its onset
  - 2. headache has not yet resolved but three months have not yet passed since its onset
- 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 skull fracture, 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:
  - i. nausea
  - ii. vomiting
  - iii. visual disturbances
  - iv. dizziness and/or vertigo
  - v. gait and/or postural imbalance
  - vi. impaired memory and/or concentration
- C. Headache is reported to have developed between seven days and three months after all of the following:
  - 1. the head injury
  - 2. regaining of consciousness following the head injury (when applicable)
  - 3. discontinuation of medication(s) impairing ability to sense or report headache following the head injury (when applicable)
- D. Either of the following:
  - 1. headache has resolved within three months after its onset
  - 2. headache has not yet resolved but three months have not yet passed since its onset
- 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 seven days of head injury (or awareness of the injury) is somewhat arbitrary. Some data suggest that headache may begin after a longer interval. In the following suggested diagnostic criteria, the maximal time interval between the head injury and headache onset is set at three months, but it is presumed that headaches that begin in closer temporal proximity to the injury are more likely to be accurately attributed to the injury. Future studies should continue to investigate the utility of these and alternative diagnostic criteria for A5.2 *Persistent headache attributed to traumatic injury to the head* that allow for headache to begin beyond seven days and up to three months 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 skull fracture, intracranial haemorrhage and/or brain contusion
- C. Headache is reported to have developed between seven days and three months after all of the following:
1. the head injury
  2. regaining of consciousness following the head injury (when applicable)
  3. discontinuation of medication(s) impairing ability to sense or report headache following the head injury (when applicable)
- D. Headache persists for >3 months after its onset
- 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 skull fracture, 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:
        - i. nausea
        - ii. vomiting
        - iii. visual disturbances
        - iv. dizziness and/or vertigo
        - v. gait and/or postural imbalance
        - vi. impaired memory and/or concentration
- C. Headache is reported to have developed between seven days and three months after all of the following:
1. the head injury

2. regaining of consciousness following the head injury (when applicable)
  3. discontinuation of medication(s) impairing ability to sense or report headache following the head injury (when applicable)
- D. Headache persists for >3 months after its onset
- 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 seven days after radiosurgery
  2. headache has resolved within three 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 three months after its onset
  2. headache persists but three months have not yet passed since its onset
- 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 its onset
- E. Not better accounted for by another ICHD-3 diagnosis.

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## **A6. Headache attributed to cranial and/or cervical vascular disorder**

### **A6.10 Persistent headache attributed to past cranial and/or cervical vascular disorder**

- A. Headache previously diagnosed as 6. *Headache attributed to cranial and/or cervical vascular disorder* or one of its types, 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:* A6.10 *Persistent headache attributed to past cranial and/or cervical vascular disorder* is poorly documented; if it exists, 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 four hours after ECT
  3. each headache has resolved within 72 hours after ECT
- D. Not better accounted for by another ICHD-3 diagnosis.

**Comments:** Clear descriptions of headache associated with electroconvulsive therapy are sparse. In a single-blind comparator trial of eletriptan and paracetamol, 20 of 72 patients (28%) complained of headaches, but these were not well characterized (only location and quality of pain were assessed).

Published data are not adequate to define A7.6.3 *Post-electroconvulsive therapy (ECT) headache* operationally and there have been no validation studies of these proposed criteria since ICHD-3 beta was published.

### 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 types, 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 non-vascular disorder
- D. Not better accounted for by another ICHD-3 diagnosis.

**Comment:** It is known from clinical experience that persistent headache can occur after a past (and resolved) non-vascular intracranial disorder; to some extent such headache has been demonstrated after 7.1.1 *Headache attributed to idiopathic intracranial hypertension* and 7.2.3 *Headache attributed to spontaneous intracranial hypotension*. However, A7.9 *Persistent headache attributed to past non-vascular intracranial disorder* is 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.

### A9.3 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 according to the latter. Headache caused by use of antiretroviral drugs should be coded as 8.1.10 *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:** The rationale for separating A9.3 *Headache attributed to human immunodeficiency virus (HIV) infection* from headaches attributed to other infections is threefold:

- 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.

Headache is reported by more than half of people infected by 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). Nevertheless, A9.3 *Headache attributed to human immunodeficiency virus (HIV) infection* remains within the Appendix 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.

In most cases, A9.3 *Headache attributed to human immunodeficiency virus (HIV) infection* is dull and bilateral, or has the features of a primary headache disorder (1. *Migraine* or 2. *Tension-type headache*). Headache severity, frequency and attributed disability seem to be 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.

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 8.1.10 *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 at least 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 has travelled 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 homeostasis**

*Diagnostic criteria:*

- A. Headache previously diagnosed as 10. *Headache attributed to disorder of homeostasis*, and fulfilling criterion C
- B. The disorder of homeostasis 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 homeostasis
- 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, electrodiagnostic 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. tenderness is elicited by pressure on the implicated cervical muscles
  - 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.

**A11.3 Headache attributed to disorder of the eyes**

**A11.3.5 Headache attributed to heterophoria or heterotropia**

*Description:* Headache caused by latent or persistent strabismus (squint), usually occurring after prolonged visual tasks.

**Diagnostic criteria:**

- A. Headache fulfilling criterion C
- B. Heterophoria or heterotropia 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 heterophoria and/or heterotropia, or led to its discovery
  2. headache has significantly improved after correction of the heterophoria and/or heterotropia
  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.
  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

**Note:**

1. 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 mostly lacking. In the vast majority of cases, probably, headache associated with these disorders reflects common underlying risk factors or aetiologies rather than a causal relationship. However, in order to make any of the diagnoses listed below, a causal relationship between the headache and the psychiatric disorder in question must be established. Thus, the headache either develops simultaneously with the psychiatric disorder or significantly worsens after the psychiatric disorder becomes evident.

Definite biomarkers and clinical proof of headache causation are difficult to obtain, and the diagnosis will often be one of exclusion. For example, in a child with separation anxiety disorder, headache should be attributed to this disorder only in those cases where it occurs *exclusively* 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 *exclusively* as one of the symptoms of a panic attack.

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 associations between headache and comorbid psychiatric disorders.

**A12.3 Headache attributed to depressive disorder****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 cavity<sup>1</sup>
- 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, or led to its discovery
  2. headache has significantly improved or significantly worsened in parallel with improvement in (with or without treatment) or worsening of the nasal lesion

**Comments:** There are a number of supportive cases for A11.3.5 *Headache attributed to heterophoria or heterotropia* but otherwise little evidence for this cause of headache. It has therefore been moved to the Appendix pending more formal study.

Patients with 11.3.5 *Headache attributed to heterophoria or heterotropia*, if it exists, are likely to seek advice from an ophthalmologist.

**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 cavity<sup>1</sup>
- 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, or led to its discovery
  2. headache has significantly improved or significantly worsened in parallel with improvement in (with or without treatment) or worsening of the nasal lesion

- 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 types of antidepressant 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 six months, although shorter durations may be consistent with 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. A 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 for six months or more, 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 exclusively when the patient is exposed or anticipating exposure to social situations
- D. Not better accounted for by another ICHD-3 diagnosis.

*Comments:* 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. The fear or anxiety is out of proportion to the actual threat posed by the social situation. 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 disorder is persistent, typically lasting for six months or more.

### 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 exclusively 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 three months or more. Symptoms may include restlessness or feeling excited, tense or nervous, 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 (PTSD)

#### 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 exclusively in the context of other symptoms of PTSD<sup>1</sup>
- D. Not better accounted for by another ICHD-3 diagnosis.<sup>2</sup>

#### Notes:

1. For example, headache occurs upon exposure to reminders of the trauma.
2. In particular, A12.3 *Headache attributed to depressive disorder*.

*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 patients whose headache is not explained by comorbid depression (i.e. cases of headache attributed to PTSD in patients *without* comorbid depression).



## Definitions of terms

*Accompanying symptoms:* Symptoms that typically accompany rather than precede or follow headache. In migraine, for example, the most frequent accompanying symptoms are nausea, vomiting, photophobia and phonophobia.

*Allodynia:* Sensation of discomfort or pain (qv) arising from a stimulus that would not normally be sufficient to have this effect. It is distinguished from hyperalgesia (qv).

*Anorexia:* Lack of appetite and dislike for food to a mild degree.

*Attack of headache (or pain):* Headache (or pain) (qv) that builds up, remains at a certain level for minutes, hours or days, then wanes until it has resolved completely.

*Attributed to:* This term in ICHD-3 describes the relationship between a secondary headache (qv) and the disorder believed to cause it. It requires fulfilment of criteria establishing an accepted level of evidence of causation.

*Aura:* Early symptoms of an attack of migraine with aura, believed to be the manifestations of focal cerebral dysfunction. The aura typically lasts 20–30 minutes and precedes the headache (qv). See also: *Focal neurological symptoms, Premonitory symptoms, Prodrome and Warning 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* signifies long-lasting, specifically over a period exceeding three months. In headache terminology, it retains this meaning for secondary headache disorders (notably those attributed to infection) in which the causative disorder is itself chronic. In this usage, *chronic* is distinguished from *persistent* (qv). 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 three 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 describes 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 (also referred to as *cluster bout*).

*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 three months.

*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*).

*Enhanced entoptic phenomena:* Visual disturbances arising from the structure of the visual system itself, including excessive floaters in both eyes, excessive blue field entoptic phenomenon (uncountable little grey/white/black dots or rings shooting over the visual field of both eyes when looking at homogeneous bright surfaces such as the blue sky), self-lighting of the eye (coloured waves or clouds perceived when closing the eyes in the dark) and spontaneous photopsia (bright flashes of light).

*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 (qv) separated by cluster remission periods (qv) rather than to attacks. Similar usage has been adopted for paroxysmal hemicrania and short-lasting unilateral neuralgiform headache attacks.

*Facial pain:* Pain below the orbitomeatal line, anterior to the pinnae and above the neck.

*Focal neurological symptoms:* Symptoms of focal brain (usually cerebral) disturbance such as occur in migraine aura (qv).

*Fortification spectrum:* Angulated, arcuate and gradually enlarging visual disturbance typical of migrainous visual aura, which can be coloured or black and white.

*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: Third Edition*, recommend as a practical solution, especially in differentiating attacks recorded as diary entries over the previous month, to count as distinct attacks only those that are separated by at least 48 hours headache-free.

*Headache:* Pain (qv) located in the head, above the orbitomeatal line and/or nuchal ridge.

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

*Heterophoria:* Latent strabismus (squint).

*Heterotropia:* Manifest strabismus (squint).

*Hypalgesia:* Diminished perception in response to a stimulus expected to be painful.

*Hyperalgesia:* Heightened perception in response to a stimulus expected to be painful. Hyperalgesia is distinguished from allodynia (qv), arising from a stimulus not expected to be painful.

*Intensity of pain:* Level of pain (qv), usually scored on a four-point numerical rating scale (0–3) equivalent to no, mild, moderate and severe pain, or on a visual analogue scale (commonly 10 cm). It may also be scored on a verbal rating scale expressed in terms of its functional consequence: 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.

*Lancinating:* Brief, electric-shock-like character of pain (qv), along a root or nerve distribution.

*Neuralgia:* Pain (qv) in the distribution(s) of a nerve or nerves, presumed to be due to dysfunction or injury of those neural structures. Common usage has implied 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, positron emission tomography (PET), single-photon emission computed tomography (SPECT) or scintigraphy, including functional modalities where applicable, usually of the brain.

*Neuropathic pain:* Pain (qv) caused by a lesion or disease of the peripheral or central 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, axonotmesis, 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

classification committee members and/or controversy in the literature.

*Nuchal region:* Dorsal (posterior) aspect of the upper neck, including the region of insertion of neck muscles on the cranium.

*Nyctalopia:* Impaired night vision.

*Pain:* According to the IASP definition: 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*).

*Palinopsia:* Visual disturbances in the form of after-images and/or trailing images of moving objects (to be distinguished from *retinal after-images*, which occur, in complementary colour, after staring at a high contrast image).

*Pericranial muscles:* Neck and occipital 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*).

*Persistent:* This term, used in the context of certain secondary headaches, describes headache, initially acute and caused by another disorder, that fails to remit within a specified time interval (usually three months) after that disorder has resolved. In many such cases, the headache is recognized as a distinct subtype or subform, with evidence of causation depending upon earlier fulfilment of the criteria for diagnosis of the acute type, and persistence of the same headache.

*Phonophobia:* Hypersensitivity to sound, even at normal levels, usually causing avoidance.

*Photophobia:* Hypersensitivity to light, even at normal levels, usually causing avoidance.

*Postdrome:* A symptomatic phase, lasting up to 48 hours, following the resolution of pain in migraine attacks with or without aura. Among the common postdromal symptoms are feeling tired or weary, difficulty with concentration and neck stiffness.

*Premonitory symptoms:* This term has been used with different meanings, often synonymously with *prodrome* (qv) but also, less specifically and somewhat ambiguously, for a range of symptoms believed to forewarn of (but possibly the initial phase of) a migraine attack. The term is better avoided.

*Pressing/tightening:* Pain (qv) of a constant quality, often compared to a tight 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.

*Primary headache (disorder):* Headache, or a headache disorder, not caused by or attributed to another disorder. It is distinguished from secondary headache disorder (qv).

*Prodrome:* A symptomatic phase, lasting up to 48 hours, occurring before the onset of pain in migraine without aura or before the aura in migraine with aura. Among the common prodromal symptoms are fatigue, elated or depressed mood, unusual hunger and cravings for certain foods.

*Pulsating:* Characterized by rhythmic intensifications in time with the heart beat; throbbing.

*Punctate stimuli:* Stimuli applied to discreet points on the skin.

*Referred pain:* Pain (qv) perceived in another area than the one where nociception arises.

*Refraction (or refractory) error:* Myopia, hypermetropia or astigmatism.

*Refractory period:* The time following resolution of an attack of pain (qv) during which a further attack cannot be triggered.

*Resolution:* Complete remission of all symptoms and other clinical evidence of disease or a disease process (such as an attack of headache [qv]).

*Scintillation:* Visual hallucinations that are bright and fluctuate in intensity, often at approximately 8–10 cycles/second. They are typical of migraine aura (qv).

*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). In migraine, scotomata are homonymous.

*Secondary headache (disorder):* Headache, or a headache disorder, caused by another underlying disorder. In ICHD-3, secondary headaches are *attributed to the causative disorder*. Secondary headaches are

distinguished from primary headaches (qv). A secondary headache may have the characteristics of a primary headache but still fulfil criteria for causation by another disorder.

*Stab of pain:* Sudden pain (qv) lasting a minute or less (usually a second or less).

*Strabismus:* Abnormal alignment of one or both eyes (squint).

*Substance:* Any of the following: organic or inorganic chemical; food or additive; alcoholic beverage; gas or vapour; drug or medication or herbal, animal or other substance given with medicinal intent although not licensed as a medicinal product.

*Tenderness:* A heightened feeling of discomfort or pain caused by direct pressure such as is applied during palpation.

*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, the term 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 (qv) or premonitory symptoms (qv), and therefore ambiguous. It should not be used.

*Withdrawal:* Interruption in use of or exposure to a medication or other substance that has lasted for weeks or months. The term embraces but is not limited to therapeutic withdrawal (cessation) of medication in the context of medication-overuse headache.

*Zigzag line:* Synonym for fortification spectrum (qv).